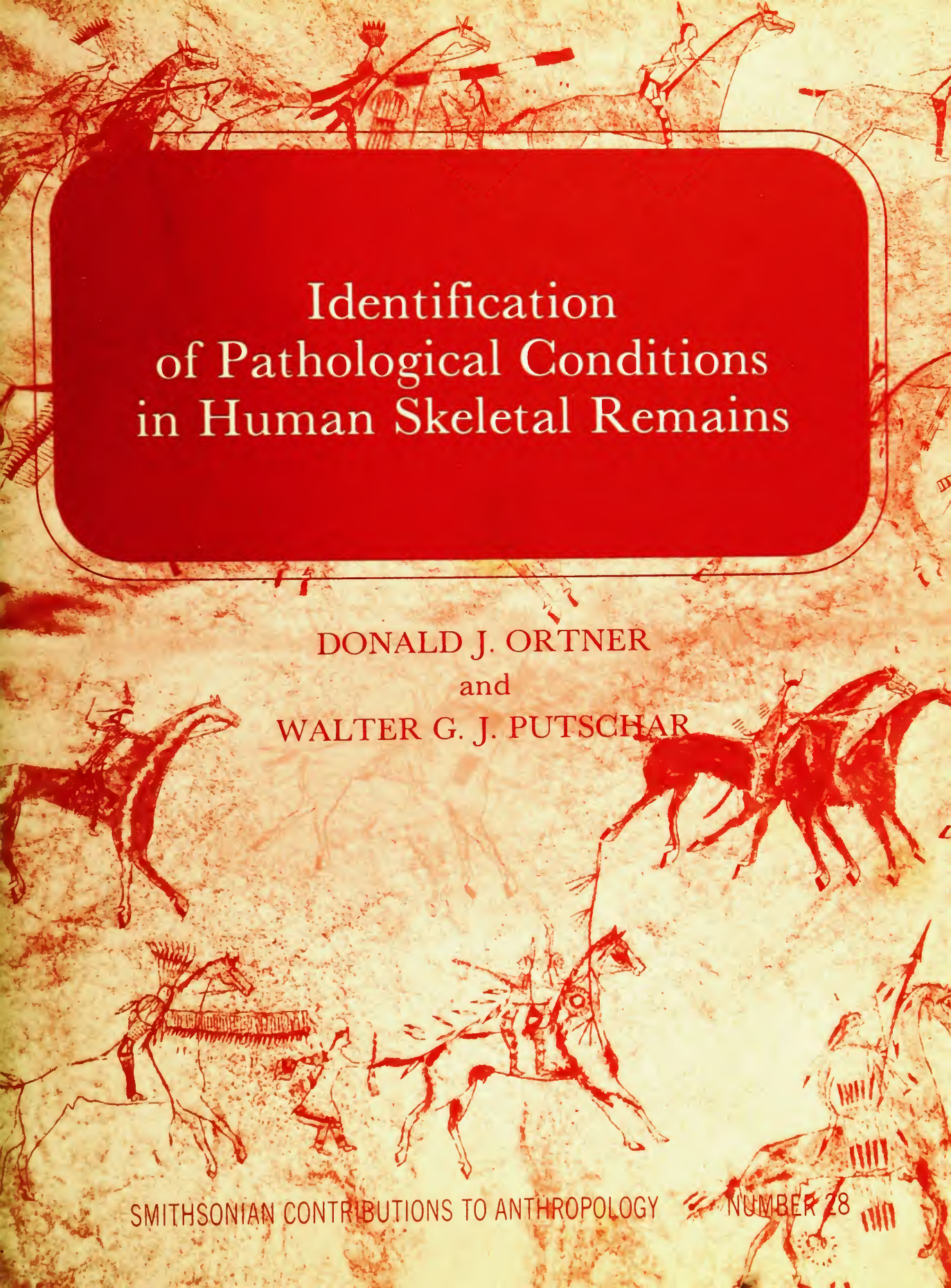

Identification of Pathological Conditions in Human Skeletal Remains

DONALD J. ORTNER
and
WALTER G. J. PUTSCHAR

SMITHSONIAN CONTRIBUTIONS TO ANTHROPOLOGY · NUMBER 28

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SMITHSONIAN CONTRIBUTIONS TO ANTHROPOLOGY • NUMBER 28

Identification of Pathological Conditions in Human Skeletal Remains

Donald J. Ortner

and Walter G. J. Putschar

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ABSTRACT

Ortner, Donald J., and Walter G. J. Putschar. Identification of Pathological Conditions in Human Skeletal Remains. *Smithsonian Contributions to Anthropology*, number 28, 479 pages, 14 tables, 765 figures, 1981.—This reference work is an attempt to provide an integrated and reasonably comprehensive treatment of pathological conditions that affect the human skeleton. The primary objective is to assist those who conduct research on archeological skeletal remains in interpreting abnormal conditions that they might encounter in the course of their research. However, there is much that ancient skeletal remains can reveal to the modern medical historian, orthopaedist, pathologist, and radiologist about skeletal diseases that are rarely encountered in modern clinical practice.

All of the major categories of disease that affect bone are reviewed from the viewpoint of the pathologist. This review is followed by a discussion of the literature on the paleopathology of each condition and the presentation of paleopathological cases thought to represent each of the morbid categories affecting bone.

This work is based on extensive individual and collaborative research by both authors on the known parameters of modern skeletal diseases and their expression in antiquity. The monograph provides essential text and illustrative materials on bone pathology, which will improve the diagnostic ability of those interested in human dry bone pathology.

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Contents

	Page
INTRODUCTION	1
Objectives of this Book	1
Acknowledgments	3
Abbreviations	4
A Brief History of Paleopathology	5
The Paleopathology Program at the Smithsonian Institution	6
THE BIOLOGY OF SKELETAL TISSUES	8
The Development of Bone	8
Cartilage	12
The Cells of Bone	13
Bone Growth	16
Osteon Remodeling	19
The Biology of Teeth	26
METHODS USED IN THE ANALYSIS OF SKELETAL LESIONS	29
Age Determination	30
Race Determination	32
Sex Determination	32
Stature Determination	33
Demographic Analysis	33
Archeological Documentation	34
Gross Study of Skeletal Lesions	36
X-ray Film Study of Skeletal Lesions	45
Chemical Analysis of Pathological Bone	51
Microscopic Study of Skeletal Lesions	52
TRAUMA	55
Fracture	55
Pathology	55
Types of Fracture	55
Fracture Healing	61
Complications Arising from Fracture	64
Paleopathology	72
History and Examples	72
Complications of Fracture	81
Dislocation	85
Pathology	85
Paleopathology	87
Deformation	90
Pathology	90
Paleopathology	90

Page

Scalping ... 92
Pathology ... 92
Paleopathology ... 93
Mutilation ... 94
Trephination ... 95
History ... 95
Paleopathology ... 96
Types of Trephination ... 97
Complications of Trephination ... 97
Traumatic Problems Arising from Pregnancy ... 100
Pathology ... 100
Paleopathology ... 101
Sincipital-T Mutilation ... 102
INFECTIOUS DISEASES ... 104
The Inflammatory Response ... 104
The Biology of Infection ... 104
Osteomyelitis ... 105
Pathology ... 105
Hematogenous Osteomyelitis ... 109
Changes in Individual Bones ... 116
Paleopathology ... 121
The Skull ... 123
Postcranial Bones ... 123
Periostitis ... 129
Pathology ... 129
Paleopathology ... 131
Brucellosis ... 138
Pathology ... 138
Paleopathology ... 139
Glanders ... 141
Pathology ... 141
Tuberculosis of Bones and Joints ... 141
Pathology ... 141
Statistical Data ... 142
General Pattern of Bone and Joint Tuberculosis ... 144
The Spine ... 145
The Pelvis ... 149
The Hip ... 150
The Trochanter ... 153
The Knee ... 154
The Ankle and Tarsal Bones ... 154
The Tubular Bones of the Hands and Feet (Spina Ventosa) ... 156
The Shoulder ... 157
The Elbow ... 157
The Wrist and Carpal Bones ... 159
The Shaft of Long Bones ... 159

	Page
The Thoracic Cage	162
The Skull	162
Paleopathology	166
Leprosy	176
Pathology	176
Paleopathology	177
Treponemal Infections	180
Pathology	180
Yaws	180
Endemic Syphilis (Treponarid)	181
Venereal Syphilis	182
Congenital Syphilis	198
Paleopathology	201
Congenital Syphilis	207
Adult Treponemal Disease	210
Actinomycosis and Nocardiosis	218
Pathology	218
Mycetoma (Maduromycosis)	222
Pathology	222
Fungal Infections	224
Pathology	224
North American Blastomycosis	224
Cryptococcosis	224
Paracoccidioidomycosis	224
Coccidioidomycosis	224
Histoplasmosis	225
Sporotrichosis	225
Aspergillosis	226
Mucormycosis (Phycomycosis)	227
Paleopathology	227
Viral Infections	227
Pathology	227
Smallpox	227
Rubella	229
Parasitic Infections (Echinococcosis)	229
Pathology	229
Paleopathology	232
Sarcoidosis	233
Pathology	233
CIRCULATORY DISTURBANCES	235
Blood Supply of Bones	235
Ischemia and Infarction	235
Pathology	235
Necrosis of Femoral Head	236
Pathology	236
Paleopathology	237

	Page
Perthes' Disease and Slipped Femoral Capital Epiphysis	238
Pathology	238
Paleopathology	239
Other Vascular Disturbances of Epiphyses and Apophyses	242
Köhler's Disease of the Tarsal Navicular Bone	242
Pathology	242
Osteochondritis Dissecans	242
Pathology	242
Paleopathology	242
Osgood-Schlatter's Disease of the Tibial Tubercle	243
Pathology	243
Paleopathology	243
Freiberg's Disease of a Metatarsal Head	244
Pathology	244
Kienböck's Disease of the Carpal Lunate Bone	244
Pathology	244
Pulmonary Osteoarthropathy	245
Pathology	245
Aneurysmal Erosion	246
Pathology	246
RETICULOENDOTHELIAL AND HEMOPOIETIC DISORDERS	248
Lipid Storage Diseases	248
Pathology	248
Gaucher's Disease	248
Niemann-Pick's Disease	249
Other Lipidoses	249
Histiocytosis X	249
Pathology	249
Anemias	251
Pathology	251
Thalassemia	251
Sickle Cell Anemia and Its Genetic Variants	254
Hereditary Spherocytosis (Congenital Hemolytic Anemia)	257
Iron-deficiency Anemia	257
Erythroblastosis Fetalis	258
Paleopathology	258
Leukemia	263
Pathology	263
Myeloma	264
Pathology	264
Paleopathology	265
METABOLIC DISORDERS	270
Vitamin C Deficiency	270
Pathology	270
Infantile Scurvy (Möller-Barlow's Disease)	270
Adult Scurvy	272

	Page
Paleopathology	272
Vitamin D Deficiency	273
Pathology	273
Rickets	273
Rachitic and Postrachitic Changes in Individual Bones	274
Late Rickets	280
Osteomalacia	280
Skeletal Changes Resembling Rickets and Osteomalacia	283
Paleopathology	283
Hypophosphatasia	287
Pathology	287
Starvation	287
Pathology	287
Fluorosis	288
Pathology	288
Osteoporosis	289
Pathology	289
Paleopathology	291
Localized Hyperostosis	294
Pathology	294
Pregnancy Osteophyte	294
Internal Frontal Hyperostosis	294
Leontiasis Ossea	294
Infantile Cortical Hyperostosis (Caffey's Disease)	294
Generalized Hyperostosis with Pachydermia	294
Pathology	294
Paleopathology	297
ENDOCRINE DISTURBANCES	298
Pituitary Disturbances	298
Pathology	298
Pituitary Gigantism	298
Acromegaly	298
Hypopituitarism	300
Pituitary Dwarfism	300
Paleopathology	300
Acromegaly	300
Pituitary Dwarfism	302
Other Endocrine Disturbances	304
Pathology	304
Hypothyroidism	304
Hyperthyroidism	305
Cushing's Syndrome	305
Hypogonadism	306
Hypergonadism	306
Hypoparathyroidism	306
Hyperparathyroidism	307

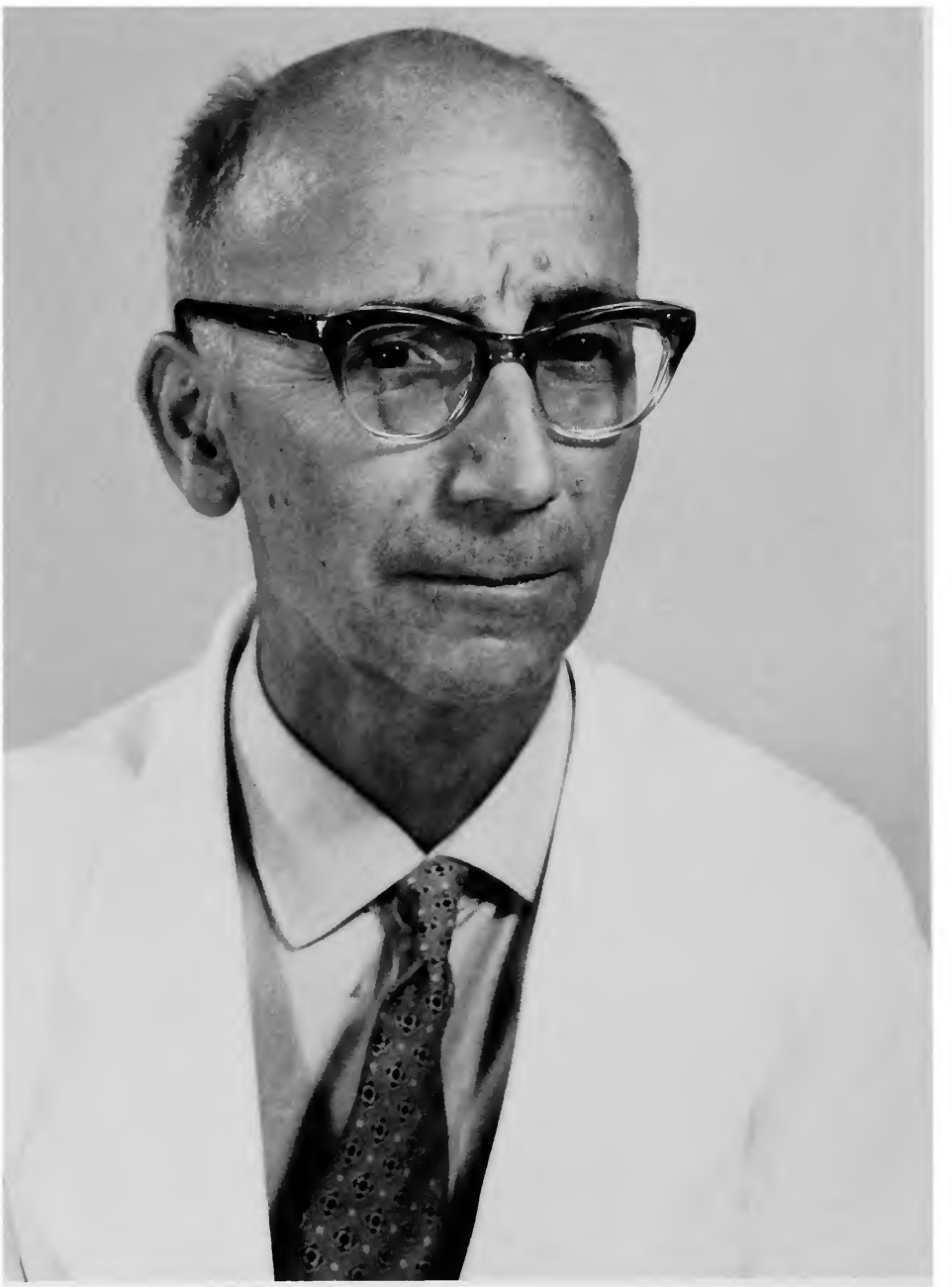
	Page
OTHER BONE DISEASES	309
Paget's Disease	309
Pathology	309
Paleopathology	315
Fibrous Dysplasia	315
Pathology	315
Monostotic Fibrous Dysplasia	317
Polyostotic Fibrous Dysplasia	317
Paleopathology	317
Myositis Ossificans Progressiva	322
Pathology	322
NEUROMECHANICAL DEFORMITIES	323
Pathology	323
Kyphosis	323
Scoliosis	324
Postparalytic Deformities	325
Paleopathology	326
Scoliosis	326
Postparalytic Deformities	328
SKELETAL DYSPLASIAS	329
Achondroplasia	329
Pathology	329
Paleopathology	331
Mucopolysaccharidoses	334
Pathology	334
Paleopathology	336
Osteogenesis Imperfecta	337
Pathology	337
Paleopathology	338
Dysostosis Cleidocranialis	338
Pathology	338
Osteopetrosis	340
Pathology	340
Pyknodysostosis	342
Pathology	342
Metaphysial Dysplasia (Pyle's Disease)	342
Pathology	342
Paleopathology	342
Diaphysial Sclerosis (Camurati-Engelmann's Disease)	344
Pathology	344
Melorheostosis (Leri's Disease)	344
Pathology	344
Paleopathology	345
Osteopoikilosis	345
Pathology	345
Osteopathia Striata	345
Pathology	345

	Page
Skeletal Malformations	346
Skull	346
Pathology	346
Paleopathology	346
Spine	355
Pathology	355
Paleopathology	356
Ribs	359
Pathology	359
Pelvis	359
Pathology	359
Paleopathology	360
Extremities	362
Pathology	362
Paleopathology	363
Tumors	365
General Discussion	365
Pathology	365
Paleopathology	365
Primary Benign Tumors	366
Pathology	366
Cysts	366
Osteoblastic Tumors	368
Chondroblastic Tumors	370
Fibroblastic Tumors	374
Giant Cell Tumor	375
Hemangioma	376
Meningioma	378
Paleopathology	378
Osteoblastic Tumors	378
Chondroblastic Tumors	380
Meningioma	382
Primary Malignant Bone Tumors	384
Pathology	384
Osteosarcoma	384
Chondrosarcoma	385
Ewing's Sarcoma	387
Chordoma	388
Paleopathology	389
Metastatic Tumors	391
Pathology	391
Paleopathology	395
Lesions of the Joints	399
Septic Arthritis	399
Pathology	399
Paleopathology	401

	Page
Rheumatoid Arthritis	403
Pathology	403
Adult Rheumatoid Arthritis	403
Juvenile Rheumatoid Arthritis (Still's Disease)	405
Psoriatic Arthritis	405
Paleopathology	405
Ankylosing Spondylitis (Marie-Strümpell's Disease)	411
Pathology	411
Paleopathology	412
Metabolic Arthritis	415
Pathology	415
Gouty Arthritis	415
Ochronosis (Alkaptonuria)	416
Hemophilic Arthropathy	417
Hemophilic Pseudotumor	418
Paleopathology	418
Gouty Arthritis	418
Ochronosis (Alkaptonuria)	418
Degenerative Arthritis	419
Pathology	419
The Appendicular Skeleton	419
Spondylitis Deformans (Spinal Osteophytosis)	420
Paleopathology	422
The Appendicular Skeleton	423
The Axial Skeleton	429
Other Lesions of the Joints	433
Pathology	433
Neuropathic Arthropathy (Charcot's Joint)	433
Pigmented Villonodular Synovitis	435
Synovial Osteochondromatosis	435
Lesions of Jaws and Teeth	436
The Jaws	436
Pathology	436
Odontogenic Cysts	436
Odontogenic Tumors	436
Nonodontogenic Cysts	437
Nonodontogenic Tumors	437
The Teeth	438
Dental Caries	438
Pathology	438
Paleopathology	439
Periodontal Disease	442
Pathology	442
Paleopathology	443
Disturbances in Dental Development	444

	Page
Pathology	444
Abnormal Quality of Teeth	444
Abnormal Quantity of Teeth	446
Abnormal Size and Shape of Teeth	447
Dental Anomalies	447
Dental Crowding	447
Paleopathology	447
Abnormal Quality of Teeth	447
Abnormal Quantity of Teeth	448
Abnormal Size and Shape of Teeth	451
Dental Anomalies	451
Dental Crowding	452
Dental Trauma	452
Pathology	452
Paleopathology	453
Dental Attrition	454
Pathology	454
Paleopathology	454
Dental Discoloration	456
Pathology	456
Paleopathology	456
LITERATURE CITED	457

This book is dedicated
to the memory of
ERWIN UEHLINGER
Emeritus Professor of Pathology
of the University of Zurich (Switzerland)
the dean of European bone pathologists
whose continued interest and advice
from the inception of this book
was a great inspiration



Erwin Uehlinger
8 August 1899 – 18 April 1980

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Donald J. Ortner
and Walter G. J. Putschar

Introduction

Human biological history involves the interaction of countless individuals and their environment. This dynamic condition includes extraordinarily complex interrelationships, not only among peoples but between individuals and a host of physical and biological factors that affect their well-being. One of the major objectives of the biologist is to discover and record this history. However, any attempt to write even a partial biological history of man depends ultimately on our knowledge of the factors that influence it.

Even the most superficial review of the history of disease reveals its enormous significance in human biological history. The terrifying plagues recorded in European history prematurely ended the lives of millions of people. The influenza epidemic after World War I resulted in the death of over twenty million people—far more than died as a direct result of that most devastating and bloody war.

Why do some people survive a morbid condition and others die? Clearly chance factors operate. Genetics and its influence on several biological factors, including the immune response, must be significant. To the extent that heritable factors play a role in resistance to disease, morbid conditions influence the continuing process of human biological adaptation. Similarly the existence of cultural mechanisms that minimize the effect of disease may be of considerable biological importance. Thus, our understanding of man's cultural and biological response to disease through time becomes an important dimension of our understanding of human history.

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Walter G. J. Putschar, Department of Pathology, Massachusetts General Hospital, Boston, Massachusetts 02114.

Objectives of this Book

The genesis of this book began in 1970, at which time plans were developed for the first Paleopathology Seminar Series to be held in 1971. This seminar series brought several leading authorities on skeletal disease, paleopathology, and related subjects to the Smithsonian Institution to present lectures to a select group of scholars interested in skeletal paleopathology.

The seminar series was held yearly through 1974. By that time the logistics of obtaining funds to offer the series, arranging for students to come

from many universities, including foreign ones, and assembling an outstanding faculty for the ten-week series became almost overwhelming. Dr. Donald J. Ortner, physical anthropologist and director of the series, and Dr. Walter G. J. Putschar, pathologist and the principal lecturer on skeletal pathology, decided that many more scholars interested in skeletal paleopathology would have access to the substance of the seminar series if the information were incorporated into a book. This book would reflect the content of the seminar series, which had included lectures on calcified tissue biology, methodology, pathology, and radiology, and laboratory sessions on paleopathology, all emphasizing the human skeleton.

There are many sources of information on the history of disease including ancient medical documents, historical records, art, and the physical remains of ancient people, both soft tissues and skeletons. Undoubtedly human skeletons represent the most ubiquitous source of information on ancient diseases. Unfortunately this fact must be tempered with the knowledge that relatively few morbid conditions affect the skeleton in a way that leaves visible changes in the dry bones. In spite of this limitation, the study of skeletal pathology in archeological materials can provide time depth to our understanding of disease and contribute to our understanding of the role of disease in human adaptation. In addition, skeletal paleopathology may also broaden our understanding of disease as it affects bone at the present time. The paleopathologist generally has access to all portions of the skeleton, a situation rarely realized in modern pathology. This means that the gross pattern and distribution of the morbid condition in all areas of the skeleton can be studied in detail. A good example of such a study is the work by Møller-Christensen (1967) on the medieval human skeletons from a leper cemetery in Denmark, which revealed hitherto not fully appreciated skeletal manifestations of the disease. Undoubtedly more careful analysis of paleopathological skeletons will reveal additional information on the biology of other bone diseases.

It is against the background of the substantial literature in paleopathology (exceeding 3000 citations) and the scholarly needs revealed by this literature that we have formulated the objectives of this book. The most fundamental objective is to provide an integrated detailed discussion of the gross pathology of the human skeleton, emphasizing those conditions that can be seen in dry specimens regardless of whether or not they have so far been identified in archeological material. In order to provide reliable standard specimens for dry bone diagnosis, we had to concentrate on the period from about AD 1750 to 1930. Earlier, the medical data are too ambiguous and, later, the pathologic manifestations are too altered by surgery, chemotherapy, radiation therapy and, above all, by the use of antibiotics. For this purpose we had to turn to the great medical museums of Great Britain and of continental Europe. The British collections proved in many ways the most useful, because they were made by physicians and surgeons, who were at all times interested in documenting clinical and historical data. We realize that even this material, which represents as closely as possible the natural history of the affections at that period in time and which are not modified by significant therapeutic procedures, is not necessarily identical with previous manifestations seen in archeological specimens. However, it is the only documented material available for comparison.

This book is mainly intended to serve as a text and atlas of dry bone pathology, regardless of whether or not each entity has been identified in paleopathology. For that reason, as many aspects as possible of documented, dry bone pathology have been illustrated, especially because the original skeletal collections can never be duplicated and may ultimately disappear. In the paleopathological discussions, emphasis is laid on careful and critical study of published reports and of actual specimens, bringing all types of evidence to bear on arriving at a reasonable diagnostic assumption. It hardly needs to be emphasized that, even so, multiple possibilities and uncertainties often remain. Not the least of these problems

is the ambiguous and confusing terminology about the chronology of archeological specimens in published reports. Chronological information on such specimens is reported in the following pages as it was reported in the sources.

The different disease categories are treated in separate chapters as is the discussion of calcified tissue biology and methodology. Within the chapters on skeletal pathology the clinical background is presented first in condensed form. This is followed by a detailed discussion of bone pathology and of the differential diagnosis. Histologic detail, since not pertinent to the study of dry bones, is kept at a minimum. The paleopathological aspects, unpublished cases, and pertinent published reports follow the discussion of the pathology within each chapter. The introduction was written jointly. The chapters on skeletal biology, methods, trauma, and dental diseases (except the section "The Jaws") were written by D. J. Ortner. In all other chapters the pathology was written by W. G. J. Putschar and the paleopathology by D. J. Ortner.

The book is written primarily with the needs of the anthropologist and archeologist in mind, with the hope that they will be able to recognize the abnormalities seen in human skeletal material. We are, however, interested in a broader readership with different backgrounds, including orthopaedic surgeons, radiologists, pathologists, and physicians who may be called upon to interpret skeletal lesions in dry specimens or who are interested in extending their understanding to the more detailed gross expressions of skeletal disease.

Acknowledgments

As a first step in preparing this book, Drs. Ortner and Putschar conducted, in 1974, an extensive survey of documented skeletal pathology in 16 of the European pathology and anthropology collections. This survey was supported by the Smithsonian Research Foundation and Hrdlička Fund and took the authors to six countries. The following list of these institutions and the staff members who gave assistance to the survey of their collections is an inadequate recognition of the many courtesies extended during our work. However, we wish by this method to express our appreciation for the cooperation we received in studying and photographing the skeletal specimens: *Austria*: Federal Pathologic-Anatomy Museum, Vienna (Dr. Karl von Portele, Dr. Alexander Müller); Pathology Museum of the University of Graz (Prof. Dr. Max Ratzenhofer); Pathology Museum of the University of Innsbruk (Prof. Dr. Albert Probst, Prof. Dr. Josef Thurner (Salzburg, Austria)). *Czechoslovakia*: National Museum, Department of Anthropology, Prague (Dr. Emanuel Vlček, Dr. Milan Stloukal, Dr. H. Hanākovā). *England*: British Museum (Natural History), London (Dr. Theya Molleson, Miss Rosemary Powers); Guy's Hospital Medical School, Gordon Pathology Museum, London; The Royal College of Surgeons of England, Wellcome Museum, London (Dr. Martin S. Israel); The Royal College of Surgeons of England, Hunterian Museum, London (Miss Elizabeth Allen); St. George's Hospital Medical School, Pathology Museum, London; Westminster Hospital School of Medicine, Pathology Museum, London. *France*: University of Strasbourg, Department of Pathology, Museum (Prof. Y. Le Gal, Prof. André Batzenchlager). *Scotland*: The Royal College of Surgeons of Edinburgh (Prof. Eric C. Mekie, Dr. Andrew A. Shivas, Mrs. Violet Tansy, Mrs. Turner, Mr. McKenzy). *Switzerland*: Anthropological Institute of the University of Zurich, (Dr. Wolfgang Scheffrahn); Historical Museum, Chur (Dr. H. Erb); Institute of Pathological Anatomy of the University of Zurich (Prof. Dr. Erwin Uehlinger, Prof. Dr. Christoph E. Hedinger, Mr. Aschwanden); Natural History Museum, Bern (Prof. Dr. Walter Huber). Dr. Cecil J. Hackett, an associate of the Royal Orthopaedic Hospital, did much to expedite our work in London, England, and offered several helpful suggestions regarding collections in other countries that proved valuable to our study.

The product of our survey was more than 1200 photographs, in both black and white and color, taken by D. J. Ortner, of approximately 500

pathological specimens jointly studied. For some cases we were able to obtain X-ray films as well. W. G. J. Putschar described the specimens in detail on tape and included original autopsy and clinical data where available. This collection of photographs, X-rays, and the transcripts of case descriptions is available for study at the Department of Anthropology, National Museum of Natural History, Smithsonian Institution, Washington, D.C. Many of them are used as illustrations in this book.

A number of people have made significant contributions in the preparation of the manuscript. Mrs. Paula Cardwell, Mrs. Eleanor Haley and particularly Mrs. Katharine Holland typed initial drafts. Mrs. Marguerite Brigida and Mrs. Elizabeth Beard typed the final draft. Mrs. Marcia Bakry prepared some of the drawings. To Ms. Jacqui Schulz goes a special note of appreciation for the many unpaid hours spent in preparing the remaining drawings and getting the photographic illustrations ready for publication. Photographic enlargements were prepared by Mr. H. E. Daugherty and Mrs. Agnes Stix. Mrs. Stix also assisted in editing and typing the manuscript. Mr. David Yong, Mr. Edward Garner and Mr. Dwight Schmidt provided valuable technical assistance. The staff of the library of the Smithsonian Institution, particularly Mrs. Janette Saquet, was most helpful. Drs. J. Lawrence Angel, T. Dale Stewart, and Douglas H. Ubelaker, Department of Anthropology, Smithsonian Institution, have made valuable suggestions, as have Dr. Saul Jarcho, New York City, USA, and Dr. George Armelagos, University of Massachusetts, Amherst, Massachusetts, USA. The staff of the Smithsonian Institution Press, particularly Mr. Albert L. Ruffin, Jr., Managing Editor, Series Publications, and Ms. Joan B. Horn, senior editor, deserve special recognition for their assistance from the conceptualization through publication of the book.

Finally the wives of both authors have been intimately involved with the preparation of the book. Mrs. Florence Putschar has spent hundreds of volunteer hours organizing photographs, typing, preparing the bibliography, editing, and otherwise making her remarkable abilities available to the project. Mrs. Joyce Ortner has also assisted in obtaining illustrative material and skeletal specimens used in the book.

Abbreviations

The illustrations in this book are of specimens from many institutions. The following abbreviations are used in the legends to avoid repetition of lengthy institutional names and locations.

AFIP	Armed Forces Institute of Pathology Washington, D. C. USA
AIAC	Australian Institute of Anatomy Canberra, Australia
AIUZ	Anthropological Institute University of Zurich Zurich, Switzerland
ANM	National Museum of Anthropology Prague, Czechoslovakia
BMNH	British Museum (Natural History) London, England
CGH	Department of Pathology Charleston General Hospital Charleston, West Virginia
DPUS	Department of Pathology University of Strasbourg Strasbourg, France
FM	Field Museum of Natural History Chicago, Illinois, USA
FPAM	Federal Pathologic-Anatomy Museum Vienna, Austria
GHPM	Gordon Pathology Museum Guy's Hospital Medical School London, England
HM	Hunterian Museum The Royal College of Surgeons of England London, England
HMCS	Historical Museum Chur, Switzerland
IPAZ	Institute of Pathological Anatomy University of Zurich Zurich, Switzerland
IPMI	Pathology Museum Institute of Pathology University of Innsbruck Innsbruck, Austria

MGH	Department of Pathology Massachusetts General Hospital Boston, Massachusetts, USA
NHMB	Natural History Museum Bern, Switzerland
NMNH	National Museum of Natural History Smithsonian Institution Washington, D. C., USA
OM	Odontological Museum The Royal College of Surgeons of England London, England
PMES	Pathology Museum The Royal College of Surgeons of Edinburgh Edinburgh, Scotland
PMSG	Pathology Museum St. George's Hospital Medical School London, England
PMUG	Pathology Museum University of Graz Graz, Austria
PMWH	Pathology Museum Westminster Hospital School of Medicine London, England
UGPM	Pathology Museum University of Göttingen Göttingen, Federal Republic of Germany
VM	Virchow Museum [destroyed WWII] Berlin, Germany
WM	Wellcome Museum The Royal College of Surgeons of England London, England

A Brief History of Paleopathology

Descriptions of dry bone lesions in anthropological or paleontological specimens go back at least two hundred years. The earliest work on paleopathology focused on nonhuman paleontological specimens (e.g., Esper, 1774; Cuvier, 1820). Warren (1822) included a discussion of artificial cranial deformation in human skulls of indigenous North Americans in his book titled *A Comparative View of the Sensorial and Nervous Systems in Man and Animals.* In 1855, Gosse published, in Paris, another study of artificial cranial deformation. Toward the end of the nineteenth century the question of the origin of syphilis began to be debated with an intensity that continues today (e.g., Jones, 1876; Virchow, 1898).

With the turn of the century there is a marked expansion of published reports on ancient disease. Particularly notable is the work of Sir Marc Armand Ruffer (1910) on Egyptian mummies, and the studies on Nubian skeletal material by Wood-Jones (1908a, 1910b) and Elliot-Smith and Wood-Jones (1910). In the United States, Hrdlička (1914) published some observations on the pathology of ancient Peruvian skulls. In 1923, R. L. Moodie's introduction to the study of ancient disease, which emphasized nonhuman paleontological specimens, appeared. A brief, general review of human paleopathology was published by H. U. Williams in 1929. This review included observations on bones and teeth, as well as on mummy tissue and ancient art. Pales (1930) followed with his book on paleopathology and comparative pathology. Most of his cases and discussions concerned European human specimens. In the same year, Hooton (1930) published his classic study of the North American Indian skeletal material from Pecos, in which he included an extensive description of pathological specimens. Hooton's study is notable in its descriptive detail, in the statistical treatment of different types of disease in the skeletal population, and in his efforts to show trends in disease frequency through the time period of human occupation at the site.

Møller-Christensen's report (1953) on the skeletal material from a medieval leper cemetery in Denmark is perhaps unique in the history of paleopathology. The historical knowledge of the existence of a leper cemetery and its location led to excavation of the skeletons. The descriptive analysis of this material has provided important information about leprosy in ancient times. It has also expanded our knowledge of the effects of the disease on skeletal tissue.

More recent general works on paleopathology include Calvin Wells' review of evidence of human paleopathology from skeletal material, mummies and art (1964a). Brothwell and Sandison (1967) brought together in a single volume many of the classic papers on paleopathology

with a broad range of new papers on numerous subjects related to the study of ancient disease. Morse's report (1969) on skeletal pathology in human archeological material from the midwestern United States provides excellent descriptive details on many types of skeletal diseases.

Throughout the development of human skeletal paleopathology as a scholarly discipline, there have been recurring problems in both theory and methodology. In the early stages of this development most of the research was conducted by physicians who had little knowledge of archeology. In more recent times most of the studies of pathological skeletal specimens have been conducted by anthropologists whose background in gross bone pathology may have been inadequate. This has led to problems in which lesions of bone were incorrectly attributed to the pre-Columbian time period by those unfamiliar with the complexities of archeological dating and in which bone lesions were incorrectly diagnosed through ignorance of anatomy and the total range of diseases that affect bone (see Stewart's comments on this problem in Jarcho, 1966:43).

Much of the emphasis in paleopathology until recently has been on descriptions of pathological specimens, and there has been little effort to relate the evidence of disease to the broader problems of human adaptation. Hints of such an emphasis exist in Hooton's Pecos Pueblo monograph (1930) and more recently in the consideration of epidemiological factors in evaluating the data on pre-Columbian tuberculosis in the New World (Morse, 1969) and in discussions on the origin of treponemal diseases (Hackett, 1963; Hudson, 1965). The trend toward population studies of ancient disease undoubtedly will become increasingly important as descriptive data accumulate and the diagnostic criteria for skeletal disease are clarified.

Much of the descriptive literature in skeletal paleopathology depends on the scholar's knowledge of gross bone pathology. Unfortunately, where this knowledge is inadequate there have been few reference sources that could be of assistance. Jarcho (1966) organized a symposium on human paleopathology, which addressed this problem among others. The participants called for the establishment of a paleopathology registry and improved diagnostic methodology to partially correct these problems. Steinbock's recent textbook (1976) on diagnosis of ancient bone disease represents the first integrated attempt to establish diagnostic criteria for the paleopathologist. As such it represents an important step in improving the knowledge regarding the types of diseases that affect bone and the morphological features associated with the disease.

The Paleopathology Program at the Smithsonian Institution

Ortner (1976) has reported the development of a program at the Smithsonian Institution, which addresses the need for a registry and includes additional reference materials. This Paleopathology Program has as its major objective the identification and classification of all pathological specimens in the vast human skeletal collections of the National Museum of Natural History. Significant progress has been made toward this objective. Another objective is to build a reference collection of clinically documented cases of skeletal disease including gross specimens, photographs, and roentgen films.

In the United States, Drs. George Huntington, College of Physicians and Surgeons in New York City, T. Wingate Todd, Western Reserve University in Cleveland, Ohio, and Robert Terry in collaboration with Mildred Trotter, Washington University in St. Louis, Missouri, developed major anatomical skeletal collections from anatomical, dissecting-room cadavers. Each of these collections contained about 2000 skeletons, among which were pathological specimens. Aleš Hrdlička, the founder of physical anthropology at the Smithsonian Institution and one of the pioneers of American paleopathology, obtained the Huntington skeletal collection for the National Museum of Natural History. This collection was accessioned in 1921, partially in the hope that it would become the nucleus of a reference

collection on pathology of the skeleton (Hrdlička letter to J. H. Kellogg, 31 January 1921). More recently the Terry collection has also been transferred to the Smithsonian. The Todd collection is currently housed at the Cleveland Museum of Natural History, Cleveland, Ohio, USA.

It is clear that collections of documented bone pathology will need to be added to these fine skeletal collections to supply adequate comparative material for the needs of the paleopathologist. In the United States, there are a few skeletal collections specifically devoted to pathological conditions including those of the Armed Forces Institute of Pathology in Washington, D. C., The Warren Anatomical Museum at Harvard University and the Mütter Museum of the College of Physicians of Philadelphia. However, direct comparison of material in these collections with archeological skeletons is awkward at best. Careful photographs and roentgen films of such materials are the most realistic vehicle for comparative studies.

The final objective in the Smithsonian Paleopathology Program is to gather together bibliographic source materials from the literature on bone and joint pathology and on paleopathology. Much of this material is in older, rather inaccessible sources, and considerable effort has been expended to obtain originals or copies of much of the relevant literature. This bibliographic material, as well as the photographs and detailed descriptions of the cases studied in our joint European museum survey, are available for study at the Department of Anthropology at the National Museum of Natural History of the Smithsonian Institution in Washington, D. C., USA.

The Biology of Skeletal Tissues

The Development of Bone

Early in the development of the fertilized human egg (zygote) the dividing cells form three distinct regions, which ultimately differentiate into the tissues and organs of the adult. These three layers are ectoderm, mesoderm, and entoderm. Of the three, ectoderm and mesoderm are relevant to our understanding of skeletal tissues. Ectoderm gives rise to structures that produce the enamel of teeth. Mesoderm differentiates into connective tissue including bone, cartilage, dentin, and cementum.

In the embryo the tissue precursor of bone results from a condensation of mesodermal cells called mesenchymal cells. At this stage, these cells are undifferentiated and can become any one of several different types of specialized connective tissue cells. The factors that induce a mesenchymal cell to become or produce one of the specialized skeletal cells remain one of the intensely interesting problems in connective tissue biology. One of the important factors appears to be the immediate vascular and cellular environment resulting from the proliferation of blood vessels and adjacent cells in the embryo. This proliferation affects the accessibility of the cell to nutrients and oxygen. Changes in these factors are known to modulate the differentiation of cells.

The specialized cells that are part of the biology of skeletal tissue induce cartilage cells and bone cells. There are two types of cartilage cells: (1) chondroblasts, which secrete cartilage matrix, and (2) chondrocytes, which maintain cartilage matrix and create conditions that lead to the mineralization of some cartilage tissue. There are three types of bone cells: (1) osteoblasts, which form bone matrix, (2) osteocytes, which maintain bone tissue, and (3) osteoclasts, which destroy bone tissue and calcified cartilage.

Classically, two types of bone formation occur during the development of skeletal tissue. In one of these (endochondral ossification), cartilage cells are an important factor. In the second type of bone formation (intramembranous ossification) only bone cells are involved. These two types of bone formation produce tissue in different ways. During cartilage matrix formation, the new matrix is secreted within existing tissue matrix (interstitial development), whereas during bone matrix formation (except for limited interstitial development during embryonic life) the new matrix is laid down on an existing tissue surface (appositional development).

Endochondral ossification is associated with the growth phase but may occur in the adult in abnormal situations, such as fracture healing. In the embryo the earliest centers of endochondral ossification are those areas that will become the major, long, tubular bones (the femur, tibia, fibula, humerus, radius, and ulna). All endochondrally derived bones begin as a condensation of mesenchymal cells which, in contrast to intramembranous ossification, develop into a cartilage model (anlage) of the future bone rather than directly into bone itself.

The cartilage model consists of the cartilage cells (chondrocytes) and a protein matrix, the latter forming the bulk of the model. However, the entire model is surrounded by a connective tissue layer called the perichondrium. As the cartilage model grows, through the proliferation of cells (chondroblasts) in the germinative layer of the perichondrium, the oldest cells near the center of the model begin to swell (hypertrophy). As this occurs the perichondrium overlying this area takes on osteogenic properties and forms a sleeve of bone around the central region of the model (Figure 1). In contrast with the cartilage model, the sleeve bone is highly vascularized and

FIGURE 1.—Histological features in endochondral ossification. Cartilage (C) at the top of the field, the zone of hypertrophic and calcifying cartilage (CC) is near the middle level of the field. Bone formation (B) on calcified cartilage cores is taking place in the lower portion of the field. Sleeve bone (SB) marking the lateral margin of the growth plate is in the left portion of the field. The section is from the femur of a 5-month human fetus. (Approximately × 50.)

with increasing hypertrophy of the cartilage cells, blood vessels invade the model. Vascularization of part of the model creates the conditions necessary for bone formation, but the initial process does not involve the synthesis of additional organic matrix as is the case in intramembranous osteogenesis.

Cartilage has many of the same protein constituents as bone and apparently the cellular hypertrophy of the cartilage is associated with a process that makes it possible to mineralize the matrix that remains between the cartilage cells. During the early stages of long bone development, the constituents for mineralizing cartilage probably arrive at the site of mineralization by diffusion through the organic matrix (Ham, 1974: 375). Vascularization makes the necessary constituents for mineralization much more accessible.

As osteoblasts are adding bone to part of the mineralized cartilage trabeculae, bone destroying cells (osteoclasts) are frequently seen in histological preparations to be removing the mineralized cartilage (Figure 2). In a longitudinal section through a developing long bone of a 20-week

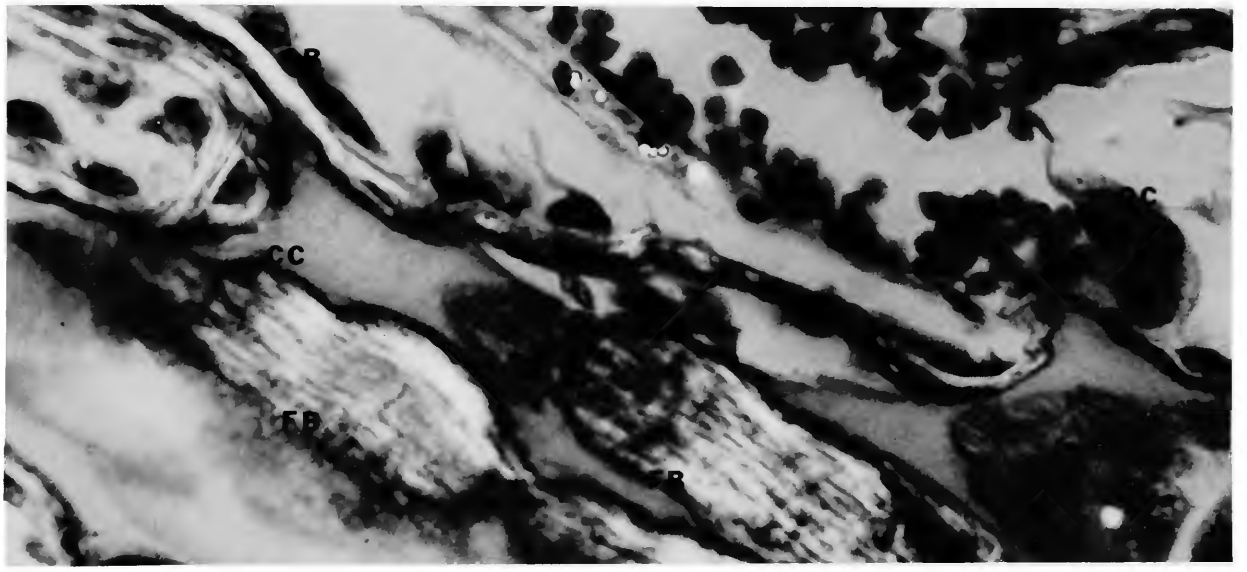


FIGURE 2.—Fiber bone formation (FB) on calcified cartilage cores (CC). Osteoblasts (OB) are active in the left of the field. An osteoclast (OC) is destroying calcified cartilage near the right edge of the field. The section is from the femur of a 5-month human fetus. (Approximately × 1000.)

fetus, it is apparent that mineralized cartilage is a very temporary tissue. Within 10 to 15 millimeters distance of the zone where the cartilage is mineralizing, most of such tissue has been replaced by the combined activity of osteoclasts and osteoblasts.

The initial site of penetration by the blood vessels into the cartilage model of a long bone becomes the nutrient foramen of the mature bone. Bone growth takes place centrifugally from this site, although not necessarily in equal amounts in all directions. As the zone of mineralizing cartilage moves away from the initial center of ossification, the sleeve bone is also growing in both directions and forms the initial compact tissue or shaft of the developing bone. Because the shaft flares near the ends, the process of growth also involves a process of remodeling in which the flared end is reduced to the appropriate dimensions for the shaft.

Toward the end of fetal life some of the long bones such as the femur establish secondary centers of ossification in the epiphyses. In these centers the process of cartilage mineralization, followed by vascular invasion and the addition of bone, in turn followed by the removal of trabeculae containing mineralized cartilage, is essentially the same as the growth of the diaphysis.

In intramembranous ossification, mesenchymal cells differentiate (probably through one or more intermediate steps) into bone forming cells (osteoblasts). This process occurs within the mesenchymal cell condensation. As cell differentiation takes place, the cells become organized and form smooth, membranous sheets. Continuing development involves the synthesis of organic bone matrix by osteoblasts. This organic matrix is in close proximity to the cell and consists primarily of collagen but contains noncollagenous, organic materials as well. In its unmineralized state the organic matrix of bone is called osteoid. Normally the mineralization of osteoid proceeds rapidly (in a matter of hours) although full mineralization of bone matrix, particularly in the adult, may take months or years.

As the long bones lengthen they also increase in diameter. Growth in diameter occurs by apposition as the result of intramembranous ossification. However, early in embryonic life most of the centers of ossification in the skull are of the intramembranous type. The development of the bones of the skull is complex and the reader should consult a standard reference on human embryology (e.g., Hamilton and Mossman, 1972) for a detailed discussion. For the purposes of this book, it is sufficient to note that the centers of endochondral ossification in the skull are primarily associated with the skull base including the sphenoid and part of the occipital (Figure 3).

It might be well at this point to emphasize that the distinction between endochondral cartilage and membrane-formed bone is more than an academic exercise. As will be emphasized subsequently, some diseases focus on skeletal tissue formed in a cartilage model (e.g., achondroplasia). Other diseases also affect the process of membrane bone formation (e.g., osteogenesis imperfecta).

The embryological development of vertebrae is also complex. There are, however, congenital de-

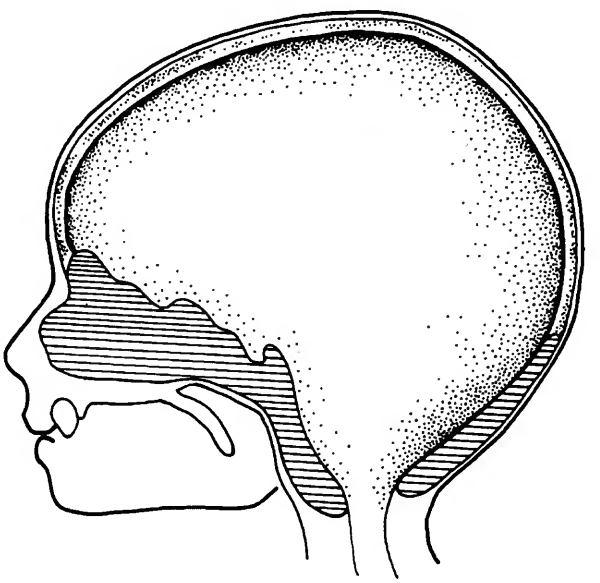


FIGURE 3.—Midsagittal drawing of the skull of a 4-month human fetus. Stippled areas indicate approximate region of intramembranous ossification, horizontal lines indicate approximate regions of endochondral ossification.

fects of the vertebral column, which can best be understood in terms of defects in embryological development. Early in the development of the embryo, cell division results in the production of separate but similar segments in a process called segmentation. One product of segmentation is the somite, which is a paired condensation of mesodermal cells on each side of the embryonic precursor of the vertebrae (notochord) and the embryonic precursor of the spinal cord (neural tube). Each of the somites will give rise to a skin segment (dermatome), a striated muscle segment (myotome), and a sclerotome that forms a portion of a vertebra and, in the case of thoracic vertebrae, a rib.

Cells in the sclerotomic condensations proliferate and completely surround the notochord, displacing the neural tube dorsally. In the dorsoventral axis the cells are distributed evenly. However, in a cranial-caudal axis this is not the case. The cranial mass of cells (the anterior sclerotome) is less condensed, while the caudal portion (the posterior sclerotome) is more condensed. The cells of a posterior sclerotome, at the boundary with the less dense cell condensation of the same sclerotomic condensation, differentiate into the intervertebral disc. The remaining cells of posterior sclerotome fuse with the anterior sclerotome of the adjacent sclerotome to form a vertebral body (Figure 4).

Within the sclerotomic cell condensations, chondrification centers develop and form the cartilaginous vertebral body. Other cartilaginous centers on either side of the neural tube begin to proliferate and form the cartilaginous precursor of the vertebral arch and spinous processes. These centers fuse with the vertebral body. While the neural tube ultimately forms the spinal cord (and in the cranium, the brain), the notochord forms the nucleus pulposus of the intervertebral disc, but it degenerates in the region of the vertebral body.

During the early growth phase, beginning with the fetus and continuing through early childhood, the growth rate is very high. For example, from the fifth fetal month to birth the length of the

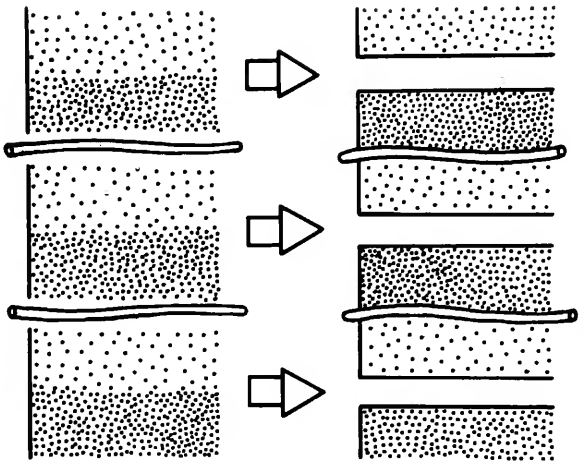


FIGURE 4.—Schematic diagram of the embryonic development of vertebrae. The early stage of development is represented by the three sclerotomes on the left. The anterior sclerotome is represented by the lightly stippled areas. The more heavily stippled areas represent the posterior sclerotome. There is a blood vessel between each pair of sclerotomes. During embryonic development the posterior and anterior portions of adjacent sclerotomes fuse to form an embryonic vertebral body as seen on the right side of the diagram.

femur doubles. However, from birth it takes approximately four years for the femur to double its length again and eight years to double a third time (Stewart, 1968:132–133).

The bone tissue associated with very rapid growth is called fiber or woven bone and is not as dense or as well organized histologically as bone associated with slower growth. In a microscopic preparation, the bundles of collagen microfibrils are irregular in thickness and do not have the highly regular organization seen in later stages of the individual's life (Figure 5). However, fiber bone may be found in later life in abnormal conditions, such as fracture healing and neoplasms.

The fiber bone of the early developmental stages is replaced by more organized tissue called lamellar bone. The histological characteristics of this type of bone are the layers (lamellae) of collagen of relatively uniform thickness and the alternating orientation of the fibers. These fea-

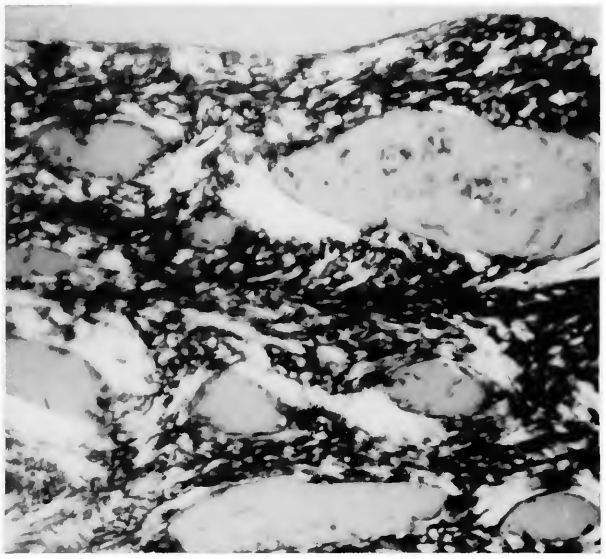


FIGURE 5.—Fiber bone in a 5-month human fetus. Note the irregular orientation of most of the fiber bundles. (Approximately × 100.)

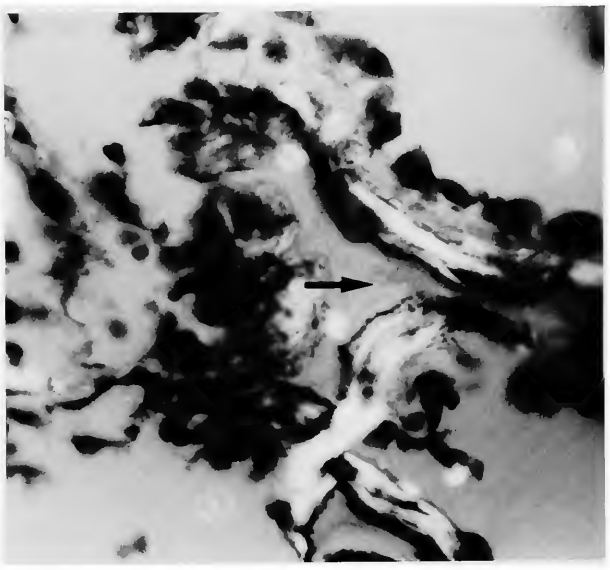


FIGURE 6.—Calcified cartilage (arrow) surrounded by developing bone in a 5-month human fetus. The light is partially polarized. Note that the calcified cartilage has optical properties similar to the background in contrast with bone, which is generally much brighter. This brightness is due to the anisotropic properties of bone. (Approximately × 1000.)

tures are easily seen in histological preparations using a polarizing microscope. The collagen of the protein matrix of bone has crystalline properties because of the periodic arrangement of the molecules of tropocollagen, which make up the major component of the matrix. This arrangement results in an optical property called anisotropy (birefringence), in which the axis of light passing through the fibers is rotated.

If two polarizing filters are adjusted so that the filter axes are perpendicular to each other, light cannot pass through the filters. If a histological specimen is placed between such filters, birefringent areas in the tissue will rotate the light permitting some of the rays to pass through the filter farthest away from the light source and be seen as a light area of tissue. Portions of the tissue that are not birefringent will not rotate light and this will appear dark in polarized light. In this context it may be well to emphasize that both fiber bone and lamellar bone have anisotropic properties in contrast with calcified cartilage, which does not rotate light and thus appears dark in a polarizing microscope (Figure 6).

Cartilage

With this background on the early development of skeletal tissue let me review in greater detail the role of cartilage and bone in the function of skeletal tissue. Three types of cartilage tissue occur in human organisms: (1) hyaline, (2) fibrous, and (3) elastic. Of these three, hyaline cartilage is germane to our discussion of skeletal tissue because of its importance in bone growth. I have already discussed the role of cartilage in endochondral bone formation in which hyaline cartilage provides a temporary mineralized scaffold for bone formation. These endochondral ossification centers or zones persist throughout the growth period and are obliterated at varying ages in different bones; the last fusion occurs by about the age of 25 years with the fusion of the two growth centers of the iliac crest.

In addition to its involvement in growth, hyaline cartilage provides the gliding surface of all true joints such as those between the long tubular

bones. Hyaline cartilage, like bone, has two basic components, cells and intercellular matrix. However, there are many features of hyaline cartilage that are in distinct contrast to bone. Although chondrocytes and chondroblasts are derived from mesenchymal cells, as are bone cells, the cartilage matrix secreted by chondroblasts contains less collagen (about 10 percent) but cartilage contains much more water and a higher concentration of chondroitin sulfate, a type of noncollagenous protein, than is found in bone. During the growth phase, chondroblasts are in direct contact with the enlarging cartilage and are contributing to the growth process by secreting protein matrix, which occasionally surrounds some chondroblasts. The chondroblasts that become surrounded by matrix differentiate into chondrocytes but retain many of the chondroblastic properties (Ham, 1974:372), such as the ability to divide, to produce new cells, and the ability to secrete matrix. Bone cells (osteocytes) do not divide and have only very limited ability to produce matrix.

Cartilage tissue is similar to bone tissue in that it has the capacity to grow by adding cells and matrix to existing surfaces (appositional growth). However, unlike bone, cartilage can expand (interstitial growth) by multiplication of cells and interstitial secretion of matrix by its chondrocytes (Ham, 1974:372–374). Cartilage cells, both chondroblasts and chondrocytes, retain optimal ability to divide and secrete matrix only during the growth phase of the individual (Ham, 1974:374).

Another contrast between hyaline cartilage and bone is that cartilage is poorly vascularized. The nutrients that are needed for chondrocyte function and the metabolites that result from cell activity must arrive and be removed while relatively isolated from vascular supply by the gel-like matrix that surrounds the cell. The minimal vascularization and the reduced ability of cartilage cells to multiply after the end of longitudinal growth appear to be related factors in greatly limiting the capacity of cartilage to repair itself if damaged by injury or disease after maturity. This fact becomes important in interpreting a number of abnormal conditions seen in the joint surfaces of bone, particularly in the arthritides.

The Cells of Bone

Two cells of bone (osteoblasts and osteoclasts) are directly responsible for its gross appearance. The third bone cell, the osteocyte, is very much part of the histological appearance of bone and plays an important role in mineralization, bone tissue maintenance, and mineral physiology. The activity of all three bone cells may be divided into five functions: (1) protein matrix synthesis, (2) mineralization of protein matrix, (3) bone tissue maintenance, (4) resorption of mineralized tissue, and (5) involvement in general mineral physiology.

The osteoblast itself contains the normal basic ingredients of a cell, a nucleus and cytoplasm containing organelles (mitochondria, endoplasmic reticulum, ribosomes, and Golgi bodies) involved particularly in the synthesis of protein matrix. The fluid phase of cytoplasm includes water, glycogen, and fat vacuoles. The cytoplasm is surrounded by a thin membrane, which has long, fine extensions filled with cytoplasmic fluid. These cytoplasmic extensions form an interconnecting network, with adjacent cells forming the main component of the germinative layer of the bone forming tissue (Figure 7).

As the osteoblast sheets, applied to an existing surface of bone or calcified cartilage, begin to secrete osteoid, a matrix is formed, which becomes interposed between them and the bone surface (Figure 8). However, some of the osteoblasts become surrounded by the matrix in spaces called lacunae (Figure 9). The cytoplasmic extensions are maintained with other nearby osteoblasts, which are also being surrounded by osteoid, and with the osteoblasts, which remain at the protein secreting front. Surrounded osteoblasts are now osteocytes. The interconnections between osteocytes surrounded by matrix are maintained after mineralization and form the little canals (canaliculi), which are a prominent part of bone histology. The interconnections between the osteoid

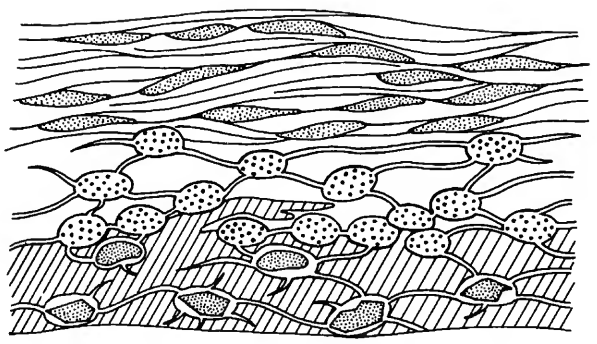


FIGURE 7.—Schematic representation of the different types of cells related to bone formation and bone maintenance. The fibroblastic layer consists of the very elongated, finely stippled cells at the top of the illustration. Adjacent to bone, the fibroblasts differentiate into osteoblasts seen as the coarsely stippled, interconnected cells on the bone surface. Osteoblasts secrete bone protein matrix and mediate the initial stage of mineralization. Some of the osteoblasts and their interconnecting processes are surrounded by bone matrix and differentiate into osteocytes represented by the fine-stippled cells. Osteocytes are involved in the physiological activities of bone including mineral exchange and final mineralization of the bone matrix.

secreting osteoblasts and osteocytes are also maintained (Ham, 1974:419) as long as the osteoblasts are active.

The protein matrix is fibrillar and in lamellar bone the collagen fibers have a highly specific orientation, with each subsequent layer having its fibers oriented at a varying angle to the preceding layer. There appears to be a fairly discrete number of osteoid layers secreted before mineralization takes place (Johnson, 1964:624). This may be related to the life span of the active osteoblast, which hypertrophies before the onset of mineralization. As with calcified cartilage, the initiation of bone mineralization is accompanied by increased alkaline phosphatase activity. With initial mineralization the death of some of the matrix forming cells occurs. With the completion of initial mineralization the organic bone matrix has achieved approximately 75 percent of maximum mineralization (Amprino and Engström, 1952: 11).

Former osteoblasts that were surrounded by protein matrix begin a process of differentiation by which they become osteocytes. While the osteocytes maintain the interconnecting network of cytoplasmic extensions (canaliculi) with other osteocytes, there is a marked reduction in the amount of cytoplasm. The space made available by this reduction appears to be filled with a special mineralized matrix, which is important in mineral physiology. The osteocytes may continue to function for years.

During the initial stage of its life cycle, the osteocyte appears to be involved in the process of final or secondary mineralization of adjacent matrix. In part, because there are many fewer cells per cubic millimeter of tissue than was the case during osteoid formation, the process of secondary mineralization is much slower than mineralization mediated by osteoblasts. Another factor may be that mineral ions, cell nutrients, and cell metabolites must pass through the canalicular system. The process of secondary mineralization not only takes longer (many months versus hours to days for primary mineralization) but becomes even slower with increasing age.

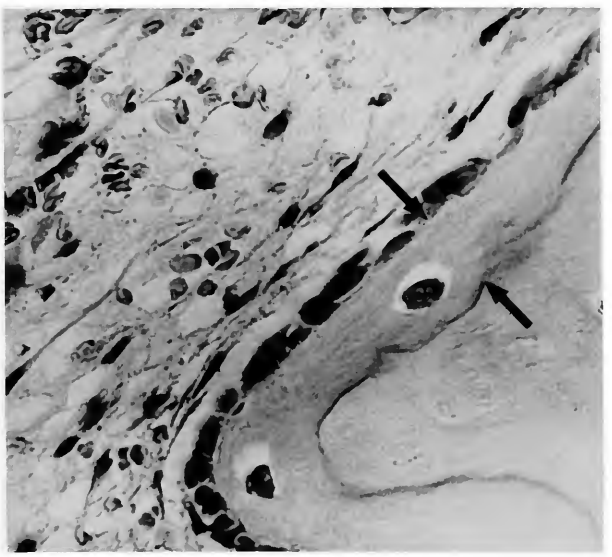


FIGURE 8.—Photomicrograph of bone cells during active matrix synthesis. The newly formed osteoid layer is indicated by the arrows. Two osteocytes are seen in the osteoid layer and osteoblasts are lining the surface of the osteoid. Fibroblasts are seen as dark elongated cells adjacent to the osteoblasts. (Approximately × 600.)

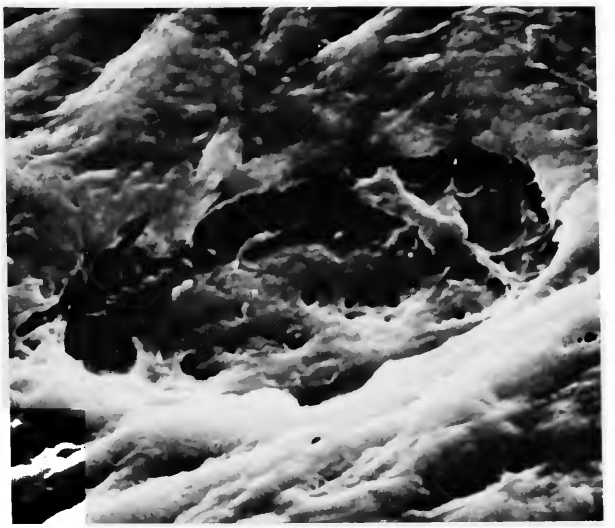


FIGURE 9.—Scanning electron micrograph of an osteocyte lacuna. Small holes at the bottom of the lacuna are opening into the canaliculi. The structure in the right portion of the lacuna may be the dried remnant of the osteocyte. (Approximately × 4000.)

The second stage of the osteocyte life cycle marks a shift away from activity involving secondary mineralization to activity in mineral physiology. Thus far I have treated bone as a tissue with primarily mechanical functions. Here I need to introduce the reader to the concept that bone is an organ involved in the physiology of the human organism. There are several ionic concentrations in blood serum, which need to be maintained at virtually constant levels. One of these is calcium which, in a healthy individual, is maintained at a concentration of about 10 milligram percent. Physiologically calcium is, among other things, a cofactor in blood clotting and involved in the contractile mechanism of muscle (McLean and Urist, 1968:259).

Serum calcium concentrations are maintained by a complex interrelationship among (1) dietary (intestinal) absorption of calcium, (2) renal (kidney) retention of calcium, (3) secretions by the parathyroid (parathormone) and thyroid (calcitonin) glands, (4) exchange with labile mineral in the skeleton (osteocyte activity), (5) bone resorption (osteoclast activity), and (6) direct exchange between bone crystal surfaces and serum. While the major factors in calcium physiology are dietary and renal, the skeleton through direct exchange and osteocyte activity provides a rapidly available (minutes to hours) finely controlled source of calcium (if there is some problem with the major mechanisms) to maintain serum calcium levels.

The final stage of the osteocyte life cycle involves senescence and death. Precisely how this affects the metabolism of the organism and the mechanical and physiological functions of skeletal tissue is unclear.

Throughout the life of the individual, there is a continuing need to remove and replace existing skeletal tissue. Earlier I have noted the need to change the contours of growing bone. The complexity of this process will become more apparent in the discussion of internal or osteon remodeling (p.19). For the moment it is adequate to note that osteoclasts are the cells that are associated with the resorptive phase of skeletal remodeling and with bone loss in morbid conditions.

Osteoclasts differ morphologically from osteoblasts and osteocytes in several aspects (Figure 10). Two characteristics that are particularly noticeable are that osteoclasts are much larger than other bone cells and that they generally have more than one nucleus. There are no cytoplasmic extensions from the cell. Structures within the cytoplasm of the osteoclast also differ from the osteoblast. There are many more lysosomes in the osteoclast reflecting its destructive potential, while mitochondria and the endoplasmic reticulum are minimal reflecting a cell function in which protein synthesis is minimal. In a single day, a single osteoclast can destroy bone that took 100 osteoblasts to form in the same time period. Grossly, all of the destructive lesions seen in skeletal disease are the direct result of osteoclastic action. Disease conditions resulting in bone destruction only stimulate osteoclastic responses and do not directly destroy bone.

Histologically the osteoclast is always associated with bone resorption at an existing bone surface. One of the distinctive features frequently

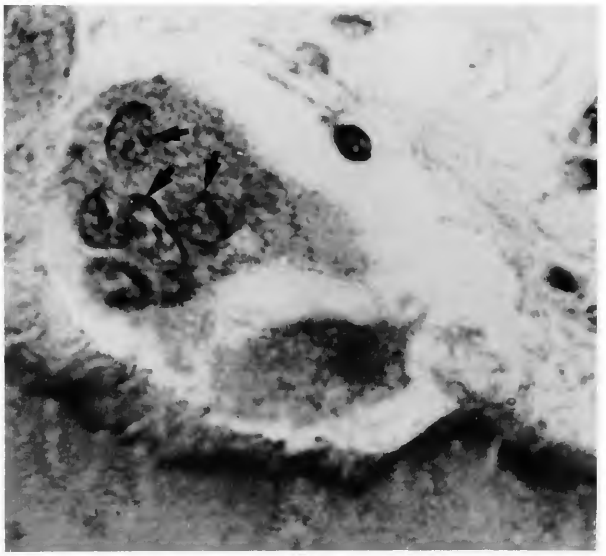


FIGURE 10.—An osteoclast in a Howship's lacuna. Arrows indicate three of the nuclei. Note the irregular edge of the resorbing bone in the lacuna. (Approximately × 1300.)

seen in bone resorption is a deep concave depression in the surface known as Howship's lacuna. Osteoclasts may be seen occupying these depressions. The side of the cell in contact with the bone surface has a striated or "brush" border, which has been shown to be the folded cell membrane that is involved in the solubilization and removal of adjacent bone tissue including both organic matrix and mineral (Hancox, 1972:123, 127).

Osteoclasts are very active cells, which are motile and thus do not necessarily resorb bone only in one place (Hancox, 1972:128). Furthermore, osteoclasts frequently form clusters of cells during resorption, such that the depressions or lacunae formed by one cell may be removed by the activity of another osteoclast leaving a relatively smooth surface. The point to be emphasized is that Howship's lacunae are always indicative of active resorption, but the absence of lacunae does not preclude the existence of resorptive activity. Since in archeological bone specimens the cells are gone, one microscopic bit of evidence of active bone resorption will be the presence of Howship's lacunae.

Bone Growth

There are two basic mechanical functions performed by bone. These are to provide a fulcrum for muscle activity and a counterforce to gravity. One should perhaps add a third function in a somewhat different context: providing space for blood-forming marrow. To accomplish these objectives it is necessary to (1) provide the surface area for joints, (2) provide attachment for muscles, (3) maximize compression, shear, and bending strength, and (4) minimize weight. All of these factors result in a general model of a long bone which is (1) hollow to minimize weight, maximize strength, and provide for space for marrow (both blood forming and fat storage), (2) flared at the ends to provide adequate surface area for joint movement, and (3) often provided with specialized areas for muscle attachment. While these criteria apply primarily to long tubular bones, to varying degrees they are also factors in the shape of other bones as well. I shall use the long bone as a model for the remodeling process.

As I have implied earlier the typical growing long bone consists of a growing diaphysis, the ends of which are flared and are called the metaphyses, a cartilage growth plate, and finally an epiphysis at each end (Figure 11). There may be other secondary growth centers (apophyses), such as the greater and lesser trochanters of the femur. To demonstrate the principles of remodeling, I will limit the discussion to the diaphysis and metaphysis of a growing long bone.

Bone growth occurs in both length and width with length growth predominating. Growth in width involves the addition of circumferential layers of bone to the external surface of the bone shaft. At the same time removal of bone is taking place on the internal or endosteal surfaces of bone. The latter is done to minimize the weight of the bone and provide space for hemopoietic (blood forming) and fatty marrow.

Growth in length is more complicated. The flared ends of the bone are growing not only in length but also in width. As the flared ends grow,

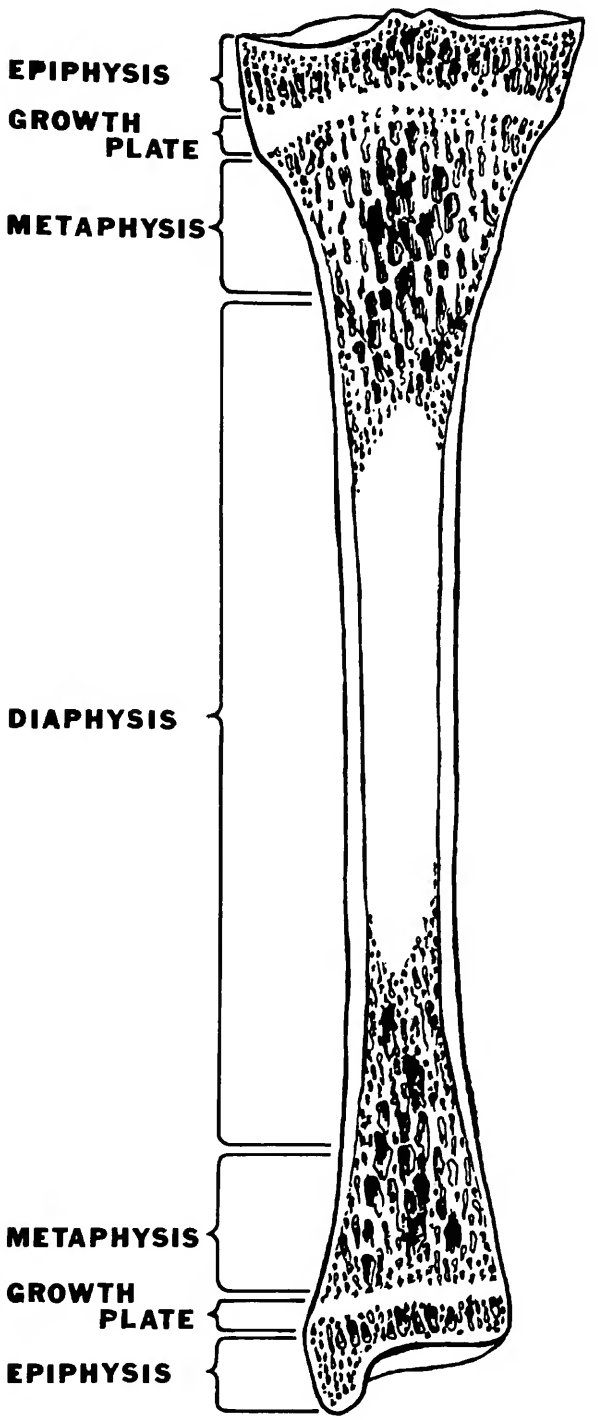


FIGURE 11.—Diagrammatic representation of the major components of a growing long bone.

what was formerly part of the metaphysis must be converted to the narrower diaphysis, so that growth in length is accompanied by the removal of bone on the external surface of the metaphysis in the region not involved in active growth (the cutback zone). This must be accompanied by the formation of bone on the endosteal surface in order to maintain the mechanical strength of the bone. Thus, while growth in the diaphysis involves addition to the external surface and removal of the inner surface, growth in the flared ends involves the reverse with the exception of growth at the very end of the flare. Here growth is occurring in length and width simultaneously (Figure 12).

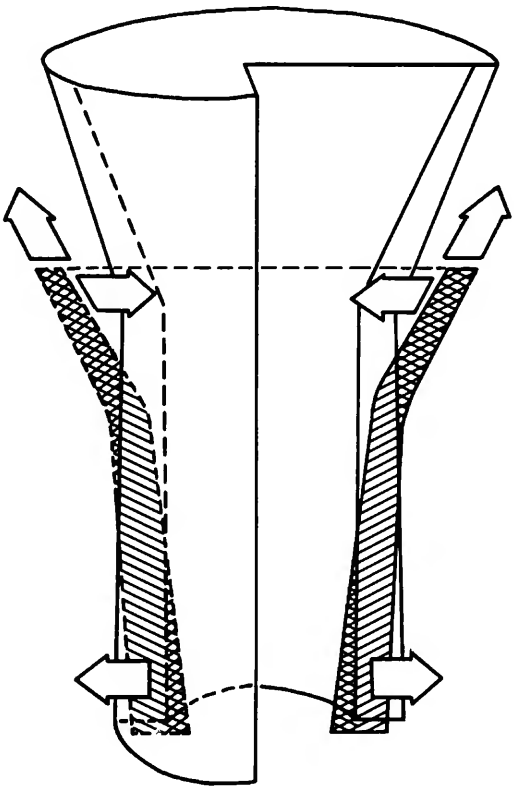


FIGURE 12.—Remodeling dynamics at the end of a growing bone. The conical end represents the metaphysis and growth plate. Hatching indicates the earlier stage in growth. The cross hatching indicates bone that must be removed as the bone grows in length. Arrows indicate the direction of bone addition during the growth process.

An understanding of the growth dynamics is very important to the paleopathologist. There are many disease conditions that produce inadequate or abnormal remodeling. Most of these are discussed in the chapter on congenital disorders (p.329). Furthermore, the very porous external appearance of the cutback zone in the growing bone can easily be misinterpreted as pathological by one who is unfamiliar with this process.

The process of growth-related remodeling stops with the fusion of the epiphysis and the metaphysis. However, appositional growth (periosteal) and endosteal resorption continue throughout life although at a greatly reduced rate. This continuous growth of bone is more than offset by the degenerative changes that are associated with aging. Removal of compact and cancellous bone in the marrow cavity (endosteal) also continues at a diminished rate. In order to maintain the mechanical integrity of the bone, the processes of periosteal deposition and endosteal removal must be in balance with respect to the particular mechanical loading of the bone. In the case of compact bone, this does not mean that the thickness of the cortex must be maintained at the level found in the young adult, since a larger external diameter greatly increases the bending strength of a tube. This means that with an increase in the external diameter there can be a considerable decrease in cortical bone thickness while still maintaining about the same bone mass and adequate mechanical strength.

However, with increasing age there is a tendancy for endosteal removal to exceed periosteal deposition. This condition is associated with a reduction in cancellous bone trabeculae with remaining trabeculae sometimes becoming thickened. The combination of abnormal thinning of compact bone and the loss of cancellous bone is called osteoporosis and is discussed in greater detail in the chapter on metabolic disorders (p.270).

Garn (1970:29) has reported the importance of endocrine secretions in controlling cortical changes. Male sex hormone (testosterone) affects periosteal deposition whereas female sex hormone (estrogen) affects endosteal deposition. Clearly other factors are involved in cortical dynamics as well (e.g., men at the end of their growth period have thicker cortical bone than women at the same stage), but the marked reduction in secretion of sex hormones occurs 10 to 15 years earlier in females than in males and appears to be a major factor in the more severe bone loss seen in the aging female skeleton.

It is important to emphasize that the process of cortical remodeling, which occurs during and after the cessation of growth, is not uniform. Enlow (1963:51–59) has documented the existence of a trend he calls drift, in which appositional growth is associated with only part of the external and internal surface. Figure 13 illustrates the process diagrammatically for the tibia. In the tibia, bone tends to be added to the anteromedial periosteal surface and the posterolateral endosteal surface. Bone is removed from the anteromedial

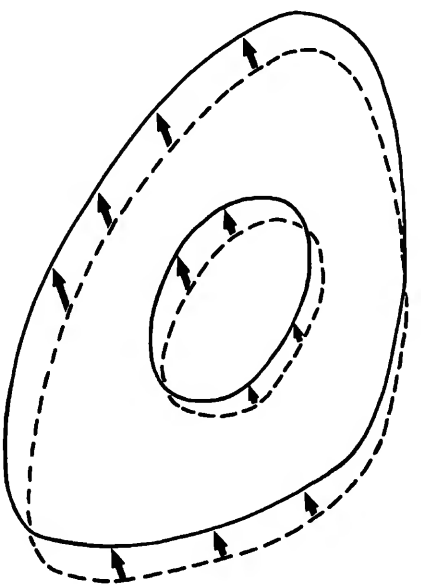


FIGURE 13.—Diagrammatic representation of the process of bone drift. During growth, the load axis of long bones may change. To compensate for the change in the mechanical loading, as in the tibia, bone is added to the anteromedial periosteal and posterolateral endosteal surface while bone is removed from the anteromedial endosteal surface and the posterolateral periosteal surface.

endosteal surface and posterolateral periosteal surface. Histologically what is apparent is that surface parallel lamellar bone characterizes those areas where bone is being added, while internally (osteon) remodeled bone and partially resorbed osteons characterize areas of bone removal. The process of drift is a response to continuing changes in the load axis of the tibia during and to a lesser extent after the cessation of growth. As such it is analogous to the remodeling of bone in fracture healing (p.61), where alignment of the broken ends is poor.

Osteon Remodeling

During the growth phase, and to a greatly reduced extent throughout the remainder of life, compact bone is added to existing external surfaces of bone by apposition. Appositional bone is deposited in layers, which give rise to the characteristic histological appearance of circumferential lamellar bone. However, like most other tissues of the body, bone needs to be renewed. The process of renewal is called internal or osteon remodeling (Figure 14). All bone remodeling reflects an obvious but important biological property of bone, namely, it is hard. This means that it cannot grow by interstitial expansion as cartilage can, and it cannot be removed without destruction by cellular activity. Because of the hardness of bone this renewal process involves the removal of existing tissue and its replacement with newly formed tissue. Throughout the skeleton much of the lamellar bone formed during growth and remodeling remains at least through early adulthood. However, in compact bone, particularly in the diaphysis of long bones, the surface parallel lamellar bone is replaced by the osteons.

The classic appearance of the osteon is seen in cross-sections of compact bone (Figures 15, 16). The basic structure of an osteon in such a preparation is circular, having a diameter that varies in size but averages approximately 300 microns. The thickness of this page of paper is about 100 microns, so it is apparent that we are talking about a relatively large histological structure. The length of an osteon is problematical since osteons are not discrete units but divide, rejoin and interconnect with other osteons to form a complex network. The order of magnitude for the length of an osteon segment from where it splits off and forms a discrete unit to the point where it joins another similar structure or terminates is from 2 to 5 millimeters. While osteons are most often viewed in two dimensions it is important to be aware of the three dimensional aspect of their morphology.

Structurally, an osteon in cross-section has a histologically discrete, round boundary. Within this boundary are circular layers of bone tissue whose collagen fibers are aligned in alternating directions like the layers in circumferential lamellar bone. Osteocytes are distributed throughout these layers and are interconnected by canaliculi in the bone, which contain the cytoplasmic extensions of the osteocytes. All viable osteons contain a central (Haversian) canal averaging about 50 microns in diameter, which includes a

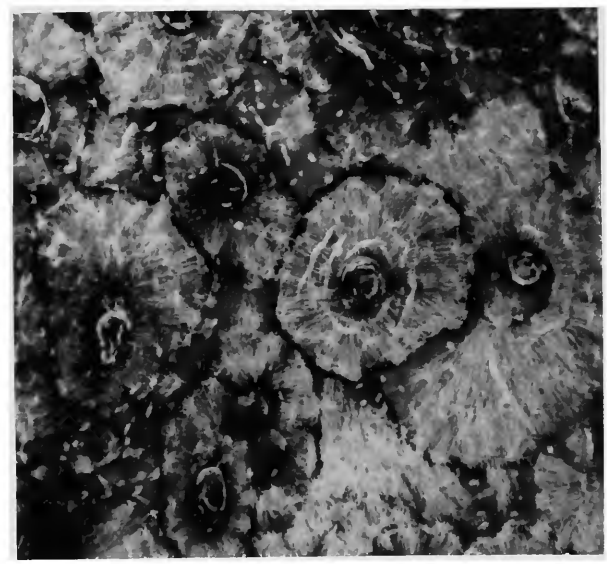
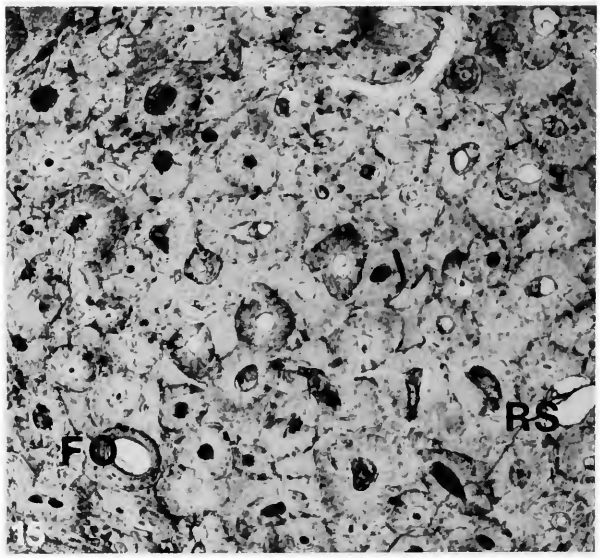
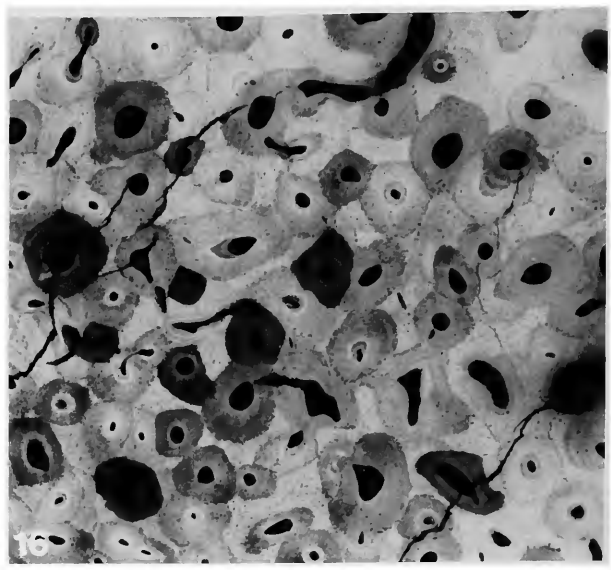


FIGURE 14.—Photomicrographic field of osteons in adult human compact bone. The elongated white structures within an osteon are lacunae. The fine filament-like structure radiating out from the central canal are canaliculi. (Darkfield illumination, approximately × 100.)





FIGURES 15, 16.—Microscopic field showing osteons in various stages of development: 15, A resorption space (RS) is seen in the lower right corner. A forming osteon (FO) is in the lower left corner. Most of the remaining osteons in the field have completed protein matrix synthesis as indicated by the small size of the central canals. (Approximately × 45.) 16, Microradiograph of field in Figure 15 showing variation in osteon density. Less mineralized osteons are darker.

minimum of one blood vessel. At least some osteons contain a nerve. Some of the canaliculi containing cytoplasmic processes of the osteocytes open into the central canal thus providing a pathway for nutrients and for mineral ions needed for continuing mineralization or ion removal in mineral physiology. The canaliculi usually do not communicate between adjacent osteons.

The process of osteon remodeling begins with the removal of bone, creating a circular tunnel (Haversian resorption space). The actual process is best visualized in its longitudinal axis (Figure 17). As an advancing wedge of osteoclasts forms the tunnel, the osteoblasts generally begin the refill process. The initial layer of bone matrix deposited on the walls of the resorption space tends to be somewhat different histologically. It is more highly mineralized and the proportion of noncollagenous protein is somewhat greater. The term "reversal line" is used to distinguish this layer from the remaining layers of bone in the osteon. The space and time sequence between the osteoclast resorbing front and the osteoblast refilling front varies, but classically it is thought to be rather small. Therefore, both resorption and formation take place at the same time, although in different parts of the tunnel. The refill of the

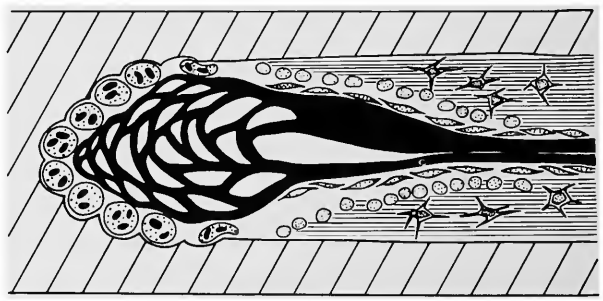


FIGURE 17.—Diagrammatic representation of osteon remodeling in longitudinal section. Osteoclastic resorption is seen to the left, with large multinucleated osteoclasts associated with the scalloped edge of bone. Osteoclastic activity is associated with active hyperemia represented by the interconnected network of capillaries adjacent to the osteoclasts. In the center and right portions of the illustration osteoprogenitor cells (spindle-shaped cells adjacent to the blood vessels) are differentiating into osteoblasts. Bone matrix synthesis and mineralization fill in the resorption space leaving an interconnected network of osteocytes seen as stellate cells in the layered matrix.

tunnel involves a process essentially identical to that described earlier for circumferential lamellar bone formation: (1) layers of protein matrix are secreted by osteoblasts; (2) cells and their cytoplasmic processes are incorporated in the protein matrix; (3) primary mineralization occurs; (4) the process is repeated in a centripetal direction leaving a central canal; and (5) osteocytes mediate continuing secondary mineralization. During osteonal refill of resorption spaces there may be temporary pauses in the refill process. This results in an arrest line, which is histologically similar to the reversal line. With the resumption of matrix synthesis and mineralization, these arrest lines are retained. The physiological factors in the temporary arrested development of osteons are not known, but could prove to be a valuable source of information on physiological conditions in archeological skeletons. The time needed for resorption and refill are important. Bone formation takes at least ten times as long as the destruction of an equal amount of bone (Johnson, 1966:127). This means that for formation-resorption to be in balance, there needs to be ten times as many sites of active bone formation as there are sites of active bone destruction. In osteon remodeling the theoretical equilibrium ratio in normal tissue remodeling is about ten osteons in the forming stage for each resorption space.

In fact some studies have shown that in compact bone of present day populations the observed ratio is very close to the theoretical ratio (Blumberg and Kerley, 1966:159; Ortner, 1970:65). Deviation from this ratio may be indicative of inadequate nutrition or other morbid conditions. A study by Richman and Ortner (1978) of osteon remodeling in Eskimos, Arikara (Plains Indians), and Pueblo Indians (Southwest U.S.) indicated that the Eskimo sample has a formation-resorption ratio of 10 to 1. However, the two American Indian samples have a ratio of approximately 6 to 1, indicating a substantial reduction in bone replacement after resorption. The meaning of this finding needs further clarification; however, nutritional factors seem a rather obvious possibility. Among the Eskimos meat constitutes the major source of food. The two American Indian groups and particularly the Pueblo Indians depended heavily on maize. Meat contains much more protein than maize and would thus be more likely to provide the amino acids needed for synthesis of organic bone matrix. Stout and Simmons (1979) review the application of histomorphometrical analysis in archeological bone specimens.

During the growth phase appositional development of circumferential bone tends to be relatively rapid. Woven bone may be deposited on the cortex as fine, cancellous bone. Subsequently, the spaces in such woven, cancellous bone are filled in by lamellar bone, creating primary osteons (Enlow, 1963:63). Although similar in appearance to secondary osteons, such structures do not represent a remodeling event. Histologically they differ from secondary osteons, tending to be smaller in cross-section. Also, primary osteons do not have a clearly defined reversal line marking the termination of bone resorption (Figure 18).

Primary osteons are generally replaced by sec-

FIGURE 18.—Primary osteon (arrow) in the long bone of an infant about 6 months of age. This structure results from the fill-in of spaces in newly formed bone. In human bone it is primarily associated with the very rapid growth occurring during fetal and infant development. (Approximately × 100.)

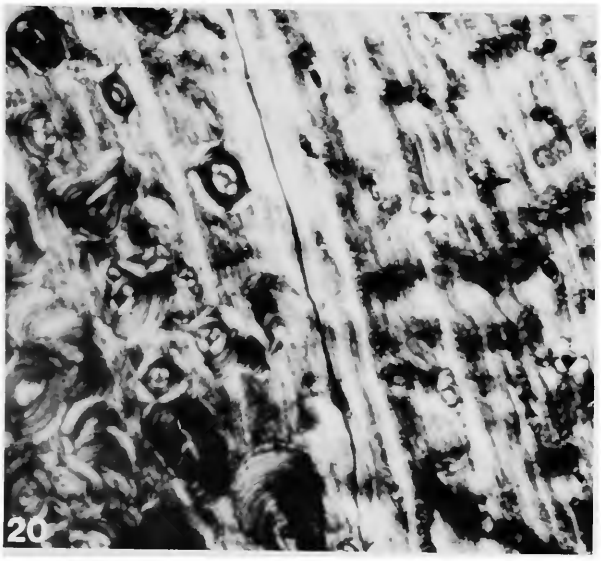
ondary osteons which, in turn, are frequently replaced by additional secondary osteon formation. Exactly what the local factors are that initiate normal remodeling within a specific osteon is not known. However, changes in the direction of force (weight and muscle activity) and the death of osteocytes are thought to play a role. The tunneling activity of the osteoclasts disregards previous boundaries of an osteon so that encroachment on the walls of adjacent osteons is a frequent occurrence and may result in the destruction of the canal of an adjacent osteon.

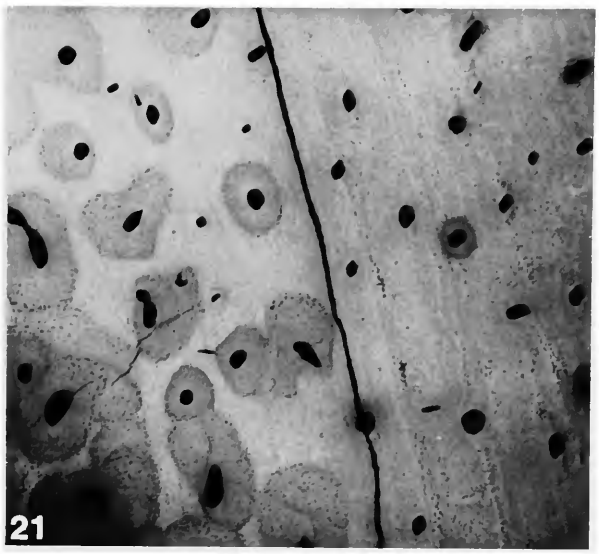
In addition to the classic osteon remodeling described above, which results in the typical appearance of the osteon, there is another type of osteon remodeling, which occurs entirely within an existing osteon. This type of remodeling (Figure 19) produces an "osteon-within-an-osteon" appearance shown in an early illustration by Tomes and de Morgan (1853, pl. 6: figs. 1,6) and is characterized by a reversal line like that found on the periphery of a classic osteon. This type of remodeling however, is intermittent along the longitudinal axis of an osteonal canal. Ortner (1975:29) has argued that the small, cross-sectional and longitudinal size of such osteons suggests a physiological rather than a mechanical function for such osteons. Richman, Ortner, and Schulter-Ellis (1979) have found differences in the frequency of this type of osteon in three archeological skeletal samples. They attribute this difference to variation in diet.

The effect of this type of remodeling on tissue in the periphery of the antecedent osteon is not known. Normally bone tissue isolated from vascular supply by the reversal line will die. However, the intermittent distribution of this type of remodeling in the longitudinal axis might make it possible for such tissue to be adequately supplied via the anastomosing network of canaliculi from areas above and below the new osteon in the antecedent osteon.

The various factors involved in osteon remodeling result in a diverse pattern in the normal cross-sectional histology of bone. The paleopathologist should be familiar with these factors in order to recognize abnormal histological patterns. This morphological diversity is illustrated in Figures 20 and 21. In Figure 20 we see a few osteons, but most of the field is unremodeled, circumferential lamellar bone. Typically the vascular supply in such tissue is poor with greater distances between blood vessels than in osteon remodeled bone. The microscopic field in Figures 15 and 20 contain: (1) osteons, (2) interstitial bone left behind after osteon remodeling of circumferential lamellar bone, (3) osteon fragments remaining after partial destruction of an osteon, (4) small osteons entirely within an older osteon, and (5)

FIGURE 19.—A small osteon (arrow) entirely within a larger osteon. The remodeling that produces small osteons is intermittent along the longitudinal axis of an osteonal canal. Such osteons appear to be a response to physiological rather than mechanical conditions affecting bone tissue. (Adult, human compact bone, approximately × 150.)





FIGURES 20, 21.—Cross-section of a compact bone from a young adult: 20, The right half of the field is primarily circumferential lamellar bone containing several primary vascular canals. The left portion of the field contains remnants of circumferential bone, but much of this bone has been replaced by secondary osteons. (Polarized light, approximately × 70.) 21, Microradiograph of field seen in Figure 20. Note that the circumferential lamellar bone is denser than that in the osteons.

osteons in various stages of development. The distribution and relative proportions of these features will vary greatly within a single bone, from bone to bone, and with the age of the individual. However, most of these features can be seen in any normal cross-section of cortical bone.

Trabecular bone is associated with the marrow and cancellous spaces of bone. In adult long bones it is found primarily in the ends of the bone. During fetal development and in infancy and early childhood, trabeculae may consist of woven fiber bone. By late childhood trabeculae are made of lamellar bone. This type of bone does include osteocytes like compact bone; however, vascular canals and osteons are rare (Figure 22).

In addition to the histological variation that can be seen in a microscopic preparation of a cross-section of bone, there is often considerable variation in mineral density of the various osteons. This aspect of bone histology can be recognized with some experience in stained or polished sections; however, it is most easily seen in a microradiograph. Briefly, a microradiograph is a high resolution X-ray image that reveals variations in the mineral density of microscopic features in undecalcified bone sections. With the completion of matrix synthesis and initial mineralization, the osteon has achieved approximately 70 percent of maximum density. Secondary mineralization is mediated by the osteocytes and occurs much more slowly, typically achieving about 90 percent of maximum mineralization within a year or two. Osteons in various stages of mineralization are easily recognized in microradiographs.

Figure 16 is a microradiographic image of the field seen in Figure 15. Several features may be seen in the microradiograph which supplement what is less apparent in the bone section itself. Osteons in the forming phase are very dark, indicating low mineralization. Osteoid seams in forming osteons that are just beginning to be mineralized are almost black. Some osteons have several light (dense) bands, which are associated

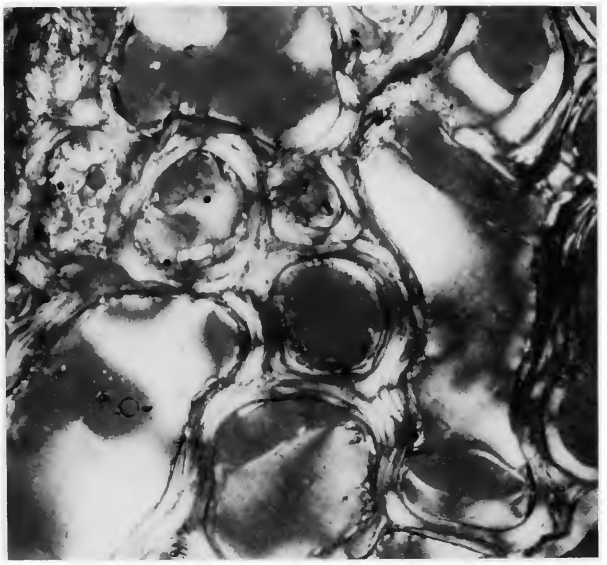


FIGURE 22.—Trabecular bone from the metaphysis of an adolescent human femur. Note the large spaces between the thin walls of the bone. (Polarized light, approximately × 40.)

with a temporary cessation of osteoblastic refill. Several osteons have a broad zone of denser bone adjacent to the central canal, which may represent a special, physiologically active, type of mineral.

The process of matrix formation and mineralization in osteon remodeling is influenced by a number of factors, including age, physical activity, and disease. From childhood through early adulthood, the process of secondary mineralization is faster than at a later age. The microradiographic picture during this period is characterized by relative uniformity in the mineral density of osteons. With increasing age both the matrix refill and secondary mineralization slow down. In the microradiograph the result is increasing variability with age in central canal size and mineral density (Jowsey, 1960:215; Ortner, 1975:33). However, vigorous physical activity will tend to minimize the effect of aging on osteon remodeling and mineralization.

The review of bone thus far has emphasized the mechanical and structural aspects. Earlier I have emphasized that bone should also be thought of as an organ with physiological functions analogous to those performed by any of the other organs of the body. However, to understand the physiological aspects of bone as an organ, the two basic components of bone, the protein and mineral phases, and their interrelationships need to be understood.

Approximately 90 percent of the bone protein matrix is collagen. The remaining matrix consists of several proteins including those subsumed under the term "muccopolysaccharides" (sugar-protein complexes) and a small amount of lipoproteins (fat-protein complexes). More recently, additional knowlege regarding the chemistry of the sugar-protein complexes has indicated that the term "protein polysaccharides" would be more appropriate for this general category of noncollagenous protein. In addition to the noncollagenous proteins, amino sugars are found in the organic matrix of bone.

It is important to emphasize that the components of the organic matrix of bone are not distributed uniformly throughout skeletal tissue. For example, the molecules of noncollagenous matrix are more highly concentrated at the boundaries of tissue, such as the reversal line and the border of the central canal in osteons. Furthermore, noncollagenous matrix is more highly concentrated during the forming and early mineralization phase of growing or remodeling skeletal tissue. Apparently the amount of collagen in bone matrix is constant once the tissue is formed. The variable components include (1) the noncollagenous organic material, (2) water (about 10 percent), and (3) the bone mineral.

The mineral matrix of bone, like the organic matrix, is complex. This complexity is still not completely understood. However, it is now generally accepted that the major mineral component is hydroxyapatite, having a chemical formula of $Ca_{10}(PO_4)_6(OH)_2$. The atoms in a molecule of hydroxyapatite are arranged in such order to give crystal properties to the mineral. In addition to crystalline calcium-phosphate, there is growing evidence for the existence of an amorphous or noncrystalline mineral phase in bone

(Posner, 1969). The precise relationship between the two types of mineral is not known. However, the proportion of amorphous mineral decreases with increasing mineralization. This suggests that the amorphous calcium-phosphate might be a transitional phase existing predominantly during the early stages of mineralization, There is some evidence that there may be localized concentrations of amorphous mineral in discrete histological zones within bone tissue (Ortner and Von Endt, 1971). These zones may be important in mineral physiology by providing a more reactive or labile source of mineral ions.

A detailed review of hypotheses regarding the chemical and structural relationship of bone protein and mineral is not pertinent here. However, bone protein precedes bone mineral and is involved in the mineralization process possibly by providing the initial site for mineralization. Two mechanisms have been proposed for this process. In one hypothesis collagen provides a template or pattern to which mineral ions are attracted and arranged in the appropriate spacial relationship for mineral crystallization. The second hypothesis involves chemical binding of one of the mineral components, such as phosphate to a reactive group of some of the amino acids of collagen. Lees (1979) has proposed a model with both inter- and intrafibril mineralization of collagen.

Another view of initial bone mineralization implicates the noncollagenous protein. However, Hancox (1972:87) notes that much of the research supporting this hypothesis is based on mineralization in cartilage. He is of the opinion that bone mineralization is different from that occurring in cartilage and questions the validity of such research in explaining mineralization in bone.

The mineral phase of bone includes calcium and phosphate. However, the crystal structure of bone mineral is such that substitutions by some elements (e.g., strontium, radium, and lead for calcium; fluorine for the hydroxyl group) can take place for elements and ions in the crystal lattice. Furthermore, many elements may be attracted to the surface of bone crystallites (adsorption) and the hydration shell surrounding the crystal. Given the small crystal size found in bone mineral, a few hundred angstroms in length and breadth and about fifty angstroms thick (Neuman and Neuman, 1958:55), there is an enormous amount of bone crystal surface and a high likelihood of considerable exchange of ions between bone and body fluids. This offers the potential for bone to contain a considerable variety of chemical elements. It has also been noted that bone mineral may be present in an amorphous phase, which may have particular significance in bone physiology.

Johnson (1966:67–68) suggests that mineral ions are almost instantly available from the labile mineral lining the canalicular system. The next most accessible source of mineral would be from the osteocyte lacunae. Neither of these sources necessarily involves the dissolution of bone matrix. The surfaces of bone including trabeculae provide a second source of mineral ions, with the Haversian canals providing the least accessible source of mineral. Johnson appears to assume that ions from the last two sources must be released by osteoclastic activity. However, radioactive tracer studies suggest that exchange (including both uptake and loss of radioactive ions) can occur without matrix destruction by osteoclasts in the tissue surrounding the Haversian canal (Rowland, 1966:243).

Regardless of the actual mechanisms involved, it is clear that bone acts as a mineral bank. Removal of mineral components from bone tissue is controlled systemically by parathyroid hormone. Jande (1972) has shown that increases in serum parathyroid hormone levels results in loss of mineral around the osteocyte. Parathyroid hormone also increases the activity of the osteoclasts in destroying bone. The hormone calcitonin is thought to act antagonistically to parathyroid hormone, although its action is less well understood and less obvious than parathyroid hormone.

The major factor that affects the secretion of parathyroid hormone is the serum calcium level. The normal range for calcium in blood serum is from 8.5 to 10.5 milligrams percent (or 8.5 to 10.5 milligrams per 100 cubic centimeters of serum).

A drop in serum calcium levels below the normal range activates the parathyroids and increases hormone levels, resulting in rapid (minutes to hours) loss of calcium from canaliculi and lacunae (Jande, 1972). If the parathyroid hormone concentration remains at the activation level for longer periods of time, osteoclastic resorption takes place. In the disease state continuing resorption may result in severe osteoporosis.

This brief review of the physiology of calcified tissue serves to highlight the complex relationship between bone structure, particularly at the microscopic and ultrastructural level, and the physiological function of bone. Abnormal physiological conditions will occasionally be associated with gross or microscopic features observable in archeological specimens. The specific details of skeletal responses to abnormal physiological conditions are treated later, particularly in the chapters on metabolic disorders (p. 270) and endocrine disturbances (p. 298).

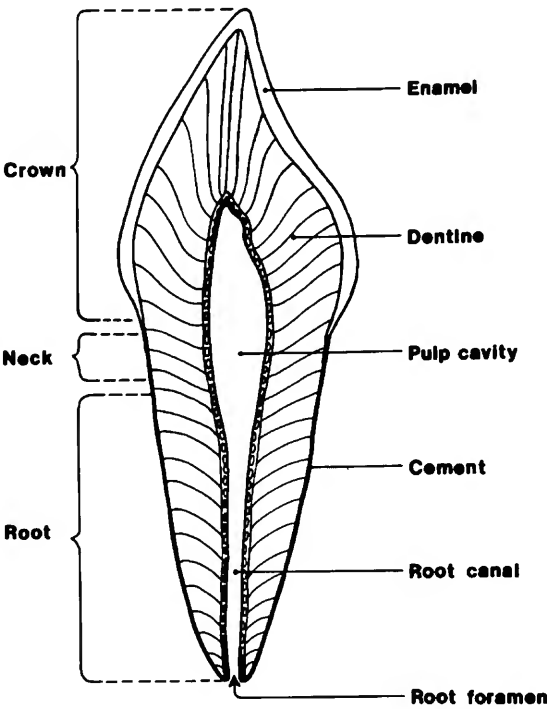


FIGURE 23.—Schematic representation of the major components of a human tooth.

The Biology of Teeth

The mature tooth consists of three components (Figure 23). The first of these is the very hard, exposed surface called enamel. Enamel has no vascular supply and no cells. Dentine is the second component and provides support for the enamel in the tooth crown and forms the hard tissue portion of the tooth root. Dentine is avascular but has cells (odontoblasts) lining its inner surface. Tubules extend from the odontoblasts to the outer surface of the dentine. The third component of a mature tooth is the pulp cavity and root canal, which contain the nerve, vascular supply, and pulp mesenchyme, which is the source of the odontoblasts.

The following review of tooth development is based largely on the figures and text of Hamilton and Mossman (1972:572–575, figs. 549–554) and Sicher and Bhaskar (1972). About the sixth week of embryonic development, ectoderm begins to invade mesoderm in the region of the upper and lower jaw. This invasion consists of an outer arch, which will result in the gum and inner arch called the dental lamina. Subsequently the dental lamina forms individual enamel organs with a lining of epithelial (ectodermal) cells called ameloblasts.

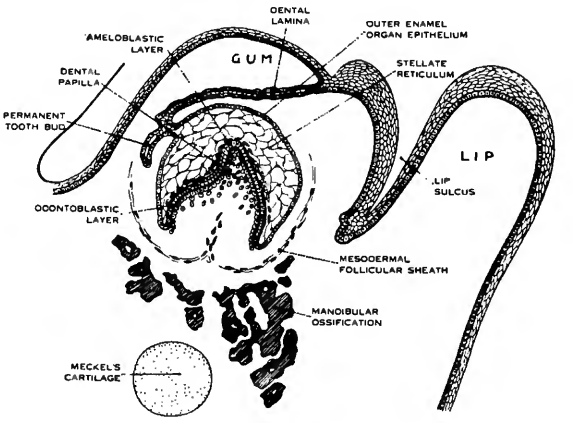
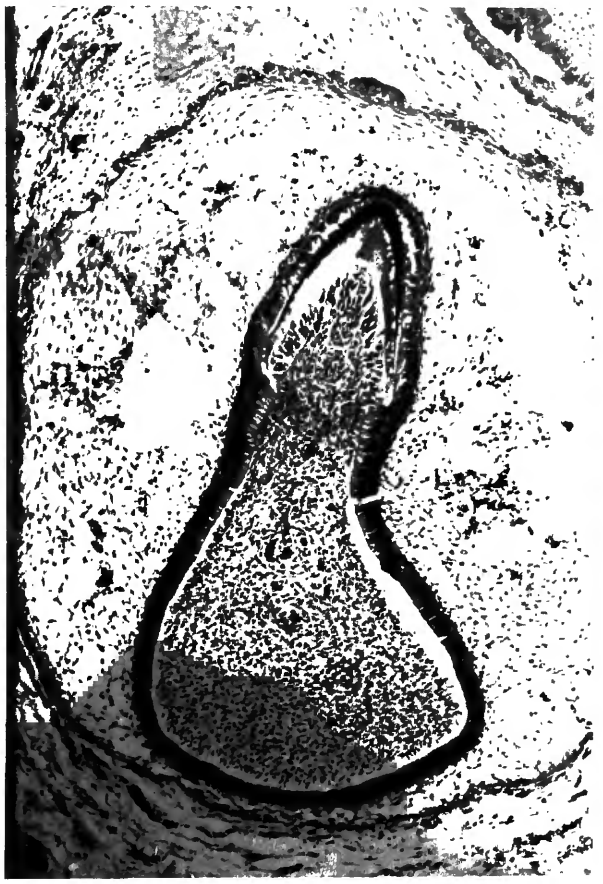


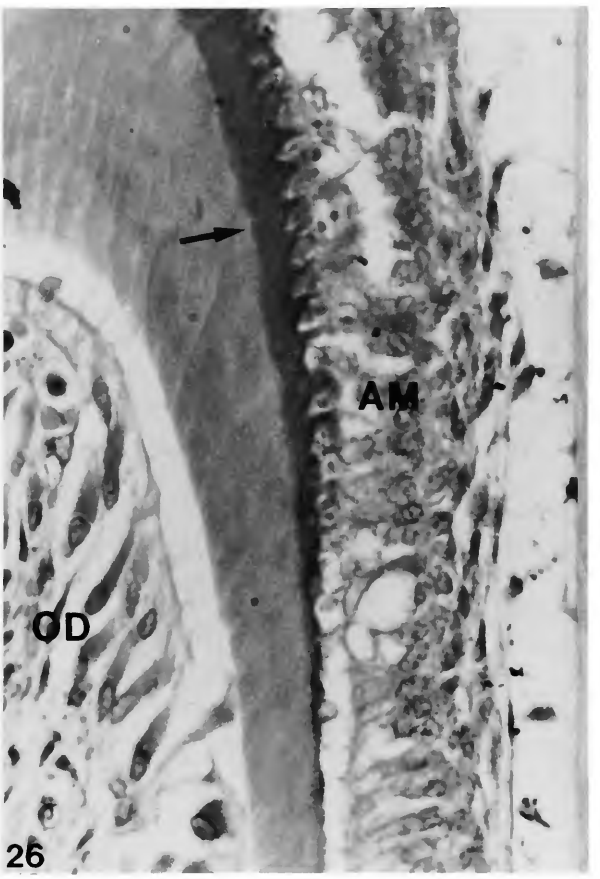
FIGURE 24.—Schematic representation of a developing human fetal incisor with a crown-rump length of 100 mm, approximately 2½ months fetal-life (Reprinted, with permission, from Hamilton and Mossman, 1972:574, fig. 552.).

This lining develops cup-like structures, which will form the enamel of each deciduous tooth. A branch of the lamina will similarly form the enamel for the permanent teeth. Filling the cup-like indentation in an enamel organ are proliferating mesenchymal (mesodermal) cells known as the dental papilla, which will produce the dentin (Figure 24).

With the establishment of a connective tissue sheath around both the enamel organ and the dental papilla the two structures are called a tooth germ or bud. At this stage of development, the tooth bud consists of (1) an outer layer of ectodermal cells (epithelium), (2) an intermediate zone of stellate reticulum, (3) an inner layer of ectodermal cells, (4) highly vascularized mesodermal cells (mesenchyme) associated with the inner layer of ectodermal cells, (5) a capillary network surrounding both the enamel organ and the dental papilla, and (6) a connective tissue sheath around the capillary network.

Later in the development of the embryo the inner layer of ectodermal cells (ameloblasts) will secrete the enamel of the tooth and is called the ameloblastic layer. Some of the mesodermal cells in the dental papilla become arranged along the





FIGURES 25, 26.—Tooth bud in a 5-month human fetus: 25, Low magnification, the enamel is the dark layer of the apex of the tooth. (Partially polarized, approximately × 85.) 26, Higher magnification showing detail of dentin and enamel in tooth bud. Boundary between dentin and enamel is indicated by the arrow. Odontoblasts (OD) are seen with long processes extending into dentinal tubules. Ameloblasts (AM) line the outer surface of the tooth bud and are actively forming the enamel. (Approximately × 400.)

ameloblastic layer. The cells in this layer are odontoblasts and will produce the dentin. The two layers of cells are separated by a membrane (membrana praeformativa) the shape of which directly defines the ultimate shape of the tooth. The formation of both dentin and enamel proceeds by addition (apposition) away from this membrane—dentin in a centripetal direction, enamel in a centrifugal direction (Figures 25, 26).

The ameloblasts ultimately disappear after enamel formation is completed, leaving behind, on the enamel surface, a thin covering or dental cuticle. The odontoblasts persist throughout the life of the tooth as an inner lining of the dentin. The remainder of the mesodermal cells in the dental papilla become richly vascularized and form the dental pulp. The tooth root begins to form before eruption but is not completely formed until some time after eruption. The root surface consists of a specialized tissue (cementum), which provides attachment for connective tissue (the periodontal membrane), which in turn attaches to bone. The sequence of tooth formation and eruption provides a useful index of age through the adolescent years. For the present discussion it is sufficient to emphasize that the timing of eruption varies with the developmental age of the individual and the tooth type.

During development, teeth require adequate nutritional input including protein, calcium, phosphate, trace elements, and vitamins A, C, and D. Inadequacies in these components may result in grossly abnormal teeth or in teeth more susceptible to wear and microbial invasion (caries). Abnormalities of teeth may also be the result of diseases occurring during development, such as rubella and syphilis, as well as of genetic problems. Various components, such as the enamel or dentin, may be differently affected by a morbid condition.

Methods Used in the Analysis of Skeletal Lesions

Today's physicians have at their disposal a vast array of data to assist them in diagnosing a morbid condition in a living patient. Most of these data are not available to the paleopathologist trying to diagnose a lesion in an archeological skeleton. In spite of the obvious limitations facing paleopathologists, it is well to remember that they have at least one significant advantage over a present day pathologist. If an archeological skeleton is recovered in a good state of preservation, the paleopathologist can examine an abnormal skeleton in its entirety and to a degree that would be quite impossible in a current patient, living or dead. In paleopathology the fundamental objective should be to obtain as much background information on the skeleton as possible and take maximum advantage of one's ability to evaluate the skeletal lesion itself by direct observation. I shall review the methods for obtaining background information on skeletal specimens first and then review the methods for analyzing a morbid condition in the skeleton.

Basic to any evaluation of pathology in archeological specimens is the following data: (1) age, (2) race or human population, (3) sex, (4) geographic location, and (5) time period or archeological horizon. A knowledge of these factors can have a substantial influence on the probabilities of various types of morbid conditions. For example, if we are able to determine that a specimen was a young adult at the time of death, this fact would greatly reduce the probability that a diseased skeleton was affected by carcinoma, since this is primarily a problem of the aged. Likewise the knowledge that a skeleton, with multiple, disseminated, destructive lesions, was male would tend to rule out metastasis from breast cancer.

It is clear that some diseases are associated with specific human populations. Sickle cell anemia primarily affects black populations while another anemia, thalassemia, is found in peoples from the Mediterranean area and in southeast Asia. While there is generally an association between racial distribution and geography, the factors affecting the individual in a specific geographic location may not be genetic as is the case in sickle cell anemia and thalassemia. For example, coccidioidomycosis is endemic mainly in dry, dusty regions of southwestern United States. This disease affects members of any human population living in this area although the incidence may vary (Miller and Birsner, 1949).

Time should rarely be a crucial factor in the diagnosis of a skeletal lesion. However, it cannot be ignored. The significance of the time factor will be more apparent in the discussion of treponemal conditions (p. 180). The problem can be stated briefly for illustrating its importance. The question is whether syphilis was introduced by people from the Old World into the New World or vice versa. This is a very complex problem but the unambiguous identification of a case of syphilis in a specimen dated before 1492 in either the Old or New World would obviously be a significant bit of information.

In evaluating all of the above factors with respect to a diseased archeological skeleton it is important to be open minded before eliminating a possible disease on the basis of any one element. For example, it would be unwise to eliminate the diagnosis of syphilis for a pre-Columbian New World skeleton simply because some scholars feel that syphilis originated in the Old World. While this is a rather obvious situation the principle applies to other variables as well, particularly since there is always a margin of error in determining any of these variables in an archeological skeleton.

Many of the criteria for determining the above variables are available in published sources and

need not be discussed in detail here. The following review is derived largely from published sources and will provide a general summary of methods for obtaining background data on archeological specimens. However, the emphasis will be on those aspects of the variables that are significant for research in paleopathology.

Age Determination

The estimation of biological age in an archeological specimen is related to several biological processes, such as growth and skeletal remodeling, which result in age related changes in the morphology of skeletal components. These changes are seen at the gross, as well as at the microscopic level. All of the methods for age determination attempt to establish the relationship between morphological changes in the skeleton and the age of the individual at the time of death. Furthermore, the methods involve the establishment of procedures for estimating age on the basis of these changes. Methods for determining age vary significantly in terms of research design, statistical methodology, appropriate age range, etc.

Through the age of 18 (subadult) three methods of aging are used. These are based on age-related variations in (1) bone length, (2) dental eruption and root development, and (3) the appearance and fusion of secondary centers of bone development (epiphyses and apophyses). Excellent and readily available reviews of these methods are found in Krogman (1962) and Stewart (1968, 1979a). It is important to emphasize that estimates of age based on these methods reflect the usual problems in standardizing such procedures. One must assume that the research design of the original study was adequate, particularly with respect to sampling the entire age distribution and population and that the morphological features can be assessed objectively. Furthermore, in applying any method of aging to an archeological skeletal sample one assumes that the sample used to establish the prediction procedure and the unknown archeological sample are from the same statistical population of individuals. These conditions, particularly the latter, frequently are not met. In such situations one hopes that the approximation is close.

There is evidence that different methods of determining age may reflect different statistical populations. Merchant and Ubelaker (1977) compared two different techniques of determining age in subadult material, both of which were based on the sequence of dental development. When both techniques were applied to a single archeological sample, they found considerable difference in the age as determined by each technique. With this and other limitations in mind let us review the various methods for estimating biological age in archeological samples.

Fetal material in archeological skeletal series is relatively rare but may be very significant, as in a situation where the mother died and was buried with the fetus. Estimation of age in such material may be a factor in inferring whether problems in birth might have been a factor in the mother's death. Stewart (1968:132) provides a regression formula based on femur length for estimating age from the fifth fetal month through birth. At birth the bony femur is about 80 mm in length, which provides a convenient criterion for distinguishing fetal from postfetal material.

Other criteria may be inferred, if the preservation and excavation has been very careful, by determining the presence or absence of centers of ossification. Standards for these features are published in Stewart (1979a) and Krogman (1962: 56–68). However, these standards are for birth onward. The absence of bones, such as the calcaneous, talus, cuboid, and the epiphyses of the distal femur, proximal tibia, and proximal humerus, would be indicative of death before birth. In archeological material identification of such ossification centers may be difficult or impossible since they may bear little if any resemblance to the shape of the bone later in development and may not be preserved. Careful excavation may permit recovery of ossification centers in association with an identifiable bone.

From birth through the growth phase, the criteria for estimating age are based on long bone

length (particularly femur), the appearance of ossification centers, the sequence of dental eruption and root development, and, during the late growth period, the fusion of secondary ossification centers. The methods of age estimation based on these variables are summarized by Krogman (1962), Stewart (1968, 1979a), and Ubelaker (1978:46–53). By the age of 18 years (earlier in females), significant growth has stopped in most individuals although some epiphyses will continue to fuse through about age 25 years.

From 18 years onward, other aging criteria become more significant. These include the continuing metamorphosis of the pubic symphysis (Todd, 1920, 1921; McKern and Stewart, 1957; Gilbert and McKern, 1973), which provides age estimates to the age of 50 years. The fusion of skull sutures has been a traditional criterion for age determination. In their study of Americans killed in the Korean War, McKern and Stewart (1957:37) found that suture closure was poorly correlated with age and of little use in age estimation. Krogman (1962:89) supports this conclusion but notes that the skull may be the only part of the skeleton present and thus the only source for estimating age in an unknown specimen. While Krogman's observations are addressed to forensic modern specimens they are also appropriate for archeological skeletal collections. At the National Museum of Natural History, for example, more than half of the collection of about 25,000 skeletal specimens consists of the skull alone. Another problem in archeological material is that the pubic symphysis and other age criteria are particularly vulnerable to destruction in the soil and may not be useable. Thus, age estimation based on suture closure may be the only criterion available, but its limitations should be recognized.

Other aging methods have been developed including those based on tooth wear (Brothwell 1965a:67–70) and the degree of arthritic degeneration (Stewart, 1958a). Without doubt these methods provide helpful corroboration for age estimates from other sources. However, they are limited by the influence of factors that are not part of the process of biological aging, such as the coarseness of the diet and the physical activity of the individual.

A recently developed method for age estimation is based on the process of microscopic bone tissue renewal (osteon remodeling), which continues throughout life. Early studies by Amprino and Bairati (1936) and by Jowsey (1960) suggested the existence of age dependent changes in osteon remodeling. Kerley (1965) was the first to apply adequate statistical samples and procedures to this problem. In his research he used four histological features: (1) osteons, (2) fragments of former osteons, (3) the amount of unremodeled bone, and (4) the number of nonosteonal vascular canals. The first two of these criteria increase with age, while the last two decrease with age. The statistical association between age and frequency of these features is the basis for estimating the age of an unknown specimen. Kerley determined this relationship for three major bones: femur, tibia, and fibula.

The error in estimating age using this method compares favorably with estimates based on changes in the pubic symphysis. However, microscopic aging methods can be applied to ages in excess of 50 years and thus, extends our ability to estimate age into the older age categories. Because there is an increasing discrepancy between biological and chronological age as one gets older, the factor of error is higher in the 50-plus age range. Regardless, this is the only method currently available that allows age estimation for skeletons over 50 years of age. Kerley and Ubelaker (1978) have published revised regression formulae for Kerley's earlier study.

As with any method there are problems in using Kerley's method. Preparation of bone sections is a time consuming and delicate procedure and even more so in archeological specimens, where the bone may have to be strengthened with plastic to prevent crumbling during processing. Soil erosion and organisms may destroy microscopic features needed for such an analysis. The procedure also assumes a fairly sophisticated knowledge of compact bone microstructure. One

modification of Kerley's method appears to offer a simpler procedure (Ahlqvist and Damsten, 1969) but does not appear to be as accurate in the lower age ranges (Bouvier and Ubelaker, 1977; Ubelaker, 1978:66).

Race Determination

Race (here used in the sense of a discrete human population) determination in archeological skeletons is rarely a significant problem for the paleopathologist, since the archeological provenience usually provides a reliable indication of at least the broad racial background. Occasionally, however, members of more than one human population will be buried in the same cemetery, and identification of this fact can provide relevant background data for the interpretation of skeletal lesions. Methods for distinguishing between broad racial categories (e.g., White, Negro, Mongoloid) are well known and have been summarized in Krogman (1962) and Stewart (1968, 1979a).

The major differences between human populations are best seen in the skull. Traditionally, physical anthropologists have characterized these differences in terms of various complexes of the skull such as skull vault shape (long headed, round headed), relative projection of the jaws (prognathism), shape of the nose (long and narrow in Whites, short and broad in Negroes), and relative position of the cheek bones (prominent in Mongoloids).

In the traditional approach to racial characterization, measurement of skull dimensions is important, but each measurement tends to be treated as a discrete feature. A more recent development has applied multivariate statistical procedures to broad racial classification (Giles and Elliot, 1962) and to more subtle skull variations within broad racial categories (e.g., Howells, 1973, Ortner and Corruccini, 1976). A multivariate statistical approach to racial variation provides greater objectivity and a measure of racial distance, which is based on many measurements.

The remainder of the skeleton does not provide unambiguous evidence of racial affiliation. Stewart (1962) has reported that the femora of Negroes tend to be straighter than those of other races. However, there is considerable overlap in the distribution of the data, making prediction of race difficult. Furthermore, the postcranial skeleton is greatly affected by nongenetic factors such as diet, making it unlikely that racial differences can be resolved on this basis alone, at least to the extent of being useful in paleopathology.

Sex Determination

Sex differences in skeletal specimens are probably the least ambiguous of all the factors basic to the paleopathologist. In the skeleton the best criteria for determining sex are in the pelvic bones. These bones reflect the fundamental differences in functions associated with reproduction, particularly childbearing, in the female. In the female pelvis the major factor influencing its shape is the necessity for the infant to pass through the pelvic canal during birth. Both the dimensions and the shape of the pelvic inlet and outlet reflect this function (Figure 27). Thus, the female inlet and outlet are relatively and usually absolutely larger and rounder than in the male. A more detailed review of the specifics of these differences is given in Krogman (1962) and Stew-

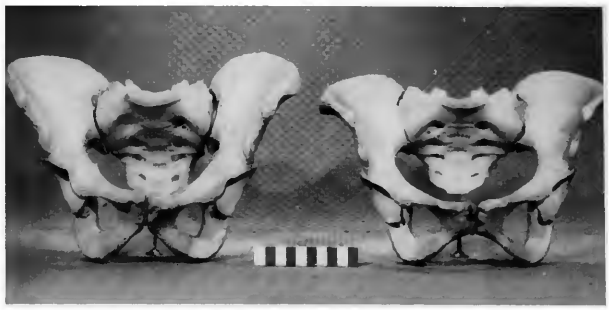


FIGURE 27.—Comparative views of the male and female pelves. Although the male pelvis is larger overall, the female pelvic inlet is larger. The female sacrum is wider and the pubic bone is longer. The female subpubic angle is larger than the male. (Male, left, NMNH 320026; female, right, NMNH 320001.)

art (1968). The problem in applying these criteria to archeological specimens is that the most distinctive differences around the pubic symphysis are also the most likely to be damaged.

Even if the pubic bone is damaged it may be possible to apply a method for determining sex as reported by Phenice (1969). This method is based on the sex differences that occur on the ischiopubic ramus involving three criteria: (1) the ventral aspect of the pubic symphysis region, which is an arch in females, (2) the subpubic portion of the ramus, which is concave in females, and (3) the medial aspect of the ramus, which is broad and flat in males, narrow and ridge-like in females. Phenice reports a remarkably good accuracy of about 96 percent in determining the sex of specimens from test samples where sex is known. The size of the angle of the sciatic notch is larger in females and is also useful in determining sex.

The sex differences that occur in the rest of the skeleton, including the skull, generally reflect the fact that females are smaller, less muscular, and more gracile. Standards for sex determination for different bones from several human groups can be found in both Krogman (1962) and Stewart (1968). It is important to emphasize that the range or distribution of this type of sex variation differs between populations, with females tending to be more male-like in some populations and males more female-like in others. Ones experience in determining sex in one archeological population is not necessarily transferable to another population. Fortunately, this caution is less significant with respect to the pelvis. However, for many sex features it is essential for the paleopathologist to adjust his criteria for sex determination as he studies different skeletal populations, by comparing nonpelvic sex features, such as those seen in the skull, with sex-related traits in the pelvis. By studying skeletons that are fairly complete first, one can adjust the male-female sex criteria for nonpelvic features toward the male or female end of the range as appropriate and thus better estimate sex when pelvic features are missing.

Stature Determination

Stature (height) is a variable that is controlled largely by genetics. However, it is also clear from numerous studies that the health of the individual is also a significant component affecting stature. General health reflects many factors, including diet and disease, so that stature comparisons, particularly in combination with other types of analysis, can provide valuable data on the general health of skeletal samples. One should, however, carefully consider whether one's research objectives can be served equally well by comparing the long bone lengths themselves rather than converting lengths to stature estimates.

Since stature is highly correlated with long bone length, it is possible to estimate stature by measuring one or several long bones and applying the appropriate regression formula. Numerous formulae for different long bones and human populations have been reported previously by Trotter and Gleser (1952, 1958). Both Krogman (1962) and Stewart (1968) summarize the work of Trotter and Gleser and other reports as well. Since the proportions of various parts of the skeleton vary in different human groups, it is important to use a formula that is based on a population similar to the archeological material being studied.

Estimation of stature in archeological skeletons is limited by the same problem of damage and missing bones as in estimating other variables. Steele (1970) has developed regression formulae for fragmentary material, which are based on modern White and Negro specimens. However, this method may be helpful with archeological samples even if not from these populations provided that one is cautious regarding conclusions based on such estimates.

Demographic Analysis

It is important in studying ancient disease not to limit one's estimate of health in skeletal populations to direct evidence of morbidity or lack of

it. Much can be learned by the analysis of mortality and the demographic aspects associated with it. Bogue (1969:549) notes that mortality exerts a varying force on different human populations. Clearly a major factor in this variation is the condition of health in these populations. Bogue (1969:549) also emphasized that "mortality has a very unequal impact on various age groups." The overall pattern of death rate in various age categories will also vary in different populations. This is clearly demonstrated in the marked reduction of infant deaths in highly developed societies as contrasted with developing societies (Bogue, 1969:556–557). Estimates of mortality in an American Indian skeletal population reveals high infant mortality but, in addition, another peak in mortality is present around adolescence (Ubelaker, 1974:60). Other estimates of mortality in ancient skeletal populations from the eastern Mediterranean have indicated higher mortality in females of childbearing age than males of the same age, which probably reflects the hazards associated with pregnancy and childbirth (Angel, 1969a; 1971:71).

Of several statistical methods treating mortality, one of the most useful is the life table (e.g., Bogue, 1969:551–552; Swedlund and Armelagos, 1976:47). In general, the life table summarizes the frequency of events and the probability of events happening within discrete intervals of time throughout the entire range of life within a population. Basically these events include (1) the number of deaths occurring per unit of population within specific age intervals and (2) the probable average number of years of remaining life for individuals in each category. Variations in these factors reflect many conditions, such as diet, disease, warfare, and other problems. As such, data in life tables may provide nonspecific but useful information on the relative health of a human population.

A discussion of the methods and limitations of constructing life tables is given in several sources (e.g., Bogue, 1969:551–555; Swedlund and Armelagos 1976:63–64; Ubelaker, 1978:91–97). Such sources should be consulted before developing a life table for skeletal material. However, it is important that one be generally aware of the assumptions and hazards in applying such techniques to archeological populations. The most obvious problem arises from the fact that modern life tables are based on events (deaths) occurring in a living population, whereas archeological reconstructions of life tables are based on a population of individuals all of whom are dead. The characteristics of the two types of populations are not necessarily the same, although this must be assumed in such applications. Another problem is the extent to which skeletons recovered from a cemetery represent all the individuals who died in a given population. Cultural practices regarding formal interment in cemeteries vary and may not include burial of infants. Furthermore smaller skeletons may not be preserved as well as larger ones. This would, for example, adversely affect the proportion of infants recovered from the cemetery during excavation. Swedlund and Armelagos (1976:47) note that the validity of developing life tables for archeological populations depends on the degree to which the population is (1) sedentary and (2) large, and (3) provides a large number of recoverable skeletons. When these three conditions prevail, or are at least approximated, life table data provide a significant supplement to analysis of bone disease.

Archeological Documentation

I cannot overstress the importance of good archeological documentation of all skeletal specimens. Resolution of interesting and significant epidemiological problems may hinge on such data. For example, one archeological skeleton from Arkansas in the collection of the National Museum of Natural History, Washington, D.C., USA, probably represents one of the most convincing examples of treponemal infection that exists. This specimen (NMNH 258778) was excavated in the late nineteenth century using methods that make accurate archeological dating impossible without considerable destruction of the specimen (Figures 329–332). This skeleton, if

dated before Columbus, would provide valuable information toward resolving the problem of the origin of syphilis. Thus, the usual elements of archeological provenience, geographical location, archeological horizon or date, and cultural association, are important variables in paleopathology.

In most cases, data obtained during excavation of an archeological specimen will supply detailed information on provenience. Although it is conceivable that the geographical location of a specimen could be lost due to a mistake in museum cataloging, we assume that data or geographical location will almost always be readily available. Very often the archeolgist can determine the date or archeological horizon of the specimen on the basis of associated artifacts. In many areas of the world the evolution of artifacts has been carefully documented and associated with time. These time sequences of artifact series frequently provide the most accurate dating of associated skeletal material. Dates based on association of artifacts may be checked with dates obtained by absolute dating methods such as ^{14}C analysis, where appropriate material such as charcoal exists. Bone itself can be analyzed, but the methods currently available on a routine basis involve the destruction of a considerable amount of bone.

Various methods of dating have been applied directly to bone including ^{14}C, nitrogen, fluorine, and amino acid dating. However, all these methods may produce spurious results because of any one of several variables that can affect the analysis. There are several sources that review the methods of dating and their inherent problems (e.g., Zuener, 1958; Brothwell and Higgs, 1970; Ortner, Von Endt, and Robinson, 1972; Michels, 1973; Hare, 1974; Von Endt, 1979). However, in view of the importance of dating in paleopathology it seems appropriate to provide a brief review of the problems with particular reference to the biology of bone as it affects dating methods.

In ^{14}C analysis of bone, the fundamental problem is bone's low carbon content, which requires the destruction of substantial quantities of bone for obtaining analytical data. Assuming that expendable skeletal material exists for such a purpose, there are many problems encountered in assuring that the ^{14}C was part of the tissue at the time of death and not added later.

Some of the carbon in bone is associated with the mineral phase of bone as carbonate. Hassan and Ortner (1977) have noted that carbonate exchange may occur after death and burial. Furthermore they point out that physical contamination with carbonaceous material also occurs. Thus, the mineral phase of bone is a poor source of ^{14}C because of contamination and recrystallization.

The protein phase in bone also contains carbon, but it is much more stable than bone mineral after death and burial. Thus, bone protein is probably the best source of radioactive carbon. It is, however, essential to remove all contaminants. In the case of bone protein, it is very much simpler to do this than for bone mineral, since bone protein is more highly resistant to demineralizing agents. Such agents would tend to remove soil contaminants and leave behind the insoluble protein containing carbon incorporated in bone tissue during the life of the individual.

In theory, at least, the protein matrix of bone should provide an accurate source for ^{14}C analysis. However, keep in mind that varying soil conditions affect the stability of bone protein. Acid soils will hydrolyze bone protein over the years, destroying the protein, and breaking down the tissue in general. This results in the loss of nitrogen and this loss has been used as an index of time (Oakley, 1950, 1963a, 1963b; Cook, 1960; Doberenz and Matter, 1965). The effect of protein degradation on ^{14}C analysis is unknown but cannot be ignored.

Nitrogen analysis does provide supporting evidence of age. However, the degradation of bone protein, on which this method depends, is affected by several factors including soil pH, temperature, and water (Ortner, Von Endt, and Robinson, 1972), which have not yet been fully clarified.

Another dating method that is based on bone protein utilizes the ratio between natural amino acids (L form) and D amino acids, which gener-

ally do not occur in biological tissue. Through time racemization occurs, in which L amino acids convert to D forms. This process is correlated with time (Hare, 1974) and has the advantage over nitrogen analysis of being affected by fewer variables. Regrettably, as of this writing, much remains to be done before amino acid dating provides a reliable method for dating archeological skeletons.

Gross Study of Skeletal Lesions

With this background on data basic to an analysis of a pathological specimen, we can focus on methods for analyzing specific morbid conditons in a skeleton. Fundamentally, there are four sources of information that may be relevant in paleopathology: (1) gross analysis of the skeleton itself, (2) analysis of X-ray images, (3) chemical analysis of bone, and (4) microscopic examination of bone.

The gross appearance and X-ray images of an abnormal bone represent overlapping features of a disease process, and both should be part of any descriptive analysis of the morbid condition (see p. 45 ff). The ultimate goal of a descriptive analysis is the identification of the disease process. It is important to recognize that the specific cause or etiology of an abnormal bone condition cannot always be derived from even the most careful analysis of the paleopathological specimen. However, even when the specific disease cannot be identified, the description of the abnormal condition may, in itself, provide important data in paleopathological research by providing information on the frequencies of cases in the descriptive categories. I cannot overemphasize the importance of careful descriptive analysis of all paleopathological specimens. This is basic to the identification of disease, but it also represents significant data in and of itself.

In a descriptive system for abnormal bone conditions, there are several essential elements. These include (1) an unambiguous terminology, (2) precise identification of the location and distribution of abnormal bone, and (3) a descriptive summary of the morphology of the abnormal bone. The remainder of this chapter focuses on the descriptive system for paleopathological skeletal specimens.

The most fundamental concept relating to the interpretation of abnormal bone conditions is that the abnormal morphology that results from pathological processes in bone is always the result of disturbances in the osteoblast and osteoclast cell populations (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952:191). Shifts in the relative and absolute activity of these cells may be associated with abnormal activity by the osteocytes, but this is a minor component of any disease process in bone. Not only can there be shifts in the relative and absolute proportions of these cells, but, at least in the case of osteoblasts, the speed or metabolic rate of the cells may change. Thus, it is variation in the cell population balance and in cell activity that accounts for all the variation in the morphology of abnormal bone. In this sense then, one of the basic objectives of a descriptive analysis of abnormal bone is to determine the actions of the cells that produced the abnormal tissue. The importance of this perspective cannot be overemphasized.

Both systemic and local factors can stimulate or inhibit the activity of osteoblasts and osteoclasts. Pathogenic organisms, including fungi, bacteria, viruses, and parasites, act on bone architecture by creating conditions that stimulate or inhibit the bone cells. While it is rather remarkable that the activity of only two types of cell can produce such a large variety of abnormal conditions in bone, it is important to remember that many distinctly different diseases produce very similar types of lesions.

As we begin to think about the description of morbid conditions in the skeleton, it is important to be aware of the fundamental ways bone cells can respond to the stimulus of disease. Schinz, Baensch, Friedl, and Uehlinger (1951–1952:194–220) provide an excellent summary of these responses, and their description has been used in the following review of bone cell reactions to disease. All abnormal conditions of bone can be

placed in one of the following general categories reflecting bone cell activity: (1) abnormal bone loss, (2) abnormal bone gain, (3) a combination of abnormal bone loss and abnormal bone gain, and (4) normal quality of bone but abnormal shape or contour.

The first step in a descriptive analysis of a pathological specimen is to determine whether the abnormality consists of (1) a solitary lesion in which there is a single focus for the disease process, (2) multiple lesions in which there is more than one focus for the disease process, (3) diffuse abnormal bone in which there is no specific focus for the disease but a general change in bone quality, and (4) a local or generalized disturbance of normal size or shape in which the quality of the bone tissue is normal. If the abnormality is a solitary lesion one should identify the bone involved and its position on the bone, and then describe the lesion itself. However, if the abnormality involves one of the other three categories the descriptive process is somewhat more complicated, largely because the distribution of abnormal bone in the skeleton is more complex. It is also important to remember that multiple lesions may not represent the same morbid process.

Regardless of which of the above categories is appropriate for a specific abnormal specimen, the location of the abnormal bone in the skeleton is a crucial descriptive parameter. Various disease processes often affect preferentially groups of bones, specific bones, and specific locations in bones. Indeed, predilection, or lack of it, is an important aspect in itself.

Functionally the skeleton may be divided into two groups of bones. The first group is the axial skeleton and consists of the skull, mandible, vertebral column, ribs, manubrium, and sternum. The second broad group is the appendicular skeleton, which includes the pectoral girdle, the arms and hands, the pelvic girdle, and the legs and feet. Within the bones of the axial and appendicular skeleton specific disease processes vary in their distribution. A descriptive analysis of an abnormal skeleton should be specific regarding the bones involved and the location within the bone. The following outline can serve as a guide to the degree of precision that should be attempted. In the discussion of particular diseases in subsequent chapters we will identify, wherever possible, the sites in the skeleton affected preferentially by a disease.

A. Location of abnormal bone(s) in skeleton
  1. Axial skeleton
    a. Skull
      (1) Vault
      (2) Face
        (a) alveolar
        (b) nonalveolar
      (3) Base (including entire sphenoid and temporal bones)
    b. Mandible
      (1) Body
        (a) alveolar
        (b) nonalveolar
      (2) Ramus
        (a) articular
        (b) nonarticular
    c. Vertebral column (except sacrum and coccyx)
      (1) Cervical
        (a) body
        (b) arch, transverse processes, and spinous process
        (c) joint surfaces
      (2) Thoracic
        (a) body
        (b) arch, transverse processes, and spinous process
        (c) joint surfaces
      (3) Lumbar
        (a) body
        (b) arch, transverse processes, and spinous process
        (c) joint surfaces
    d. Ribs
      (1) Near vertebrae (proximal)
      (2) Near sternum (distal)
      (3) Intermediate
    e. Sternum
      (1) Manubrium
      (2) Corpus
      (3) Xiphoid process
  2. Appendicular skeleton
    a. Pectoral girdle
      (1) Scapula
      (2) Clavicle
    b. Arm and forearm
      (1) Humerus

(2) Radius
(3) Ulna
c. Hand
(1) Carpals
(2) Metacarpals
(3) Phalanges
d. Pelvic girdle
(1) Innominate
(a) Ilium
(b) Ischium
(c) Pubis
(2) Sacrum
(3) Coccyx
e. Thigh and leg
(1) Femur
(2) Patella
(3) Tibia
(4) Fibula
f. Foot
(1) Tarsals
(2) Metatarsals
(3) Phalanges

B. Distribution of abnormal lesions within affected bones
1. Long bones
a. Longitudinal axis
(1) Proximal joint surface
(2) Proximal epiphysis
(3) Proximal apophyses
(4) Proximal metaphysis
(5) Diaphysis
(6) Medullary
(7) Distal metaphysis
(8) Distal epiphysis
(9) Distal apophyses
(10) Distal joint surface
b. Cross-sectional axis
(1) Extracortical
(2) Periosteal (Parosteal)
(3) Cortical
(4) Endosteal
(5) Medullary
2. Skull/Mandible
a. Outer table (Outer cortex)
b. Diplöe (Medullary)
c. Inner table (Inner cortex)

The above outline provides a systematic nomenclature for identifying the location of abnormal bone tissue. With the spatial parameters of the disease clearly identified, the next step is a careful description of the abnormal bone tissue itself. Keep in mind that the objective of any description of the abnormal conditions is to clarify the activity of bone cells with respect to the abnormal morphology. In the chapter on the biology of skeletal tissue (p. 13) I note that bone cell populations can destroy bone or form bone and may do both in fairly close proximity in some disease processes. Let me review the morphological manifestations associated with each of these possibilities.

Abnormal bone loss can be the result of four types of cellular disturbances: (1) an abnormal increase in osteoclast activity with normal osteoblast activity, (2) an abnormal decrease in osteoblast activity with normal osteoclast function, (3) a combination of increased osteoclast activity and decreased osteoblast activity, and (4) normal osteoclast function with normal osteoblast matrix synthesis but an inability to mineralize the matrix. Since unmineralized protein matrix will rarely survive in archeological material the last category will probably appear as increased osteoclast activity.

Abnormal bone loss is seen macroscopically as bone atrophy. Schinz, Baensch, Friedl, and Uehlinger (1951–1952:197–200) divide bone atrophy into two general categories: osteoporosis and osteolysis. Osteoporosis involves a loss of bone in which much of the spongy bone is resorbed, leaving behind a few, sometimes thickened, trabeculae and an increased porosity of compact bone. There may be a thinning of the compact bone as well. Osteolysis is localized bone destruction, which results in a specific defect in the morphology of bone. When osteolysis occurs in a limited area on the outer surface of bone it is called erosion; if erosion is more extensive the term "caries" is more appropriate. Osteolysis within compact bone or marrow may result in a bone cyst or cavity.

While osteoporosis will usually involve more than one bone, osteolysis may be solitary or multiple, and, if multiple, it may be seen in one bone or several. Furthermore, the osteolytic lesions may be discrete or coalescent. At this stage of the discussion it is appropriate to introduce the concept of time in the development of an osteolytic lesion (Figure 28). If the lytic disease process is

FIGURE 28.—Schematic representation of bone tissue responses to osteolytic activity. If the lytic process is slow or stops after an acute phase, the bone tissue will circumscribe the lesion with dense, compact bone as seen in the left example. If the disease process is more active there will be little or no circumscribed, sclerotic reaction, but the boundary of the lytic lesion is sharply defined and there may be slight evidence of some bone formation. This type of lesion is associated with lytic processes having moderate destructive activity and speed as represented by the middle example. If the lytic activity is very rapid there tends to be a gradient of destruction at the margin of the lesion as represented by the right example. In such lesions the destructive boundary is not sharply defined.

rather slow and chronic, there will be a tendency to circumscribe the disease process by surrounding it with newly formed bone, which is frequently normal in mineral density. As the speed of the disease process increases there is a decreased tendency to wall off the disease, resulting in sharply defined borders with very little or no bone circumscription in the lytic area. If the speed is very great, there is a zone of partial lytic activity surrounding the central lytic focus—in other words, a gradient of lytic activity from complete bone destruction to intact bone on the extreme periphery of the disease process.

Bone hypertrophy contrasts with bone atrophy in that the activity of osteoblasts predominates in the reaction to disease. This means that abnormal bone formation and increased bone density are part of the gross and radiographic picture. Bone hypertrophy can occur in any part of the skeleton but the descriptive terminology associated with this process focuses on its cross-sectional distribution (Figure 29). Abnormal bone formation on the outer surfaces of bone is often referred to as periostitis in the American literature. This term means peripheral bone inflammation and as such implies more than is appropriate in the diagnosis of dry bone lesions. Periostosis means periosteal bone formation and precisely describes what is going on without implying the mechanism and is thus a more appropriate term. However, in view of current usage, both terms will be considered as synonyms. Abnormal bone formation can also occur on the endosteal, medullary surface of compact or cortical bone and is known as endostosis. Bone hypertrophy of cancellous bone is called spongiosclerosis.

The morphological variants of bone hypertrophy are greater than for bone atrophy but are basically the result of an imbalance in the activity of osteoblasts and osteoclasts in which three relationships exist: (1) increased bone formation with increased bone resorption (2) increased bone formation with normal bone resorption, and (3) normal bone formation with decreased bone resorption.

Periostosis is expressed in many ways and is frequently associated with changes in the endosteal and cancellous parts of bone (Figure 30). One of the most common examples of periostosis in archeological skeletons is fusiform bone hypertrophy on the diaphysis of long bones. This lesion involves an increase in the diameter of the cortex which may result from several abnormal conditions. In the paleopathological literature fusiform

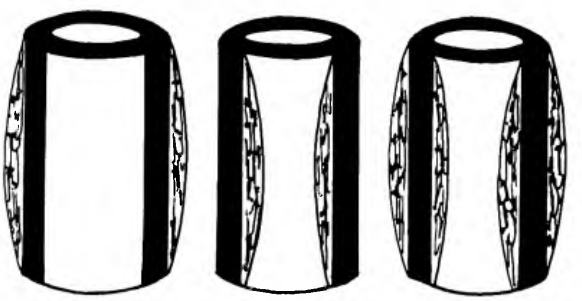


FIGURE 29.—Schematic representation of the basic types of abnormal bone formation. In the example to the left abnormal bone is added to the periosteal surface of bone. In the middle example, abnormal bone is added to the endosteal surface. The third possibility is depicted in the right example, in which abnormal bone is added to both periosteal and endosteal surfaces.

FIGURE 30.—Diaphysial, periosteal, and endosteal bone hypertrophy. Note the lamellar nature of the compact bone on the cut surface. Specimen of a left femur of a child about 12 years of age. The lesion is attributed to syphilis. (12-year-old child, ANM 2999.)

periostosis has been attributed most often to infectious diseases, such as osteomyelitis. Schinz, Baensch, Friedl, and Uehlinger (1952–1952:201) indicate that inflammatory reaction to a pus-forming disease organism is a common cause of periostitis, but that trauma can result in periostosis. These fusiform lesions may be a solid mass of new bone or they may be layers of dense bone with intermediate, less dense or spongy layers in between, creating an onion skin effect in cross- or longitudinal section.

While fusiform bone hypertrophy of a localized variety is very common in skeletal material, more extensive hypertrophy of diaphyseal bone is also often seen. These more extensive lesions frequently appear to be a more severe manifestation of the disease conditions that produce fusiform lesions. However, infectious disease rather than trauma is the most likely factor. Infectious disease may result from local conditions, such as an infected cut, but may be disseminated by the blood from a focus elsewhere in the body. The more extensive, diaphyseal, hypertrophic lesions of bone may also have the lamellar appearance seen in some fusiform bone lesions.

The periosteal lesions described thus far are generally the reaction to low grade or chronic disease conditions in bone. Other morbid conditions can result in more exuberant development of abnormal bone. Neoplasms, both those that arise primarily in bone and those that metastasize to bone, may result in the most dramatic periostosis. Within neoplastic conditions of bone there is considerable morphological variation depending on the type of tumor.

Some primary neoplasms of bone, that is those which arise in skeletal tissue, produce dramatic, exuberant growths of bone. The lesions tend to fall into two categories. The first of these lesions consists of spicules or plates of bone arranged perpendicular to the surface of the bone (Figure 31). This arrangement creates a sunburst effect. These rays may fill in, ultimately creating a solid mass of bone, but the ray-like development of the periostosis is still apparent. The second type of lesion has the appearance of a head of cauliflower (Figure 32) and may reflect a more slowly developing course of the disease.

The secondary neoplasms of bone, by which one means those resulting from metastasis from other parts of the body, may resemble the ray-like conditions of some primary neoplasms but can also result in a fairly low-grade diffuse periostosis similar to that seen in infectious diseases.

Endostosis results in partial to complete occlusion of the marrow space in some cases, with little or no change in the existing cortex (Figure 33). This type of bone hypertrophy illustrates an imbalance between osteoblasts and osteoclasts that results in increased bone. One of the diseases discussed in skeletal dysplasias (p. 340) is osteopetrosis (Albers-Schönberg's disease). This disease is the result of normal osteoblast activity and greatly depressed, or nonexistant osteoclast activity. Since osteoclasts do not enlarge the medullary

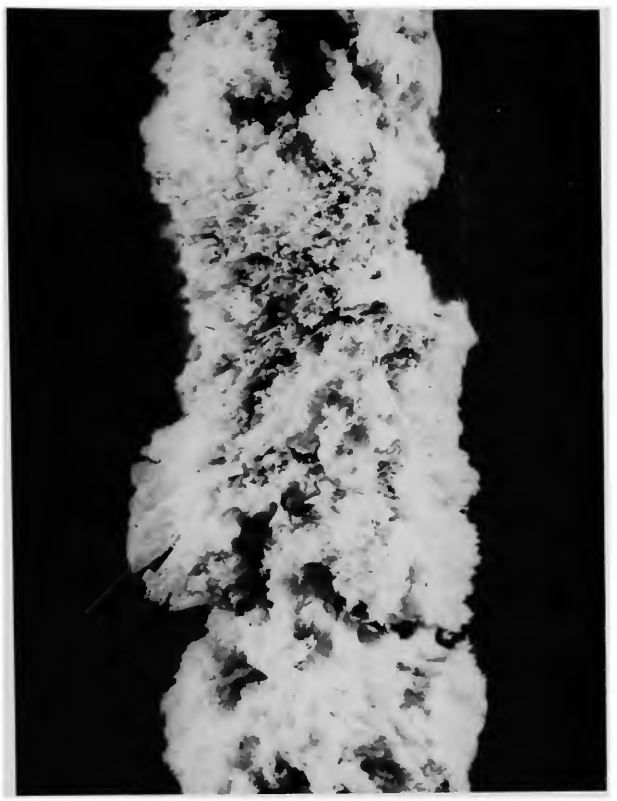


FIGURE 31.—Bone hypertrophy producing spicules or plates of bone that are perpendicular to the bone surface. Specimen is the right femur of a child about 7 years of age. Osteosarcoma, Ewing's sarcoma, and metastatic neuroblastoma are possible morbid conditions stimulating this bony reaction. (7-year-old child, WM S72 a.3.)

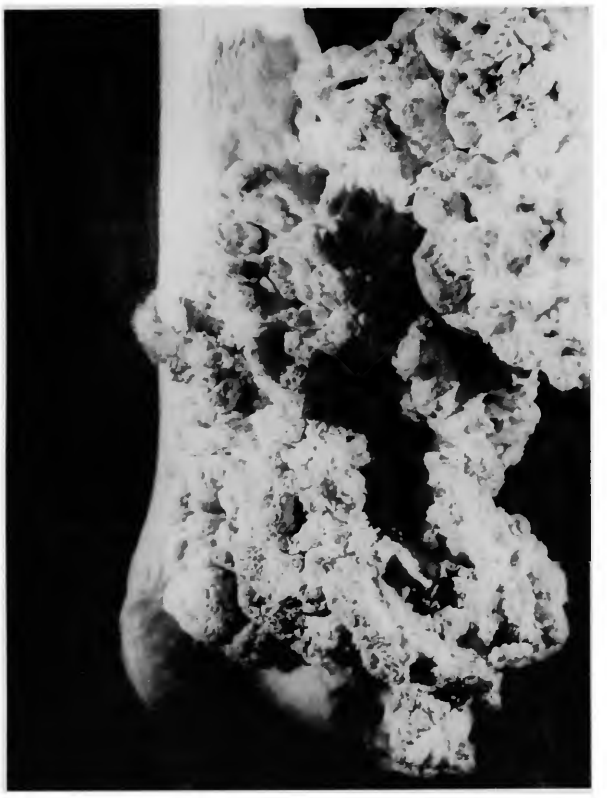
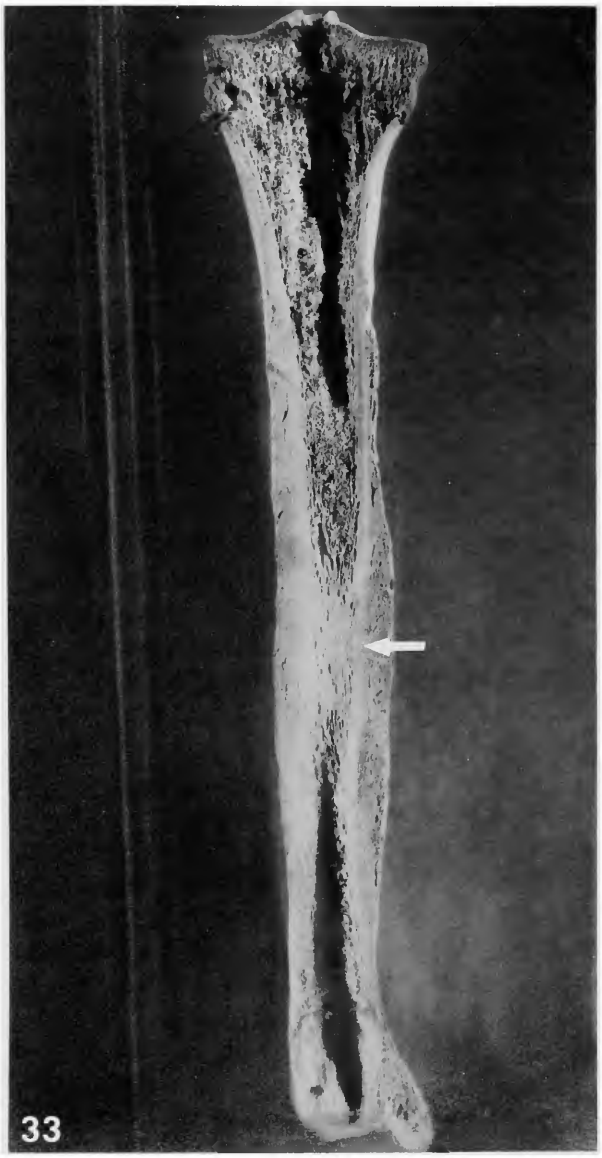


FIGURE 32.—Bone hypertrophy in which the new bone has the appearance of cauliflower. The lesion is attributed to osteosarcoma, but periosteal chondrosarcoma should also be considered. (Distal right femur, 31-year-old male, WM S72 a.2.)

cavity as the bone grows, the marrow space is filled with bone. Here we have endostosis in the sense that bone remains where it does not belong but there is no abnormal bone formation, only the retention of bone tissue that should have been removed. Both endostosis and periostosis may occur as responses to the same disease process (Figure 34). Osteomyelitis, in which the original focus of the disease is in the marrow, can be limited to the marrow and produce only endostosis or spongiosclerosis; more often, however, it also involves the outer surface of bone and stimulates periostosis.

Abnormal bone formation can occur in response to an infectious disease process, such as osteomyelitis, in which newly formed bone tends to wall off the infectious focus. Neoplasms may stimulate spongiosclerosis and one can find this condition associated with other types of disease, such as osteopetrosis mentioned above. Radiologically, a bone with spongiosclerosis in its pure form will show relatively normal outer contours but increased radiodensity.

Thus far in the descriptive review of bone lesions I have indicated that bone disease can be characterized as a process of abnormal bone destruction, bone formation, or both. I have further characterized bone disease by referring to the cross-sectional manifestations of disease. It is important to know that most bone disease involves several, if not all, of these descriptive categories. In osteomyelitis, for example, we may have general thickening of the compact bone and cortex



FIGURES 33, 34.—Endostosis and periostosis of the midshaft of a left tibia: 33, Internal view. The arrow indicates the original cortex. 34, External view of periostosis in the diaphysis; note the porous nature of the thickened diaphysis. (PMWH W0721.)

(bone hypertrophy) but with the compact bone becoming quite porous (bone atrophy) and have periostosis, endostosis, and spongiosclerosis in the same bone. Here, I have introduced one, new, descriptive element and that is the quality of the compact bone.

Changes in periosteal, endosteal, or cancellous areas of bone are often associated with changes in the compact bone. In general, more bone tissue cannot be incorporated within existing compact bone since the only place for such addition would be vascular spaces. The increase in radiologic compact bone density is a function of new bone added to existing surfaces. Thus, the only abnormal development that can occur within previously normal, compact bone is rarefaction or

osteoporosis. This can be accomplished, as in hyperparathyroidism, by an increase in bone resorption by osteoclasts, in which the osteoblast cell population cannot keep up with abnormal resorption or, as in osteomalacia, the protein matrix produced during osteon remodeling is not mineralized.

In Figure 33 we have seen that rarefaction is associated with the thickened cortex in one case of purported syphilis. Similar thickening of the cortex and increased porosity of the original cortex are associated with other disease conditions, such as Paget's disease. The point to be made is that the addition of new bone to existing compact bone is often associated with osteoporosis of the original compact bone.

The mixed type of bone lesion involving both bone hypertrophy and bone atrophy can be found anywhere in the skeleton. A relatively common expression of this type of lesion is found on the skull vault and has been variously referred to as porotic hyperostosis, spongy hyperostosis, cribra cranii, and symmetrical osteoporosis. There are many manifestations of this general lesion and it provides a useful example of the complexity and pitfalls in the descriptive analysis of a bone lesion. For this reason I will review the morphological variants here although they will receive a more detailed treatment in the chapters on infectious diseases (p. 104), reticuloendothelial and hemopoietic disorders (p. 248), and metabolic disorders (p. 270). Porotic hyperostosis basically is characterized by bone hypertrophy of the skull vault. In some cases the abnormal bone is not found in the areas of major muscle attachment. In other cases this limitation does not prevail. Grossly the lesion results in thickened bones of the vault, but the added bone is not smooth (Figure 35). Its external gross appearance is usually porous but may also consist of labyrinth-like plates of bone. The porous nature of the abnormal bone varies from fine, pumice-like pores to large pores in excess of 2 millimeters in diameter. A single lesion may contain all variations of porosity.

The X-ray film appearance of porotic hyperostosis frequently suggests a ray-like arrangement

FIGURE 35.—Porous hypertrophy of the right parietal bone in a 1-year-old girl. Arrows indicate the thickness of the porous hypertrophic bone. Note that orientation of the bone tends to be perpendicular to the surface of the skull. The outer table is almost completely remodeled away. (FPAM 3874.)

of the abnormal bone, which has been called the hair-on-end or hair brush appearance. Often, the original outer table of bone has been destroyed so that the bony rays are continuous with the diploë.

Porotic hyperostosis also varies in its cross-sectional morphology. As suggested by the X-ray film appearance, the disease process may destroy the outer table creating the appearance of a greatly thickened diploë. However, in some examples of gross porotic hyperostosis the cross-sectional morphology indicates that the lesion is entirely superficial to the outer table and thus is a periostosis. In such a case there is no significant change in the diploic space or the outer table. It seems likely that superficial, porous periostosis of the skull vault represents a disease process quite distinct from porous bone hypertrophy associated with enlargement of the diploë and destruction of the outer table. The above review illustrates the importance of careful descriptive analysis of gross lesions, which must include gross and X-ray film morphology, but also the cross-sectional ap-

pearance if this cannot be determined by the X-ray film. The attempt should always be made to determine if the bone abnormality is the result of abnormal osteoblast or osteoclast activity or an abnormal combination of both. It is also important to indicate whether the disease process is accomodating some other body function, as is true in some cases where the development of abnormal bone tends not to affect areas of muscle attachment.

In archeological skeletal material one is also confronted with abnormal-appearing bone tissue that is not the result of antemortem biological processes. Such conditions have been called pseudopathologies and considerable experience with pathological conditions may be necessary to make a distinction. However, an understanding of general principles of bone biology will help to resolve many such problems.

Pseudopathologies are the result of postmortem destruction of bone tissue and result from two basic conditons: (1) the immediate burial environment and (2) careless excavation techniques. While the identification of pseudopathology as pathology is to be guarded against, it is important to remember that many pathological conditions will result in weakened bone tissue that may deteriorate more rapidly after burial than adjacent normal bone. Thus, destruction of bone tissue by components of the soil does not always mean that such abnormal bones represent only pseudopathologies.

The preservation of bone tissue is generally a function of soil pH, temperature, and ground water volume. However, local conditions immediately adjacent to a specific bone may produce marked changes in bone preservation. Copper containing ornaments buried with the individual will greatly enhance the preservation of bones located next to the ornament. In contrast, bone resting on wood, as in a wooden coffin, deteriorates more rapidly than parts of the skeleton not in direct contact with wood. Earlier I have noted changes at the microscopic level, produced by fungi, which could be confused with osteoclastic activity.

Damage by careless excavation can easily be confused with antemortem trauma. Indeed, the distinction between injury immediately before death and that occurring after death, including that occurring at the time of excavation, cannot always be made. In general, trauma will result in inflammation, within a few days, followed by bone remodeling, which is frequently apparent in archeological specimens. Depressed fractures occurring during life will generally have less sharply defined borders than those resulting from postmortem damage. There may be bone fragments still attached at the edge of the defect. This is almost certainly indicative of an antemortem lesion.

Careless handling of a shovel can remove a slab of bone from a skull, the cut surface of which will resemble a sword wound. However, with sufficient experience with pathological specimens and the knowledge of how bone reacts to disease, the paleopathologist should be better able to identify pseudopathology in specimens.

Assuming that paleopathological conditions can be described and classified in most cases, the information from such classifications needs to be integrated with the broader biological framework. In this context human group dynamics become the focus rather than individual cases of disease. Having determined the frequency of different types of disease in a group, such frequencies can be compared between groups. Inferences about demographic or environmental effects may be possible, as in Angel's research on malaria in the eastern Mediterranean areas (Angel, 1964, 1966).

In the broader paleopathological context of group responses to disease, we need to be careful in making inferences about the relative health of a population on the basis of the incidence of skeletal disease. Skeletal manifestations of morbid conditions tend to be associated with chronic responses to disease. Skeletal paleopathology, particularly infections, tend to be viewed as evidence of high morbidity and, indeed, this may be the case. The converse, however, is also possible. Skeletal paleopathology may indicate a better host

response to a disease since the host lived through the acute stage when other afflicted individuals may have died. Thus, absence of skeletal disease implies death due to acute conditions, while evidence of skeletal disease indicates a sufficiently adequate immune response to insure survival to the chronic stage.

A final cautionary note is that skeletal diseases, particularly those caused by infectious agents, may not be the same today as they were in ancient times. Selection and adaptation undoubtedly improve the immune response to disease. At the same time infectious agents tend to become less virulent since the organism typically dies with the host making reduced virulence selectively advantageous. This means, of course, that an increase in the frequency of skeletal manifestations of disease after mutation of an existing disease organism or the introduction of a new infectious disease may mean only that the disease is becoming more chronic in nature and thus, is more likely to affect the skeleton.

X-ray Film Study of Skeletal Lesions

Analysis of X-ray films of gross pathological bone specimens is one of the most important tools for the paleopathologist and should be part of almost all descriptions of such material. The preparation of X-ray films has the advantage over many other methods in being completely nondestructive and thus, should always be used before chemical or histological methods are applied. In view of the importance of X-ray films in paleopathological studies I will review, in considerable detail, the methods involved in producing good X-ray films as well as the principles in interpreting such films after they are prepared.

The correct exposure of an X-ray film depends on several factors including (1) electrical current flow measured in milliamperes, (2) energy of current measured in kilovolts, (3) time of exposure usually measured in seconds, (4) the distance between the source of X-rays and the film, (5) the speed of the film emulsion which may be enhanced by the use of phosphorescent screens, and (6) the density of the specimen. In actual practice most of the above variables are standardized, making the preparation of most X-ray films routine. Current, energy, and time are variables that are limited by the X-ray machine. In general, varying either current or time will produce the same result. Thus, if, as is often the case, the current is fixed on a particular machine, time is adjusted to meet the requirements of different exposures. The energy of the X-rays affects their potential to penetrate dense materials. The greater the energy the greater the penetrating potential.

One tries to select a voltage that will penetrate the thickest or densest part of the specimen enough to expose the film somewhat, but not so high that it is not partially absorbed by the thinner or less dense areas of a specimen. For most archeological specimens the energy used to generate X-rays is between 60 and 80 kilovolts. However, if the specimen is fossilized, higher kilovoltages may be necessary. For delicate or small bones (e.g., hands, feet, ribs, or fetal bone) lower voltages may be needed.

Radiology shares with photography many of the problems inherent in projecting the image of a three dimensional object onto a two dimensional plane. Like photography, it is possible to minimize the effect of inevitable distortion by keeping in mind certain principles. The first of these is primarily inherent in the X-ray equipment and is a function of the area of the target which produces X-rays (the focal spot). X-rays are generated from the entire surface of the focal spot and radiate out in a cone from each discrete point in the spot. For illustrative purposes I have taken the extreme edge of the focal spot, which, in effect, produces overlapping but slightly different images and results in reduced sharpness of the X-ray image (Figure 36). In selecting X-ray equipment it is important to obtain one with a small focal spot. Most manufacturers produce equipment with a focal spot of 0.5 millimeters or smaller.

The effect of distortion produced by the geometry of the focal spot can be minimized by

increasing the distance between the focal spot and the film plane. This also minimizes parallax distortion, which results when three dimensional objects are projected onto a plane (Figure 37). However, there are practical limits on the distance between the focal spot and film plane. X-rays, like light energy, diverge from the source so that the same number of X-rays are spread over a larger area as the distance increases. This principle is known as the inverse square law and means that one must increase the number of X-rays (by increasing time and/or current) if one increases the distance. In my experience an optimal distance from the focal spot to the film plane is between 3 and 4 feet (91–122 cm). However, for critical work on bones such as the skull, where parallax is a greater problem than in long bones, this distance may need to be increased.

The orientation of the specimen with respect

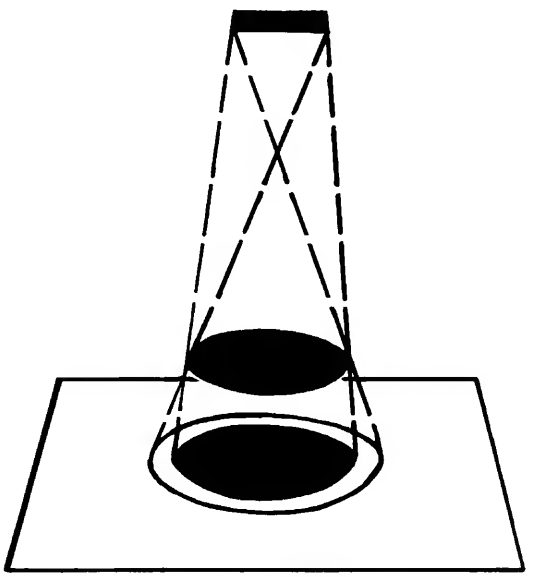


FIGURE 36.—Effect of focal spot size on X-ray image sharpness. X-rays are generated on the surface of a focal spot, represented by the solid bar at the top of the diagram. X-rays emanating from two different points on the focal spot (dashed lines) intercept the object, represented by the black disk, at different angles and project the images of the disk to overlapping but different areas of the film plane. The white area within the circle represents the area of reduced image sharpness resulting from the geometry of X-rays generated from different areas of the focal spot.

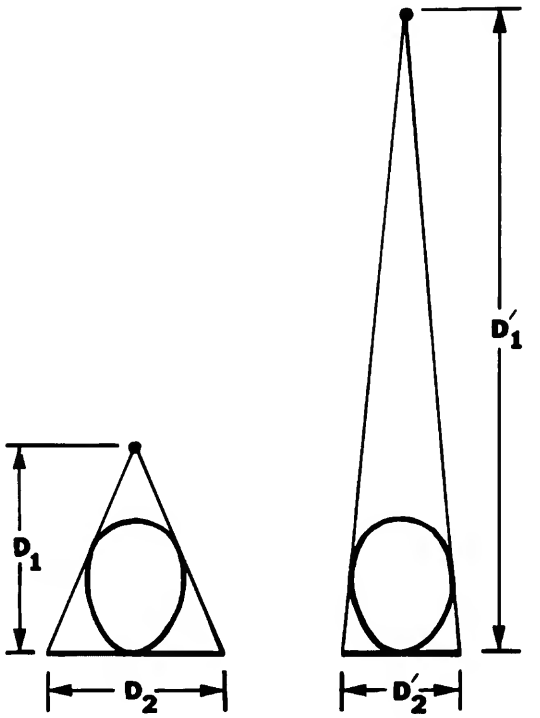


FIGURE 37.—Variation in parallax distortion (D_2) of X-ray images due to differences in focal spot to film plane distances (D_1).

to the X-ray source and the film plane is also a significant factor in image quality and accuracy. As a general rule one should be consistent in the placement of the specimen with regard to the source and film plane. Most often the film plane is perpendicular to the central axis of the X-rays. In routine roentgen procedures the specimen should be positioned to have the same anatomical relationship to the central axis. For example, in a lateral view of the skull one might wish to position the skull with external auditory meatus corresponding to the central axis of the X-rays; for long bones the midshaft might be appropriate.

In producing an X-ray image of a specific lesion, we are generally less concerned in producing a relatively undistorted image of the lesion than in producing an image that helps us to characterize the pathology of the lesion. Thus, we may want to know if the marrow has been encroached upon or whether a lytic process has been

circumscribed by dense bone. In these and other related situations one positions the specimen so that these factors can best be visualized. In general one should have lateral and front views of a lesion on diseased bone, so that can at least partially reconstruct the three dimensional appearance of the lesion. It is important to try to avoid the superimposition of other parts of the bone on the diseased part if possible. In some bones, such as the skull, this may be very difficult, but an effort should be made to minimize the effect of superimposition. This can be done by careful positioning.

There are several advantages in producing X-ray films of archeological specimens in contrast with those of a living patient. Obviously, we are not concerned with radiation hazard to a skeleton, so long exposures and film speeds are not a critical consideration. Furthermore, if carefully positioned and supported, the bone specimen will not move. In addition, there are no soft tissue features to interfere with the image of the skeletal tissue. So many of the variables of concern to the clinical radiologist need not concern the paleopathologist.

These advantages need to be considered in selecting a film. Film speed is, in general, inversely related to film resolution. Thus, the faster the film the poorer the quality of the image. Since high film speed is not an advantage with paleopathological specimens, image quality and convenience of processing become important factors in selecting a film. In my experience I have found Kodak Royal Blue Medical and Kodak X-Omat G film to be satisfactory. I have evaluated some slower, high resolution industrial films, such as Kodak Industrex M film, and found them to produce excellent image quality. These should be considered in situations where fine detail is needed to bring out all the radiographic features of a lesion.

X-ray film is available in many sizes. Many of these sizes are appropriate to specific applications in modern clinical radiology and only two are needed for general radiography in paleopathology. The most useful size for skulls is 24 × 30 centimeters. In my experience, this size will accommodate the largest skulls, including the mandible, where this is desired. For the bones of the postcranial skeleton, film 35 × 43 centimeters is the largest standard size available from Kodak and is satisfactory for most applications. This size film is frequently not long enough to contain an entire femur on its long axis. In such a situation the diagonal position may be adequate. However, there will be femora for which even this orientation will be inadequate. In such situations one eliminates the least important end or exposes two films. Longer films are available commercially but create problems in processing and storage.

In choosing a film one needs to keep in mind the method which will be used for processing. In most clinical laboratories, X-ray films are developed in high speed processors, which produce a dry film in one to two minutes. Special films have been created to accommodate this rapid processing. Such equipment has high initial and maintenance costs and will generally not be appropriate for the intermittent use needed by most paleopathologists. It may be possible to have film processed by a clinical facility, but frequently the more conventional and slower manual processing procedures will be used. Film not adapted for high speed processing must be processed by hand. However, the Kodak X-Omat G film can be processed manually or in a high speed processor. A decision on the method for processing should be made before X-ray film is purchased.

While film can be purchased in a light proof envelope, it is considerably more expensive than film packed for loading in a film holder or cassette. For most applications in paleopathology a simple cardboard film holder, such as the Kodak X-ray Exposure Holder, is adequate. These holders are reasonable in cost. The purchase of several for each of the film sizes to be used will be a convenience. Film cassettes are much more expensive but last longer. Recent developments in cassette design, such as the Kodak X-Omatic cassette make loading very simple and offer the option of using phosphorescent screens to shorten exposure times. These screens are placed in close

contact with the X-ray film and are excited when exposed to X-rays. Those areas of screen that are excited by X-rays give off light (fluoresce), and since the film is sensitive to both X-rays and light the effect of the screen is to enhance the effect of the X-rays and decrease the exposure time. While shortened exposure times are not a crucial factor in paleopathology, they do have two advantages: (1) they lessen the effect of vibration on the X-ray image, and (2) they lessen the risk of exposure to X-rays for persons exposing the X-ray film. Recently, I have begun the routine use of the Kodak X-Omatic cassette with fine, intensifying screens. There is a slight loss of resolution when these screens are used. However, the contrast may be enhanced and the overall X-ray film appearance of a lesion may be as good, or better, than the same film exposed without a screen.

Basically, two types of situations exist in the radiology of paleopathological specimens. In the first of these we may have some external evidence of disease and want to clarify the nature of the lesion or see if there is any additional evidence of disease not apparent externally. To accomplish the latter objective we will need X-ray films of the bones of the entire skeleton. A second objective of radiology in paleopathology would be to conduct a general survey of nonspecific bone responses to disease (such as Harris' lines) in an archeological skeletal sample. In both these applications it is important to prepare a standard set of films for the entire skeleton with views that can be compared with X-ray films of normal skeletons.

In doing this, attention to a few details will make comparison and interpretation much simpler. It is very important to include some type of identification number (museum catalog number or field number) on the film and to indicate from which side of the skeleton the bones are taken. Lead letters and numbers are available from distributors of X-ray supplies and should be part of an X-ray facility. If they are not available, process each film separately and write the information on the film when processed and dry.

Common sense dictates that the orientation of the bones conform as much as possible to the anatomical position and relationships. It is frequently possible to group the bones of the arm and entire pectoral girdle (scapula and clavicle) on a single 35 × 43 centimeter film. The proximal end of the long bones should be positioned at the top of the film, bones from the right grouped together and, likewise, for bones from the left side.

The vertebrae pose special problems for the paleoradiologist because of their irregular shape. One solution is to obtain some soft, flexible, polyurethane foam and cut it into strips about 2 × 2 × 35 centimeters long. After assembling the vertebrae in their correct anatomical relationship the foam can be threaded through the neural canal of the individual vertebrae. The vertebrae can be adjusted by sliding or twisting on the foam to achieve correct anatomical relationship. The foam will hold the vertebrae in the correct relationship and will have negligible effect on the X-ray image. Since a complete vertebral column is usually too long for a single 35 × 43 centimeter film, I divide the vertebrae into anatomical groups: cervical, thoracic, and lumbar vertebrae.

In the standard radiography of all bones it is important to orient the bones in the anteroposterior (A-P) axis or mediolateral (M-L) axis. To facilitate this, it is very helpful to have several wedge-shaped pieces of polyurethane foam. For most purposes these need not be large, the longest edge being about 5–8 centimeters long. A few pieces two or three times this size are useful for skull radiography.

The skull poses the most difficult problems in the radiography of skeletal material. Because of the globular nature of the bone, it is impossible to avoid superimposition of images of one part of the skull on other parts. For this reason the positioning of the skull is more critical than that of other bones and should be done carefully. In a complete skull, the most useful landmarks for orienting the skull are the mastoid processes. In a lateral view the tips of the mastoid processes should be perpendicular to the film plane (Figure 38). In an anteroposterior view the tips of the mastoid processes should be parallel to the film plane (Figure 39). The skull should be rotated on

an axis through the mastoid processes to minimize the superimposition of the occipital bone on the maxillary bones and teeth.

The second type of problem in the radiography of paleopathological specimens is preparing films of specific lesions in order to get a better understanding of the gross lesion and to determine to what extent tissue that cannot be seen externally is involved in the disease process (e.g., medullary involvement in a periosteal lesion). In many cases the standard orientation, anteroposterior (A-P) and mediolateral (M-L) of the specimen will provide the information needed. However, with some lesions better results are obtained if the orientation point is the lesion itself rather than the whole bone. In such situations A-P and M-L views of the lesion, irrespective of the orientation of the rest of the bone, should be obtained. Generally, the lesion should be as close to the film as possible. For example, if a lesion is on the right parietal of the skull, this side should be placed on the film cassette or holder.

In general radiography of a specific lesion one attempts to demonstrate two features: (1) involvement of bone tissue in areas that cannot be observed externally and (2) the pattern of bone density in the lesion and the area immediately surrounding the lesion. For illustrative purposes let me review the features of an expansive, diaphyseal lesion of a major long bone. The gross appearance of such a bone indicates that the diaphysis is abnormally thick. The bone tissue itself may be somewhat porous and have large openings or cloacae. We are unable, however, to determine from external inspection if the marrow space is normal, enlarged, or smaller than normal, if there are unusual features within the cortex, or if the abnormal tissue was laid down evenly or in alternating high and low density layers.

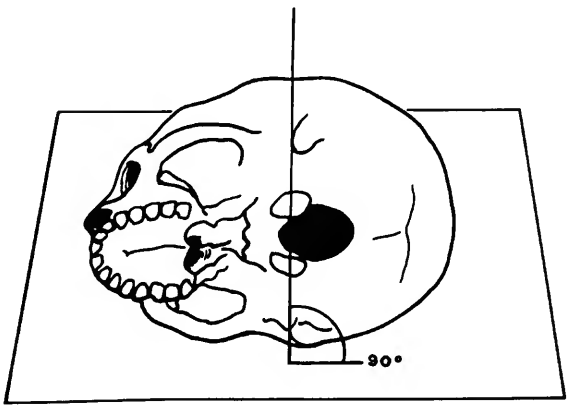


FIGURE 38.—Orientation of the skull relative to the film plane. The axis through the mastoid processes should be perpendicular to both the *x* and *y* axes of the film plane when preparing a standard lateral X-ray film of the skull.

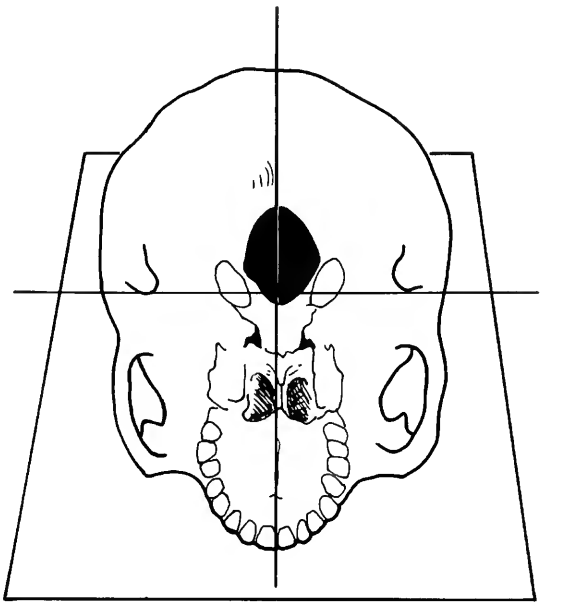


FIGURE 39.—Orientation of the skull relative to the film plane in an anteroposterior view. The axis through the mastoid processes should be parallel to the *x* axis of the film.

An X-ray film, in which the axis of the X-rays is perpendicular to the plane of the lesion, may reveal increased bone density due to increased thickness but will not demonstrate marrow space encroachment or any lamellar arrangement of the abnormal tissue. Such information requires that the axis of the X-rays be parallel to the plane of the lesion. While the possible combinations of density patterns are almost infinitely large, common sense application of the above principle will permit optimal radiographic resolution of most abnormal conditions in skeletal tissue. A more comprehensive review of radiographic techniques is found in most radiology reference sources. The

first English edition of *Roentgen-Diagnostics* (Schinz, Baensch, Friedl, and Uehlinger 1951–1952:3–72) and the first chapter in *Anatomy Basic to Radiology* (Meschan, 1975:1–23) are excellent examples of such sources.

After exposure of the X-ray film, it must be processed to make the image visible. The principles involved are very similar to processing photographic film and involve immersing the film in (1) a developer solution that darkens the exposed silver halide crystals on the film and (2) a fixing solution that removes the unexposed crystals (Figure 40). The film must be washed to remove the processing solutions and then dried before viewing and storage. A detailed review of these procedures is available from X-ray film suppliers (e.g., Eastman Kodak publication, *Radiography in Modern Industry*, 1969).

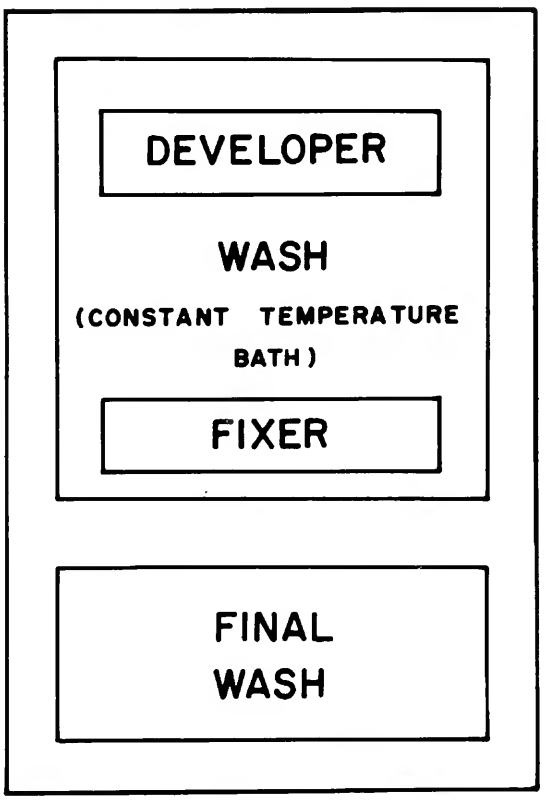


FIGURE 40.—The processing sequence in the preparation of X-ray films. The first solution is developer, which darkens the silver halide crystals in the exposed portions of the film emulsion. The developed film is washed for about two minutes before immersion in the fixer solution, which removes the unexposed silver halide crystals. A final wash removes all the processing solutions.

In my brief discussion of film selection I have already noted that a decision regarding automatic versus hand processing needs to be made before the purchase of film. Using the automatic processing facility of a local clinical X-ray laboratory is a great convenience and saves a considerable amount of time. For most paleopathologists the number of X-ray films processed simply does not justify the establishment of a special laboratory, and arrangements to take and process films should be made with a clinical facility. Often this will be done as a professional courtesy, but even if charges are made for this service it is probably very much cheaper than establishing one's own laboratory.

In the event that the volume of X-ray film preparation justifies the establishment of an X-ray laboratory the major expenses will be (1) the construction of a special room to protect the technical staff from radiation during exposure, (2) the purchase of an X-ray machine, (3) the construction of a dark room, and (4) the purchase of film processing tanks. Many educational or research institutions have regulations regarding the safety of such a facility, and the official responsible for implementing these regulations should be consulted in the planning stage. The operator of X-ray generating equipment must minimize personal exposure to X-rays and insure that others in the vicinity are protected as well.

Given the existence of an adequate facility for preparing X-ray films, one should consult standard sources for the specific details on taking and processing such films. I have already indicated the approximate parameters for exposing X-ray film. Hand processing the exposed film involves attention to the processing specifications that are appropriate for a given film. The most critical stage in processing is development, where time and temperature are the most important factors. In general, the temperature of the developer should be between 60° and 75°F. Exceeding this

temperature range on either end will result in poor image quality and should be avoided. In view of the importance of temperature the processing tanks should be in a water bath in which the temperature can be controlled. Cold tap water can be mixed with hot water to produce the optimum temperature by using a temperature control valve. This valve is generally part of a commercial hand processing tank system. Unfortunately, tap water temperature during the summer months may exceed the upper limit for development. If you anticipate processing film during such periods you will need to install a water cooling system as part of a processing system. Such equipment should be compatible with the water flow through the tank and can generally be purchased as part of the system. The temperature of the fixative is less critical, but since both developing and fixing tanks are usually in the same water bath, correct temperature for the developer automatically results in appropriate temperature for the fixer. Films such as Kodak X-Omat G are adapted for high temperature automatic processing and have greater temperature latitude when hand processed.

In hand processing, the exposed film is attached to special, stainless steel wire, developing frames and immersed in the developing tank. A typical developing tank contains 5 gallons of reagent and can develop 5 X-ray films at one time. Practical experience has shown that the risk of scratching the film surface greatly increases if this number is exceeded. It is also important not to process different sizes of film at the same time. The almost inevitable result is scratches on some of the film surfaces. After development for the appropriate time (usually about 5 minutes) the film is washed for about 2 minutes. The film is then immersed in the fixer solution for about 5 minutes, then washed for 30 to 45 minutes in running water and hung up to dry. Most X-ray films are made to be insensitive to a fairly bright amber light so that the work in the dark room is done in relatively light conditions. However, safelight specifications should be followed carefully,

Once the film is dry, it should be stored in standard X-ray envelopes, available through any X-ray supply dealer. These envelopes have special areas for recording the specimen number and additional information that cannot be put on the film itself. X-ray films are much more useful when this information is part of the record. Considerable care should be taken in handling the dry X-ray film. The image is part of a thin emulsion and is easily scratched. As with any photographic film, X-ray film should be handled by the edges both before and after processing. As the number of X-ray films accumulate, a wise investment is a film storage file that will keep the films in order and make them more accessible.

Chemical Analysis of Pathological Bone

In modern medical practice, tests of biological functions and products have become a routine part of diagnostic procedures. However, most of these tests utilize materials, such as blood or urine, which are not available in archeological material. Diseases that affect protein synthesis, such as osteogenesis imperfecta, may result in abnormal bone proteins. Eastoe, Martens, and Thomas (1973) have determined the amino acids in biopsy specimens from recent cases of this disease and found a relative increase in the neutral amino acids compared to normal bone protein. In research conducted in my laboratory (unpublished), I have found abnormal amino acid residues in a rare bone disease, known as fibrogenesis imperfecta ossium.

In archeological skeletons the application of chemistry in the diagnosis of disease is very limited but should not be overlooked where appropriate. Such analyses will be limited to situations where bone preservation is very good, since degradation of bone protein by the soil introduces an uncontrolled variable making interpretation of data difficult, if not impossible. Von Endt and Ortner (1977) have applied amino acid analysis of bone to a suspected case of iron deficiency anemia in a prehistoric child's skeleton from the American Southwest. In this study, samples from

two prehistoric skeletons were analyzed. One specimen (NMNH 308697) had no evidence of disease affecting the skeleton. The other specimen (NMNH 327107) had marked, gross, porotic hyperostosis of the skull and in the postcranial skeleton, thinned compact bone and expanded marrow spaces. This combination of lesions is strongly suggestive of anemia. The amino acid residues from the abnormal skeleton were different from the normal skeleton and from bone of a modern child. These results are discussed in greater detail in the chapter on reticuloendothelial and hemopoietic disorders (p. 248) and suggest the potential for the study of protein chemistry in archeological specimens.

The mineral phase of bone may also be affected by disease. In rickets, the protein matrix does not mineralize. In fluorosis, abnormally high levels of fluorine can result in gross and microscopically abnormal bone. It may also increase the concentration of fluorine in bone mineral. I have used the electron probe microanalyzer to investigate the toxic effects of fluorine on bone (unpublished data). This research demonstrated almost twice the normal concentration of fluorine in affected bone mineral. Analysis of strontium levels in bone have been used in dietary studies of archeological specimens (Schoeninger, 1979). Strontium levels in bone are lower with greater emphasis on meat in the diet and higher if plant sources account for more of the caloric intake.

While chemical analysis of archeological skeletons may provide valuable data on a disease condition, considerable caution is needed in interpreting such an analysis. The process involves at least a small amount of destruction of valuable bone. Furthermore, one must insure that data from chemical analysis reflect conditions during the life of the individual and not one or more variable changes of bone in the soil. Data obtained by comparison of normal and abnormal tissue from the same site may not be reliable, since soil conditions may vary within a cemetery site. Indeed, a complete understanding of the chemical composition of normal and abnormal modern bone tissue remains an unachieved goal.

Microscopic Study of Skeletal Lesions

Microscopic study of bone, like chemical analysis, is limited in its application. As a general rule, in archeological skeletal material, microscopic data add little to what can be seen grossly or on X-ray films. Because microscopic examination does involve destruction of some of the bone, it should only be applied when diagnosis is likely to be aided. It should not be considered a routine procedure.

If, after careful analysis of gross and X-ray film data, it seems likely that microscopic data would be valuable, several factors need to be considered. The first step in any analysis is the removal of a sample of the abnormal bone. Every effort should be made to minimize the amount of tissue taken for microscopic study. Very thin, dental sawblades are available, which destroy only a minute amount of bone in the cutting process. Furthermore, there is usually no need to remove a sample more than ½ centimeter thick since the sample will usually be reduced to a thinness that will permit transmission of light.

In removing a specimen of tissue for microscopic analysis, include in the section some of the normal bone adjacent to the morbid tissue and enough of the abnormal tissue to insure that different aspects of the pathological process will be included. For example in some disease processes the edge of a lesion may reveal bone resorption while the more central part of the lesion may show bone formation. A clear understanding of the lesion requires that tissue from both areas be examined.

Once an appropriate thin slab of bone has been removed for study it can be viewed in reflected or incident light using either a dissecting microscope or incident light optics on a research microscope. However, one of the cut surfaces of the specimen should be prepared to insure reasonable resolution. On most cut surfaces, microscopic detail is obscured by the irregular groves from the cutting action. If the bone is fragile it should first be infiltrated with or imbedded in some plastic material before cutting to strengthen it. Ubelaker

(1978:99–101) provides a brief review of this procedure.

After strengthening, one surface can be ground on fine, water-resistant, carborundum paper to remove the rough scratches. It can then be polished slightly by rubbing the surface on good quality bond paper. Repeated microscopic inspection during processing will indicate when grinding and polishing is adequate but the order of magnitude is a few minutes.

In most cases a bone section (75–100 microns) is the most satisfactory preparation for microscopic study. Elaborate equipment and procedures exist for making such preparations (e.g., Jowsey 1955; Moreland, 1968; Ortner and Yong, 1975); however, a few inexpensive supplies will make it possible to produce a satisfactory thin section in most cases (Frost, 1958).

In general the preparation is an extension of the procedure for polishing a single surface except that the grinding stage continues until the specimen is very thin (about 100 microns) before the polishing begins. Usually it is important to grind both cut surfaces and polish at least one surface, although the latter may not be needed if the section is mounted on a glass slide and coverslipped. If the specimen is large (2 cm or more) it may be difficult to grind the surface uniformly. Mounting one ground and polished surface on the glass slide and then grinding down the free surface to the appropriate thinness may help, but specially designed cutting and grinding equipment will produce the best results. Such equipment is available at many universities in the departments of geology or mineral sciences.

Additional data may be obtained from a contact microradiograph of a bone section. Techniques for preparation of microradiographs are well-known (Amprino and Engström, 1952; Sissons, Jowsey, and Stewart, 1960; Ortner, 1975). In essence the technique involves placing a bone section on a high resolution X-ray film or plate and exposing the plate to low energy (10–15 Kv primary voltage) X-rays for a relatively long time (up to 1½ hours). Special equipment is necessary for this, and consultation with one of several laboratories doing such work would be appropriate.

Having obtained a bone section and perhaps a microradiograph, the next step is to evaluate the findings. One should be familiar with the microscopic appearance of normal bone at different ages, for it is against this background that an understanding of the abnormal condition must be obtained. Given this background there are several aspects of disease that should be considered. However, with the exception of slight changes produced by osteocytes, all the abnormal changes seen in bone microstructure are the result of abnormal osteoclastic or osteoblastic activity.

Microscopically, one can, in theory, see decreased, normal, or increased osteoblast activity in combination with either decreased, normal, or increased osteoclast activity. Within a single lesion there may be one or more combinations of cell activity, and multiple lesions may vary in their expression.

In compact bone, disease is expressed as an abnormal quantity of resorption spaces and/or forming osteons. The latter can be identified by the presence of osteoid seams or, if these are missing in archeological bone, as low density osteons (in a microradiograph) with large central, or Haversian, canals. Let me illustrate these principles with a few concrete examples. I have already noted that Paget's disease involves an increase in the activity of both osteoclasts and osteoblasts with an increase in the internal or osteon remodeling of compact bone. This results in a microscopic appearance of many resorption spaces and many forming osteons and many osteon fragments. In hyperparathyroidism, there is a marked increase in osteoclast activity but little or no effect on osteoblasts. Here the microscopic picture is one of abnormal resorption occurring within existing bone. In compact bone there is a dramatic increase in the number of resorption spaces. The trabeculae of spongy bone will frequently be tunneled out by osteoclastic activity. This contrasts with severe osteoporosis, in which there is increased osteoclastic activity and/or possibly decreased osteoblastic activity in which the

overall cortex becomes thinned (involution) in addition to the increase in resorption spaces.

Another aspect of the overall bone cell response to disease is the speed of the activity, particularly with respect to bone formation. If the process driving osteoblasts to produce bone is very rapid, the result will be poorly organized fiber bone rather than lamellar bone. Such bone will also tend to be poorly mineralized, a feature that would be apparent in a microradiograph.

There is an additional problem to keep in mind in archeological specimens. Microorganisms in the soil have the ability to cause local destructions of bone and may give at least a superficial appearance of osteoclastic activity (Hamperl, 1966: 82–83). Marchiafava, Bunocci, and Ascenzi (1974) report that fungi are one type of organism responsible for such activity. In general the spaces produced by destruction of bone microorganisms are much smaller than those produced by osteoclasts.

Trauma

Fracture

Pathology

Next to the almost ubiquitous degenerative changes seen in archeological specimens, the most common pathological condition affecting the skeleton is trauma. In the most general sense, trauma affects the skeleton in four ways: (1) partial to complete break in a bone, (2) an abnormal displacement or dislocation of bone, (3) a disruption in nerve and/or blood supply, and (4) an artificially induced abnormal shape or contour of bone. There are other traumatic conditions that occasionally can be inferred from circumstances of burial, such as death from complications of pregnancy.

It is important to remember that trauma generally represents extrinsic influences on the skeleton, which result from many factors. Clearly the incidence and location of traumatic events is greatly influenced by culture. Hunters undoubtedly have a different pattern of traumatic conditions in the skeleton than sedentary farmers. Women generally differ from men, and children differ from adults. Of course, there are physiological factors to be considered as well, such as senile osteoporosis and other morbid conditions, which greatly increase the vulnerability of the skeleton to trauma. The various types of trauma that affect the skeleton include (1) fracture, (2) dislocation, (3) post-traumatic deformity, and (4) miscellaneous traumatic conditions, including those which do not affect the skeleton directly but can be inferred by the position or association of skeletal specimens. I will emphasize the biology and modern expression of each of these conditions and then review the archeological evidence for each type of trauma.

In this discussion the term "fracture" is used in its broadest sense: any traumatic event that results in partial or complete discontinuity of a bone. Thus, traumatic conditions, such as a sword or axe wound, an injury to bone from a spear or arrow, or a surgical procedure such as trephination, are considered to be fractures, as well as the more conventional discontinuities of bone. If the break is incomplete, meaning that it does not go through the entire bone, it is called infraction; complete separation is called fracture.

Fracture most often is the result of abnormal stress applied to one or more bones. This stress can be dynamic, meaning sudden high stress, or it can be static in which the stress is low initially but gradually increases until the break occurs. If a bone is exposed to excessive but intermittent stress over a fairly long time (several weeks), a fatigue fracture may develop. A final category of fracture occurs when bone is weakened by a morbid condition such that minimal stress results in a break. This is called a pathological fracture.

Types of Fracture

Stress in bone results from the application of one or more of the following types of force: (1) tension, (2) compression, (3) torsion or twisting, (4) flexion or bending, and (5) shearing. Figure 41 illustrates the direction of force for these types of stress.

Dynamic stress fracture is the most common traumatic condition in skeletal material. Indeed, it forms the bulk of cases in modern orthopaedic practice. Each type of dynamic stress produces a different type of fracture. However, many fractures are the result of more than one type of stress. Although it is not always possible to identify the type of stress from the resulting fracture after callus formation and healing, attention to this aspect of fracture may provide significant insights regarding common traumatic stresses in archeo-

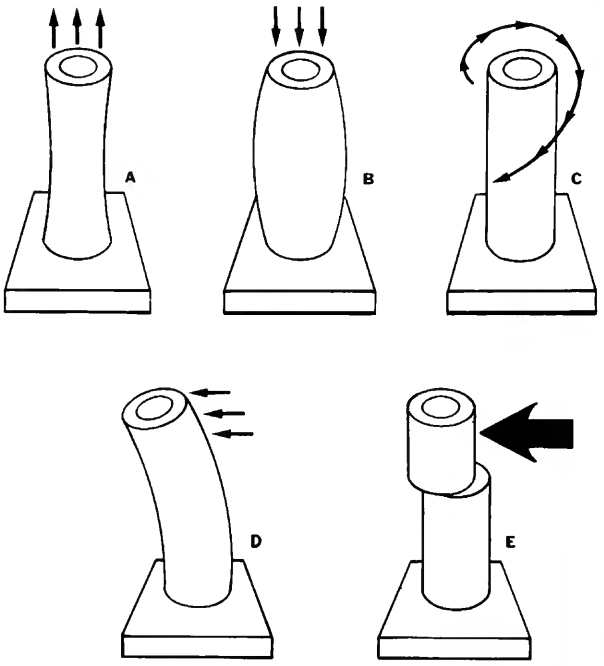


FIGURE 41.—The types of stress in bone that can result in fracture: A, tension, B, compression, C, twisting, D, bending, and E, shearing.

FIGURE 42.—Transverse, partially healed, fracture of the patella (arrow). This fracture is attributed to trauma to the patella resulting from a fall on the knee from a great height. The morphology of the fracture is also compatible with fracture resulting from sudden tension of the quadriceps tendon and the ligamentum patellae. (WM S33 A1.)

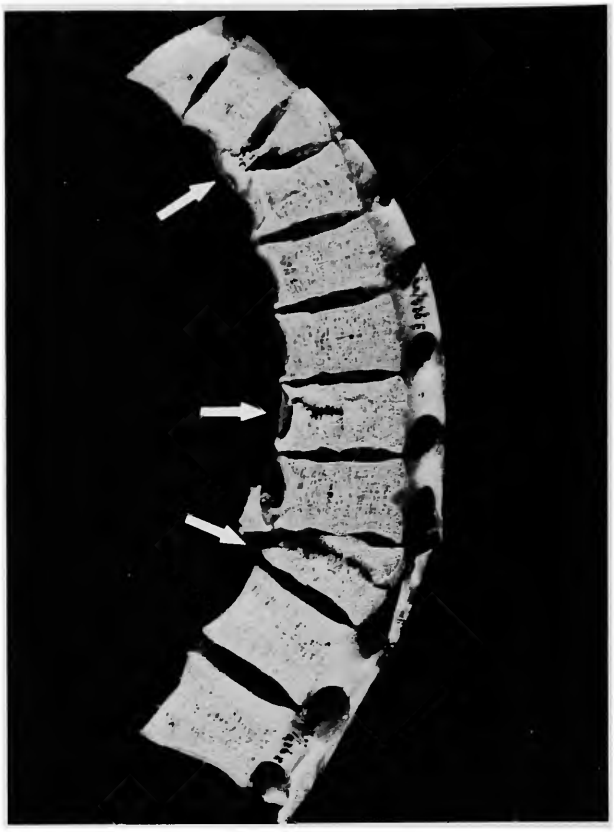
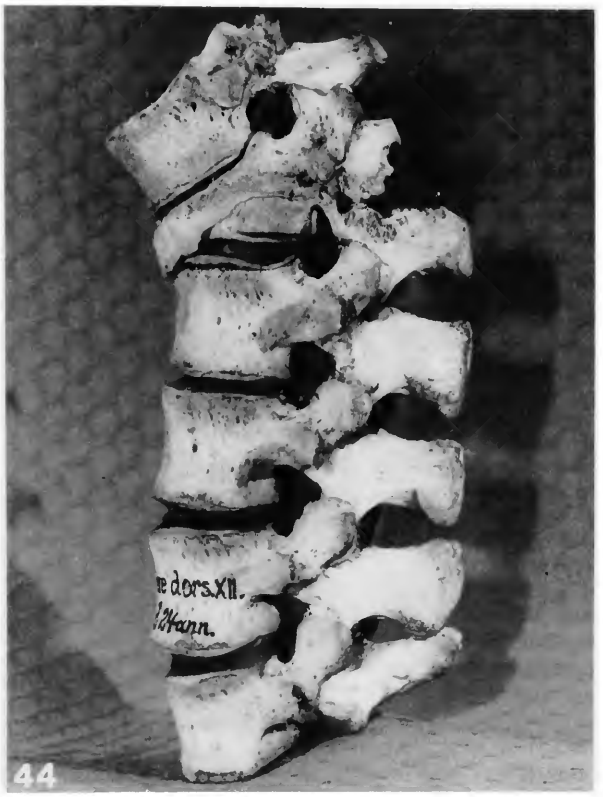


FIGURE 43.—Multiple compression fractures of the thoracic (T) vertebrae. T6 (upper arrow) was fractured about one year before death. Callus is evident on the anterior portion of the vertebral body. T10 and T12 (lower arrows) were fractured in a fall occurring shortly before death. No callus is evident. (64-year-old male, IPAZ 8596.)

logical skeletal samples. For this reason I provide a brief review of the relationships between a specific type of stress and the resulting fracture. A more detailed discussion is found in Schinz, Baensch, Friedl, and Uehlinger (1951–1952:270–280).

Tension fractures are generally associated with tendinous attachments to bone. In this type of fracture the tubercle or process to which the tendon is attached breaks off as a result of excessive tension from the tendon (Figure 42). This type of fracture is often associated with dislocation of a joint.

Compression fractures are the result of sudden excessive impaction and result in a variety of patterns. Bone can split in the same axis as the



FIGURES 44, 45.—Healed fracture of the twelfth thoracic vertebra (second vertebra from the top): 44, Lateral view. The vertebral body has collapsed because of compression; however, the spinous process has separated from tension produced during the compression of the anterior portion of the body; the fracture resulted in angular deformity. 45, Anterior view. (24-year-old male, FPAM 5085.)

direction of the force. In long bones excessive compression may produce a fracture in which the cortex buckles and bulges outward. Pure compression fractures are best illustrated in the spine where most fractures of the vertebral body are the result of compression (Figures 43–46). One also sees compression fractures resulting from a blow to the skull (Figures 47, 48). Many of the fractures of joint surfaces are the result of compression.

In torsion fractures the force is directed in a spiral or twisting direction as when one end of a limb is fixed and the other rotates. In modern times this type of fracture is frequently associated with skiing accidents, in which the lower leg is rigidly fixed and the body twists during a fall. Because the stress occurs in a spiral direction the fracture line also spirals. Torsion fractures can be confused with compression fractures in long bones, which follow a natural spiral cleavage plane in the bone. Torsion fractures always involve abnormal rotation of the bone.

Bending fractures are the most common type of bone separation and are often complicated by other types of stress. Abnormal stress can result when the bone is bent as the result of a fall or in response to a blow, as in the "parrying" fractures of the forearm. In either case the maximum stress often occurs at a discrete spot in the bone and may result in a simple transverse separation of the bone. The force can also radiate from the impact point in a conical shape with the fracture

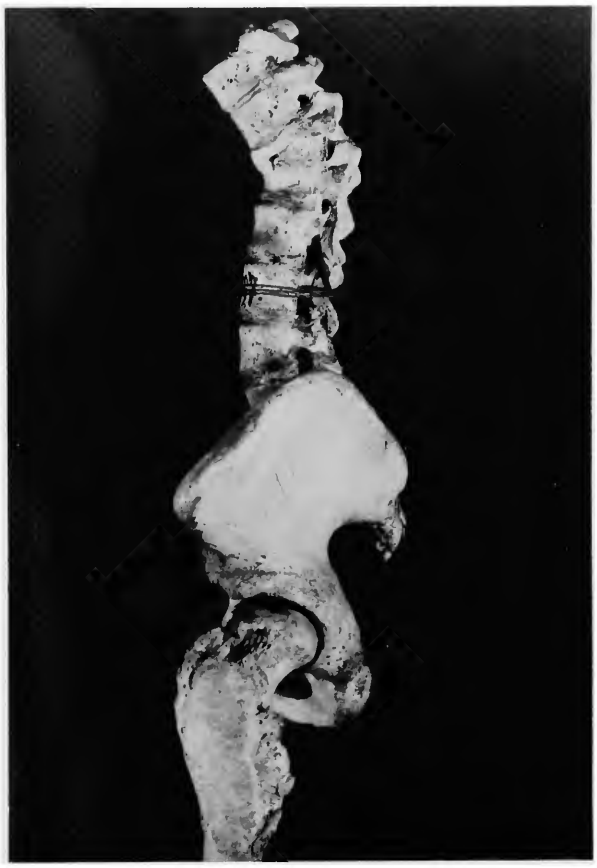
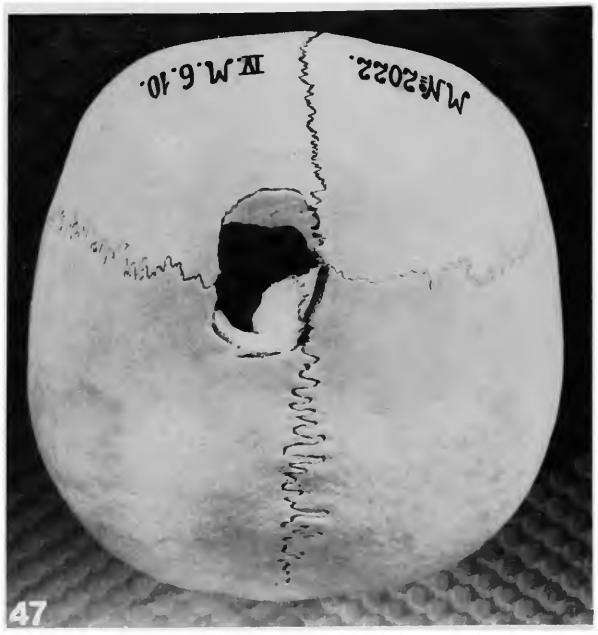
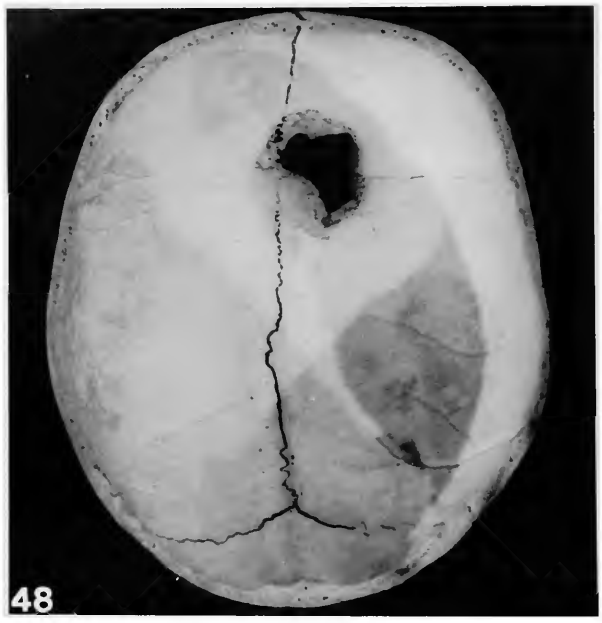


FIGURE 46.—Healed compressed fracture of the twelfth thoracic vertebra (third vertebra from top). Both femora show marked, periosteal, bone hypertrophy over the trochanters, suggestive of myositis ossificans or a reaction to decubital ulcers secondary to the interruption of the spinal cord. A bone fragment had been pushed into the spinal cord adjacent to the fracture. (PMUG 4697a.)





FIGURES 47, 48.—Depressed fracture of the skull vault: 47, Some broken fragments are still attached to the skull, which is indicative of fracture occurring at or near the time of death. 48, Internal view. (Calvarium from a 19-year-old male, FPAM 2022.)

developing along the surface of the cone. This produces a triangular fragment of bone (Figure 49). In a young person bending fractures may produce an incomplete transverse break in a long bone with longitudinal splitting. This condition is known as a "green stick fracture" (Figure 50).

Shearing fractures result when opposite forces are applied to bone in slightly different planes. The opposing forces need not both be dynamic. The bone may be supported by a static force in one axis with dynamic force applied in the opposite direction (Figure 51). One example of shearing trauma is Colles' fracture of the distal radius. This is a fracture resulting from a fall forward, in which the individual reacts by extending the arms to minimize the impact. In this case the dynamic force is the falling body, the static force is the ground or floor. The result is a

fracture in which the distal end of the radius is sheared off and displaced backward. Schinz, Baensch, Friedl, and Uehlinger (1951–1952:276) note that in Colles' fracture the shearing stress rapidly changes to bending stress, and it may be impossible to distinguish the effects of the two types of stress in the resulting fracture.

A fracture varies not only in terms of the type of stress but also with respect to its severity, which is related to the extent of the fracture. A simple fracture is one in which there is only one separation of the bone. This contrasts with a more severe fracture with many broken fragments, which is called a comminuted fracture. In either of these types of fracture the traumatic event may result in the broken bone being exposed through the skin. This is called a compound fracture and can greatly complicate healing because of the possibility of infection (Collins, 1966:50).

While fractures brought about by sudden overwhelming stress are the most frequent of traumatic problems, other types of fracture occur in which other patterns of stress or pathological conditions are contributing factors. In modern populations unusual and continued stress over a period of weeks may result in a fatigue fracture. In fatigue fractures the stress applied to bone does not immediately result in fracture. In all cases such fractures are associated with the onset of intense physical activity of a type not engaged in previously. Fatigue fractures are often seen in military recruits during their initial training period (Schinz, Baensch, Friedl, and Uehlinger 1951–1952:285). In fatigue fracture of the tibia, for example, the predilected site is the proximal, medial metaphysis, particularly in the region of active cutback, where cancellous support is poor. Such fractures are often incomplete, involving only a portion of the cross-sectional plane of a bone. They are also associated with unosteonized circumferential lamellar bone (Johnson, 1964: 607). The physical activity triggers an attempt by the bone to increase its mechanical strength by osteon remodeling. However, the first step in

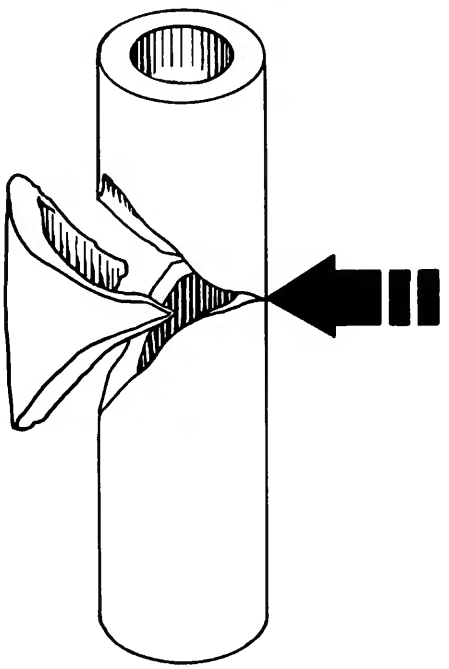


FIGURE 49.—Schematic representation of bending fracture and the triangular fragment that may occur in such fractures.

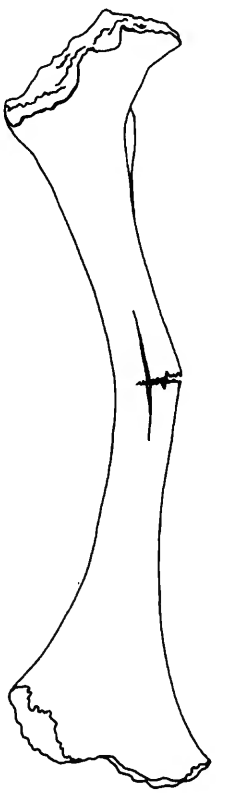


FIGURE 50.—Schematic representation of bending fracture, which produces a green stick separation of part of a bone. Both transverse and longitudinal breakage may occur.

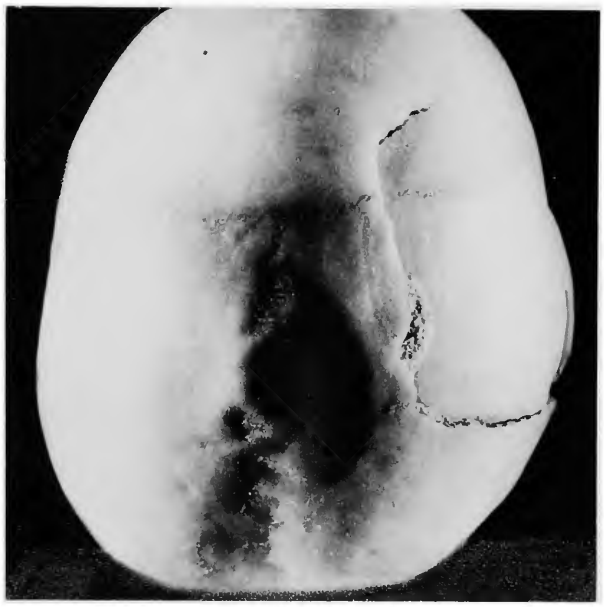


FIGURE 51.—Depressed fracture of the skull of sufficient force to produce shear between the large fragment and the rest of the skull. (Calvarium of an adult male, WM S19.4.)

osteonization is the formation of resorption spaces, which initially weakens the bone. Normally resorption space formation would be accomplished without a serious reduction in the mechanical integrity of the bone. Abnormal physical activity, however, greatly accelerates bone resorption and, if continued, results in callus formation and fracture. Part of the problem of increased osteon remodeling is the fact that osteonal refill of a resorption space occurs at about one-tenth the speed of resorption.

Whether or not the somewhat unusual conditions of physical activity that result in fatigue fractures in modern populations could have existed in archeological skeletons is problematical. Detection of such fractures may also be more difficult since the bone may not be completely broken, and even when it is, good alignment of the broken ends is much more likely in fatigue fracture than in violent fracture. This means that the repair process may leave little trace of the fracture. It is, however, important for the paleopathologist to be aware of such fractures and attempt to make the distinction, since the stress involved is quite different from violent fractures and could contribute misleading data in frequency tables of violent fractures in various parts of the skeleton.

The capacity of bone to resist stress depends on the quantity, quality, and architecture of bone. Conditions that adversely affect one or all of these factors can create a situation in which bone is unable to withstand a moderate force that normally would be tolerated easily. When fracture occurs in such situations it is called a pathological fracture meaning that some morbid process has weakened the bone to the point that it cannot resist relatively normal stress. Many diseases can produce abnormal bone loss and result in pathological fracture (Figure 52). Collins (1966:52) lists many of these which include congenital, metabolic, and infectious diseases, as well as neo-

FIGURE 52.—Pathological fractures of the left femur. Unhealed fractures are seen in the neck of the femur and the proximal metaphysis. The fracture of the upper diaphysis has healed but with angulation. (Adult female with carcinoma of the breast, WM H578.1.)

TABLE 1.—Distribution and types of fractures in cases reported by Hamilton (1853:33)

Bone(s) affected	*Compound & comminuted*	*Compound*	*Simple*	*Total no. of cases*
Nose and face	6	6	12	24
Clavicle	0	4	37	41
Scapula	0	0	3	3
Humerus	2	1	36	39
Radius	0	2	25	27
Ulna	0	2	20	22
Radius and ulna	0	2	31	33
Femur	3	1	67	71
Patella	0	0	7	7
Tibia	0	6	13	19
Fibula	1	4	11	16
Tibia and fibula	13	19	40	72
Bones of the hand	1	2	2	5
Ribs	0	0	4	4
Vertebral column	0	0	3	3
Pelvis	0	0	1	1
Total number of cases	26	49	312	387

plasms. Collins also reports that pathological fractures from different diseases are associated with rather specific age categories. To this list should be added aging, which is generally associated with gradual bone loss and which can contribute to pathological fracture (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952:256).

Previous studies have shown that the location of fractures in the skeleton varies with the age and sex of the individual (e.g., Buhr and Cooke, 1959). Furthermore, there are differences in the frequencies of fracture in various locations in the skeleton (Table 1), which may be associated with cultural differences (Hamilton, 1853; Schinz, Baensch, Friedl, and Uehlinger, 1951–1952:249; De Souza, 1973; Iqbal, 1974). It is apparent that different cultural and environmental conditions create different fracture hazards. The potential fracture hazard of someone negotiating an icy sidewalk is quite different from a person living in the tropics. Similarly, the fracture hazard of the ancient hunter is likely to be different from that of the ancient city dweller. Differences in fracture pattern have considerable significance for paleopathology in part because of these cultural variations and the inferences that can be made regarding the characteristic hazards encountered by different groups.

Fracture Healing

The process of fracture healing in some respects recapitulates events associated with growth. There are, however, significant differences in which the conditions associated with the fracture event itself are significant. Traumatic, fatigue, and pathological fractures will also vary in the healing process. In the following review of fracture healing the major emphasis is on the biology of traumatic fracture repair.

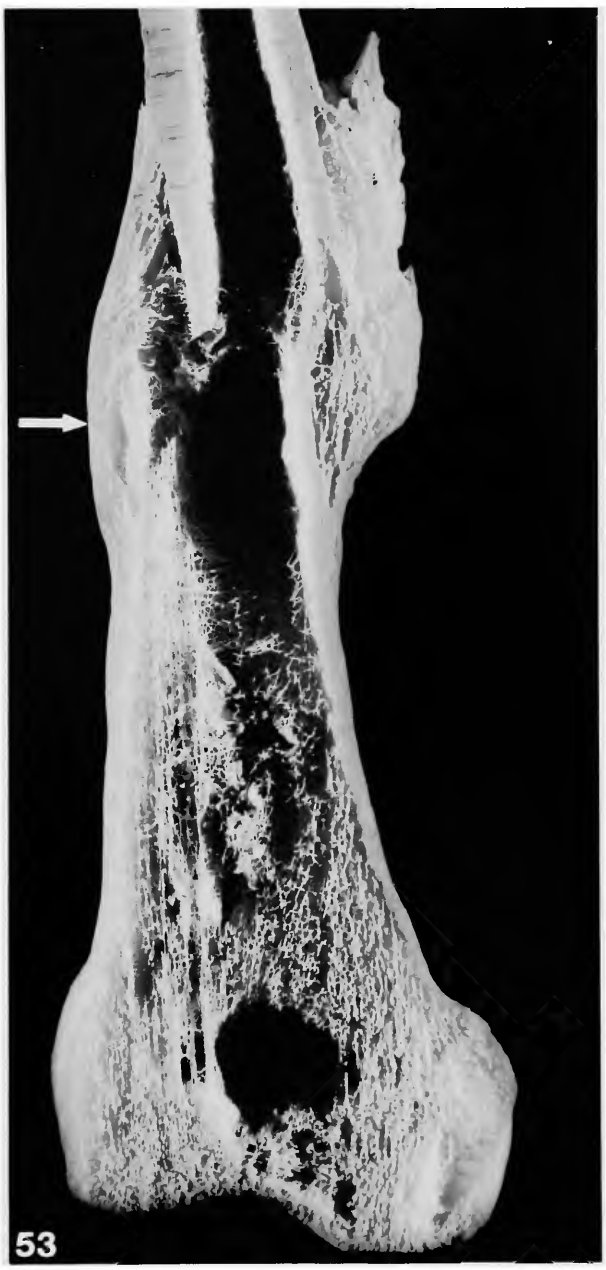
Several events occur with the fracture of a bone. Blood vessels are ruptured both in the cortex (in Haversian canals) and also in the periosteum and marrow. Blood vessels in overlying muscle may also be torn. With the rupture of the blood vessels, blood flows into the fracture region forming a hematoma, which coagulates. There is some debate regarding the value of the hematoma in the healing process. Collins (1966:50) suggests that the clot interferes with healing. More recent research (Schenk, 1973:xv) suggests that hematoma is not essential to fracture healing and indirectly supports Collins' opinion. Regardless of its value in fracture repair a hematoma is an almost inevitable result of fracture. The broken ends of the blood vessels contract and the open ends are sealed by clotted blood. However, the traumatic event stimulates an inflammatory reaction in which intact blood vessels and the undamaged portions of broken vessels dilate and release a plasma exudate, which adds to the fluid at the fracture site (Collins, 1966:44).

The periosteum will usually be broken during fracture, although this is not inevitable, particularly in young individuals. The stress that breaks the periosteum also tends to strip it from the surface of the bone for a few millimeters adjacent to the fracture site. This appears to activate the bone-forming potential of the osteogenic layer in the periosteum and initiates the formation of

callus (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952:378).

Breakage of the blood vessels at the fracture site also results in the death of tissue and cells including the osteocytes supplied by such vessels and any bone fragments resulting from the breakage. This bone will ultimately have to be removed and replaced.

In classic fracture healing, the formation of a blood clot is followed by the second step in fracture repair, in which the blood clot is permeated by fibrous connective (granulation) tissue. Collins



FIGURES 53, 54.—Healed fracture with slight angulation in an adult right femur: 53, Anterior view, arrow indicates callus. 54, Posterior view. (IPAZ 1780/62.)

(1966:50) notes that the hematoma, if not extensive, can assist in initial union by providing a matrix for the advance of granulation tissue. However, granulation tissue can penetrate hematoma only to a limited depth; thus, extensive hematoma between broken ends seems to delay the healing process.

Schinz, Baensch, Friedl, and Uehlinger (1951–1952:378) indicate that the fibrous callus that unites the ends of the fragments is derived from the granulation tissue. This fibrous union of the broken ends is the third stage of fracture healing. While the fibrous callus is unmineralized, it provides the matrix for the formation of fiber bone and the primary bony callus, which is the fourth stage in fracture healing. As I have indicated previously (p. 11), fiber bone is associated with rapid bone formation. In normal conditions it occurs only during the growth phase. Fiber bone is a temporary tissue, replaced by lamellar bone, which has much greater mechanical strength.

Collins (1966:47) notes that primary callus formation generally begins about one week after fracture in human bone. Initially, primary callus consists of coarse, fibrillar trabeculae within the fibrous callus. Primary callus is associated with different areas of the fracture site and three basic types have been defined for conceptual purposes (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952:378). The intermediate or sealing callus joins the broken ends of the bone. The endosteal callus unites the opened marrow spaces forming a plug between the two ends, while the periosteal or bridging callus arches over the fracture site. The bridging callus provides the externally visible evidence of healed fracture in an archeological specimen. One other component of primary callus is the occasional occurrence of cartilage in the region of the callus. Collins (1966:48) expresses the opinion that cartilage is incidental to the overall process of fracture healing. In support of this conclusion, he cites the work of Ham (1930), who suggested that cartilage formation is associated with poor vascular supply. Aegerter and Kirkpatrick (1975:237) challenge this concept but offer no alternative.

It is important to remember that bone formation requires an adequate blood supply. Intense vascular proliferation begins with the formation of granulation tissue and is associated with callus formation. The blood vessels supplying the formation of callus also create conditions in which resorption of bone fragments and dead bone ends at the fracture site can occur.

The fifth stage in fracture healing involves the conversion of the fiber bone callus to lamellar bone. This includes the apposition of lamellar bone to existing fiber bone surfaces, the removal of fiber bone by osteoclasts, and internal osteon remodeling. The result is a much stronger union of the broken ends. This in turn means that less bone is needed for optimal mechanical strength. In the final, sixth stage of fracture healing the callus is reduced to a minimum (Figures 53, 54). If the orientation of the broken ends is good, the callus may no longer be apparent. If angulation has occurred there is compensatory remodeling (Figure 55).

The time for fracture healing varies based on several variables including (1) the bone involved, (2) the severity of the fracture, (3) the apposition of the ends, (4) the stability of the fractured ends, (5) the nutritional state of the individual, and (6) the age of the individual. Collins (1966:52) indicates that in a typical bone under ideal conditions the primary callus takes about six weeks to develop. The secondary callus becomes significant after this time but bony union and return of adequate function is too variable to predict.

Cruess and Dumont (1975:97), rather than assigning a specific timetable, indicate the approximate percentage of the total healing time spent in the three major phases of fracture healing. These phases include the inflammatory phase (10 percent), the reparative phase (40 percent) and, the remodeling phase (70 percent). The phases do not total 100 percent because of the overlap in time between the phases. In any case the time of reduced mechanical function is certainly a matter of weeks even in an ideal situation.

The significance of this period of reduced activity is difficult to assess. Schultz (1939:978)

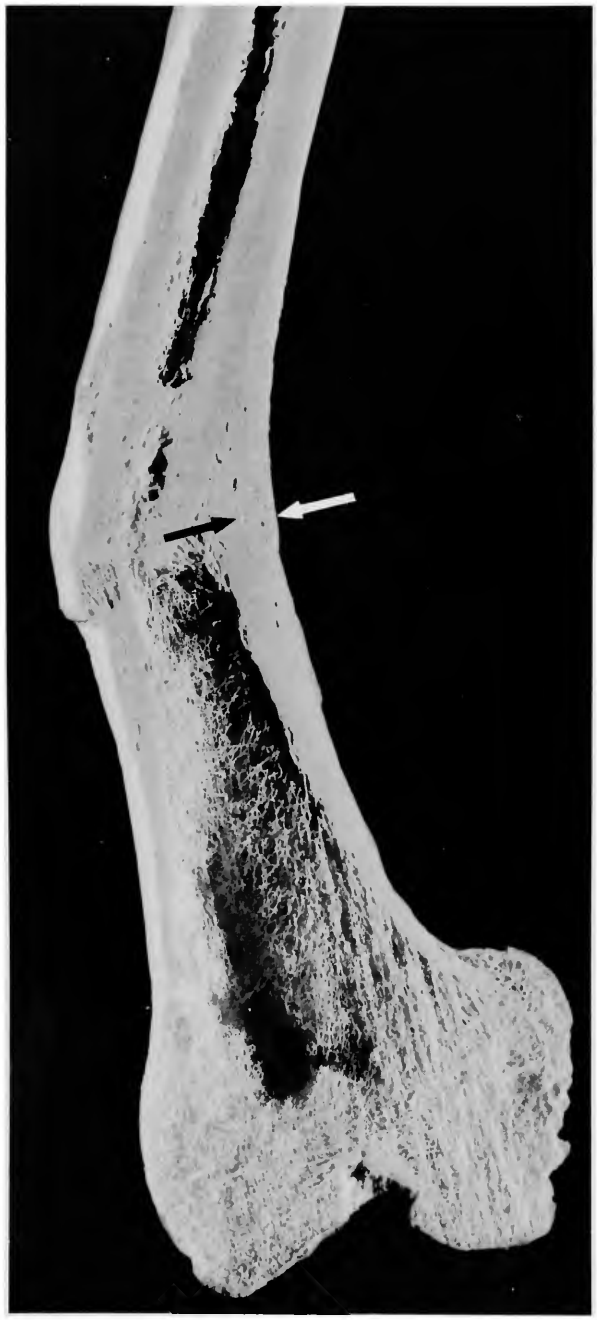


FIGURE 55.—Compensatory remodeling in angulated healed fracture of the right femur. Arrows indicate the thickness of new cortical bone added to the concave side of the angulated femur shaft. On the opposite side some of the cortical bone of the broken ends has been removed. (Adult, IPAZ 192/60.)

reports that the percentage of fracture cases for free-ranging nonhuman primates varies between 12 and 34. Primates shot in the wild present numerous examples of well-healed fractures (Duckworth, 1912; Schultz, 1939), indicating that fracture need not be a serious threat to life. In human populations the threat is likely to be even less, but greatly dependent on the environment of the group. For an adult member of an Eskimo family, such an injury could be very serious indeed.

Complications Arising from Fracture

The complications of fracture include (1) infection, (2) tissue necrosis and loss of innervation, (3) inadequate fusion of the fracture, (4) bone deformity, (5) traumatic arthritis, (6) joint fusion, and (7) traumatic myositis ossificans. I shall provide a brief review of each of these conditions.

Infection is a serious complication of fracture and can occur whenever infectious agents become associated with the fracture site. This event can occur if the soft tissue in the fracture area is penetrated by one or both of the broken ends of the bone. It can also occur if the agent producing the fracture also penetrates the skin, as would be the case with a bullet or other projectile. Infection is also possible if bone penetrates any portion of the digestive track, or any body cavity where infectious organisms exist.

Böhler (1935:65) indicates that open (compound) fractures with direct injury to the skin (crushing or penetrating wounds) have a much greater risk of infection than those due to indirect injury (as when a broken bone penetrates the skin). Two major infectious conditions pose a serious threat to life. The first of the conditions is gas gangrene arising in necrotic tissue by action of organisms, such as *Clostridium*, which do not require oxygen (anaerobic). The other major threat to life is pus-forming (suppurative) organisms, which can spread from the fracture site and can produce general infection of the blood (septicemia) and ultimately death. Böhler (1935: 64) reports statistics for 1887, in the pre-antibiotic

period, in which the estimated mortality from open fractures in German hospitals was 40 percent. The data reported in Table 1 indicate that compound fractures (compound-comminuted and compound only) occur in about 19 percent of all fracture cases.

Another complication of local infection at the fracture site is disruption of the blood supply to bone leading to death of the tissue (necrosis). At the boundaries of the dead tissue osteoclasts may isolate the dead bone through osteoclastic resorption. Such an isolated segment of dead bone is known as a sequestrum. Often, if the blood supply to the bone itself is cut off, the periosteum remains intact and produces a new sheath of bone called involucrum around the necrotic tissue. A more detailed review of infection in bone is presented in the chapter on infectious diseases (p. 104). With respect to fracture the significant point is that infection, certainly in pre-antibiotic times, is a most serious complication, having a significant risk of mortality. However, in archeological skeletons it will require very careful and observant excavation technique to distinguish an unhealed fracture resulting in or associated with death from that of postmortem damage from soil movement or careless excavation.

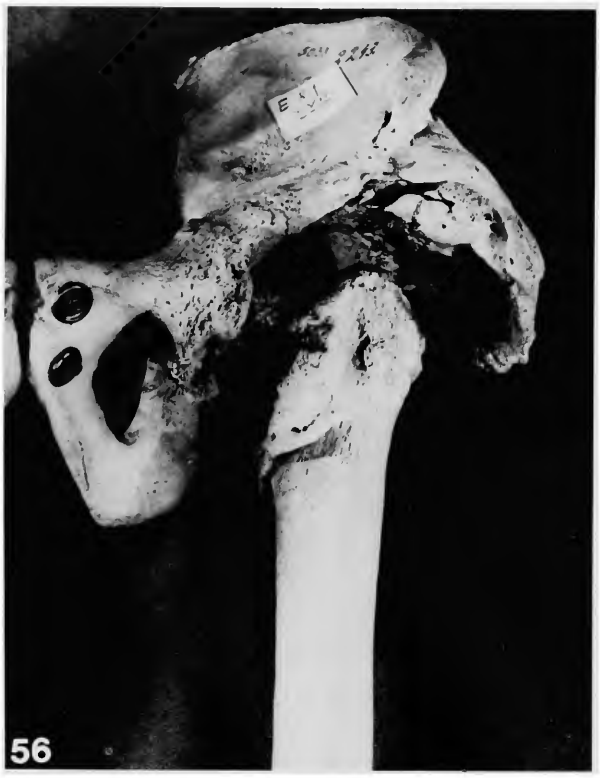
I have already noted that rupture of some blood vessels is an inevitable result of all fractures (p. 61). Damage to major blood vessels and nerves is not inevitable because of the flexible and relatively tough nature of these structures. Hazard to major blood vessels and nerves is greatest in crushing injuries resulting in fracture. However, even in simple fractures, the displacement of bone ends can compress or twist blood vessels or nerves (Böhler, 1935:73) and lead to ischemia and loss of nervous stimulation. Loss of blood supply will lead to delayed fracture healing and, if uncorrected, can result in bone necrosis. Loss of nerve supply does not appear to be as serious. However, if sensory nerve supply is lost to the fracture site, the lack of pain may allow continued use of the broken bone and prevent healing or, in the case of a fracture of an articular surface, lead to a neuropathic or Charcot joint, in which there will be exuberant bony response to unrelieved trauma to the joint (Figures 56, 57).

In some cases the distribution of dynamic forces immediately after trauma may not break the bone but will disrupt the blood supply, lead to ischemia, and may result in tissue necrosis. Three such cases may be seen in the Hunterian Collection in London (Figures 58–60). All three cases are attributed to a blow on the head. If the blood supply in the immediate region of the blow is disrupted, the tissue becomes ischemic or necrotic, and there is an inflammatory response adjacent to the affected bone.

Traumatic interruption of blood supply in a growing bone can produce abnormal shortening of bone. The growth plate in such bones is particularly vulnerable to trauma because of the relative weakness of calcified cartilage. Trauma to the ends of growing bone cannot only break the cortex but, also, can shear the epiphysis (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952:422). In both cases, the blood supply is disrupted and growth is retarded. Such injuries to the epiphysis may also result in premature fusion of the growth plate. The result of any of these complications is a shortened bone. If only one of the two bones of the forearm or lower leg is involved, abnormal angulation of the hand or foot ensues. If both the radius and ulna or the tibia and fibula are affected the entire forearm or lower leg will be shortened. Similarly, fracture at the growth zone of other long bones can produce reduced length of the bone.

Serious disruption of nervous supply to other parts of the body may be associated with fracture of the vertebral column, depending on the vertebrae involved, since either the spinal cord or spinal nerves can be affected. Böhler (1935:109–145) reviews these fractures and their complications. The most serious complication occurs in complete severance of the spinal cord. The permanent paralysis below the level of the injury, which inevitably results, affects various parts of the body depending on where the break occurs.

The comminuted fragments can put pressure on the spinal cord or spinal nerves producing



FIGURES 56, 57.—Charcot joint resulting from traumatic fracture of the left femoral neck: 56, Anterior view of innominate and femur; note the large mass of bone over the acetabulum creating a new joint, which comes in contact with the greater trochanter. 57, Inferior-lateral view. (ANM 2243.)

paralysis, which probably will not heal spontaneously. Since medical procedures are not likely to have been available in archeological populations, evidence of a partially occluded neural canal above the second lumbar vertebra should be suggestive of paralysis. Evidence of this should be sought in the limbs likely to have been affected (e.g., Figure 46). In general, paralysis prevents effective use of the paralyzed limb and will result in bone atrophy (p. 325).

A less serious result of spinal fracture is a blood clot or callus producing pressure on the spinal cord or nerve root. The blood clot will disappear in time if the individual is kept quiet and the callus may be partially remodeled away after the break is healed. Dysfunction in the form of paralysis in such cases may be temporary.

In some fractures the healing process is delayed or incomplete. This condition is almost always the result of inadequate immobilization of the fracture during the healing phase. Two complications can arise. In the first of these there is inadequate mineralization of the callus allowing the fractured ends continued movement. Such a condition is called pseudarthrosis. The second condition is called nearthrosis and differs from pseudarthrosis in that an actual joint space is formed (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952:425–430). In the American literature both these terms are frequently used synonymously and with the term "nonunion." However, each condition produces a slightly different result in bone and thus, should have distinguishing terms.

The factors that produce nonunion remain somewhat obscure. Schinz, Baensch, Friedl, and

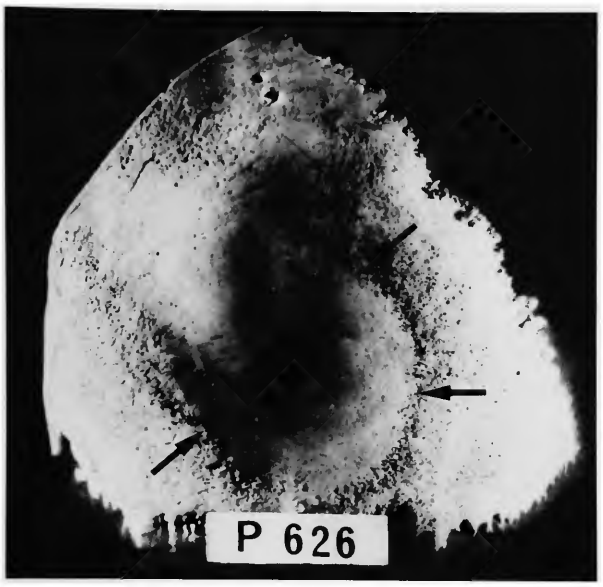


FIGURE 58.—Central bone necrosis surrounded by a zone of hypervascularity (arrows) following a blow to the head. (Portion of the left frontal bone, HM P626.)

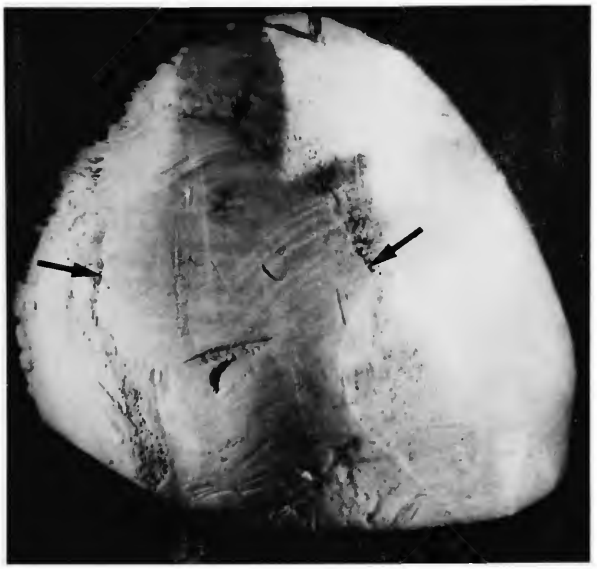


FIGURE 59.—A large area of bone necrosis (arrows) surrounded by a zone of hypervascularity following a blow to the head. (Frontal bone, HM P627.)

Uehlinger (1951–1952:379) indicate that repeated disruption of the callus results in increased hyaline cartilage formation instead of the mineralizable osteoid normally expected in callus. This may be a response to reduced blood supply caused by breakage of developing vessels during movement of the broken ends (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952:427). Another possibility is the interposition of other tissue (e.g., muscle) between the broken ends.

In pseudarthrosis resulting from fracture, the ends of the broken bone are joined by connective tissue and, indeed, extensive callus may form. However, this tissue does not mineralize properly due to a lack of osteoid, and the union remains flexible. With time the broken ends of the bone will be sealed off by new bone formation and will be connected by flexible fibrocartilage. With continued movement, hyaline cartilage may develop near the middle of the fibrocartilage mass and a cleft develop forming a true joint. Ultimately, a joint capsule may surround the joint space, thus approximating the anatomy of a normal joint. The result is usually severely diminished function

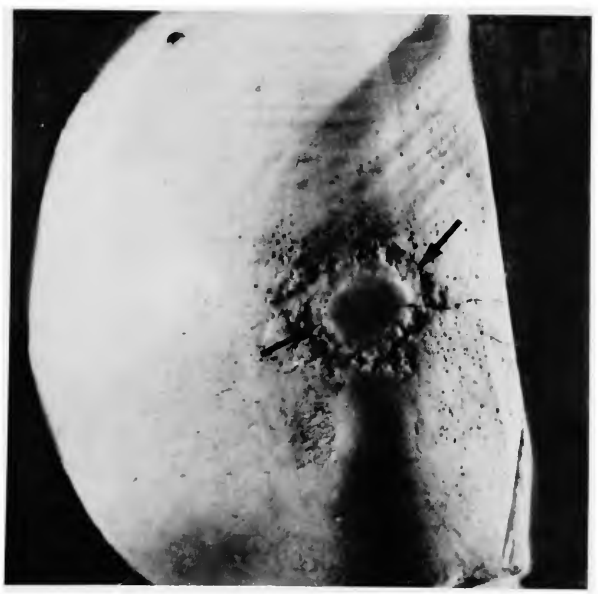
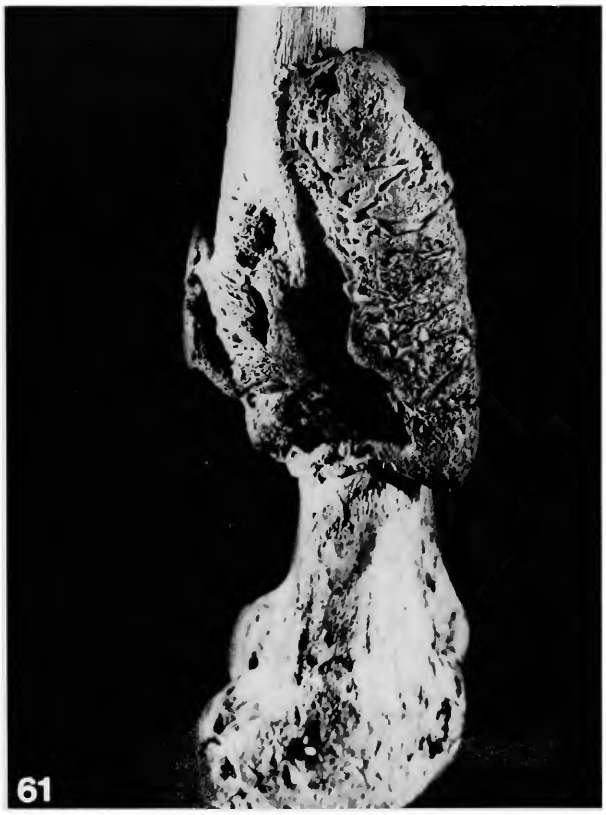


FIGURE 60.—A small area of bone necrosis surrounded by a zone of hypervascularity and bone resorption (arrows) following a blow to the head. The traumatic nature of the lesion is demonstrated by the partially healed bone fragment occurring internally. (Portion of the frontal bone, HM P628.)



FIGURES 61, 62.—Healed diagonal fracture of the distal, left femur with marked mediolateral and longitudinal displacement of the broken ends: 61, Medial view. 62, Posterior view. The displaced ends are joined by abundant callus. There has been considerable cortical remodeling of the broken ends, which have been sealed off with compact bone. (WM S32.7.)

of the limb. However, in the case of the forearm or lower leg, nonunion in one bone may be compensated by extensive hyperplasia of the other bone and adequate function can result (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952:428).

Problems in mechanical function can also be caused by misalignment of the broken ends. Two factors contribute to this. The stress producing the fracture may produce misalignment. At least equally significant, however, is the fact that the sensory stimuli triggered by the fracture produce a virtually instantaneous and powerful contraction of the muscles in the fracture area. Subsequent movement of the limb will repeat this stimulus if pain results (Böhler, 1935:58–59). With such muscle contraction the likelihood of misalignment is high. In modern clinical practice initial management of fracture involves minimizing muscle contraction and careful realignment of the broken ends so that the bone approximates, as near as possible, its original shape.

Even when this procedure is attempted, however, misalignment can occur as is illustrated in a modern case (Figure 55). The effects of poor alignment vary with the bone involved and the severity of the fracture. Rarely, if ever, does it produce a direct threat to life. However, it can lead to premature and severe degeneration of a joint because of the abnormal stress on the joint brought about by misalignment. In the long bones this can result in reduced locomotion and manipulative ability. Misalignment of a vertebral

fracture can result in varying degrees of angulation in the spine since the vertebrae are all closely linked to each other in mechanical function.

Fracture can, directly or indirectly, produce premature osteoarthritic degeneration of joints. If the joint surface is broken, the cartilage and subchondral bone are disrupted. Because of the absence of blood supply, the repair of cartilage depends on nutrients from the joint capsule, and any repair is a slow process. A break of the subchondral plate will usually result in permanent discontinuity of the bony joint surface. This complicates the healing process of the articular cartilage, since it results in poor contact between the gliding surfaces of the joint. This creates serious problems in dissipating the stress applied to the joint and results in premature deterioration of the articular cartilage.

The fracture need not involve the joint itself to produce premature degenerative changes in the joint. As I have already noted (p. 68), a change in the axis of a long bone can shift the major focus of mechanical stress to a portion of the joint less well adapted for such stress. Similarly, abnormal shortening of a long bone after fracture (Figures 61, 62) can produce uneven locomotion, which results in unusual stress to the joint and premature degenerative change (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952:430). In trying to distinguish between premature degenerative change resulting from fracture and degenerative change from aging, it is helpful to make a comparison with the joint of the corresponding limb.

Fracture of the joint surfaces can also result in destruction and fusion of the joint. This problem is associated with comminuted fractures of the joint where more than one joint surface is involved and callus formation joins two or more bones. In the limbs this means loss of movement for the joint. In the vertebral column such a fusion is seen in Figure 46 and is a common sequela of compression fractures of one or more vertebral bodies (Böhler, 1935:142–143). In such cases, the mechanism is activation of the periosteum of the body with callus formation extending to adjacent vertebrae. The effect is a rigid segment of spine, perhaps associated with angulation, resulting in little overall loss of spinal movement. Fusion of fractured joints can easily be confused with fusion following infection of the joint (p. 113). In some cases a distinction may not be possible; however, traumatic fusion may retain evidence of the fracture line.

The treatment of many fractures can be accomplished on the basis of intuitive knowledge and common sense. The frequent occurrence of well-healed fractures in archeological skeletons testifies eloquently to this fact. Böhler (1935:566) lists three fundamental laws for fracture treatment. They are (1) reposition and realignment of the broken ends (reduction of the fracture), (2) immobilization (fixation) of the broken bone, and (3) maintenance of circulation and muscle tone through careful exercise of the affected limb.

Obviously, in comminuted fractures, compound fractures, or simple fractures where marked displacement has taken place, proper reduction and restoration may be difficult, if not impossible, without modern techniques, including traction and surgical intervention. However, it is also apparent in archeological specimens that badly misaligned bones do heal and, judging from the lack of disuse atrophy, provide at least limited use of the limb. Perhaps such fractures heal spontaneously with the pain associated with movement being the immobilizing force.

I have noted earlier (p. 61) that trauma to bone can also damage the overlying muscle. Johnson (1964:610) notes that muscle tissue can provide cells that contribute to callus formation around the fracture site. As with bone, the trauma to muscle tends to produce hematoma. With time the hematoma is usually dissolved. Occasionally, however, the muscle tissue will respond to the trauma by producing bone directly in the muscle tissue itself, often in association with the hematoma. This condition is known as traumatic myositis ossificans. This excessive formation of bone by muscle can be entirely separate from the bone (Figure 63) or it can become part of existing bone tissue (Figures 64, 65). The most common areas are the extensors and adductors of the thigh, the

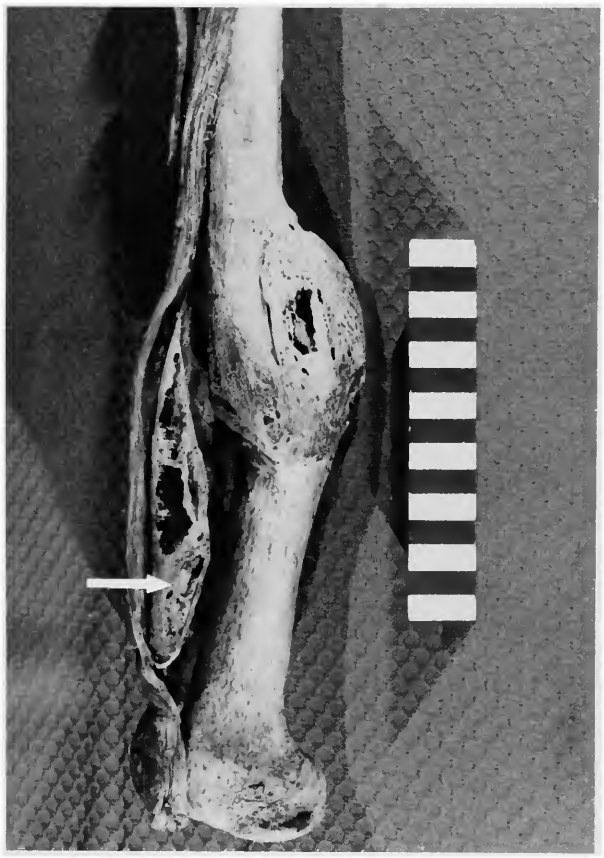
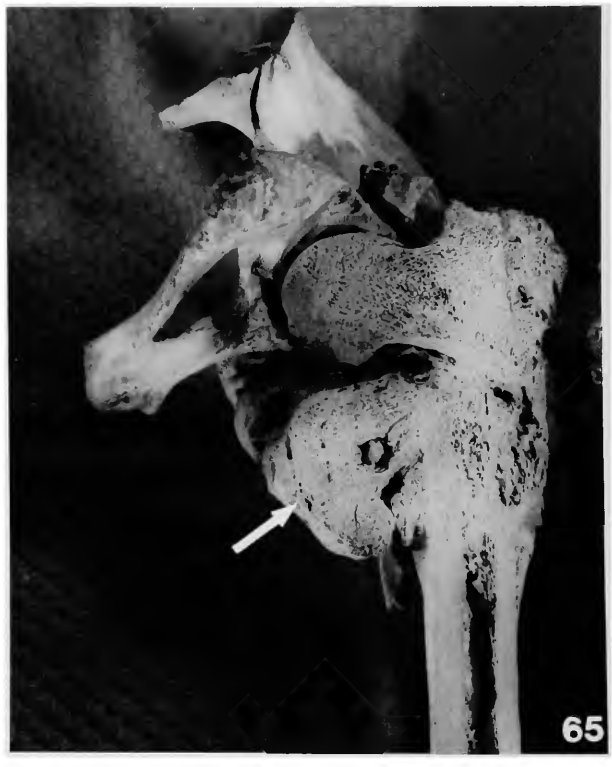
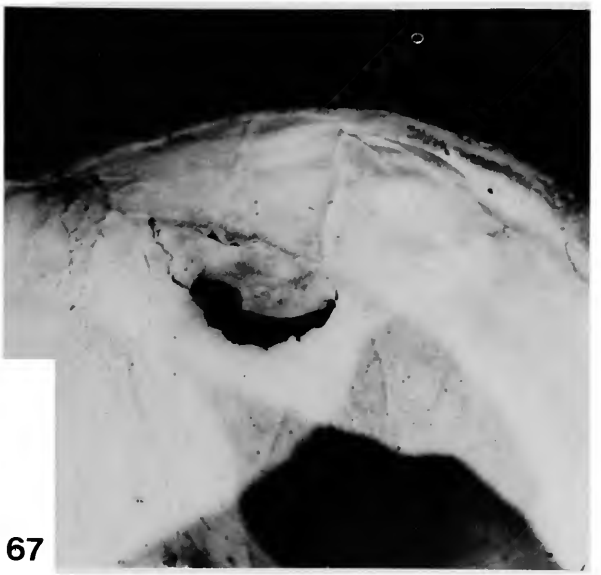


FIGURE 63.—Traumatic myositis ossificans (arrow) within muscle tissue overlying a healed fracture of the left femur. (40-year-old male, FPAM 4751; scale in cm.)

deltoid and the pectoralis. In paralyzed patients minor trauma of the insensitive muscles may lead to metaplastic bony bridging of joints, especially the hip.



FIGURES 64, 65.—Traumatic myositis ossificans of the right innominate and proximal femur of an adult male: 64, Anterior view of hip; massive development of hypertrophic bone is seen in the area of the iliacus muscle (arrow) with exuberant bone development in the area of the intertrochanteric fracture of the proximal femur. 65, Inferior view showing the intertrochanteric fracture and development of hypertrophic bone in the region of the adductor muscle insertions on the femur (arrow). (FPAM, no number.)



FIGURES 66, 67.—Saber wounds of the left parietal: 66, There is a large, displaced fragment on the lateral portion of the left parietal; the wound healed with the fragment in the displaced position leaving an opening in the skull; the second healed saber wound is seen superior to the large defect. Only the outer table was affected. 67, Internal view. (Adult male, ANM 2069.)

Thus far in the review of fracture and its complications I have made only limited reference to the skull. Fracture of the skull presents an additional potential complication not associated with fracture in other bones. The most immediate problem in skull fracture is the hematoma and edema, both of which invariably result from such trauma. An unrelieved major increase in pressure on the brain from hematoma or edema will force the brain tissue into the foramen magnum, disrupting the blood supply to the brain and damaging the brain stem. If the skull is fractured by a blow, the pressure buildup may be relieved through the fracture site. Obviously if the blow is too severe, death will result from direct injury to the brain.

The extent of injuries to the skull that an individual can survive is often remarkable. Figures 66 and 67 show views of a saber wound to the skull, in which a piece of the left parietal was completely dislodged with healing of the fragment in the displaced position. In this specimen (ANM 2069), currently located in the National

FIGURE 68.—Healed wound from battle axe or saber in the left parietal of an adult male skull. Note the fracture lines radiating from the main defect. Evidence of healing and long term survival is seen in the smooth, rounded edges of the major defect. (FPAM 5719; scale in cm.)

Museum of Anthropology, Prague, the edges of the wound are well remodeled, indicating considerable time after the injury before death occurred due to an unrelated cause. In this specimen the glancing nature of the sword wound may have prevented any damage to the brain. Another case of severe trauma to the skull is seen in a specimen (FPAM 5719) from the Federal Pathologic-Anatomy Museum in Vienna (Figure 68). This specimen shows massive fracturing of the left parietal and occipital bones with fracture lines radiating from the major fracture site. The size of the gap is suggestive of a wound from a saber or axe. The edges of the break are well remodeled indicating a long-standing recovery.

Surgical intervention in pathological conditions that affect the skull or its contents is a common practice today. Such a procedure carries with it the risk of infection and damage to the brain seen in traumatic injury of the skull. In view of the risk, evidence of surgical treatment of the skull (trephination) in antiquity is a remarkable phenomenon and will be discussed as part of the review of the paleopathology of trauma.

Paleopathology

History and Examples

In the following review of fracture in archeological skeletal material I use published source materials but emphasize skeletal specimens from the collections of the National Museum of Natural History, Smithsonian Institution, Washington, D. C., USA, and my own experience and observations on paleopathology. It is important to emphasize, at the outset, that the advent and expansion of civilization has not eliminated trauma, either of the accidental or intentional type. What has happened is the introduction of new factors producing trauma, such as the substitution of the automobile for the horse in transportation, and the use of guns and bombs instead of clubs, axes, swords, arrows, and spears in warfare.

While the human species is still subject to trauma, treatment has greatly improved. This improvement took a quantum leap forward with the development of antibiotics, which have greatly reduced the possibility of infection complicating the traumatic event.

These introductory comments serve to highlight the importance of a reference point for the paleopathology of trauma, in which modern medical concepts and treatment, to a large extent, did not exist. It is important to add that, in the treatment of fracture, ancient populations often did a remarkably good job (e.g., Elliot-Smith, 1908:734).

Fracture and the complications arising from fracture most often provide the paleopathologist with easily interpretable lesions in archeological skeletons. However, there are some problems in the interpretation of this type of trauma of which the paleopathologist should be aware. First, evidence of healed fractures occurring in children can be completely obliterated by the skeletal remodeling that occurs with growth. Second, it is difficult and often impossible to make a distinction between fractures occurring at the time of death and those that occur subsequent to death and burial. Wood-Jones (1908b:736) indicates that in Egyptian skeletons, fractures occurring at the time of death produced blood staining adjacent to the fracture site. However, such staining may survive only under conditions of minimal rainfall and soil water content, so that this criterion may be of little value in most geographical areas.

In fracture occurring while the periosteum and other soft tissues are still intact, small bone fragments in the fracture site may still be adherent to adjacent bone (Figure 69). This condition is very helpful in distinguishing between fracture at the time of death and damage to the skeleton after interment resulting from shifting soil or damage during excavation. Also surfaces of bone tend to be stained a somewhat different color from bone not in direct contact with the soil. Thus, breaks occurring during excavation or in subsequent processing may have a surface coloration quite distinct from surfaces in contact with the soil for a long period of time.

A third factor of significance to the paleopath-

ologist is the distinction between accidental fracture, as from a fall, and fracture resulting from intentional violence either in single combat or warfare. Here, both the direct evidence in the skeleton and the circumstances of burial are significant variables.

Occasionally one finds evidence regarding the causative agent in fractures or infractions imbedded in the skeleton. Metallic or mineral fragments of projectile points have been found in archeological skeletons (Figures 70–72). The injury may clearly indicate that a sword, axe, or similar cutting instrument produced the wound (Figure 73). Similarly, a mass burial containing many, unhealed, broken bones may be evidence of warfare or massacre (e.g., Wood-Jones, 1908b; Turner and Morris 1970).

One usually cannot be certain that a healed or unhealed fracture was caused by an accident. However, the location of the lesion in the skeleton is indicative. Wood-Jones (1910c:297) notes a high incidence of defensive (parry) fractures of the ulna in Egyptian materials in comparison with its low incidence in modern city populations. This contrasts with fractures of the humerus, which have virtually the same incidence, suggesting that fractures of the humerus are more likely to be produced by accidental factors that are similar across cultural and time boundaries. There are distinctive accidental fractures (such as Colles' fracture of the distal radius) which are well known in modern clinical practice and are identifiable in archeological specimens (Figure 74).

FIGURE 69.—Adherent bone fragments at multiple fracture sites (arrows) in the left parietal of an adult skull. The adherent nature of the fragments is indicative of trauma occurring while the periosteum and other soft tissues are still intact. (NMNH 266571.)

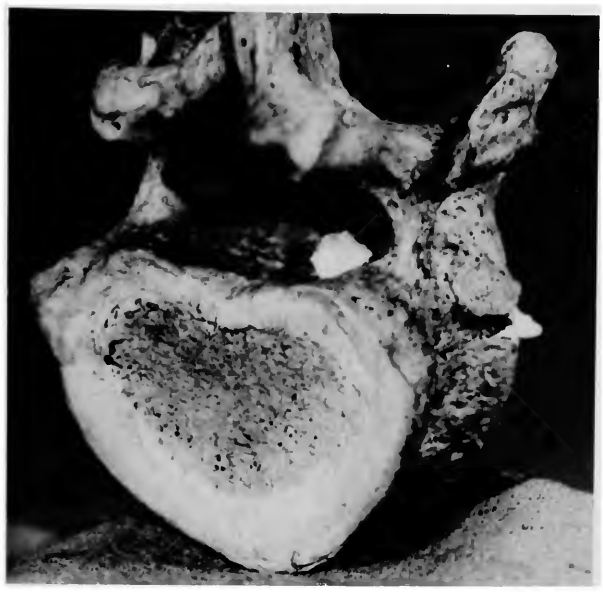


FIGURE 70.—Stone projectile point embedded in the posterior portion of the body of the twelfth thoracic vertebra. (Adult male skeleton from an archeological site in Illinois, USA, NMNH 379841.)

FIGURE 71.—Broken stone projectile point embedded in the anterior portion of the body of a fifth lumbar vertebra. (Adult female skeleton from an archeological site in Illinois, USA, NMNH 380118.)

FIGURE 72.—Tip of stone projectile point embedded in the inner portion of a right innominate bone. (Adult male skeleton from an archeological site in Illinois, USA, NMNH 380120.)

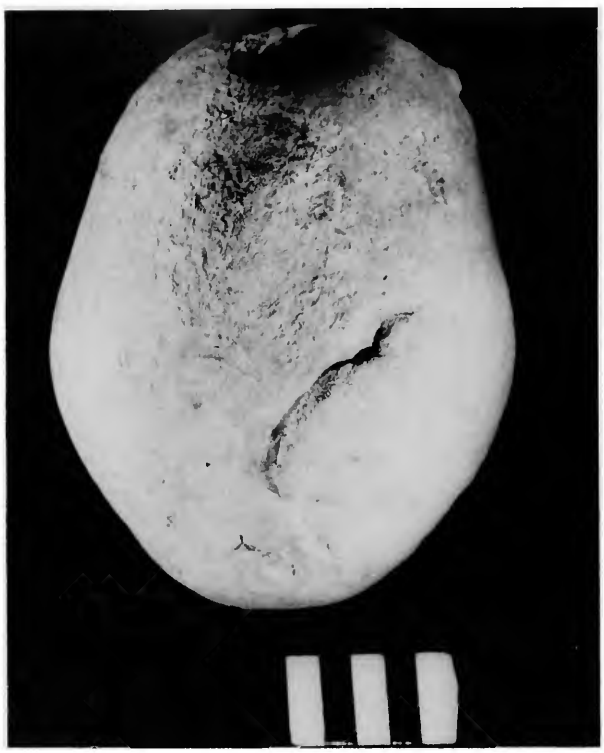


FIGURE 73.—Healed sword wound in the posterior portion of the right parietal. (Adult male, BMNH CG69, XXXIX, 280, Burial 1132.)

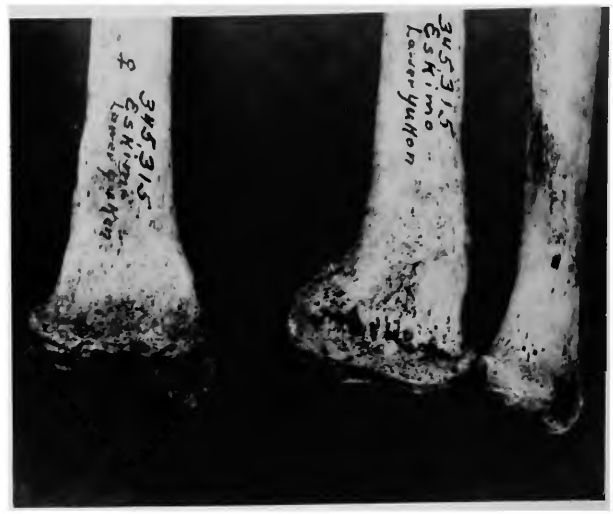
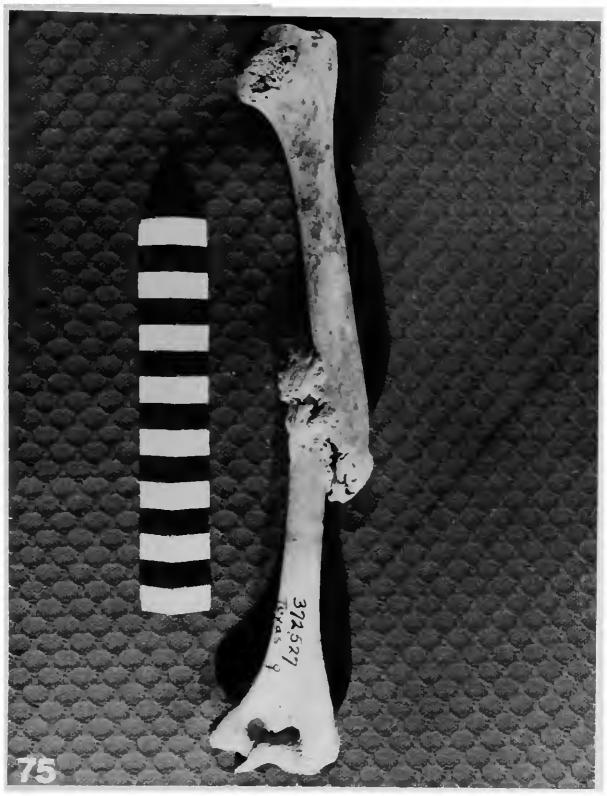


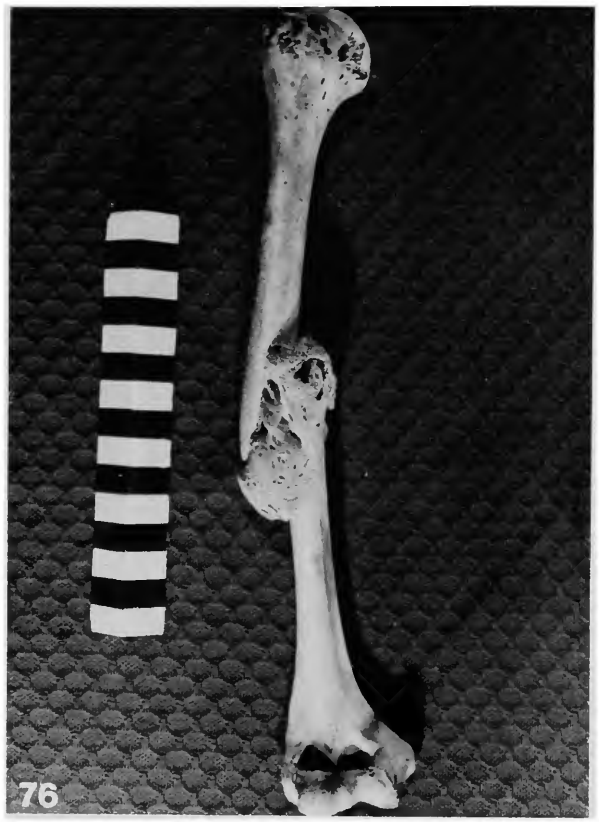
FIGURE 74.—Healed Colles' fracture of the distal left radius (center), shown with the unaffected left ulna (right) and compared with the normal right radius (left). (Female skeleton excavated from an Eskimo burial ground in Alaska, USA, NMNH 345315.)

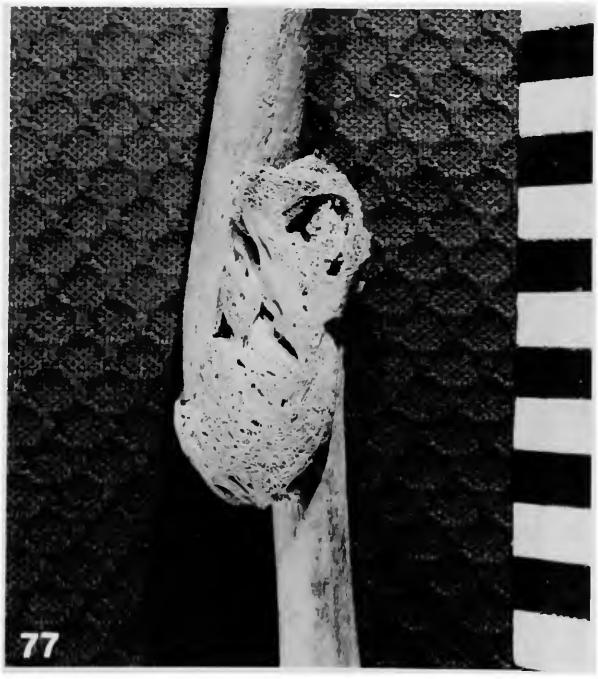
A comprehensive review of the problems in identifying and interpreting traumatic lesions in archeological skeletons is beyond the scope of this book. However, the above observations should serve to make the paleopathologist aware of the types of variables that are important to consider in interpreting traumatic lesions.

Fracture has been documented in some of the Early Paleolithic skeletal remains of *Homo erectus* from Choukoutien cave in China (Weidenreich, 1939:37). Fracture has also been reported in *Homo sapiens* specimens from the Middle and Upper Paleolithic (Moodie, 1922:393; Weidenreich, 1939:38; Roper, 1969). Angel (1974a) surveyed the incidence of fractures from the Neolithic to modern times in skeletal samples from the Eastern Mediterranean. Wood-Jones (1910c:293–340) provides a very detailed report on fractures in ancient Nubian skeletal materials. These reports demonstrate, as one might expect, that fracture is as old as the vertebrates. It is significant, however, that the incidence and distribution of fractures in the skeleton appears to have changed.

While accident undoubtedly was a significant cause of fracture in ancient times, it is clear from the high incidence of fractures attributable to







FIGURES 75–77.—Healed but malaligned fracture of the right humerus: 75, Posterior view. 76, Anterior view. 77, Detail of anterior callus; note remodeling of diaphysial ends. (Adult, female skeleton from Goat Cove, Texas, USA, NMNH 372527.)

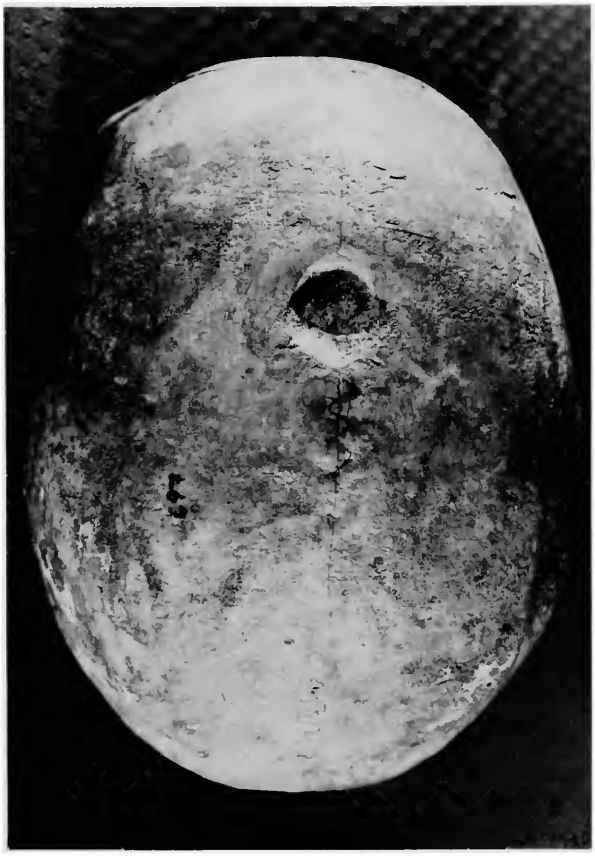


FIGURE 78.—Healed, depressed fracture in an adult male skull. (BMNH Winchester-Saxon pathology collection, CG69, 697, 280 XXXIX.)

violence that intentional violence was a major cause of fractures. Wood-Jones, for example, reports an incidence of fractures of 3 percent in a series of 5000–6000 Nubian skeletons (1910c:294). However, 31 percent of all fractures occur in the bones of the forearm. Wood-Jones found that the ulna is broken more often than the radius and the injury is more frequent on the left side. This led him to conclude that most of these fractures were the result of an attempt to fend off a blow using the forearm (1910c:297). In support of this conclusion he sites the modern habit in Nubia of using a heavy staff for offense and defense. This habit appears to have an antiquity at least going back through dynastic Egypt. Wood-Jones (1910c:295) also reports that the incidence of forearm fractures is about twice that found in

FIGURE 79.—Two healed depressed fractures in the right parietal of an adult female skull (arrows). Note lateral fracture lines radiating from the medial depression. (Specimen from Cinco Cerros, Peru, NMNH 293818.)

FIGURE 80.—Probable healed, glancing, sword wound of the right parietal bone from a Twelfth Dynasty rock tomb at Lisht in Upper Egypt. Note central area of partially filled-in, spongy bone of the exposed diploë. Arrows indicate the porous, reactive margin of the wound (NMNH 256414a.)

early twentieth-century, Western city populations. Other comparisons indicate fewer fractures of the lower leg in the ancient Nubian material in contrast with modern city dwellers. Wood-Jones attributes this difference to modern footwear, which increases slipping and which was not worn by the Nubians (1910c:296). Another marked contrast with modern populations is the incidence of hand fractures, which is about 10 times greater in modern city populations. Injuries from machinery appear to be a major factor in this difference (Wood-Jones, 1910c:297). In the New World, Hooton (1930:312) found a rate of 3.96 percent fractures of long bones or pelvis in a skeletal series of 503 pre- and post-Columbian skeletons from the Pecos site in the North American southwest. When fractures of the skull are added, the total percentage increases to 7.23.

Angel's findings (1974a:13) in skeletal material of various time periods from the Eastern Mediterranean suggest an inverse relationship between cultural level and fracture incidence. However, this trend does not occur in modern populations in the United States, where the fracture incidence is almost twice that of ancient Eastern Mediterranean peoples. Like Wood-Jones, Angel finds that the most common site of fracture is the forearm, which he also attributes to an effort to parry a blow. This suggests that intentional violence is a major causative factor of fracture in the Eastern Mediterranean, as well as in Nubia. Courville (1967:621) reaches a similar conclusion with respect to cranial injuries in antiquity, noting that most wounds and fractures of the skull are the result of "blows of malicious intent."

Earlier (p. 69), I have indicated that the basic concepts of fracture treatment do not require sophisticated knowledge and indeed reflect common sense procedures. It is not surprising then to find that immobilization of fractured bones with a splint was practiced at least as early as the Fifth Dynasty in Egypt (Elliot-Smith, 1908). Although much later in time, there is evidence of similar sophistication in treating fracture among the Incas of Peru (Daland, 1935:550). Wells (1974a) finds evidence for both reduction and splinting in Anglo-Saxon England (ca. eighth century A.D.).

While recognizing that fracture treatment may have been fairly sophisticated in regions of ancient high civilization, it is equally apparent that many fractures, even in these areas, received little, if any, helpful treatment (Wood-Jones, 1910c: 294). The large number of badly deformed, healed fractures from many archeological skeletons testifies to this (Figures 75–77). The fact that fractures heal spontaneously, allowing at least some continued function of the affected bone, is not surprising in view of the reports of healed fractures in wild shot primates (Duckworth, 1912; Schultz, 1939, 1956) where, obviously, there was no treatment.

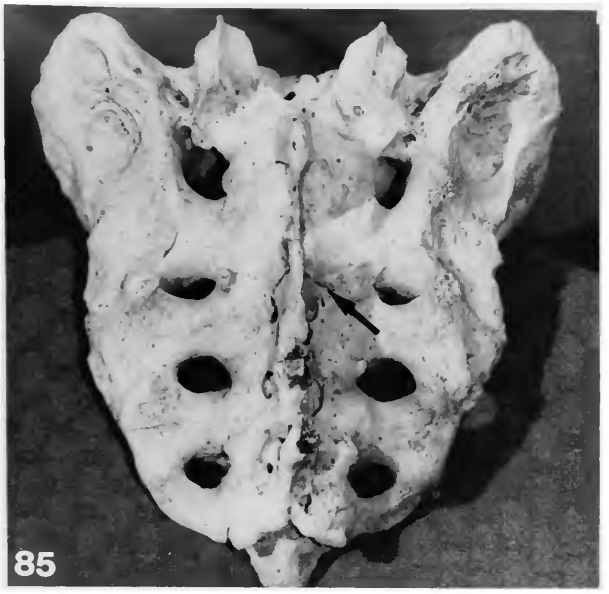
Evidence on the methods for treating fractures in archeological populations is scanty. However, among primitive and peasant groups living in historic times, there are records of fairly sophisticated techniques to reduce fractures (bring about optimal realignment of the broken ends) and immobilize the broken bone until healing can take place (Elliot-Smith, 1908:734; Daland, 1935: 550). There is convincing evidence for the use of splints, pads to absorb blood in compound fractures, and linen bandaging by the Fifth Dynasty (2730–2625 B.C.) in ancient Egypt (Elliot-Smith, 1908:732–733). Freeman (1918:445) reports that manipulation and splinting of fractures occurs among the indigenous peoples of North America as well. Elliot-Smith (1908:733) notes one case where the splinting did little to stabilize the fracture. This is not a serious indictment of their skill since, without X-ray examination and in the presence of considerable pain, it could be very difficult to identify the location of the fracture. With or without effective reduction and immobilization, most often the healing of fracture was uneventful. Elliot-Smith (1908:733) notes evidence of infection in only one case out of 100 fractures in ancient Egyptian material.

Injuries to the skull are rather common in archeological specimens. One type of skull injury is trephination, but because of its importance in antiquity this topic will be treated as a separate category of the paleopathology of trauma

FIGURES 81, 82.—Sword or axe wound in an adult male skull from a rock tomb at Lisht in Upper Egypt dated between the Twentieth and Twenty-fifth Dynasty: 81, The point of contact is the left parietal near vertex; the blow produced a large bone fragment largely in the parietal. 82, Skull with bone fragment reflected. (NMNH 256384.)

FIGURE 83.—Extensive radiating fractures from a central impact point (arrow) in an adult female skull from Pachacamac, Peru. (NMNH 266584.)

FIGURE 84.—Compression fracture of the second lumbar vertebral body in an adult male skeleton from the Hawikuh archeological site in New Mexico, USA. (NMNH 308653)



FIGURES 85, 86.—Partial fracture of the third sacral segment (arrow) of specimen from the rock tombs at Lisht, Upper Egypt, dated between the Eighteenth and Twenty-first Dynasty: 85, Posterior view. 86, Detailed view; note partial bony fill-in of fracture gap and porous reactive bone (arrow) adjacent to the fracture line. (NMNH 252874.)

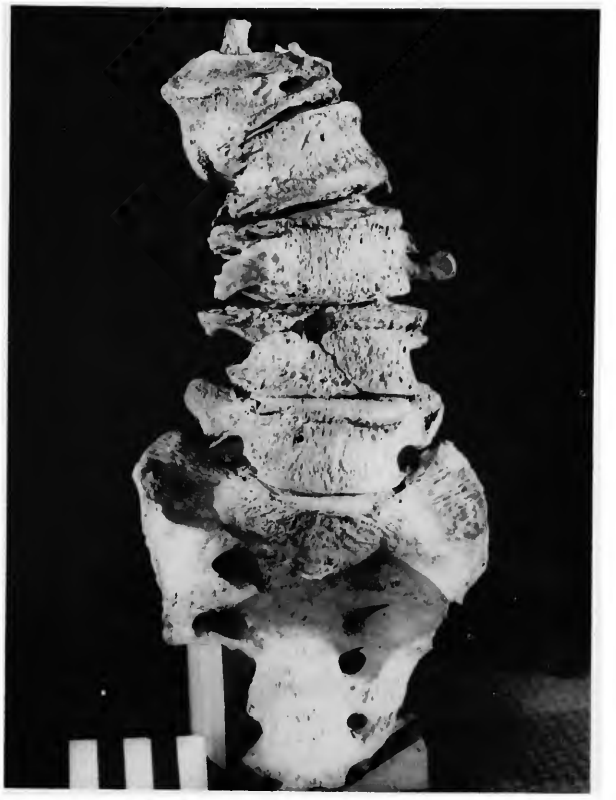
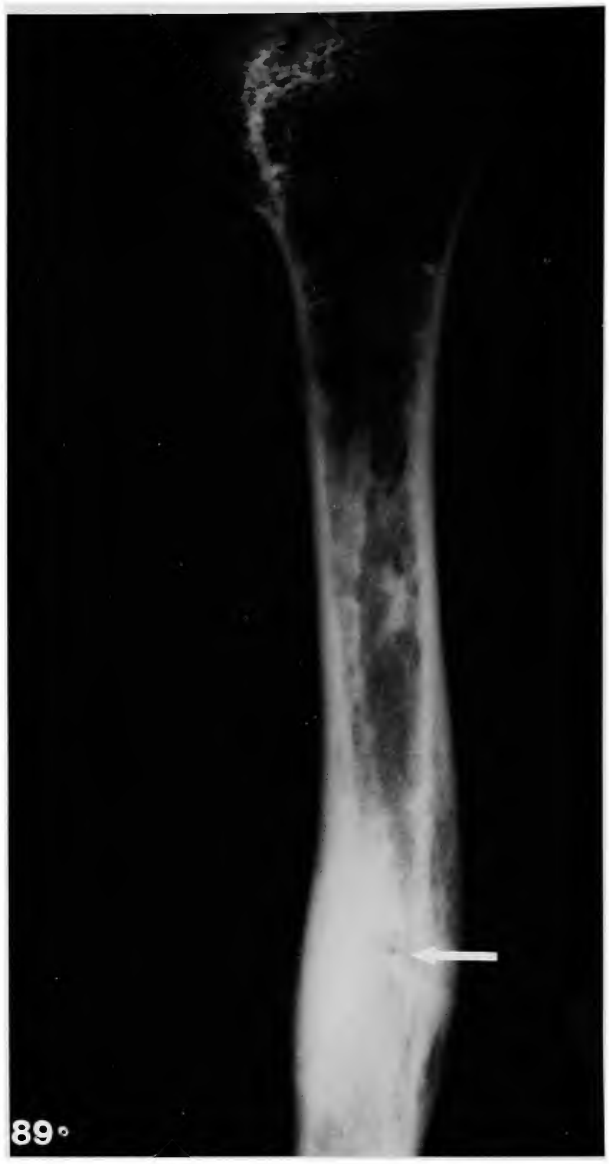


FIGURE 87.—Compressed fracture of a first lumbar vertebra in a male specimen from the Saxon period at Cathedral Green, Winchester, England. Anterior view of lumbar vertebrae and sacrum. Note evidence of periosteal reactive bone on vertebral bodies. (BMNH CG66, XXIII, layer IV.)

(p. 95), although fracture is often the event that initiates the surgical procedure of trephination. Most of the fractures to the skull appear to be related to intentional violence rather than accident. An example of a depressed fracture in an adult male skull vault is seen in Figure 78. This specimen (CG69, 697, 280 XXXIX) is from the medieval site at Winchester, England, and is stored at the British Museum (Natural History). The lesion is well healed, indicating long survival after the injury.

An example of a more massive, healed fracture is seen in an adult female skull from Cinco Cerros, Peru (NMNH 293818). The archeological age is unknown. There are two large, depressed fractures both of which are on the right parietal (Figure 79). One of these is located near the sagittal suture on the posterior portion of the right parietal. Radiating out from this lesion are two fracture lines, one of which crosses the sagittal suture well into the left parietal bone. The other depressed fracture is located in the central portion



FIGURES 88, 89.—Early stages of fracture healing and callus formation in the midshaft of an isolated left humerus from an Eskimo cemetery on St. Lawrence Island, Alaska, USA: 88, Note the porous nature of the recently formed callus. 89, X-ray film; note the radiolucent line (arrow) indicating the fracture line. (NMNH 364816-32)

of the right parietal, superior to the squamous portion of the right temporal bone. This lesion is deeper and more extensive than the first lesion, but it does not exhibit the long, radiating fracture lines. Both lesions are completely healed, indicating long survival after the injury.

A right parietal bone (NMNH 256414a) from the rock tombs at Lisht in Upper Egypt exhibits a circular lesion suggestive of a healed, glancing, sword wound (Figure 80). This specimen is dated to the Twelfth Dynasty. The outer boundary of the lesion is pitted, perhaps indicative of an inflammatory response. The sword wound apparently removed the outer portion of the parietal in

the region of the parietal boss, exposing the diploë in the central portion of the lesion and creating the porous appearance, which was partially filled in by subsequent remodeling.

Another specimen from the rock tombs at Lisht and dated between the Twentieth and Twenty-fifth Dynasty (NMNH 256384) illustrates a lesion resulting from a sword or axe wound, which occurred at the time of death. The wound penetrated the left parietal bone near vertex (Figure 81). The blow removed a portion of the right parietal bone (Figure 82). No postcranial bones are associated with this adult male skull.

An adult female skull from Pachacamac, Peru (NMNH 266584) has an extensive unhealed fracture of the left parietal bone (Figure 83). The archeological age of this skull is unknown. The focus of the trauma is the anterior portion of the left parietal, which exhibits a crushing injury with fracture lines radiating down the left parietal along the coronal suture. A fracture of the left zygoma could have occurred at the time of the skull fracture. Another fracture line proceeds posterolaterally across the sagittal suture and into the right parietal. There is no evidence of healing, indicating that the blow came at the time of death.

Three examples of fracture of the vertebral column serve to illustrate the essential features of such trauma in archeological specimens. The first of these is seen in the second lumbar of an adult male from the Hawikuh site in New Mexico, USA. This is a multicomponent site dating from the late prehistoric to early historic period. The anterior portion of the second lumbar vertebral body is crushed (Figure 84). There is fusion with the left lateral and central portions of the inferior border of the first lumbar vertebra and contact between large, bony projections from the inferior L2 and superior L3 vertebral bodies. The second example of fracture of the vertebral column is a male sacrum from Lisht in Egypt dated between the Eighteenth and Twenty-first Dynasty (NMNH 252874). The fracture occurs in the third sacral segment and radiates laterally from the superior portion of the spinous process (Figures 85, 86). The fracture involves only a portion of the posterior segment and was in the early stages of healing at the time of death. Evidence of early callus formation can be seen in Figure 86. The third example of vertebral fracture is from the Saxon period at an archeological site in Winchester, England (BMNH CG66, XXIII, layer IV). The first lumbar vertebra is wedge-shaped on the left portion of the body (Figure 87). There is bony bridging between the right sides of the first and second lumbar bodies and considerable rotation of the axis of L1. The fracture is well healed indicating survival after the trauma.

Fracture in the remaining postcranial bones is common in archeological specimens and should offer few problems in correct diagnosis. Evidence of a fracture line seen in an X-ray film will confirm such a diagnosis. This line will often be seen as a rather sharply defined radiodense line in the fracture callus. However, in some cases, particularly if the individual died within a few months following the trauma, the line may be radiolucent. An example of the latter is seen in an isolated left humerus from an Eskimo burial on St. Lawrence Island off the coast of Alaska (NMNH 364816-32). The archeological age of this specimen is uncertain. The fracture is located near midshaft with evidence for the fracture seen, externally, as a porous bony callus (Figure 88). The X-ray film appearance shows a general increase in radiodensity resulting from the callus added to the cortical bone. However, the radiolucent fracture line is apparent in Figure 89.

Complications of Fracture

Severe pain undoubtedly accompanies any attempt to reduce a fracture. Thus, even in those time periods and areas where fractures could be treated, it is not surprising to find numerous examples of malaligned, but healed, fractures in archeological skeletons. Wood-Jones (1910c) reports many such examples in his review of fractures in ancient Nubian skeletons. Wells (1974a) finds similar evidence in Anglo-Saxon England. Perhaps the important point to be made regard-

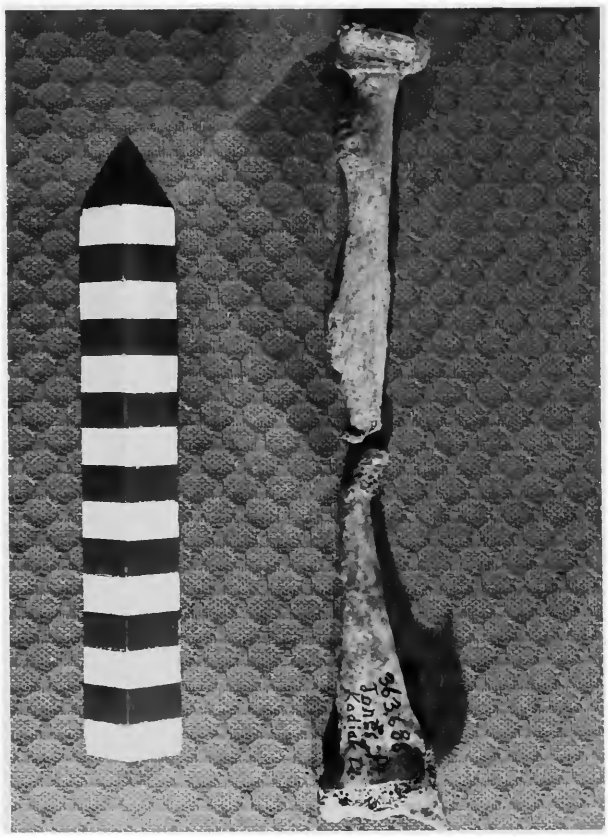


FIGURE 90.—Nonunion of a midshaft fracture of an isolated left radius. Specimen is from an archeological site at Jones Point on Kodiak Island, Alaska, USA. (NMNH 363688; scale in cm.)

FIGURES 91, 92.—Fracture and nonunion of the left femoral neck in an adult male skeleton from an Eskimo archeological site on Golovin Bay, Alaska, USA: 91, The fractured femoral neck has been pushed into the proximal metaphysis. 92, Detail of sclerotic reaction of the broken femoral neck and the corresponding cavity of the femoral metaphasis in above specimen. (NMNH 333453.)

ing these deformations is that individuals having them generally continue to live and somehow compensate for the dysfunction that results.

The failure of the broken ends to unite at all is more serious than malunion, primarily because of even greater malfunction, particularly if this condition occurs in a major long bone. Stewart (1974) has summarized the evidence for nonunion in archeological populations and published five cases from the ancient New World. He concludes (1974:878) that in both Egypt and America the forearm is the most common site of nonunion. This probably reflects the high frequency of fractures at this site. He notes that ulnar nonunion is higher in Egypt but nonunion of the radius is more common in New World material (Figure 90). In both geographical areas the incidence of nonunion is low.

In Stewart's survey all the examples of nonunion are from the upper extremity. While this probably approximates real conditions, it is important to note that nonunion does occur in the lower limb. In two cases I have studied (NMNH

FIGURE 93.—Fracture and nonunion of the left femoral neck in an adult male skeleton from Lisht in Upper Egypt dated from the Twelfth Dynasty. The fragmentary remains of the femoral head and neck are seen to the left. Note the extensive development of callus and the sclerotic remodeling in the cavity of the proximal femur. (NMNH 256580.)

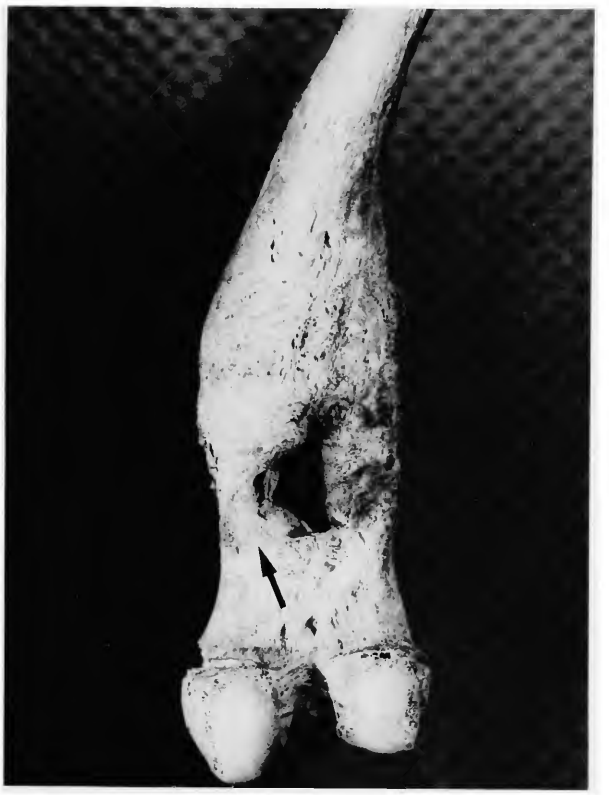


FIGURE 94.—Healed compound fracture complicated by infection in the right femur from a young adult female skeleton excavated from an archeological site near Akron, New York, USA. Note the deviated axis of the distal shaft at the fracture site. There is a large cloaca and reactive bone (arrow) probably associated with a fistula. (NMNH 326853.)

333453 and 256580) the fracture and nonunion occurred in the neck of the femur. In both cases there is evidence of continued use of the limb with the fractured end of the femoral head and neck becoming embedded in the marrow space of the greater trochanter and proximal shaft (Figure 91). One of these two examples (NMNH 333453) is from an Eskimo site in Golovin Bay, Alaska. The skeleton is that of an adult male between 20 and 25 years of age. The archeological age is uncertain. The fracture occurred through the neck of the left femur. The X-ray film of both femora reveals considerable osteoporosis of the left femur shaft and diminished density of the nonunited femoral head, particularly in the medial half of the femoral head. The latter is suggestive of impaired circulation. There has been considerable remodeling activity on both sides of the fracture site (Figure 92). The second specimen (NMNH 256580) is from the site of Lisht in Egypt and is dated to the Twelfth Dynasty. The fracture occurred in the femoral neck of the left femur. There is exuberant callus formation around the fracture site (Figure 93) with extensive remodeling in the bone adjacent to the fracture. The individual was an old male and exhibits excessive osteoporosis of the fractured femur. The nonunited femoral head is damaged by postmortem breakage. In these examples of nonunion the weight of the body tends to force the broken end

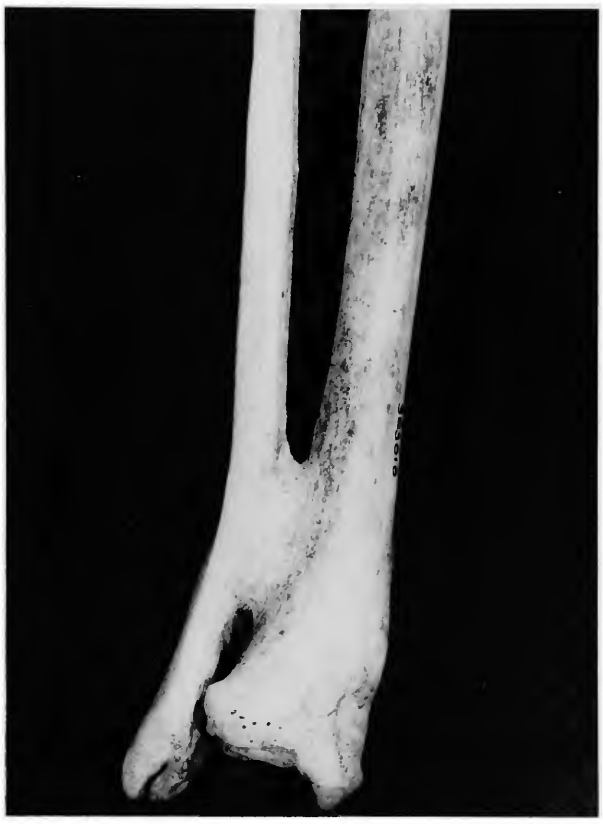


FIGURE 95.—Bony fusion of the distal tibia and fibula following fracture. (NMNH 383010.)

FIGURE 96.—Hypertrophic bone development, probably due to traumatic myositis ossificans, on the left femur of a *Homo erectus* (Pithecanthropus) skeleton from Java. (NMNH 362452, cast of original specimen.)

of the femoral neck into the proximal shaft of the femur. This condition and the fibrous connective tissue undoubtedly present in the living individual permitted some use of the limb, but some disuse atrophy is evident in both specimens.

Of the fractures occurring in archeological skeletons, very few show evidence of infection. In part, this observation must reflect the low incidence of compound fractures that would permit the passage of infectious organisms to the fracture site. Hamilton's data (1853), from a 19th-century American city population, summarized earlier in Table 1 indicate that 19 percent of the fracture cases in his sample are compound. Of all the compound fractures of the bones 63 percent involve the tibia and fibula. Thus, the lower leg is by far most vulnerable to compound fracture in the population.

Infectious complication of fracture is not limited to compound fracture nor will all compound fractures become infected. However, the incidence of compound fractures gives some idea of the probabilities for infection of fractures in archeological populations. Wood-Jones (1910c) cites two examples of fracture complicated by infection. One is a clavicle (1910c:306), the other an ulna (1910c:313). He makes no mention of infection in fractures of the tibia or fibula. Indeed, fractures of the tibia of any kind are less common in archeological populations than modern civilized groups. Hooton (1930:312) found, among the prehistoric Pecos skeletal sample from New Mexico, USA, that the humerus and femur were the most commonly fractured long bones. He states that some fractures show a lack of union,

although he does not indicate whether this is due to death occurring shortly after fracture or to nonunion. Hooton also indicates that some fractures seem to have suppurated, indicating that they were complicated by infection. In the collections of the National Museum of Natural History, Smithsonian Institution, USA, there are several examples of fracture complicated by infection. These cases tend to show marked deformation suggesting considerable traumatic force, possibly indicating penetration of the skin by the fractured bone. One example of this complication of a compound fracture is seen in the skeleton of a young adult female from an archeological site near Akron, New York, USA (NMNH 326853). The archeological age is uncertain. The fracture is located in the distal shaft of the right femur. There is considerable axial deviation, as well as some rotation. The callus is massive with considerable porosity. On the posterior portion of the callus there is a large, oblong cloaca measuring 15 by 25 millimeters (Figure 94). There is bony evidence of a fistula, which would have extended to the skin surface. The tibia of the same leg was also fractured but with good alignment and no evidence of infection.

In archeological populations one finds arthritic deterioration occasionally associated with fracture. Although not specifically addressing this problem, Wood-Jones (1910c:308, 312, 315, 317, 318, 322) cites several examples of fracture contributing to the development of arthritis. Morse (1969:100, 104) cites similar conditions in archeological specimens from the midwestern United States. Making the distinction between traumatically induced arthritis and degenerative arthritis depends on two factors: (1) demonstration of fracture in the affected bone and (2) lack of bilateral symmetry in the pattern and degree of arthritis (Morse, 1969:13).

Trauma to adjacent bones may also result in ankylosis. An example of this condition is found in an historic period archeological specimen from South Dakota, USA (NMNH 383010), in which the distal tibia and fibula are fused in the area of a fracture (Figure 95). Clearly such an injury and its sequelae could have affected locomotion. However, the long-standing callus and extensive remodeling indicate long survival and an active life after trauma.

The last factor complicating fracture to be discussed in this review is myositis ossificans. Perhaps the most famous example of this condition is the bone excrescence on the fossil left femur from Java (Figure 96) associated with *Homo erectus* (Pithecanthropus). In this specimen there is an exuberant growth of bone on the posterior aspect of the femur extending from the insertion of the adductor muscles. Wood-Jones (1910c:317) has published a line drawing of a right femur from Nubia with fracture of the femoral neck. There is an extensive, bony spur projecting medially, which Wood-Jones attributes to extensive callus. The extent and shape of the bony mass suggests ossification in muscle tissue rather than exhuberant callus formation. However, the distinction between the two may be academic. While the Java and the Nubian specimens have the bony growth attached, the paleopathologist should be aware that ectopic bone can develop in muscle tissue and be completely unattached to any bone.

Dislocation

Pathology

The second major category of trauma involving the skeleton includes luxation, in which the joint constituents are out of contact and the joint capsule is disrupted. Subluxation is a less severe condition characterized by partial loss of the contact between joint components, but no disruption of the capsule. Both these conditions can be associated with fracture but often occur without fracture. Luxation and subluxation can occur in most joints. However, to be detected in an archeological specimen there must be permanent change in the skeleton. This means that subluxation will rarely be identified and luxation will be seen most often in joints where the anatomy makes spontaneous correction of the problem difficult. The two joints particularly prone to

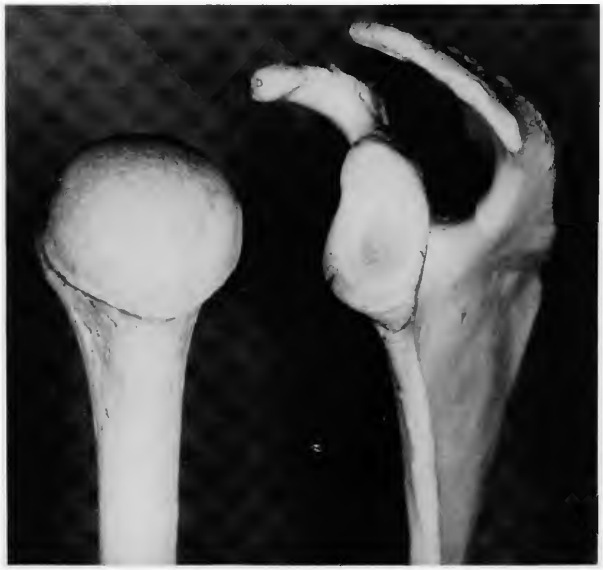


FIGURE 97.—Morphology of the components of a normal left shoulder joint. (Specimen from the Terry Collection (NMNH 382085-1600).

FIGURE 98.—Long-standing inferior dislocation of the right hip. A new acetabalum has been formed in the area of the obdurator foramen. The original acetabulum (arrow) shows considerable atrophy. (FPAM 3939.)

chronic or permanent luxation are the shoulder and hip and I will focus on these to illustrate the problems associated with this type of trauma.

The anatomy of the shoulder joint permits maximal movement of the arm in virtually all axes (Figure 97). This degree of movement is highly useful for the manipulative functions of the arm and hand. However, the biological trade off for this mobility is a relatively unstable joint. Rockwood (1975:624) notes that the glenohumeral joint is the most commonly dislocated major joint in the body.

The bony components associated with the shoulder joint are the scapula, humerus, and clavicle. The round head of the humerus articulates with the glenoid cavity of the scapula. However, unlike the acetabulum of the hip, the glenoid cavity is little more than a slight depression, so that the humeral head rests against the joint surface but receives little support from it. The major support for the glenohumeral joint is the ligament and capsule attaching the humeral head to the scapula. Overlying these are the muscles of the rotator cuff and the deltoid, which also help to stabilize the joint. However, all these supporting structures are loose to permit optimal movement and thus can do little to prevent traumatic dislocation.

Rockwood (1975:641) indicates that indirect force is the most common cause of glenohumeral dislocation. The mechanism by which the force is applied involves sudden, extreme movement of the upper arm away from the body (abduction) in association with external rotation as could happen in a backward fall in which the hands are used to break the impact. The effect is to drive the humeral head into the anterior capsule. Forces associated with this traumatic effect frequently produce a compression fracture as the edge of the glenoid cavity is forced into the head of the humerus (Rockwood, 1975:642). According to Rowe (cited in Rockwood, 1975:641), 96 per-

cent of dislocations were traumatic in origin, 4 percent are nontraumatic. Congenital defects in the bony structure or supporting ligaments and muscles appear to play a minor role in traumatic and nontraumatic dislocations.

Because of the anatomy of the glenohumeral joint, spontaneous reduction of the dislocation often will not occur. Rockwood (1975:625–627) notes that the methods for reducing shoulder dislocations go back at least to the time of Hippocrates (ca. 430 B.C.). Thus, it seems possible that the treatment of dislocations could have existed in many archeological populations. However, it is also clear that unreduced dislocations do occur in archeological populations. In such situations a new joint surface is created where the humeral head comes in contact with the scapula and provides clear evidence of dislocation.

While the anatomy of the hip permits considerable movement, its structure is influenced by its major weight-bearing function. In contrast with the glenohumeral joint, the hip is a classic ball and socket joint with the head of the femur deeply extended into the hip socket or acetabulum. While the ball and socket arrangement of the bony components suggests a more stable arrangement than is found in the glenohumeral joint, it is important to remember that the ligaments and muscles associated with the hip joint provide relatively less support than analogous structures in the shoulder.

Unlike the glenohumeral joint, the hip joint components are joined directly by a ligament (ligamentum teres), which also contains the vascular supply for that portion of the femoral head adjacent to the ligament. Dislocation of the femoral head thus threatens the vascular supply of part of the femoral head.

In congenital hip dysplasia, the acetabulum is abnormally shallow, creating a tendency for the femoral head to slip out of the socket in response to slight trauma or even walking. Such a condition may be recognizable in skeletal material due to the abnormally shallow acetabulum, although the tendency for the acetabulum to be remodeled after traumatic dislocation may make it difficult to make this distinction. The mechanism for traumatic hip dislocation varies, depending on the position of the leg and the direction of the force. Depending on these factors, dislocation of the femoral head may occur in an anterior or posterior direction with the latter being the most common in modern orthopedic practice (H. Epstein, 1973:116). Reduction of hip dislocations is more difficult than reduction of the shoulder. The failure to reduce the dislocation will result in the formation of a secondary articular surface on the innominate (Figure 98). Since spontaneous reduction is less likely, identification of this type of trauma to the hip in archeological material should be fairly easy and should more accurately reflect the actual incidence of dislocations in the archeological population than evidence of shoulder dislocation.

I have reviewed dislocation of only two of the major joints for illustrative purposes. The student of paleopathology interested in this subject should consult the orthopaedic source materials for a more comprehensive treatment. However, these two examples of dislocation reveal several principles that are associated with all dislocations: (1) Although there may be congenital factors involved, trauma is by far the most common cause of dislocation; (2) dislocation may be associated with fracture; (3) unreduced dislocation creates a secondary joint surface; (4) the evidence for dislocation in archeological samples reflects only a portion of the actual incidence of this type of trauma; (5) the blood supply to the bony part of the joint may be disrupted by dislocation, resulting in tissue death (aseptic necrosis); and (6) the trauma associated with dislocation may also tear the ligaments and muscles associated with the joint.

Paleopathology

Dislocation of the shoulder and hip should come to the attention of the paleopathologist more often than other types of dislocation, because of high frequency and problems in reduc-

FIGURES 99, 100.—Anterior dislocation of the left shoulder in an archeological specimen from Norton Bay, Alaska, USA: 99, The humeral head articulates with a new joint on the anterior surface of the scapula; there is some degeneration of the anterior glenoid fossa. 100, Secondary joint formation on the scapula; note porous surface of new joint surface and arthritic degeneration including eburnation (arrow) on the humeral head. (NMNH 346205.)

FIGURE 101.—Anterior dislocation and secondary joint formation of the left scapula in a male skeleton from Kodiak Island, Alaska, USA. Part of the glenoid fossa has been remodeled away and a new joint surface has been formed on the remodeled surface (arrow). There is considerable degenerative arthritis on the humeral head. (NMNH 363615.)

tion, which make permanent dislocation more likely. In his survey of the Nubian skeletal materials, Wood-Jones (1910c:341) reports that evidence of dislocation is rare, with only one hip dislocation in several hundred skeletons. He cites other cases but admits that the diagnosis is dubious. One case described by Wood-Jones is of an abnormal forearm (case 7:6) but probably is not a dislocation but a congenital malformation. Elliot-Smith appends to Wood-Jones report (1910c: 342) a case of a traumatic knee dislocation associated with a fracture.

I have studied several cases of dislocation found in the collections of the National Museum of Natural History. Most of the shoulder dislocations involve anterior luxation. One example of this type of luxation is from an adult male skeleton from Norton Bay, Alaska, USA (NMNH 346205). The archeological age is uncertain. The left humeral head is completely displaced and articulates with the anterior surface of the left scapula (Figure 99). Continued use of the arm has resulted in the formation of a secondary joint on the scapula and arthritic degeneration and eburnation of the humeral head (Figure 100). Another example is from an adult male skeleton from Kodiak Island off the coast of Alaska, USA

(NMNH 363615). The archeological dating is uncertain. The dislocation in this specimen also involves the left shoulder. However, the luxation was not as severe, having stimulated a secondary joint on the anterior portion of the glenoid fossa (Figure 101). There is considerable arthritic degeneration on the humeral head. One example of posterior dislocation of the shoulder was found in a disturbed burial from an Early Bronze Age cemetery associated with the town site of Bab edh-Dhra in Jordan (Figure 102). A secondary joint was created on the posterior half of the glenoid fossa. There is evidence of arthritic degeneration and eburnation on the secondary joint. The humerus was not recovered.

Figures 103 and 104 illustrate fairly typical changes associated with hip dislocation. The skeleton is from Kodiak Island, Alaska, USA (NMNH 372897), its archeological age is uncertain. Figure 103 shows the effects of the dislocation on the right innominate, comparing it with the normal left innominate. In both innominates the acetabulum is somewhat shallower than normal, suggesting a possible congenital predilection for dislocation. Figure 104 provides a detailed view of the secondary joint on the innominate

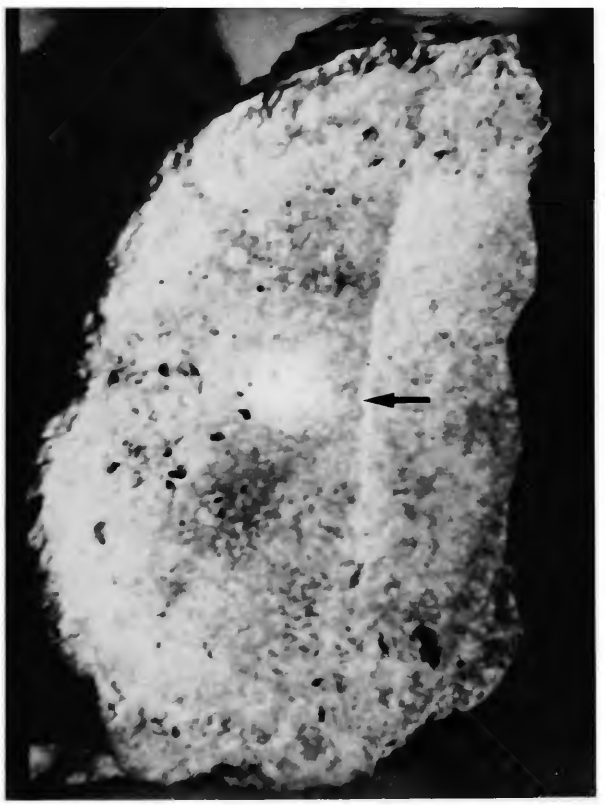
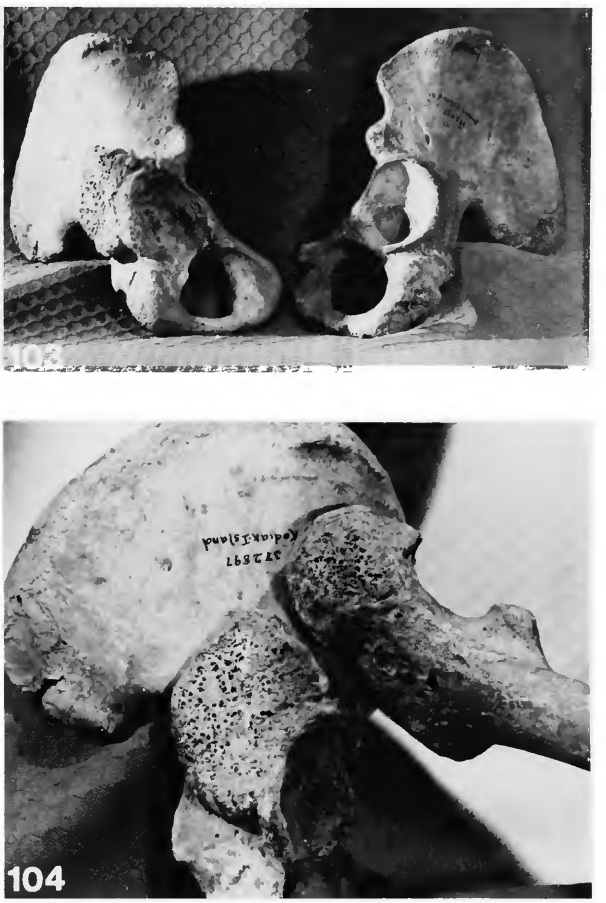


FIGURE 102.—Posterior dislocation of the right shoulder in an isolated scapula from the Early Bronze Age site of Bab edh-Dhra in Jordan. The arrow indicates the ridge between the original joint surface to the right and the new joint formed after luxation. The new joint is porous and eburnated. (NMNH, no number.)



FIGURES 103, 104.—Luxation of the right hip (left side of figure) with secondary joint formation in an adult male skeleton from an archeological site at Jones Point, Kodiak Island, Alaska, USA: 103, The acetabulum is shallower than normal in both innominates. 104, Detailed view of porous, secondary joint surface on the right innominate of above specimen; note arthritic change on associated femoral head. (NMNH 372897.)

and the degenerative changes on the femoral head.

Deformation

Pathology

Thus far, I have largely focused on the effects of sudden trauma on the skeleton. However, chronic, low-grade trauma over an extended period of time can also modify the normal shape of bone. Such deformities are often, but not always, intentional. The basic principal to be emphasized is that any long-term, deforming pressure brought to bear on a bone or group of bones can result in permanent deformity, particularly if it occurs during the growth period.

In relatively recent times perhaps the best known intentional deformity was the foot-binding practice of upper class Chinese women. In this practice the feet of female children were bound in such a way that the metatarsals were bent downward and compressed toward the heel of the foot. This created abnormal angulation of the tarsal bones and abnormal relationships between all the bones and joints of the foot (Figure 105).

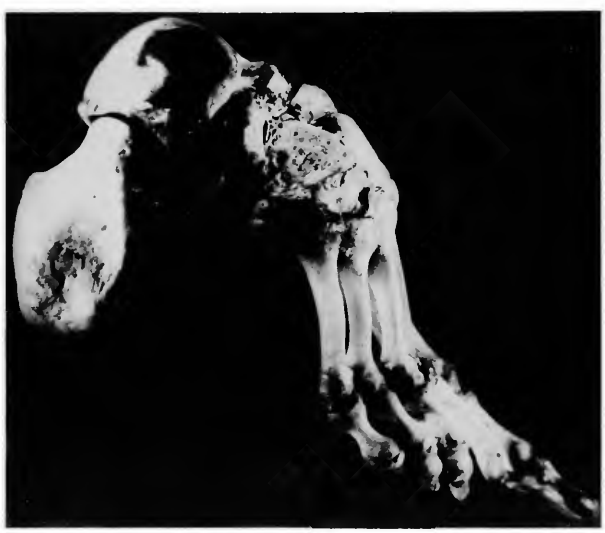


FIGURE 105.—Lateral view of artificially deformed, right foot in an adult, Chinese female resulting from foot-binding. The arch of the foot is greatly exaggerated with major changes occurring in the metatarsals. (WM S17.1.)

In modern skulls one occasionally finds the skull vault to be asymmetrical or the occipital bone flattened; such deformation may be intentional. In the modern skulls, however, there is no evidence of intentional pressure on the skull. Probably this deformity is the result of the individual sleeping in the identical position on a relatively hard surface during infancy and childhood. Such deformation illustrates the fact that slight, intermittant pressure over a long time is adequate to produce a gross deformity in a bone.

Paleopathology

Dingwall's review (1931) of cranial deformation indicates that artificial modifications of the skull was one of the most ubiquitous cultural practices in antiquity, being found on every continent except Australia. A comprehensive review of cranial deformation and other forms of artificial modification of bony structure is beyond the scope of this book. It is important, however, that cultural deformation not be confused with pathological processes, such as osteomalacia, which also can result in bones of abnormal shape.

While several methods of producing cranial deformation exist, most modifications of skull shape are the result of applying pressure to various areas of the skull, namely: (1) the occipital region, (2) the frontal region, (3) both frontal and occipital regions (Figure 106), and (4) along a transverse axis, approximately passing through vertex, the mastoid region, and the region just above the insertion of the nuchal ligament on the occipital bone. This last type of pressure, utilizing circular bands, produces the circular or Aymara type of deformity (Stewart 1941:343) and proportionately lengthens the skull (Figures 107, 108).

The reason for the practice of cranial deformation is universally cosmetic and is frequently associated with physical manifestations of high social status. The origins of such practices are buried in the unknown past of the people who induced the deformation. Snorrason (1946) hypothesizes that abnormal, artistic representations

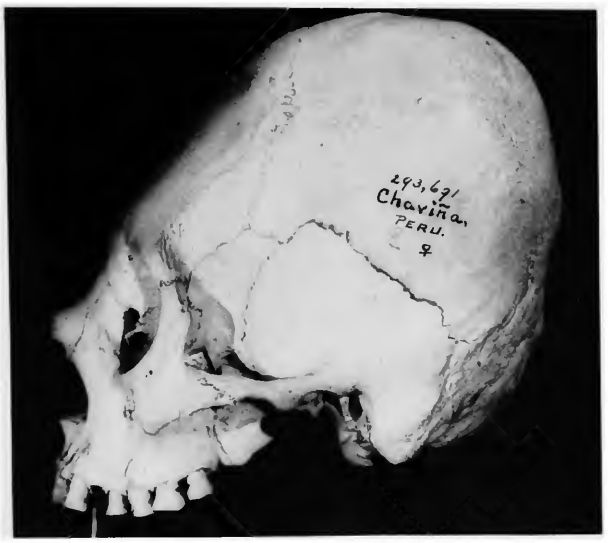
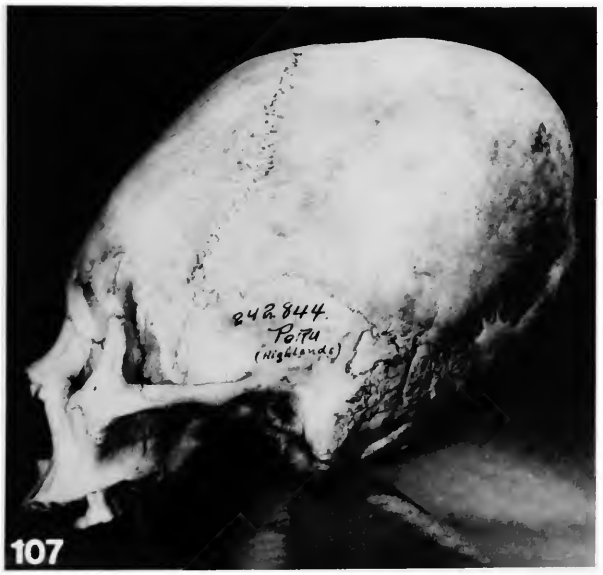
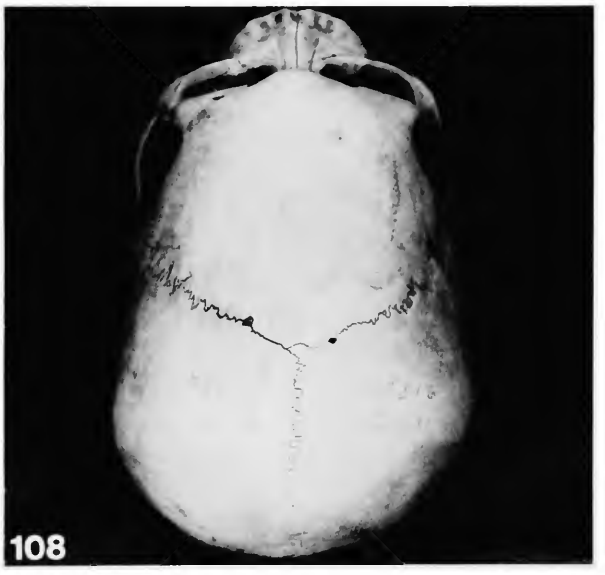


FIGURE 106.—Fronto-occipital artificial skull deformation in an adult female skull from an archeological site near Chavina, Peru. (NMNH 293691.)

of heads during the reign of Akhenaten was due to the abnormal shape of the pharoah's head, perhaps resulting from acromegaly. Dingwall (1931:102) reports that cranial deformity in Egypt is unknown prior to the Eighteenth Dynasty and the reign of Akhenaten. The purported skull of Akhenaten (Dingwall, 1931, pl. VIa) does show a rather long, low profile, particularly with regard to other skulls of that period from Egypt. Dingwall (1931:104) cites the opinions of Elliot-Smith and Ferguson that the skull shows signs of hydrocephaly.

The suggestion is that the unusual morphology of the pharoah's head led to the establishment of a new and temporary fashion, and that people deformed their childrens' heads to conform with this fashion (Dingwall, 1931:110). It must be emphasized that the evidence for this cranial deformation is limited to artistic representations





FIGURES 107, 108.—Circular or Aymara type of artificial skull deformation, which results from circular constricting bands wrapped around the posterior half of the skull during the early growth phase of the individual: 107, Lateral view. 108, Top view showing somewhat elongated aspect of the deformed skull. (Specimen from Ancon, Peru, archeological age unknown, NMNH 242844.)

and not observed on skulls. In spite of the speculative nature of this hypothesis the general influence of Akhenaten's appearance certainly influenced at least the portrayal of people during his reign—a practice that soon disappeared after his death (Snorrason, 1946:601).

The broader implication of this account is the possibility that leaders with unusual skull morphology from natural causes might have given rise to a fashion in some societies which, unlike the case of Akhenaten, became a permanent part of the cultural tradition.

The complications of cranial deformation are mainly cosmetic. However, Stewart (1948:71) suggests that compression may disrupt the normal growth that takes place at the skull sutures and can give rise to a depression in the sagittal region. This depression creates a bi- or tri-lobed appearance in the skull and could be confused with the results of a therapeutic practice producing the sincipital-T deformation (p. 102). McGibbon (1912) and Moss (1958) report minor anatomical abnormalities that are associated with cranial deformation. However, Moss (1958:248) emphasized that the magnitude of growth is not reduced. This suggests that the practice probably did not produce any serious problems.

Scalping

Pathology

Traumatic avulsion of the scalp may be the result of accident or intentional violence. A well-documented case of accidental scalp avulsion illustrates the gross morphological features of this trauma. The specimen is from the Pathology

Figures 109, 110.—Accidental scalping in a female industrial worker from the early twentieth century: 109, The irregular external surface of the parietal bones is the bony response to infection and proliferation of granulation tissue during a 8-month period after the accident and before death due to septic meningitis. The arrow indicates the anterior margin of the lesion. 110, The inner table showing the region of the sagittal suture. Note the porosity indicative of an inflammatory response in this area. (PMES 1.EB.1.(6).)

Museum of the Royal College of Surgeons of Edinburgh (PMES 1.EB.1.(6)) and dates from the early twentieth century. The specimen consists of the skull and mandible of a female jute weaver, aged 55 years. Her hair had been caught in the machinery, tearing the scalp from the parietal region but leaving it suspended by its frontal attachment. The scalp was replaced and sutured but became infected, leading to extensive sloughing and proliferation of granulation tissue. The infection led to septic meningitis and death occurred eight months after the injury. The outer table of the skull exhibits the irregular surface typical of the bony response to granulation tissue, as well as marginal porosity indicative of hypervascularity and inflammation. These changes are more extensive on the left parietal bone (Figure 109). On the inner table there are many small pores along the sagittal sulcus, reflecting hypervascularity in this region as well (Figure 110). This case provides evidence on the extent of changes that can occur in eight months. Thus, relatively short-term survival after scalping might be expected to produce changes in the skull that could be identified in paleopathological specimens.

Paleopathology

Reese's (1940) review of the history of scalping indicates considerable antiquity for this practice, with the earliest reference found in the fourth book of Herodotus (485–425 B.C.). The second book of the Maccabees (ca. 67–37 B.C.) describes the practice of scalping of living captives in Palestine. Reese reports that the Visigoths, Anglo-Saxons, and Franks practiced scalping, and it is noted among the North American Indians in 1520 just 28 years after the European discovery of the New World.

The question of whether the practice of scalping was introduced into the New World by the European colonists remains unresolved. Neumann (1940) reports a skull from central Illinois dated around A.D. 1000–1500, which has cut marks suggestive of scalping. Hamperl and Laughlin (1959:81) conclude that "there is little evidence for the antiquity of the practice in the United States."

FIGURE 111.—Probable example of scalping in a pre-Columbian adult female skull from the Sea Island Mound in Georgia, USA. The irregular surface of the skull vault is virtually identical with that seen in Figure 109. The anterior margin of the injury is indicated by the arrow. (NMNH 379000.)

Possible evidence for the practice of scalping in pre-Columbian North America is seen in an adult female skull from the Sea Island Mound in Georgia, USA (NMNH 379000). The mound from which this skull was excavated is dated about A.D. 1000. Although the possibility that the burial was intrusive cannot be excluded, such a possibility does not appear likely. The skull is well preserved and normal except for a large lesion involving the frontal and both parietal bones (Figure 111). The boundaries of the lesion are most pronounced on the frontal and anterior parietals. There is no evidence of active inflammation. All aspects of the lesion suggest a response to granulation tissue, but with good recovery and no infection at the time of death. There is no evidence of inflammation on the inner table. The morphology of

the lesion is compatible with a diagnosis of scalping.

Regardless of the origin of scalping, it is well documented for many of the Indian tribes in various geographical regions of North America in early Colonial times. Catlin (1844:239–240) reports that the Colonists would pay for scalps of enemies both White and Indian. Indeed, he notes that a knife commonly used for scalping was made in Sheffield, England, and traded to the Indians for horses (Catlin, 1844:236).

The scalp was removed to provide evidence of having killed an enemy. Nadean (1941:181–182) indicates that some tribal variation existed regarding the amount of scalp taken. Catlin (1844: 30–31, 238) indicates that the scalp taken from "the place where the hair radiates from a point" was most important but that scalp from other areas would be used to decorate clothing in some tribes (e.g., Blackfoot). In removing the scalp, the skin would be cut around the desired area and pealed off. The initial cutting may have left marks on the skull whether or not the victim was dead (Owsley, Berryman, and Bass, 1977).

Catlin (1844:239), Reese (1940:16), and Nadean (1941:193) report that survival after scalping did occur. Reese (1940:18–19) included in his paper a report by a physician who treated a case of scalping by Cheyenne Indians in 1867. The report indicates that the scalp was removed, the periosteum covering the skull cut and badly damaged. The outer table was exfoliated (shed), after which the wound healed. Hamperl and Laughlin (1959:88), in commenting on this case, suggest that the destruction of the periosteum deprived the outer table of its superficial blood supply. This resulted in necrosis with granulation tissue in the diploë eventually separating the necrotic tissue from the skull. An archeological specimen showing a lesion probably reflecting this same process is reported by Hamperl and Laughlin (1959).

Mutilation

Among historical preliterate groups mutilation of the body involves the soft tissues, teeth, and bone. In this section we shall only review mutilations of bone. Dental mutilations will be discussed, but soft tissue mutilations are beyond the scope of this book.

Mutilation affecting bone may be voluntary, in which the victim either performs the mutilation on himself or willingly allows it to be performed by someone else. Involuntary mutilations are usually associated with punitive action against a criminal or a captive. Moodie (1920: 1300) has published Aurignacian (ca. 20,000 B.P.) cave art from Gargas, Spain, suggesting finger amputations in that period. He further reports (1920:1303) the existence of finger mutilation throughout the world. In the Old World, the most commonly reported reason for finger mutilation is as a symbol of mourning. Among the North American Indians it was part of initiation rites.

Major portions of one or more limbs have also been removed. Brothwell and Møller-Christensen (1963b) report a case of amputation with subsequent healing of the stump of the distal right forearm from Egypt dated 2000 B.C. They offer three possibilities for the cause of the mutilation: (1) injury during warfare, (2) punitive action against a criminal or captive, and (3) surgical amputation for therapeutic reasons. While punishment for criminal behavior remains a possibility, Aldred (1964:56) challenges the likelihood that the Egyptians would have cut off the hand of a captive. He notes that to do so would have deprived the captors of a valuable spoil of war: the slave labor. He further notes that amputation of the hand was limited to the dead and dying victims of warfare and was used as a method of recording the dead from the field of battle. In another report Brothwell and Møller-Christensen (1963a) described an amputation of the distal left forearm in a skeleton from Sicily dated to the seventh century A.D. The ends of the radius and ulna have fused, indicating healing. They conclude that the amputation was punishment for a criminal act—a practice common also in that age. Stewart (1950:43) reports the existence of bones showing amputations from Peru but does not indicate which bones are involved.

While there seems to be convincing evidence that both voluntary and involuntary mutilation of bony parts of the skeleton did occur in antiquity, a word of caution is appropriate regarding unequivocal interpretation. With respect to missing fingers or toes in an archeological specimen, at least two other conditions could be implicated. These are infectious disease (particularly leprosy) and frostbite. Amputation of the distal forearm or distal leg should also be reported with caution. Stewart (1974:889–890) calls attention to the morphological similarity between the skeletal manifestation of pseudarthrosis and what might be expected in an amputation. He suggests that bony fusion of the distal radius and ulna in the case reported by Brothwell and Møller-Christensen (1963b) might be more reasonably explained by fracture and nonunion. Diagnosis in this case is complicated by the fact that the fused radius and ulna are isolated bones, raising the possibility that the distal portion of the specimen, if it was a case of nonunion, had not been recovered.

Trephination

History

Perhaps the most remarkable trauma seen by the paleopathologist is trephination or the surgical removal of a portion of the skull. The earliest written accounts of trephination are found in the Hippocratic writings (460–377 B.C.). However, the evidence of trephining goes back at least to the Neolithic period (Moodie, 1919:484; Guiard, 1930; Parry, 1931; Hrdlička, 1939; Stewart, 1958b:470; Oakley, Brooke, Akester, and Brothwell, 1959:93). It continues to be practiced among many recent, primitive peoples (Ruffer, 1918a: 99–102; Freeman, 1918:445; Moodie, 1919:485; Hrdlička, 1939; Oakley, Brooke, Akester, and Brothwell, 1959). The geographical distribution of the practice is extensive, including Europe, the Pacific, North Africa, the Middle East, and South America (Stewart, 1958b:470–480). Wood-Jones (1910c:299) states that trephining has not been reported in ancient Egyptian skulls. However, he saw no reason why trephined skulls should not have been found in Egypt in view of the contact with Greece where it was practiced in antiquity. Ruffer (1918a:95) published a photograph of a purported trephination from Egypt. Moodie (1919:485) has expressed reservations regarding Ruffer's case and further states (p. 494) that trephining did not occur among Greeks, Romans, Phoenicians, Babylonians, or Egyptians. More recently Oakley, Brooke, Akester, and Brothwell (1959:95) report a case of trephination from Tarkhan, Egypt, dated to early dynastic times. Clearly the Greeks of the Periclean Age performed trephination (Withington, 1927:3) and the strong evidence of trephination in Palestine suggests that the practice occurred in Middle East countries. Mallin and Rathbun (1976) have reported a case of trephination in ancient Iran. In any case, the practice in the Middle East appears to be relatively rare, certainly by comparison with South America, where over 1000 cases of trephining have been found (Stewart, 1958b:474).

There is debate regarding the practice of trephination in pre-Columbian North America. Stewart (1958b:476–479) surveyed 17 purported cases and concluded that evidence for trephination in North America had not been demonstrated. One of the factors contributing to the controversy is the difficulty of conclusively identifying a trephination in an archeological specimen. Given a partial or complete hole in a skull there are several possibilities for the cause. Goldsmith (1945:348) suggests that some of the so-called examples of trephination are, in reality, cases of congenital, bilateral openings of the parietal bones (p. 352). This condition is a hereditary enlargement of the parietal foramina (Goldsmith, 1945:350). Other congenital conditions such as dysraphism (p. 351) can be confused with trephination. Both infection and neoplasm can produce holes in the skull, which might be confused with trephination. Injury such as a glancing sword or axe wound can remove a portion of the skull and produce a healed lesion similar to trephination. Furthermore, a trephine-like wound may not have been part of a surgical procedure on the living. Some groups apparently practiced postmortem removal of circular pieces of skull

(Ruffer, 1918a:91, Parry, 1931:1388–1389; Stewart, 1958b:475). Obviously, no healing would have occurred, but polishing of the edges by intentional or unintentional rubbing can create a superficial appearance of healing (Stewart, 1958b:477).

It is clear that the identification of the cause of holes in the skull cannot always be made. However, there are some criteria that will be important in any analysis. The first question to be resolved is whether the hole is the result of mechanical intervention. Evidence for cutting or scraping may still exist even if healing has taken place. The next problem to be clarified is if the cutting was done before or after death. The significant question here is whether we are seeing the product of a surgical procedure or the result of some postmortem ritual. Any evidence of healing or inflammatory reaction to the cutting is indicative of trephination. However, it is possible that a patient can die during the trephination or so soon after it that no reaction takes place. Such a situation in an archeological example of trephination may be indistinguishable from postmortem ritual removal. In such situations the demonstration of unambiguous trephination in the same geographical area would certainly be significant in interpreting the equivocal cases. Another important criterion is the presence of fracture in association with evidence of cutting. Many, if not most, trephinations occur in association with skull fracture. As with most other problems in paleopathology careful observation combined with a comprehensive knowledge of the options is the most important prerequisite in determining the presence of trephination.

In trephination, four basic responses can occur. First, in those cases where trauma preceded the trephination procedure, death may be due to the initial trauma. Second, the surgical procedure itself may cause death. Third, the surgical procedure may not directly cause death but may introduce disease organisms that cause infection and possibly death. Fourth, there may be no complications resulting from surgery, in which case the individual survives with varying degrees of repair to the surgically induced defect. If there is no healing, as evidenced by the lack of remodeling of the cut edges or fill in of exposed, diploic spaces, the reasonable assumption is that death occurred at the time of, or shortly after, surgery. A zone of porous, reactive bone surrounding the surgical area suggests survival for some time after surgery, but with the possibility of infection complicating the healing process and causing death. Partial to complete refill of the surgical defect is indicative of recovery and long-term survival after surgery.

Paleopathology

Freeman (1918:445) remarks, somewhat whimsically, that trephination in ancient populations was probably no worse "than the torture occasionally inflicted on us by our dentist." Nevertheless, the pain involved, as well as the threat to life inherent in the procedure, raise inevitable questions regarding the biological or cultural reasons for trephination. Broca (1876:572) suggested that trephination was performed as a cure for mental disorders. Ruffer (1918a:91) states that in Europe until the Middle Ages, powdered cranial bone was thought to possess curative properties. He also states that roundels (circular pieces of human skull) were worn as ornaments as late as Gallic times. Parry (1931:1388) asserts that Neolithic man performed trephination "as a religious rite ridding an affected individual of some demon or demons. . . . " He also indicates (1931:1388–1389) that amulets were made from the trephined skulls of deceased individuals, which included healed edges of the trephination. Stewart (1958b:481) suggests that this practice is limited mainly to Europe. More recently Oakley, Brooke, Akester, and Brothwell (1959:93) have supported the concept of the ritual significance of trephination, citing Albert Schweitzer's observation in modern African tribes that trephination is used to obtain a powerful fetish object.

While the evidence for religious implications in trephination appears convincing, it is equally clear that trephination was also a surgical procedure done to correct a serious medical problem.

The Hippocratic writings (Withington, 1927:3, 35) indicate that trephination was well known and practiced in the Golden Age of Greece (ca. 500 B.C.) for relief of the effects of skull fracture. The expertise of the technique revealed in the discussion suggests even greater antiquity for trephination as a treatment of skull trauma.

It should not be surprising to find trephination practiced for therapeutic treatment of skull fracture in other geographical areas and time periods. Moodie (1919:484) suggests that this is the reason for trephination in primitive societies. Hrdlička (1939:175) notes that "a very large proportion of aboriginal trephinings in America were true surgical or curative operations for the relief of the effects of head wounds or for that of actual or supposed pathological conditions within the skull." Stewart (1958b:481) concludes that relief of pressure from skull fracture was the most frequent reason for trephination in Peru and Melanesia. Oakley, Brooke, Akester, and Brothwell (1959:93) indicate a similar rationale for trephination practiced by medicine men in Bolivia in the 1950's.

Types of Trephination

Trephinations vary in terms of the extent of bone removed, the depth to which bone is removed, and methods used. The extent or area of bone removal varies with the size of the injury and the judgment of the surgeon. The depth of bone removal appears to reflect two possibilities: (1) the surgeon can remove the outer table of bone and diploë or (2) he can remove both tables of bone, exposing the dura. In ancient Greece, surgeons were conscious of the dangers of penetrating into the dura (Toole, 1964:91–92), and it is likely that such knowledge would quickly be acquired and disseminated.

The methods of trephination have been reviewed by several scholars (e.g., Parry, 1931, 1936; Hrdlička, 1939; Stewart, 1958b; Oakley, Brooke, Akester, and Brothwell, 1959). From these reports it is clear that the most widely used method was careful scraping through the bone with a sharp implement to the desired depth (Figures 112–114). This method undoubtedly involved the lowest risk of damaging the brain and is found in all geographical areas where trephination occurs. Other methods included drilling a circle of small holes and breaking or cutting the intervening bone and sawing or cutting directly into the bone (Figures 115, 116). Both these methods involved considerable risk to the brain. Eloquent testimony to this risk is seen in unhealed trephination of this type indicating that death occurred during, or shortly after, surgery (Figure 117). Initially the direct cutting type of trephination was thought to occur only in the New World; however, Parry (1936:170–171) reports this type of trephination in Palestine.

The surgical approach for trephination required that the overlying skin be cut, unless it was already cut by injury. Stewart (1958b:483) suggests that in some cases a portion of the scalp would be removed entirely. In modern cases from Melanesia the skin is reflected back, the bone removed and the skin replaced over the trephine. He notes that the incision into the scalp may produce scratches on the underlying bone and can occasionally be seen peripherally to the trephine indicating the extent of scalp removal.

Complications of Trephination

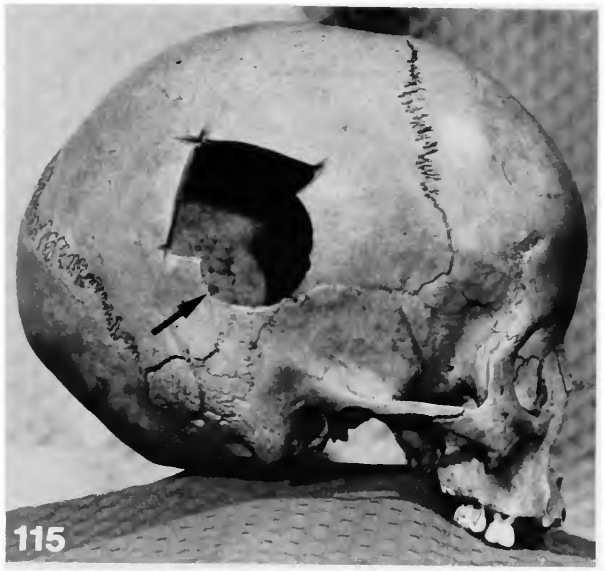
There are two major potential problems arising from trephination: (1) direct injury to the brain resulting from improper method or poor technique, and (2) infection. Either of these complications can cause death. Moodie (1919:481–485) estimates the total mortality rate from trephination at 50–75 percent as compared with a rate of 15–18 percent in civilized modern populations in the early 1900's. Hrdlička (1939:175) is much more optimistic regarding the outcome of trephination, expressing the opinion that complications, including infection, were rare. Piggott (1940:122) and Stewart (1958b:486) support this opinion. Stewart reports that of 214 operations he studied, 55.6 percent show healing, 16.4 percent show slight healing, and only 28 percent show no

FIGURES 112–113.—Trephination by scraping in an adult male skull from Lupo, N. Huarochiri, Peru: 112, The lesion is confined to the frontal bone; note the marks of the scraping at the margin of the lesion (arrow)) and that most of the inner table was left intact. 113, Detail of trephination. This case shows evidence for the association of trephination and previous trauma to the skull; note the edge of fractured bone (arrow). The large hole is probably due to the removal of a fractured fragment during surgery. There is no evidence of healing, indicating that the patient died during or shortly after trephination. (NMNH 293643.)

FIGURE 114.—Healing after trephination by scraping in an adult female skull from Cinco Cerros, Peru. Arrow indicates the outer margin of the lesion. (NMNH 293794.)

healing. With respect to the slightly healed and unhealed cases, it is important to note that they may have died from causes other than the surgery. This is particularly possible if the trephination was done to alleviate the complications of skull fracture.

A case of partial trephination from Peru (NMNH 293782) illustrates the problem of determining the sequence of events that produced a specific condition on the skull. The large lesion is primarily on the left portion of the frontal bone (Figure 118). The lesion consists of a central bony sequestrum containing a single trephine cut, which extended through both tables. The sequestrum is surrounded by an almost purely lytic zone, which grades into the large porous area characterizing most of the lesion. On the left boundary of the lesion, the margin between normal and abnormal bone is sharply defined. In the remainder of the lesion the boundary is not well defined. The fundamental question is what produced the marked inflammatory reaction in the bone? The lesion could be a response to infection following the aborted attempt at trephination. In my opinion, a more likely possibility is an injury to the skull that resulted in tissue death and



FIGURES 115,116.—Trephination by cutting in a child of about 8 years of age from Cinco Cerros, Peru: 115, There is clear evidence of association with cranial trauma. Note the long circular fracture (arrow) adjacent to the trephine and the radiating fracture line extending inferiorly and posteriorly from the circular fracture. 116, Radiating fracture line in the base of the skull continuing the fracture line seen on the lateral aspect of the skull. There is no evidence of healing of the fracture or the trephination indicating death during or shortly after surgery. (NMNH 293315.)

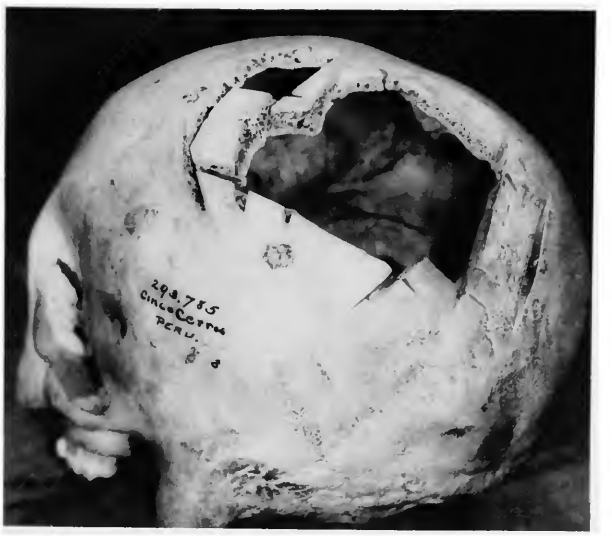


FIGURE 117.—Extensive trephination by cutting in an adult male skull from Cinco Cerros, Peru. There seems to have been some indecision on the part of the surgeon about the appropriate place to cut. There is no evidence of healing, suggesting that the patient died during or shortly after surgery. The somewhat indecisive nature of the trephination suggests the possibility of experimentation, perhaps on a cadaver. (NMNH 293785.)

FIGURE 118.—Injury from blow to the skull followed by necrosis and sequestration of bone (arrows) near the point of impact. There is hypervascularity peripheral to sequestrated bone, which contains evidence of an aborted trephination with a single cut through the sequestrated bone. (NMNH 293782.)

sequestration of bone near the point of impact and a marked, inflammatory reaction to the trauma in the surrounding tissue. This process may have been complicated by infection. Surgical intervention was not completed, perhaps because of the poor condition or death of the patient.

Traumatic Problems Arising from Pregnancy

Pathology

Pregnancy and particularly traumatic complication of pregnancy that threatens the life of a mother is difficult to document in skeletal remains. Evidence for pregnancy, or at least parturition, is thought to be seen in the pitting and erosion of the pubic bones adjacent to the pubic symphysis (Putschar, 1931, 1976b; Angel, 1969a; Stewart, 1970; Gilbert and McKern, 1973). Even demographic data suggesting increased female mortality (e.g., Acsádi and Nemeskéri, 1970:251) does not provide unequivocal evidence of increased death directly due to pregnancy and childbirth. Wells (1975) argues that death due to pregnancy may have been high in Europe from the seventeenth to the early twentieth century when widespread rickets produced unusually high mortality from obstructed parturition. He states that rickets was not widespread before the seventeenth century and that complication from pregnancy may have been too rare to account for the high female mortality. Wells attributes differential female mortality to the cultural practice of males having nutritional needs met first, with females getting less and poorer quality food, particularly during periods of famine.

In contemporary developing countries, death due to complications of pregnancy accounts for about 10 percent of female deaths during the childbearing period from 15 to 44 years of age (World Health Organization, 1976). Thus, while it may be inappropriate to attribute all the difference between male and female life expectancy to the hazards of pregnancy, it does constitute a significant threat to life today and probably did so in antiquity.

Although death during pregnancy may occur due to any number of reasons, including obstructed labor, infection, or hemorrhage, it is difficult to estimate how these hazards relate to ancient populations. Indeed, it is likely that the knowledge and experience needed to minimize mortality varied significantly in ancient societies.

In obstructed labor the problem may be the result of the pelvic outlet being too small for the infant or an abnormal presentation of the infant during delivery. In modern clinical practice the physician has several options available to alleviate this problem and complications arising from it, including surgical intervention and forceps delivery. These options were not generally available in populations whose skeletons are studied by the paleopathologist. Therefore, obstructed labor would have been a serious threat to life, although midwives or their analogue in modern primitive and peasant societies, show rather remarkable skill in successfully assisting a complicated delivery.

Implantation and development of the fetus in any place other than the uterus is known as ectopic pregnancy. Willson, Beecham, and Carrington (1971:180) indicate population differences in frequency. However, in the general U.S. population incidence is one in about 80 normal intrauterine pregnancies. Termination generally occurs in 6 to 16 weeks and is accompanied by a rupture of the fallopian tube, where the fetus is most often implanted in ectopic pregnancy. The rupture is associated with profuse bleeding which, without surgery, almost always will cause death. In the early 20th century, the mortality was between 10 and 15 percent for such pregnancies (Willson, Beecham, and Carrington, 1971:188). However, much higher mortality would be expected in more primitive societies. If the fetus continues to full term there is a serious problem in delivery which must be done by surgical removal of the infant, an option not available in most ancient societies.

Pregnancy increases the risk of infection, which represents a serious hazard to both mother and the developing fetus. There are, for example,

some urinary tract infections that seem to be exacerbated by pregnancy (Willson, Beecham, and Carrington, 1971:272). The childbed fever scourge, with its high mortality in the obstetric hospitals of the eighteenth and nineteenth centuries, may not have caused significant mortality in ancient populations (Wells, 1975:1240). However, women are particularly prone to infection through the genital area immediately following childbirth.

Hemorrhage during pregnancy or childbirth can be caused by several conditions. I have already noted this problem in association with ectopic pregnancy. Postpartum hemorrhage occurs in about 5 percent of normal deliveries. It is also associated with improper implantation of the placenta in the lower region of the uterus (Willson, Beecham, and Carrington, 1971:318).

Willson, Beecham, and Carrington (1971:306) report that 7 to 10 percent of all pregnancies in the United States are complicated by toxemias. One-half of these is due to high blood pressure, the other half is due to a poorly understood condition called preeclampsia and its more serious form eclampsia. Eclampsia is characterized by convulsions or coma. Mortality in the United States for advanced stages of preeclampsia and eclampsia is 10 percent, making it a fairly serious threat to pregnant women. Presumably the mortality would be even higher in populations that do not have access to modern therapeutic means.

Paleopathology

The conditions that cause death during pregnancy rarely leave any evidence in the skeleton. However, this brief summary of the more serious complications of pregnancy should alert the paleopathologist to these conditions when there is evidence of fetal or newborn skeletal materials in association with an adult female burial.

Elliot-Smith and Wood-Jones (1910:260) report from ancient Nubia five cases of female skeletons with deformed pelves, in which death occurred during pregnancy and possibly labor. Acsádi and Nemeskéri (1970:253) report twelve similar cases from Hungary, although Wells (1975:1238) challenges their interpretation and suggests that only two of these cases were deaths resulting from childbirth itself. Wells, however, does not rule out other complications of pregnancy, such as erysipelas or meningitis as a cause of death. Sjøvold, Swedborg, and Diener (1974) report a case of death due to childbirth in a woman having multiple exostoses blocking the birth canal. Hawkes and Wells (1975) report a case of a female burial with a baby's skeleton still partially in the pelvis. They conclude that death took place during childbirth.

Unambiguous archeological examples leave little doubt that awareness of this problem and attention to careful excavating technique make it possible to reconstruct a traumatic event. However, Wells correctly cautions against the assumption that all examples of infants buried with a female skeleton or fetal bone lying inside the abdominal region are indicative of death due to pregnancy and childbirth. A pregnant woman can die of infectious diseases and several other conditions not necessarily related to pregnancy.

One should also keep in mind that labor and childbirth may be traumatic to the infant even when delivery is completed. Evidence for such trauma will probably be uncommon in archeological materials, since severe trauma would result in the immediate death of the infant. In modern clinical practice difficult labor may be assisted by instrument delivery, in which the infant is physically drawn out of the mother with large forceps. Such intervention may injure the tissues of the infant and produce hematoma. When this occurs on the skull, the outer portion of the hematoma may ossify, creating a thin shell of bone. While the events leading to this specific problem are unlikely to occur in archeological populations, the morphological features seen in such problems serve to illustrate what might be seen if an infant were injured in some other way during delivery.

Two cases from recent times provide examples of ossifying hematoma in infant skulls. Figure 119 shows a large hematoma on the left parietal in a case from the Federal Pathologic-Anatomy Mu-

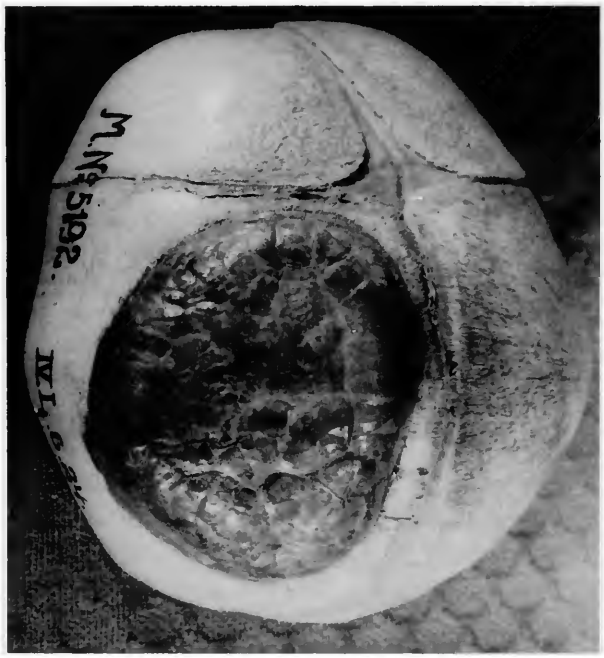


FIGURE 119.—Ossified hematoma on the left parietal of an infant who died several weeks after birth. Note that the shell of the hematoma does not cross the suture. (FPAM 5192.)

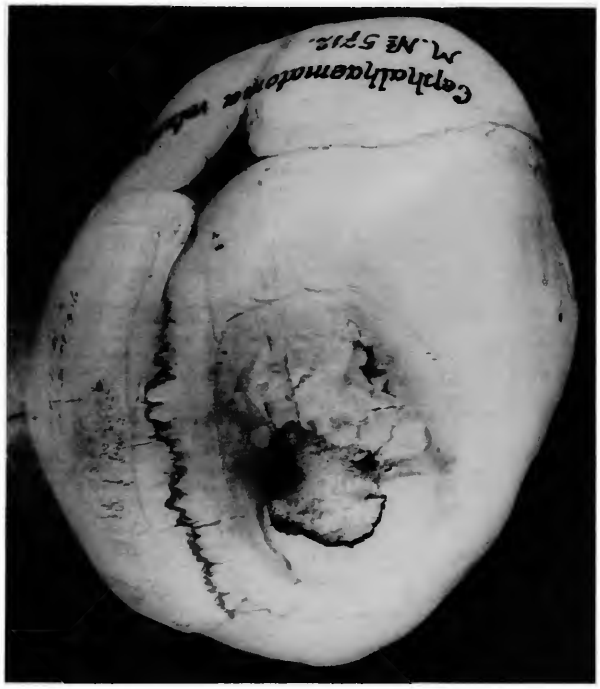


FIGURE 120.—Ossified hematoma on the right parietal of an infant about 4 weeks of age. Hematoma is the result of trauma during birth. Note the delicate nature of the outer shell of bone and the porous appearance of the underlying cortical bone. (FPAM 5712)

seum, Vienna (FPAM 5192). The skull is from an infant who died shortly after birth in 1890. Note that the hematoma does not cross the suture lines. The second case is from the same museum (FPAM 5712). The skull is from an infant about four weeks of age at the time of death. The trauma giving rise to the ossifying hematoma over the right parietal occurred during delivery (Figure 120). The delicate nature of the ossifying outer shell is apparent. The underlying bone is porous, indicating change in the cortical bone in response to the injury and overlying hematoma.

Sincipital-T Mutilation

The skull lesions thought to be associated with cautery in the treatment of mental illness were first brought to the attention of anthropologists by Manouvrier (1895). He used the term "T-sincipital" to describe the lesion. What appears on the skull is a depressson in the shape of a T or a cross in which the vertical portion of the T is associated with the sagittal suture while the cross bar on top of the T is often associated with the lambdoid suture.

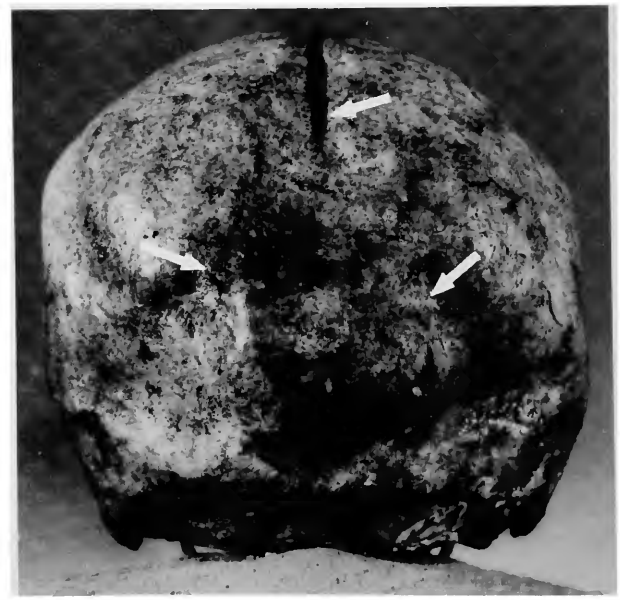


FIGURE 121.—Atrophy of the skull in the region of the sagittal and lambdoid sutures (arrows). The inner table is normal (NMNH, no number.)

In Europe, skulls with these lesions were found in skeletons of the Neolithic period (Manouvrier, 1895). Zaborowski (1897) reports similar findings in central Asia. Moodie (1921) and Weiss (1955) report the sincipital-T pattern in ancient Peruvian skulls. However, Stewart (1958b:480) cautions against interpreting any large bone scar as the result of cautery.

Virchow (cited in MacCurdy, 1905:20) suggested that a tartar-emetic ointment smeared on the scalp could produce scarring and affect the bone. However, weight is lent to the hypothesis of cautery being the cause of the lesion by Avicenna's medieval prescription of melancholia in which "the head is to be cauterized in the form of a cross" (MacCurdy, 1905:23). Most often the lesion resulting from cautery did not expose the dura (MacCurdy, 1905:17–18), although at least one specimen exists in which the sincipital-T defect in part penetrated through both tables of the skull.

There is also a possibility that senile atrophy can occur in the sagittal and lambdoid sutures. Atrophy in this location would produce a depression very similar to the changes reported for sincipital-T mutilation. One skull from a prehistoric site (Irene Mound) in Georgia, USA (NMNH unumbered) exhibits a depression in the posterior sagittal suture and much of the lambdoid suture (Figure 121). The inner table is normal. The skull appears to be that of a female over 50 years of age with considerable antemortem tooth loss. There is evidence of senile atrophy medial to both temporal lines suggesting that senile atrophy is a likely explanation for the atrophy of the sutural areas.

Infectious Diseases

The Inflammatory Response

The inflammatory response to infection begins as a vascular phenomenon. The capillaries dilate and their walls begin to allow the escape of large molecular proteins and cells normally retained in the circulation. Albumins, globulins, and fibrinogen enter the tissue where fibrinogen is converted into fibrin. This is followed by active emigration of neutrophilic leucocytes, which are capable of amoeboid motion and thereby can squeeze through moderately enlarged pores in the vascular wall. In the tissue they migrate, by chemotactic attraction, to the bacterial focus. It is these leucocytes, in connection with proteins and fibrin, which make up the pus of acute infections. Many leucocytes ingest bacteria by phagocytosis and may inactivate or destroy them; others succumb to bacterial toxins and disintegrate. The severity of inflammatory response is proportional to the number and virulence of infecting organisms. If the capillary leakage becomes gross, erythrocytes, which have no active mobility, may also enter the tissue in large numbers. Thus, it is customary to classify the inflammatory response by the type of exudate as serous, fibrinous, hemorrhagic, or purulent, reflecting the increasing severity of the infection. After the acute phase other cells, from the blood and from the tissue, participate in the inflammatory response. Lymphocytes and plasma cells particularly, involved in the cellular and humoral immune responses, appear in increasing numbers in and around the lesion in the subacute and chronic phases of infections.

Histiocytes and macrophages of the tissue are mobilized and serve in cleaning up infected areas by means of phagocytosis of debris and of organisms. Multinucleated symplasts, so-called giant cells, may appear in this phase. The healing is mainly accomplished by proliferation of fibroblasts laying down collagen, which is the main constituent of scar tissue. In infections that have a component of allergic reactivity, as, particularly, in parasitic diseases, eosinophilic leucocytes may be prominent in the immediate inflammatory response. In viral infections, where the organisms propagate intracellularly and offer no opportunity for phagocytosis, the initial leucocytic response as well as the formation of pus is usually lacking. Lymphocytes, plasma cells, and macrophages dominate the picture even in the acute phase, mainly in response to the breakdown of infected tissue cells.

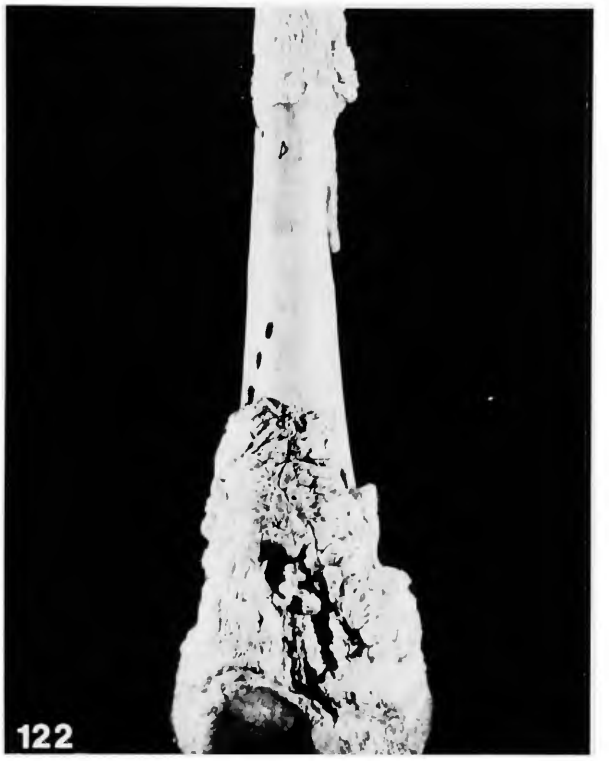
Organisms that are difficult or impossible to control by the ordinary cellular defense mechanisms discussed above produce the so-called granulomatous response to infection. Such organisms are the acid-fast mycobacteria of tuberculosis and leprosy, many of the fungi, and treponemal infections. In these infections histiocytic proliferation around organisms form nodular foci. Multinucleated giant cells phagocytose organisms but often are not able to kill them. Thus, the center of the granuloma may undergo necrosis but harbor confined, viable organisms for a long time. Lymphocytes and plasma cells border the granulomas, indicating the continued response to the infection.

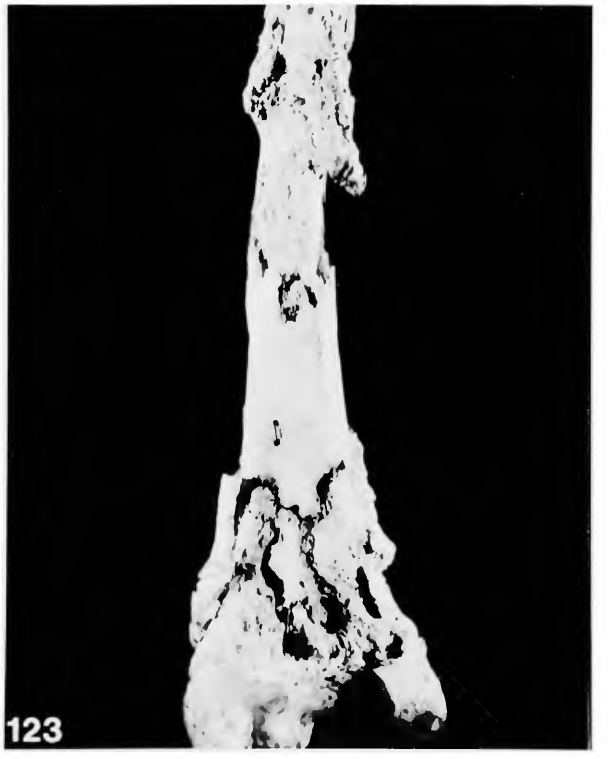
The Biology of Infection

In human populations of antiquity, generally about half the individuals born died during infancy or childhood. The major cause of this mortality was infectious disease, the single greatest threat to life. Of those living to adulthood, many died of the direct or indirect effects of infectious disease, but trauma becomes increasingly significant. Even among hunters and gatherers living today, infection of the digestive system results in the death of many of the infants and children.

With the development of agriculture and its associated sedentary life many more infectious diseases became endemic in human populations. Civilization, with its high concentration of people in a limited area, brought with it the spector of death on a massive scale, with epidemics killing half or more of the inhabitants of a city.

For the paleopathologist, one of the great sources of frustration is the fact that infectious diseases, particularly those that result in death, rarely leave behind any direct evidence of their existence in the skeletons of the individuals who die of them. This places great restrictions on the ability to investigate the biological effects of these diseases on human populations. Despite these limitations, it is apparent that some infectious conditions do affect the skeleton and that careful analysis and appropriate investigations can reveal much about human adaptation in response to disease. It is important to emphasize that infectious conditions affecting the skeleton tend to be subacute, chronic diseases and may not be the immediate cause of death. Many of the chronic infectious diseases produce morphologically overlapping responses in skeletal tissue. These responses make specific diagnosis difficult even in modern clinical cases where many more of the variables of the disease state are known.



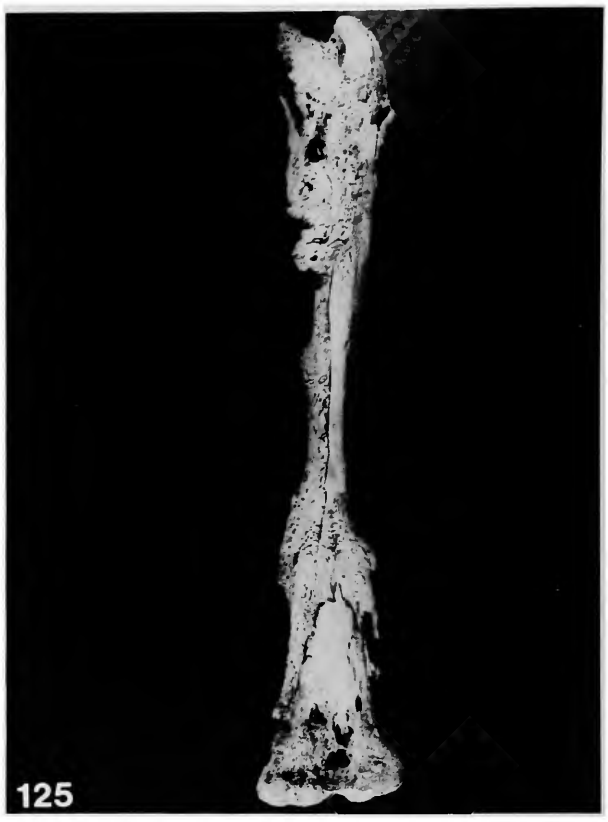


Osteomyelitis

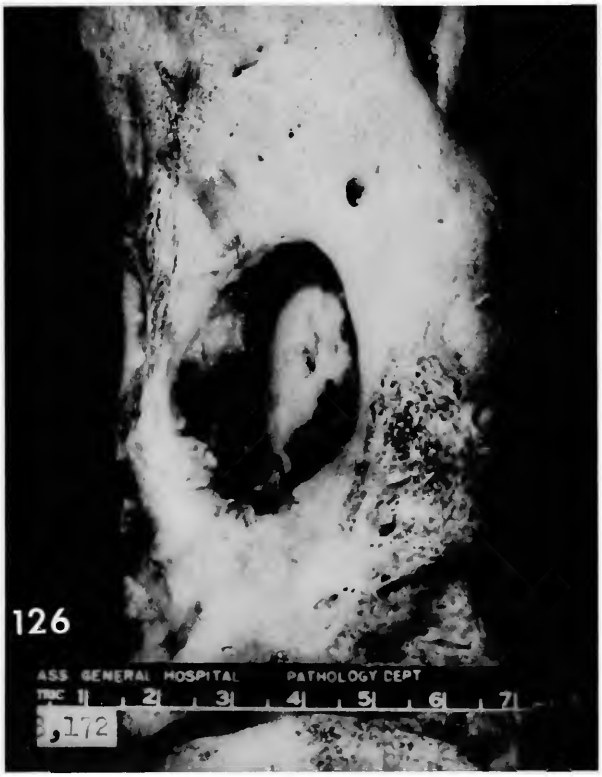
Pathology

Osteomyelitis is the result of introduction of pyogenic bacteria into bone. The bacteria may reach the skeleton by several different routes: (1) by direct infection through traumatic or surgical wounds, (2) by direct extension from adjacent soft tissue infections, or (3) by the hematogenous

Figures 122, 123.—Osteomyelitis of distal right femur: 122, Anterior view showing smooth total sequestrum with involucrum above and below. 123, Posterior view showing destruction of metaphysis and proximal diaphysis; notice the jagged border of the sequestrum at the junction with the living bone. (About 10 years, WM HS 44.5 from 1841.)



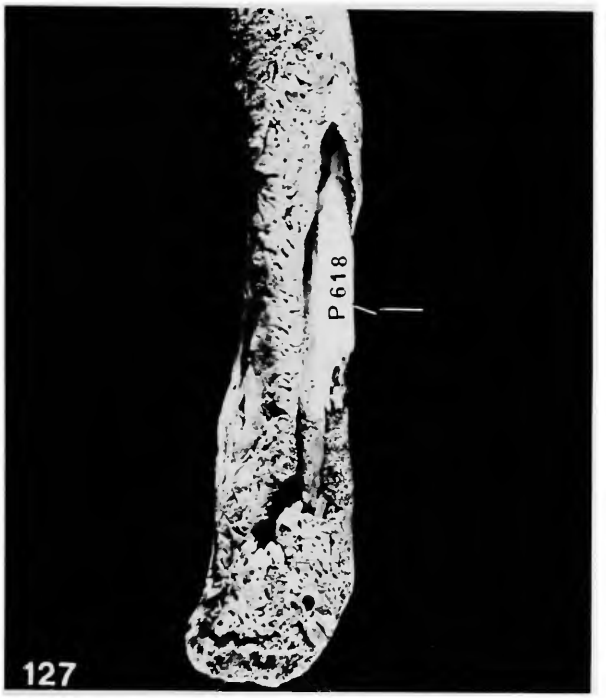
FIGURES 124, 125.—Osteomyelitis of right femur with total diaphyseal sequestrum, involvement of both metaphyses and involucrum; epiphyses missing: 124, Anterior view. 125, Posterior view. (About 14 years, FPAM 837.)



route from a remote septic focus. The causative organism, in close to 90 percent of the cases, is *Staphylococcus aureus*; the second in frequency is *Streptococcus*, with other cocci and bacilli making up a small percentage.

The osteomyelitis secondary to wounds, compound fractures, or surgery obviously can occur at any age and in any part of the skeleton. However, since injuries to the limbs and, if not penetrating, to the skull are more likely to be survived than injuries to the thorax or abdomen, posttraumatic osteomyelitis will be found more often in the bones of extremities and the cranium than in those of the trunk. Although this route of

FIGURE 126.—Chronic osteomyelitis of femur after fracture, 10-years duration. Involucrum with large cloacal opening exposing cortical sequestrum. (33-year-old male, MGH autopsy 33172.)



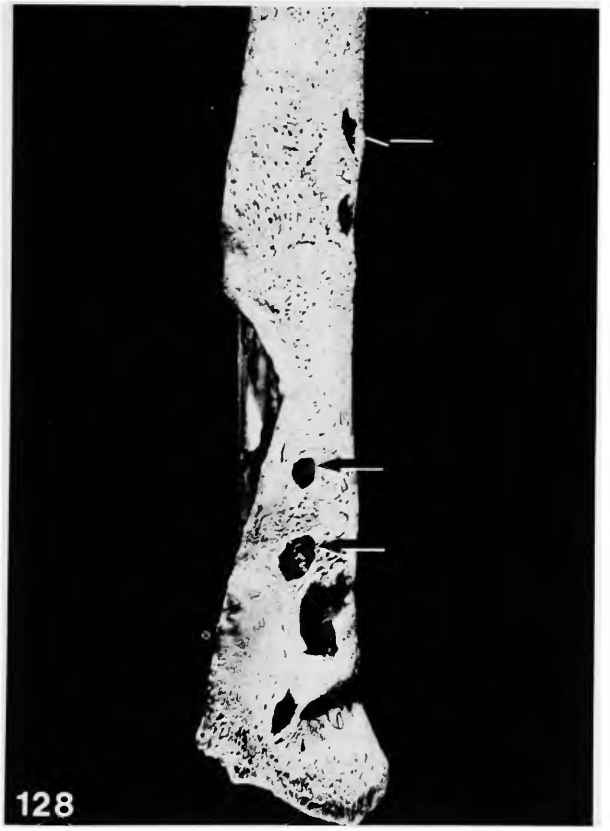


FIGURE 129.—Osteomyelitis of left tibia with massive involucrum and numerous cloacal openings, anterior view, 3-years duration. (20-year-old male, PMES 1.E.B.16(12) before 1931.)

infection may result in the full blown picture of acute and chronic osteomyelitis, it may more often be more limited and localized. This is also true of osteomyelitis secondary to adjacent soft tissue infection. In such cases, infection may be limited to the periosteum and cortex and not spread through the medullary cavity. The result of such localized infections would be focal periosteal bone deposition around a partial cortical defect, with or without a small sequestrum, and with some sclerotic response in the vicinity. Such local infections may heal with sclerotic scarring

FIGURES 127, 128.—Osteomyelitis of distal right tibia: 127, Massive involucrum exposing sequestrum (arrow) in large opening. 128, Porous involucrum with multiple cloacal openings (arrows). (HM P618.)

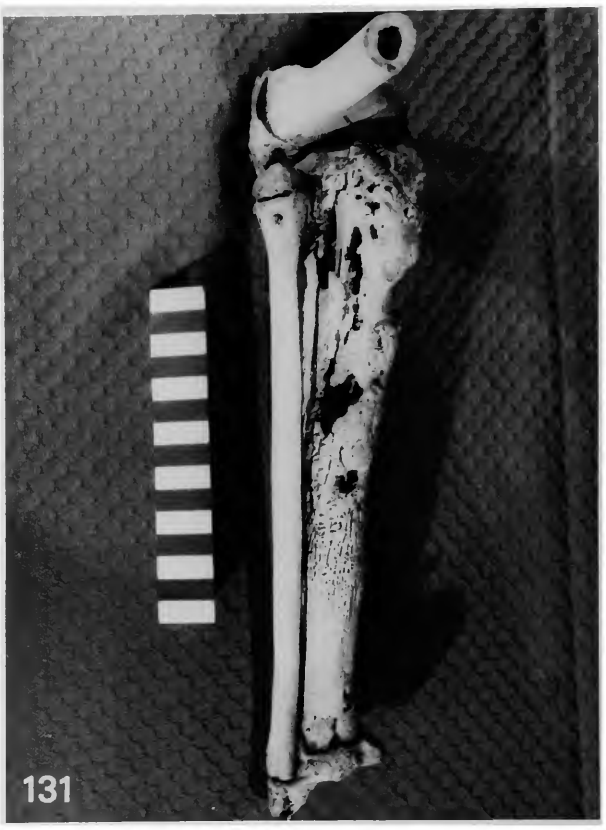


FIGURE 130.—Osteomyelitis of left tibia following slight injury, 18-months duration. Notice massive hypervascular involucrum exposing large sequestra in large cloacal openings, periostitis of fibula. (Adolescent, WM HS44.6 from 1851.)

FIGURE 131.—Chronic osteomyelitis of left tibia with involvement of the proximal epiphysis and partial bony ankylosis of knee in flexion. Notice cloacal openings, involucrum, and sequestrum. (12 years, FPAM 4948 from 1888; scale in cm.)

FIGURE 132.—Chronic osteomyelitis of right tibia with ankylosis of ankle joint, involvement of talus and periostitis of fibula. Notice massive involucrum with multiple cloacae exposing sequestra. (About 20 years of age, FPAM 358 from before 1817.)

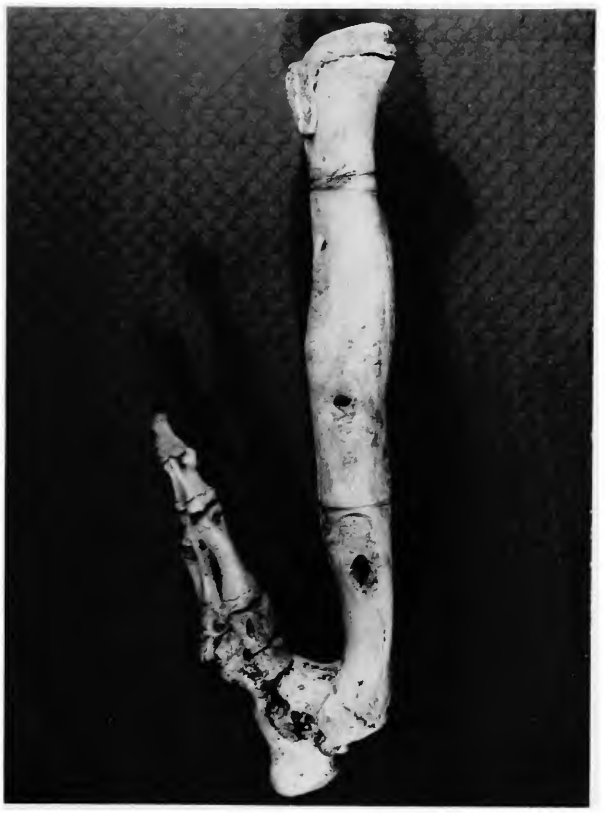


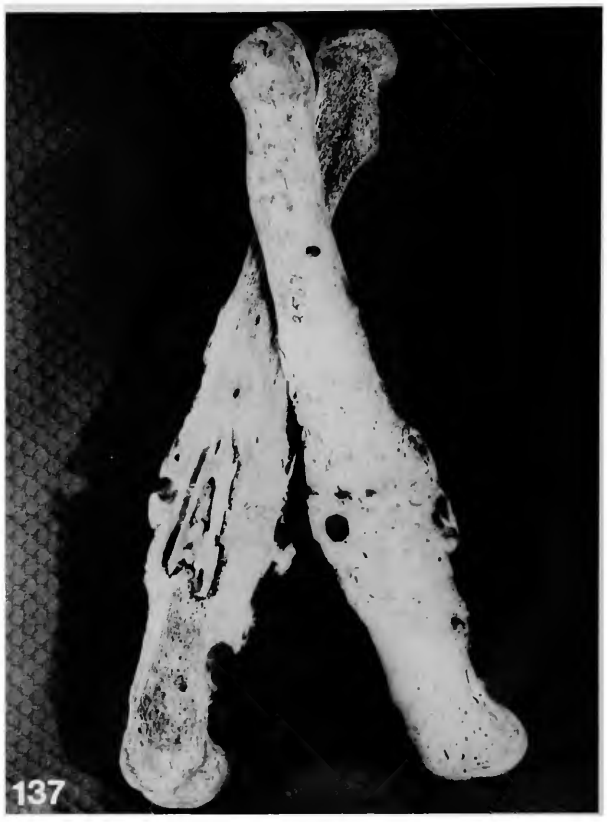
FIGURE 135.—Chronic osteomyelitis of right tibia with slipped distal epiphysis and ankylosis of ankle joint resulting in pes calcaneus deformity of the foot. (17-year-old female, PMUG 3239, autopsy 8278 from 1877.)

around a depression. These scars may subsequently be greatly effaced by remodeling.

Hematogenous Osteomyelitis

Hematogenous osteomyelitis runs a fairly typical course and shows distinct variations in frequency and appearance in different age groups (Trueta, 1959:671–680; Putschar, 1976a:41–60). In Trueta's (1959) series of over 200 cases, 7 percent were infants, 80 percent children, and 13 percent adults. In preantibiotic days the hema-

FIGURES 133, 134.—Chronic osteomyelitis of right femur with bony ankylosis of knee and disuse osteoporosis of tibia; two fistulae draining a large periosteal abscess were found at autopsy: 133, External view. 134, Cut surface. (58-year-old male, IPMI KM180, autopsy 4802, annual 249 from 1898.)



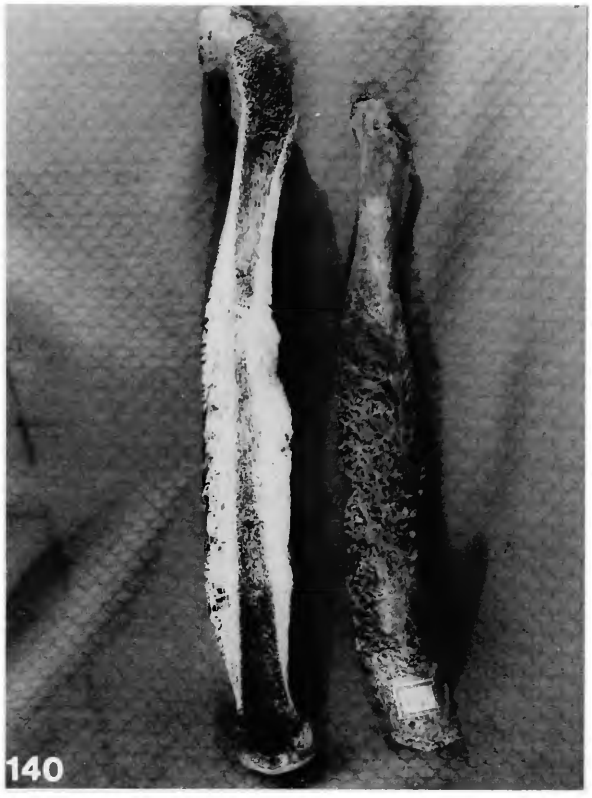
FIGURES 136, 137.—Chronic osteomyelitis of left femur: 136, Medial, outside view, showing remodeled periosteal reactive bone and cloaca. 137, Cut surface and lateral view, showing sclerotic remodeled involucrum, central sequestrum, and multiple cloacae. (ANM 2950.)

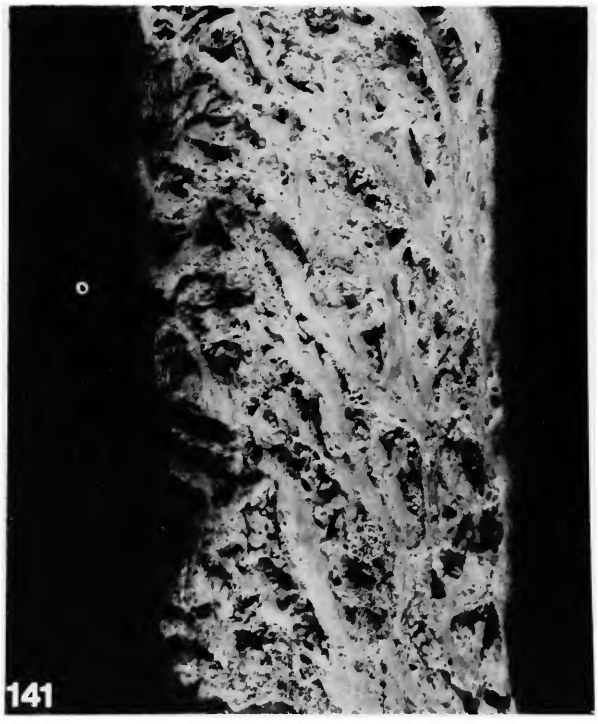
FIGURE 138.—Chronic osteomyelitis and periostitis of left femur, showing cancellous bone filling the medullary canal around residual abscess cavities. (34-year-old male, IPAZ 3796, surgical specimen MB 1515 from 1936.)

togenous osteomyelitis of childhood and adolescence made up an even greater percentage of the total. In this age group there is a marked predominance of males over females at a ratio of at least 3:1. The sex difference in infant osteomyelitis is not marked and in adult osteomyelitis is less than in the juvenile form. Hematogenous osteomyelitis greatly predilects the long bones of the extremities. It is limited to one bone in 80 percent of the cases; two or, rarely, more bones are affected in only 20 percent (Garrè, 1893:276). The localization of juvenile hematogenous osteomyelitis is intimately related to the rate and time of growth of the various growth plates. The infection almost

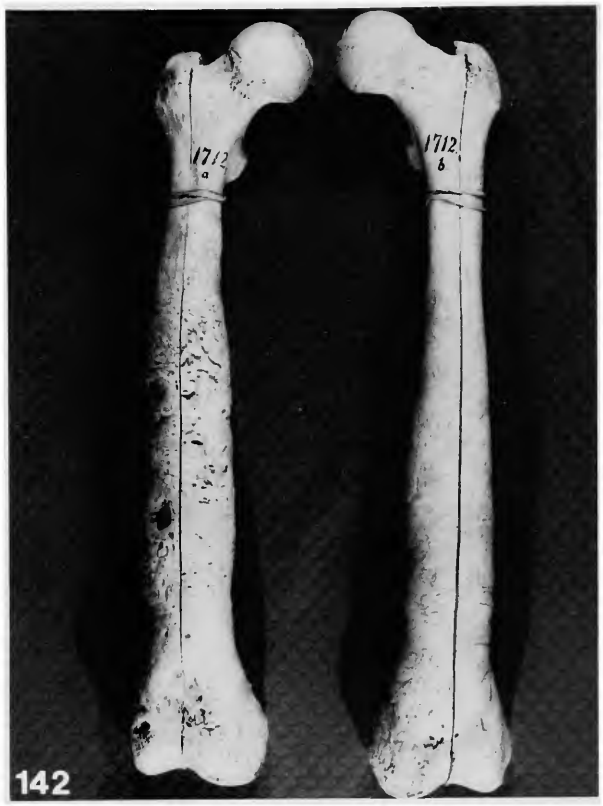
FIGURE 139.—Chronic sclerosing osteomyelitis of left tibia, showing central lytic cavities and medullary sclerosis; marked, mostly anterior, periosteal bone deposition. Notice absence of sequestra and cloacal openings. (About 17 years old, IPMI KM 187; scale in cm.)

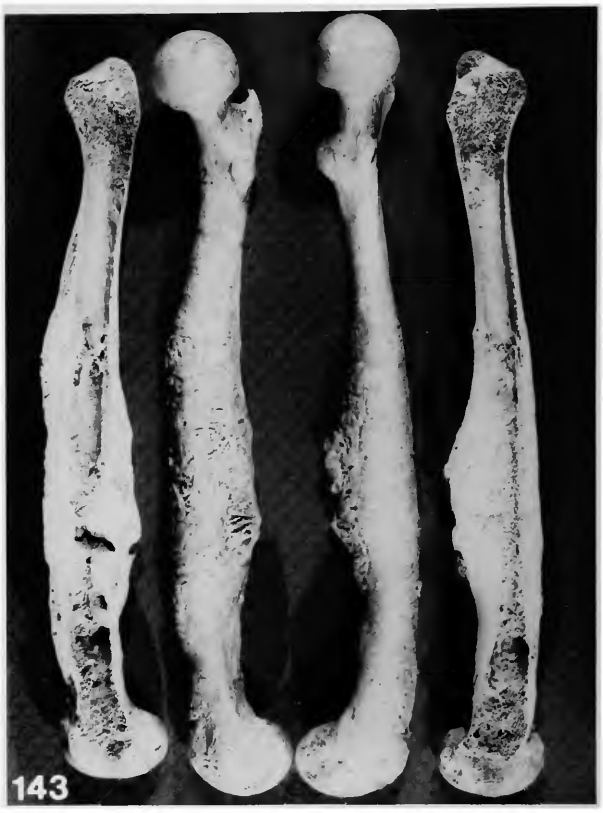
always starts in the metaphysis near an actively growing plate. The greatest frequency involves the distal femoral metaphysis, the proximal tibial metaphysis, the distal tibial metaphysis, and the proximal femoral metaphysis in decreasing order. Femur and tibia together account for close to 80 percent of locations, followed by the humerus with about 10 percent. Other long bones of the extremities are less often affected, especially those of hands and feet. The cancellous bones and the





FIGURES 140, 141.—Sclerosing osteomyelitis and perostitis of left femur, probably nonspecific but, in absence of sequestra and cloacae, syphilis cannot be excluded: 140, Cut surface (KM 133c) and outside surface (KM 133c′). 141, Detail of periosteal surface (KM 133c′). (Adult, no data, IPMI.)

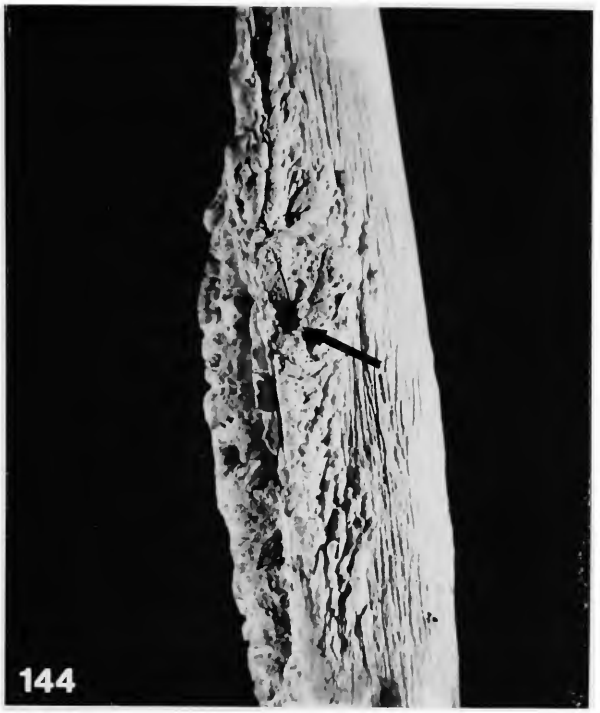


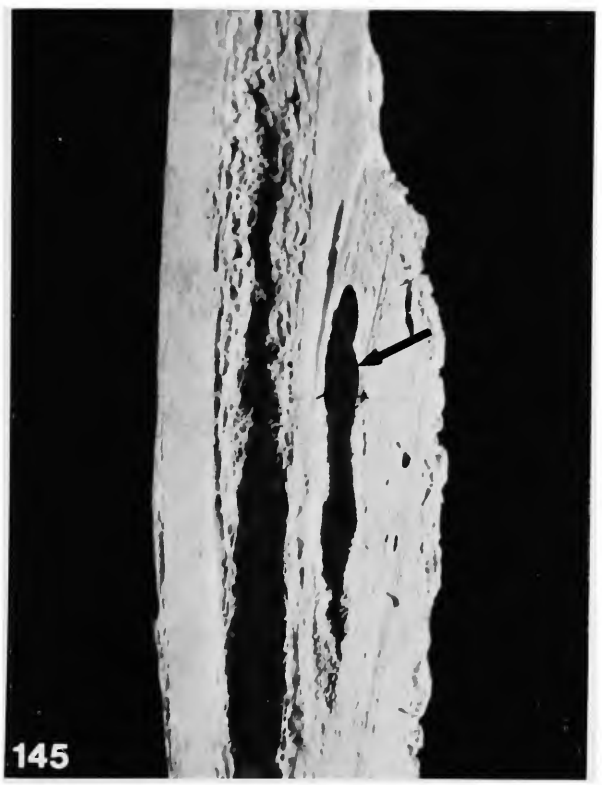


FIGURES 142, 143.—Chronic sclerosing osteomyelitis of both femora; notice the similarity of the periosteal bone pattern to tertiary syphilis: 142, Anterior view. 143, Interior and medial view. (23-year-old male with periosteal abscess and skin fistula over both femora; died from amyloidosis; PMUG 1712 a and b, autopsy 3260 from 1869.)

skull participate least in hematogenous osteomyelitis (Wilensky, 1934:276, 277).

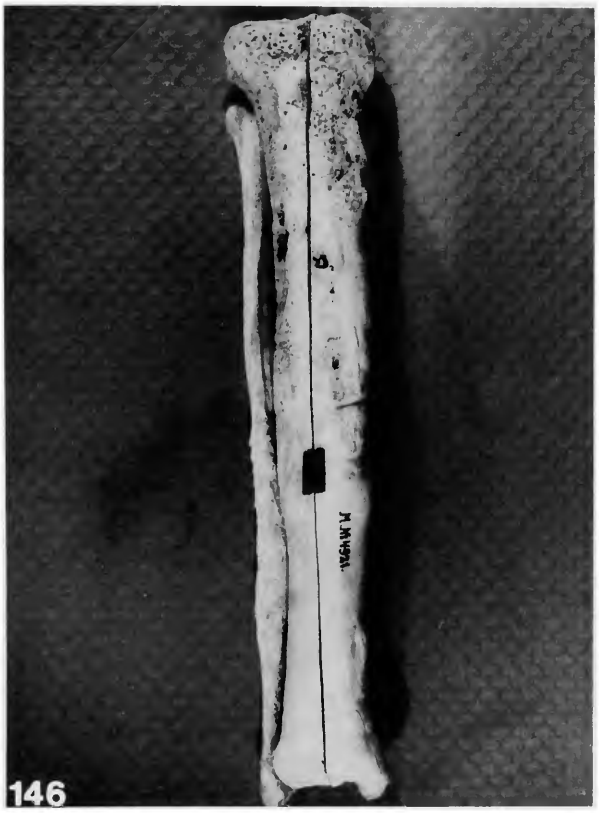
The acute hematogenous osteomyelitis begins with one or several purulent foci in a metaphysis, leading to lytic destruction of the newly formed spongiosa. The exudate spreads through the marrow cavity, increasing the pressure within the rigid confinement of the diaphysis and resulting in more or less extensive necrosis of the cortex by means of vascular compression. In the metaphysis, near the growth plate, the thin remodeling cortex permits easy extension of the exudate under the periosteum. The active osteogenic periosteum of the child readily strips from the cortex, permitting the formation of a subperiosteal abscess and depriving the diaphysial cortex of its external blood supply. This results in formation of large, often circumferential (Figures 122, 123) and sometimes total necrosis of the diaphysis (Figures 124, 125). The dead bone becomes the sequestrum, which can only be resorbed or remodeled if small. Large sequestra remain and maintain the infection if they are not extruded through fistulae or removed by surgical intervention. The elevated periosteum continues to produce bone, forming a shell of hypervascular bone surrounding the sequestrum (Figure 126). This enveloping reactive bone formation is called involucrum and serves as a scaffolding in ultimate repair. The metaphysial bone usually does not undergo total necrosis because of the easy escape of the exudate through the thin, porous cortex. The separation of living bone and sequestrum takes place in this area by means of osteoclastic





resorption (Figures 127, 128). The subperiosteal pus seeks escape through the involucrum and through the surrounding soft tissue to the skin surface, causing a large, round cloacal opening in the involucrum (Figure 129), often exposing parts of sequestra to view (Figure 130). Skin fistulae, which discharge pus and fragments of dead bone, develop. The epiphysis in the growing child is relatively protected by the growth plate and by the fact that its vascular supply does not communicate with that of the metaphysis and diaphysis. For this reason extension into the epiphysis and the adjacent joint is uncommon in that age group. An exception is the hip and, to a lesser extent, the shoulder joint, where part of the metaphysis is located within the articular space. However, in long standing or recurring osteomyelitis the adjacent joint may become involved at any age and terminate with bony ankylosis (Figures 131–134). In the active phase of the disease the growth cartilage may be partly destroyed and epiphysiolysis (Figure 135) used to occur in up to 15 percent of the cases (Garrè, 1893:287). Pathological fractures, mostly through the area of demarcation between living and dead bone, rarely through the involucrum, used to be observed in 8 percent of the cases (Garrè, 1893:293). The endosteal process of lytic destruction was followed by reactive sclerosis, limiting the intraosseous infection to local areas (Figure 136–138). The combination of these various phenomena over a period of months, years, and, sometimes, decades results in a great variety of pictures, which defy detailed description. However, the common characteristics are presence of sequestra, porous hypervascular periosteal bone, and cloacal openings. Since dead bone is not easily attacked by osteoclasts, large sequestra may exhibit a smooth surface but with a jagged border at the line of

FIGURES 144, 145.—Osteomyelitis of femur with subperiosteal abscess surrounded by a thick layer of reactive bone. The abscess cavity contained a cortical sequestrum: 144, Outside view, showing massive reactive bone with cloacal opening (arrow). 145, Cut surface, showing elongated abscess cavity (arrow). (20-year-old male, WM S.48.2 from 1882.)



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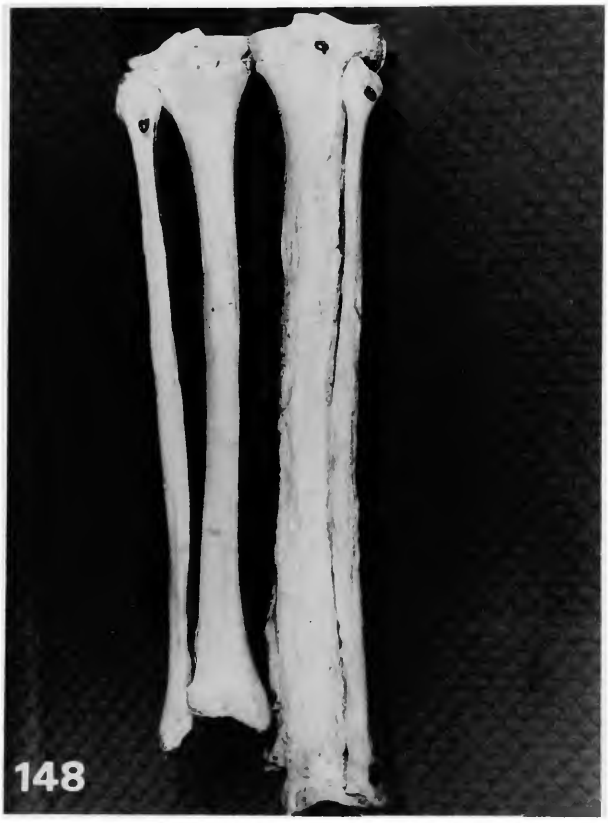

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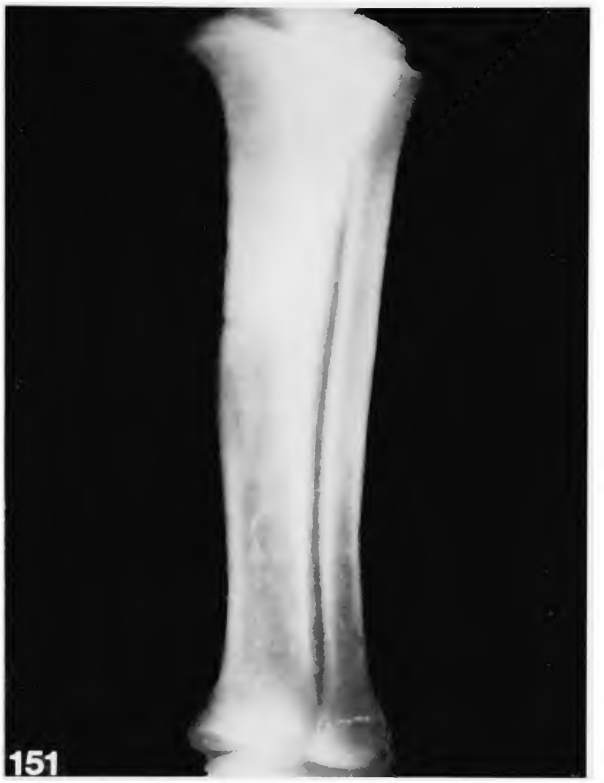

FIGURES 150, 151.—Acute osteomyelitis of left tibia: 150, Tibial diaphysial sequestrum, extracted from the fistula. 151, X-ray one year later, showing complete regeneration of the tibia. (2-month-old African negro, WM S44.6, from 1963.)

separation. The mortality of this disease used to be at least 20 percent. In many instances the disease remained active for a long time; in others it healed temporarily to recur after years; others healed permanently. Other cases showed a less severe course with fewer or no sequestrations and more sclerotic response (Figure 139) (Garré, 1893: 257–263). In the absence of sequestra and of cloacal openings entering the medullary canal, differentiation from tertiary syphilis is difficult or impossible (Figure 140–143). Some cases result only in a local abscess formation (Brodie's abscess) (Figures 144–147), usually near the growth plate, in the metaphysis, surrounded by sclerotic bone. The collateral hyperemia, not uncommonly, results in excessive, longitudinal growth of the affected bone if the growth plate (Figure 148) is not destroyed. In rare cases of chronic osteomyelitis, carcinoma may develop from the epithelium of the fistulous tract (Figure 149).

INFANTILE OSTEOMYELITIS.—This disease exhibits some differences from juvenile osteomyeli-

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FIGURES 146, 147.—Chronic recurring osteomyelitis of right tibia with involvement of the proximal epiphysis and elongation. Notice the low position of the proximal end of the fibula. Reactive periostitis of fibula: 146, Anterior view. 147, Cut surface; notice abscess cavities with perifocal sclerosis. (FPAM 4921, autopsy 86827 from 1888.)

FIGURE 148.—Both lower legs. Chronic osteomyelitis of right tibia with elongation and with ankylosis of ankle joint, periostitis of fibula. (53-year-old female, FPAM 5217, autopsy 91835 from 1892.)

FIGURE 149.—Squamous cell carcinoma of left tibia secondary to chronic osteomyelitis. Notice the extensive destruction and pathological fracture of the tibia and periostitis of fibula. (45-year-old male with osteomyelitis since childhood, PMES IVNa 16(4).)

tis. It used to be uncommon in the preantibiotic period, because most infants succumbed to the septicemia before osteomyelitis was established (Green and Shannon, 1936). In infantile osteomyelitis massive sequestration is unusual because the thin, loosely structured cortex permits escape of pus without raising the intramedullary pressure to critical levels. Involvement of the epiphysis and concomittant septic arthritis are common because, in this age group, the epiphysial blood supply is not yet separated from that of the metaphysis (Trueta, 1957:360–366). If the osteomyelitis heals, rapid remodeling removes much of the evidence of the disease, for, in the rapidly enlarging bone, the entire old cortex is removed as the bone grows in diameter. This is true even after elimination of a large sequestrum (Figures 150, 151).

ADULT OSTEOMYELITIS.—Adult hematogenous osteomyelitis is rare and may often represent continued or recurrent juvenile osteomyelitis. New infections also predilect the metaphysial areas of long bones, especially femur and tibia. The inflammation is less acute and less extensive. Sequestra tend to be small. The firm attachment of the periosteum prevents extensive stripping but local subperiosteal abscesses and fistulae are common. Since, after closure of the growth plate, communications of metaphysial and epiphysial blood vessels are reestablished to some extent, involvement of the epiphysis and of the adjacent joint is not unusual.

Hematogenous osteomyelitis caused by bacteria other than *Staphylococcus* and *Streptococcus* should be briefly discussed. About one percent of patients with typhoid fever, regardless of age, develop osteomyelitis (Murphy, 1916). The main locations are ribs, tibia, and spine. The tibial lesion often is a diaphysial subperiosteal abscess involving the underlying cortex. The spinal lesion predilects the lumbar segment and often involves two adjacent vertebrae and the intervertebral disc between them. In various hemoglobinopathies, especially in sickle cell anemia, complicating salmonella osteomyelitis is not uncommon (Hook, Campbell, Weens, and Cooper, 1957:403–407; Golding, MacIver, and Went, 1959:711–718). Multiple bones are affected in about two-thirds of these cases. Long bones are predilected, but about 25 percent of the patients show involvement of bones of hands and feet. There are usually no major sequestra and the periosteal reactive bone is sparse (Putschar, 1976a:49).

Changes in Individual Bones

The overall description of hematogenous osteomyelitis given above mainly focuses on the changes in long bones of the extremities. A discussion of the pecularities of osteomyelitis in different other parts of the skeleton is therefore indicated.

SHORT TUBULAR BONES.—Phalanges, metacarpals, and metatarsals are uncommon sites of osteomyelitis. In young children hematogenous infections in these rapidly growing bones do occur and lead to rapid destruction of the thin cortex and formation of a massive expanded involucrum, resembling the spina ventosa in tuberculosis and similar changes in congenital syphilis (Figure 152). In adults phalanges, especially of toes, are more often involved in cases of peripheral vascular disease secondary to skin ulcerations. These cases resemble the features of chronic adult osteomyelitis in other bones (Figure 153).

THE SPINE.—Vertebral osteomyelitis is uncommon, amounting to about 2 percent of all cases (Schmorl and Junghanns, 1971:389). It occurs more often in adults than in children (Kulowski, 1936:343–344). In contrast to tuberculosis, only one vertebra is affected in 75 percent of the cases (Donati, 1906:1132–1133). The frequency of location in the spine increases distally. Lauche (1939:39) estimates that of cervical, dorsal, and lumbar involvement at a ratio of 1:2:3. The first and second cervical segments are very rarely affected. As far as the location within the vertebra is concerned, in contrast to tuberculosis, the spinous process and neural arch often participate in the lesion. The infection may even be limited to the neural arch and/or spinous process (66 percent (Donati, 1906:1133) 58 percent (Volkmann, 1915:485)). The transverse process is rarely affected. In destructive spinal osteomyelitis, col-

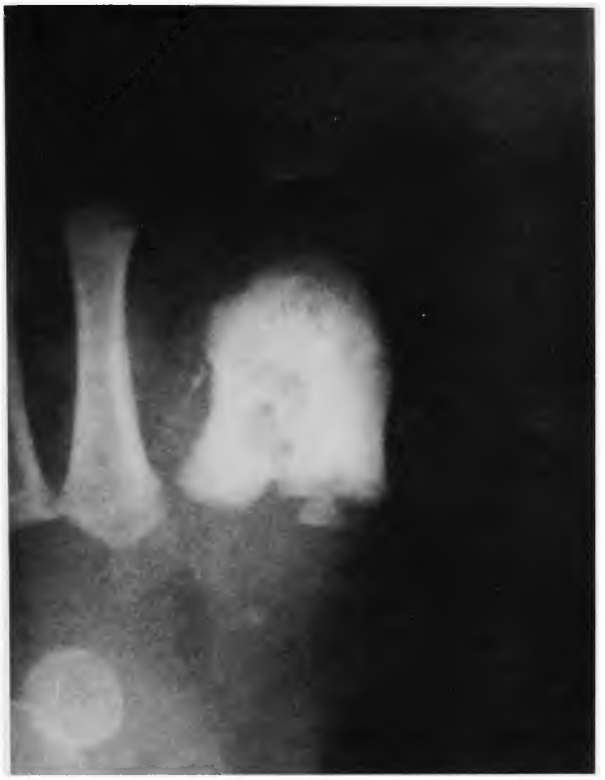


FIGURE 152.—X-ray of hematogenous staphylococcic osteomyelitis of first metatarsal, massive involucrum of diaphysis. (3-year-old male, surgical specimen 7637-1970, MGH.)

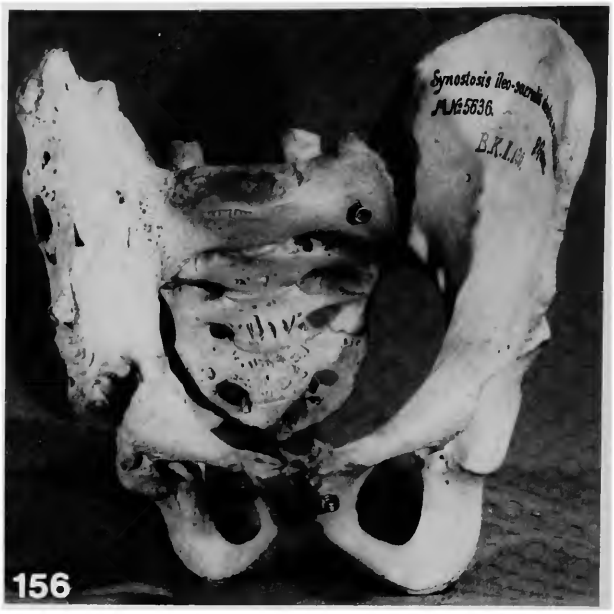
FIGURE 153.—Osteomyelitis of basal phalanx of right big toe with involucrum and sequestrum, periostitis of terminal phalanx, secondary to infected wound. (Adult, PMES 1 EB 18 (100).)

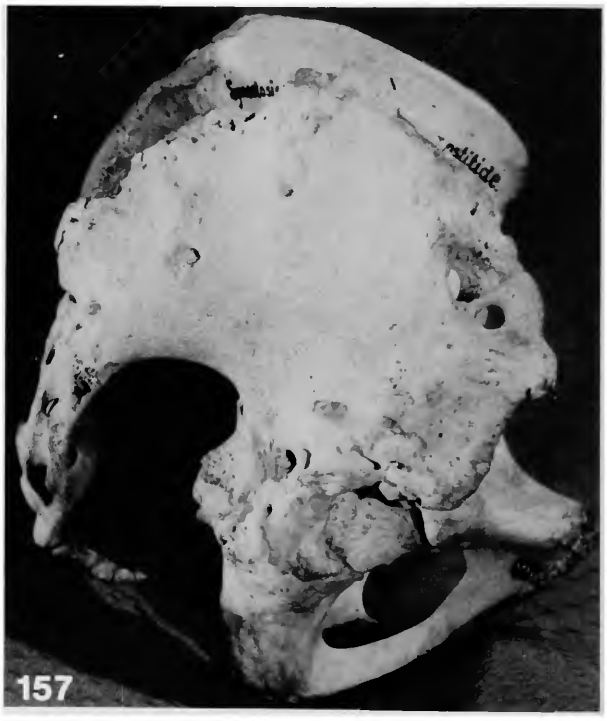
lapse of a vertebra is not uncommon. In multiple involvement of adjacent vertebrae, some of the intervertebral discs are usually better preserved than in tuberculosis but may show bony bridging (Junghanns, 1939:325–326). Destruction of several adjacent vertebrae may result in an angulation of the spine, which may be indistinguishable from tuberculous gibbus. Paravertebral abscesses can occur and may elicit reactive periosteal bone formation.

CANCELLOUS BONES.—These bones are rarely involved, but when they are affected, the process locates in the area with most cancellous bone: In the scapulae, in the spina (Figures 154, 155); in the pelvis, in the iliac crest (Figures 156, 157); in the sacrum, in the lateral wing. Pelvic osteomyelitis may localize in the acetabular roof, secondary to septic arthritis of the hip or around a ruptured symphysis pubis in puerperal septicemia (Putschar, 1931:91, 92). Ribs account for about 2 percent of osteomyelitis. The infection is usually located near the junction with the cartilage or posterior, at the angle. There is usually no sequestration (Lauche, 1939:39).

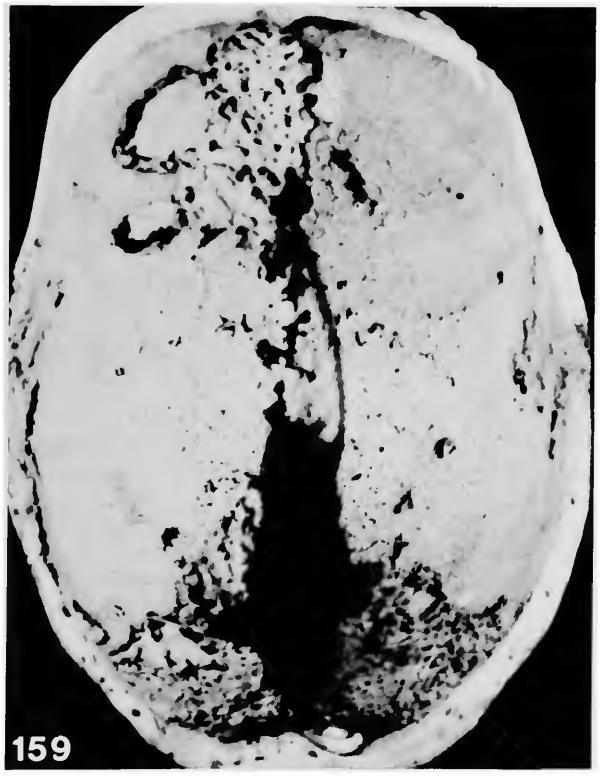
SKULL.—Primary osteomyelitis of the skull is rare (Figures 158, 159). The most usual is extension of an empyema of the frontal sinus into the frontal bone. The structure of the diploë with its interconnecting large vascular channels permits spreading of the infection through the cranial vault. Sutures may act as a temporary barrier, but extension into the parietal bone is not uncommon. The occipital bone is usually spared. Osteomyelitis of the skull base, originating from an empyema of the sphenoid sinus, is even more rare. Of more importance is traumatic osteomyelitis of the cranial vault following open wounds

FIGURES 154, 155.—Osteomyelitis of left scapula removed surgically, showing multiple cloacae over sequestral defects and massive involucrum: 154, Posterior view. 155, Anterior view. (14-year-old female, WM S45.1, from 1858.)





FIGURES 156, 157.—Chronic osteomyelitis of right ilium with right sacroiliac ankylosis and septic arthritis of right hip, 3-years duration: 156, Frontal view. 157, Lateral view. (17-year-old female, FPAM 5636.)



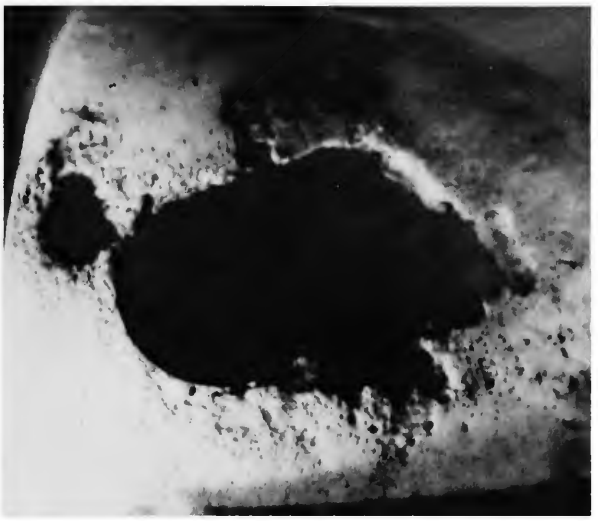
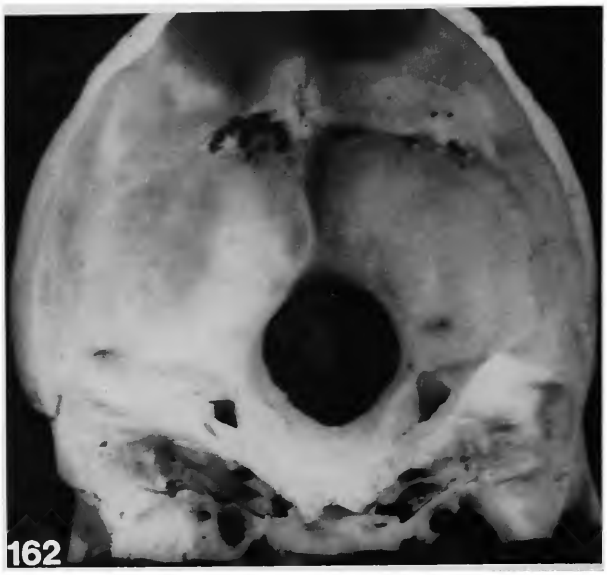


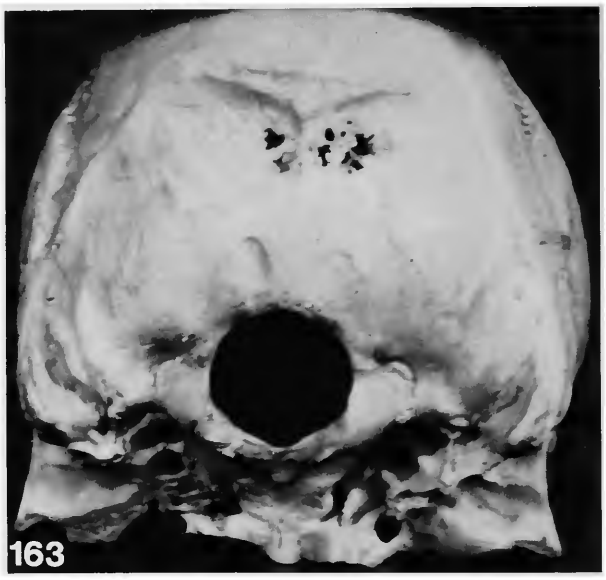
FIGURE 160.—Osteomyelitis of right parietal bone secondary to infected scalp wound and removal of sequestrum by trephining. (Adult, before 1850, PMES 1E.B.1(1).)

or blunt trauma, with or without fracture and surgical intervention (Figures 160, 161). This is particularly pertinent to the problems of archeological trephination and the custom of scalping. Infection, spreading from the intact scalp or through an open wound into the periosteum and bone of the cranial vault, tends to remain localized. The course is often protracted, eliciting a sclerotic response around a central, partly lytic, area around a sequestrum. The lesion usually is larger on the outer than on the inner table. Even direct infections of the opened diploë tend to remain circumscribed and take a chronic course with considerable perifocal sclerosis. Infected compound fracture may become the source of spreading osteomyelitis of the cranial vault, extending along the many venous channels of the diploë. In some instances, avascular necrosis of fracture fragments is followed by infection in and around the dead bone. For a detailed discussion of traumatic osteomyelitis of the skull, see Adel-

FIGURES 158, 159.—Osteomyelitis of skull vault, probably pyogenic, but syphilis with superimposed osteomyelitis cannot be excluded: 158, Outside view. 159, Endocranial view, showing marked involvement of inner table and sequestra. (WM S 43.1, from 1869.)

FIGURE 161.—Osteomyelitis of frontal bone with early sequestration, probably secondary to scalp infection. (Adult, HM P625.)





FIGURES 162, 163.—Osteomyelitis of posterior portion of skull secondary to chronic left middle ear infection and streptococcic meningitis: 162, Internal view, showing lytic foci in sulcus transversus. 163, External view, showing corresponding small defect of outer table. (67-year-old male, PMES 1E.B.1(2a).)

stein and Courville (1933). Localized, often chronic, and sclerotic osteomyelitis secondary to middle ear infections occurs in the mastoid process, the temporal, and the petrous bone. Occasionally ear infections spreading to the venous sinuses or meninges may lead to cranial osteomyelitis (Figures 162–164). The jaws of infants and small children, which are crowded with dental buds, may become infected from the mucosal surface or through the bloodstream. In the course of the infection, dental follicles may be sequestrated and extruded through mucosal ulcerations. Extension of the infection into the floor of the orbits before formation of a maxillary sinus is not uncommon. After 2 years of age, the infection is much more common in the mandible (Lauche, 1939:37). Baranoff (1934) reports that the mandible is affected in 91 percent and the maxilla in 9 percent of the cases in a Chinese series, most of which are in the young adult age range—16 to 30 years. The preponderance of the mandibular infections remains throughout the adult period, due to extension from oral, dental, and periodontal infections. The course is usually very chronic, revealing little pus formation and no sequestrum. The condition is dominated by osteosclerotic condensation, with or without thickening of the mandible (Panders and Hadders, 1970). The changes can simulate Paget's disease; however, the typical mosaic pattern of Paget's is not found microscopically. Other facial bones may be affected by

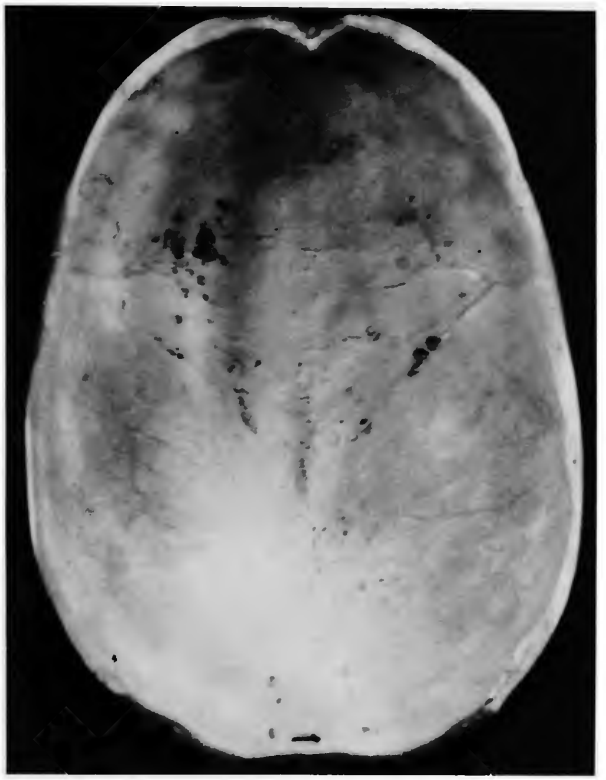


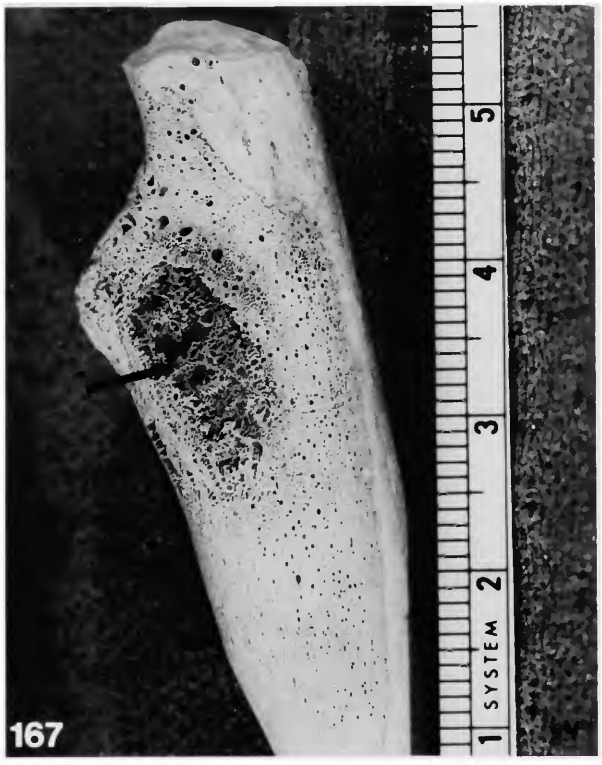
FIGURE 164.—Osteomyelitis of cranial vault secondary to chronic ear infection and streptococcic meningitis, showing disseminated lytic foci of the inner table along branches of both middle meningeal arteries. (67-year-old male, PMES 1E.B.1(2).)

extension of infection of the paranasal sinuses or of the facial soft tissues.

PALEOPATHOLOGY

In historical times, before the availability of antibiotics, osteomyelitis was a common clinical condition well known to physicians of the period. This would suggest that osteomyelitis might be common in archeological skeletons as well. However, there are few references to this disease entity in the literature on paleopathology. Hooton (1930:309) reports four cases of osteomyelitis in a total sample of 503 pre- and post-Columbian New World Pueblo Indians (0.8 percent). Wood-Jones (1910b:283), in his report of disease in ancient Egyptian skeletons, states that inflammatory conditions of bones are very rare. Even in open compound fracture, where osteomyelitis was such a dreadful sequela in historical times, evidence for purulent infection in Egyptian skeletons is very limited. Wood-Jones (1910b:283) concludes that the ancient Egyptians had a much greater resistance to infectious diseases of bone than the Europeans of his day. However, this conclusion may reflect inadequate or biased sampling of the European population, because Wood-Jones, as an anatomist, probably would have based his observation regarding the incidence of European osteomyelitis on its frequency in dissecting room cadavers. This source does not provide a human sample comparable with the ancient Egyptian skeletal sample. Although osteomyelitis may have been a common ailment in early twentieth century medical practice, particularly in the poorer, working-class people, this does not mean that the incidence of the disease in the general population was high. This is evidenced by the League of Nations epidemiological report for 1921 (1922), which does not include osteomyelitis as a significant disease entity. Thus, in an archeological skeletal sample, the incidence of osteomyelitis might be low but not differ significantly from a primitive or peasant population of today.

Both osteitis and periostitis are descriptive terms in the paleopathological literature to describe inflammatory conditions that could include osteomyelitis. By definition, osteomyelitis is an infection of bone involving the marrow. While this may be a helpful concept in the differential diagnosis of osteomyelitis in archeological specimens, it should be emphasized that clear evidence of marrow involvement may not be revealed in a gross specimen. It is due to such problems in diagnosis that scholars, reporting on lesions in archeological skeletons that could be osteomyelitis but are not clearly so, tend to avoid identifying a lesion as osteomyelitis, unless there is other evidence such as a cloaca or sequestrum to support this conclusion. Thus, many gross periosteal lesions possibly arising from osteomyelitis will not be attributable to this specific disease.



FIGURES 165–168.—Possible disseminated hematogenous osteomyelitis: 165, Large abscess of the first deciduous upper left molar in the skull of a child about 6 years of age; note the presence of a broad zone of reactive bone (arrow) on the maxilla peripheral to the abscess. 166, Periosteal reactive bone predominantly in the metaphyseal areas of the major long bones (scale in cm). 167, A central lytic focus (arrow) surrounded by periosteal reactive bone in the proximal right ulna. 168, Extensive accumulation of periosteal reactive bone in the metaphyseal region of the posterior, distal, left humerus. (NMNH 379345.)

The Skull

Wood-Jones (1910b:283) notes that dental disease accounts for almost all of the infectious conditions seen in the skull and mandible. This conclusion is supported in the clinical studies of Wilensky (1932:197) and Blum (1924:802). In ancient Nubian skulls, Wood-Jones (1910b:283–284) reports a case of bony destruction in the nasal and palatal regions of a young female who had worn and carious teeth. He attributes the destructive process to chronic rhinitis. Hooton (1930:310) reports three crania with inflammatory lesions, in which osteomyelitis is possible; however, syphilis is also thought possible. Roney (1966:102) reports two cases of osteomyelitis in a sample of 44 skeletons recovered from the pre-Columbian Pueblo Indian site at Mesa Verde. The bones involved include the mandible, ulna, femur, and tibia. Infectious conditions of the mastoid region have been reported in ancient Nubian skulls by Wood-Jones (1910b:284).

While a more detailed treatment of infection arising from dental disease is presented (p. 438), brief mention is appropriate here regarding complications of caries that can affect bone and possibly be a focal point for systemic infection. One possible example of this condition occurs in the skeleton of a young child in the skeletal collections of the National Museum of Natural History, Smithsonian Institution, Washington, D.C. (NMNH 379345). The specimen is from a site in Virginia, USA, and is dated about A.D. 1550. In the left maxilla there is a periapical abscess, which has destroyed bone adjacent to the root of the first deciduous upper left molar on the buccal side (Figure 165). A zone of periosteal, reactive bone extends beyond the lytic focus for 1–2 centimeters. There is clear evidence of caries in the deciduous second molar, leaving no doubt regarding the initial focus of the infectious process. The long bones of the skeleton show extensive development of periosteal, reactive bone indicative of a disseminated, inflammatory condition (Figures 166–168). While it could be that the infectious condition of the maxilla and the inflammatory reaction in the long bones are unrelated, it is possible that the infectious condition in the maxilla was the initial focus for hematogenous dissemination of the infectious organism. Such a condition could have stimulated a general, periosteal response before the child succumbed to this or other problems.

Postcranial Bones

The best diagnostic evidence for osteomyelitis in skeletal material is a drainage canal in bone (cloaca) or sequestration in association with periosteal bone formation (involucrum). Lacking this, inflammatory lesions of the long bones are difficult to attribute to osteomyelitis.

Wood-Jones (1910b:287) reports an ancient Nubian example of a humerus with a necrotic distal portion associated with a superficial layer of inflammatory bone. He attributed this to an infectious complication of an injury.

Hooton (1930:308) found 13 out of 503 Pueblo skeletons that had nonspecific periostitis, some of which could have been osteomyelitis. However, he did identify four cases that he judged to be osteomyelitis (Hooton, 1930:309). Cressman and Larsell (1945:332) reported a case of probable osteomyelitis in a prehistoric Indian skeleton from Oregon, USA. The right arm was fused at the elbow and the left tibia, tarsals, and metatarsals were fused together. They attribute this condition to osteomyelitis acquired during childhood. The infection resulted in septic arthritis. Jarcho, Simon, and Bick (1963) have published their observations on a pre-Columbian, Pueblo skeleton from Arizona, USA, in which the left femoral head was fused to the innominate. They suggest the possibility that this condition is septic arthritis with ankylosis following infection, possibly of pneumococcal, gonococcal, staphylococcal, or streptococcal origin.

In the skeletal collections of the National Museum of Natural History, Washington, D.C., USA, there are several archeological skeletons with lesions, which could appropriately be attributed to osteomyelitis. Perhaps the most clearly identifiable example is a case of a juvenile from Peru (NMNH 378243). All that was recovered of

this skeleton is the right and left tibia, both of which are in an excellent state of preservation. The estimated age, based on the length of the normal, right tibia, is 9 years. Unfortunately, the archeological provenience is poorly documented. However, it seems likely that the specimen is pre-Columbian.

Grossly the two tibiae differ dramatically in size (Figures 169–175). The measurements in Table 2 indicate the marked increase in size of the pathological left tibia in contrast with the normal right tibia. Osteomyelitis results in hyperemia, which may stimulate excessive growth in the affected bone. This effect of the disease is clearly seen in the two tibiae of this case. The excessive growth of periosteal involucrum is the result of pathological stimulation in response to the death of the cortex. This problem does not exist at the growth plate where excessive, longitudinal growth is stimulated by the general hyperemia.

TABLE 2.—Metric comparisons of the right (normal) and left (pathological) tibia in osteomyelitis from a 9-year-old child from an archeological site near La Oroya, Peru (NMNH 378243)

Measurement	*Left* (mm)	*Right* (mm)
Maximum length	219	196
Maximum midshaft diameter	34	17
Mediolateral diameter of proximal growth plate	50	45

While the proximal growth plate of the pathological, left tibia is enlarged, the surface appears normal, although there is a postmortem break in the surface. There is a cloaca one centimeter below the growth plate, but it does not penetrate it. The distal medial cortex of this bone has a large, cloacal opening with a channel penetrating the distal growth plate. There is also some postmortem damage. The cloacal openings penetrate into the marrow space and are lined with smooth compact bone indicating a long-standing chronic condition.

The proximal metaphysis of the left tibia contains a cloaca in the anterior cortex. There is a small, bony spur on the lateral anterior cortex, which is a localized periosteal reaction. The distal metaphysis has a cloaca on the medial aspect, which is continuous with the opening in the growth plate mentioned above.

The diseased shaft contains a sequestrum, which projects through a large cloaca on the anterior shaft (Figure 170). There is a smaller (5 mm diameter) cloaca inferior to the large cloaca. Both of these openings are part of a single channel that contains the sequestrum. There is a small, lytic pit near the sequestrum and a cloaca in the posterior portion of the distal shaft.

The roentgen-film appearance of the diseased left tibia shows a greatly thickened, but porous, involucrum. The old sequestrated shaft is still apparent, although the cortex of the metaphyses has been removed by remodeling. The diameter of the shaft is compatible with an age of a 4- to 5-year-old child. Since sequestration would most likely have occurred shortly after the onset of the disease, this suggests that the child suffered from osteomyelitis for about 5 years before death.

In the lateral roentgen view of the osteomyelitic tibia, there appears to be another, much smaller, sequestrum in the distal anterior cortex (Figure 175). Grossly, there is a bulge in this area but no cloaca. There are other roentgen features also suggesting sequestra; however, their identification on the X-ray film is less clear cut.

A comparison of Harris' lines also offers some insight into the disease process. In the normal (right) tibia there are four distinct lines in the distal and three in the proximal ends. The osteomyelitic tibia contains two distinct Harris' lines and two much less distinct lines in the distal tibia, which may correspond to the four distal lines seen in the normal tibia. There is no evidence of Harris' lines in the proximal end of the pathological tibia. If they existed the disease has obliterated the roentgen evidence.

Table 3 shows the distances between the Harris' lines in both distal tibiae. Since the distances between all Harris' lines are greater in the osteomyelitic tibia it would appear that accelerated growth has characterized all stages of the disease process. Table 4 shows the distances between the distal Harris' lines and the growth plate. In every

TABLE 3.—Distance (in millimeters) between the Harris' lines for the right (normal) and left (pathological) tibia of NMNH 378243 (lines 1 and 4 are indistinct in the left tibia, making the associated measurement somewhat speculative as emphasized by the question marks)

Harris' line	*Left* (mm)	*Right* (mm)
First to second	20 (?)	18
Second to third	27	21
Third to fourth	6 (?)	3
Total	53	42

TABLE 4.—Distance (in millimeters) between the distal Harris' lines and the distal growth plate of the right (normal) and left (pathological) tibiae of NMNH 378243 (lines 1 and 4 are indistinct in the left tibia, making the associated measurement somewhat speculative as emphasized by the question marks)

Harris' line	*Left* (mm)	*Right* (mm)
First to growth plate	63 (?)	50
Second to growth plate	42	32
Third to growth plate	15	10
Fourth to growth plate	8 (?)	3

case the distance is greater in the pathological tibia.

If we assume that growth in the distal tibia accounts for approximately two-thirds of total tibial growth, we can get an estimate of the age of the individual when the first Harris' line was formed. Doing this for the normal tibia only we find that there is 50 millimeters from the first Harris' line to the growth plate. This translates to about 75 millimeters total growth for the tibia from the first arrested growth line. Extrapolating from Stewart (1968:133) the tibia grows about 17 millimeters per year between the ages of 4 and 9 years. At this rate, it would take approximately 4½ years to add 75 millimeters to the total length of the tibia. Since the child was about 9 years old at the time of death, this suggests that the first Harris' line was formed between the ages of 4 and 5 years. Because this age is in agreement with the age estimated on the basis of the midshaft diameter of the sequestrated tibial shaft, this first Harris' line may coincide with the time of onset of the osteomyelitis.

The second case I shall review in considerable detail is an Eskimo specimen from Shishmaref, Alaska (NMNH 332611). This case is illustrative of the problems encountered in identifying the specific cause of an infectious condition. The archeological age is uncertain, but a bullet wound in the skull indicates a date in the postcontact period.

The specimen is clearly male with an age at death estimated to be about 23 years on the basis of pubic symphysis morphology. The skull and long bones both suggest a muscular, robust individual.

There are two pathological conditions in this specimen. Undoubtedly, the immediate cause of death was a bullet injury to the skull, in which the bullet entered the occipital region and exited, with a typical fracturing pattern, through the left parietal (Figure 176). The other pathological condition is a largely destructive process limited to the lumbar vertebrae and sacrum (Figures 177, 178).

The first lumbar vertebra is only slightly affected by the disease process with barely visible, lytic foci on the lateral parts of the vertebral body. The second lumbar vertebra has large, lytic, scalloped lesions in the posterior part of the vertebral body. These lesions are continuous with the spinal canal and on the right side extend into the transverse process. The proximal and distal endplates of the vertebral body and the true joints of the superior and inferior processes are unaffected in the first two lumbar vertebrae. There are three lytic depressions approximately one centimeter wide on the left lateral and anterior cortex of the second lumbar vertebra. In all of the lytic depressions there is a varying amount of bone remodeling in the spaces of the spongy bone in the vertebral body adjacent to the disease process. This is a typical response by bone, particularly spongy bone, to a destructive disease process. In slowly developing, chronic, destructive conditions, there may be a continuous wall of bone adjacent to the disease focus. There has been a limited, but incomplete, osteoblastic response to

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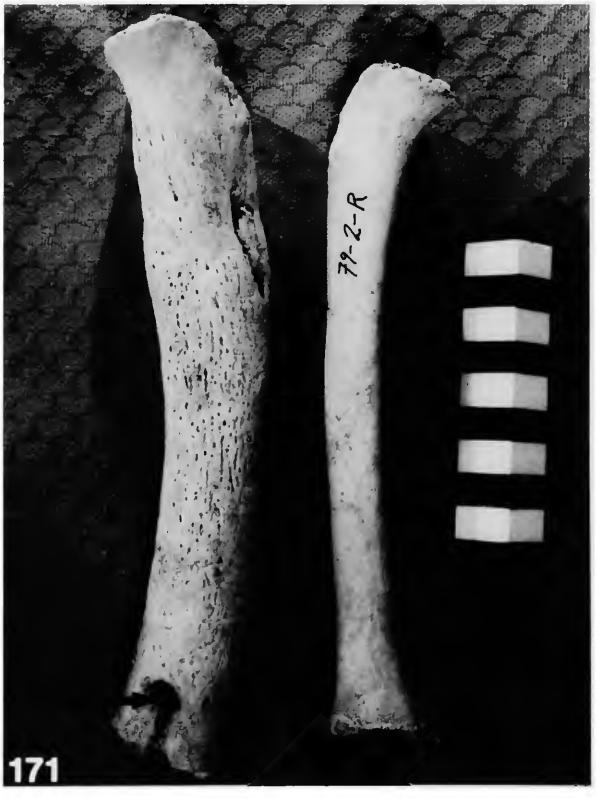

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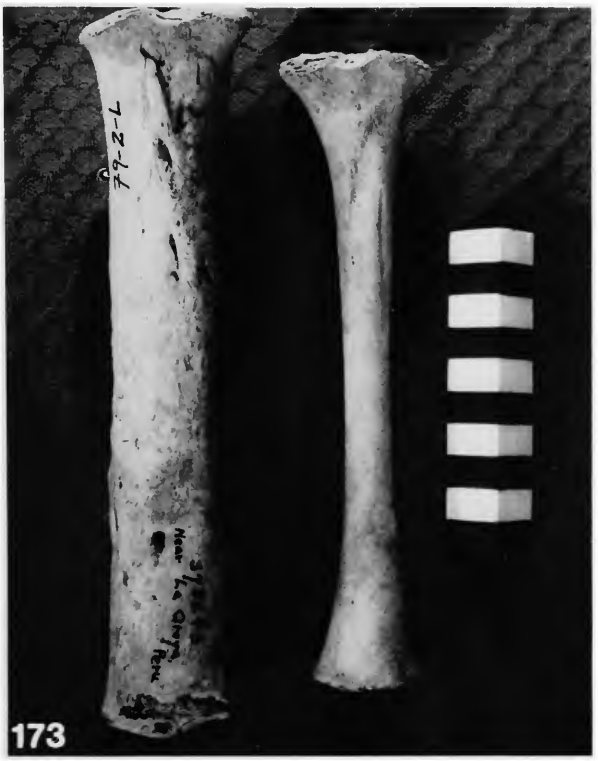
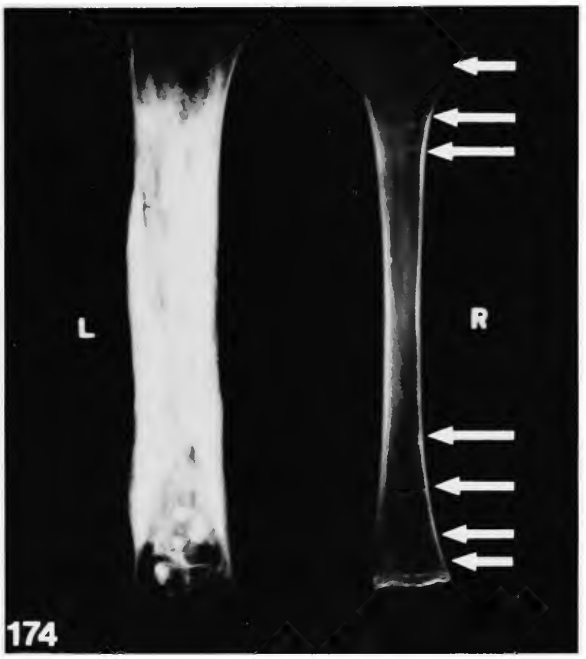
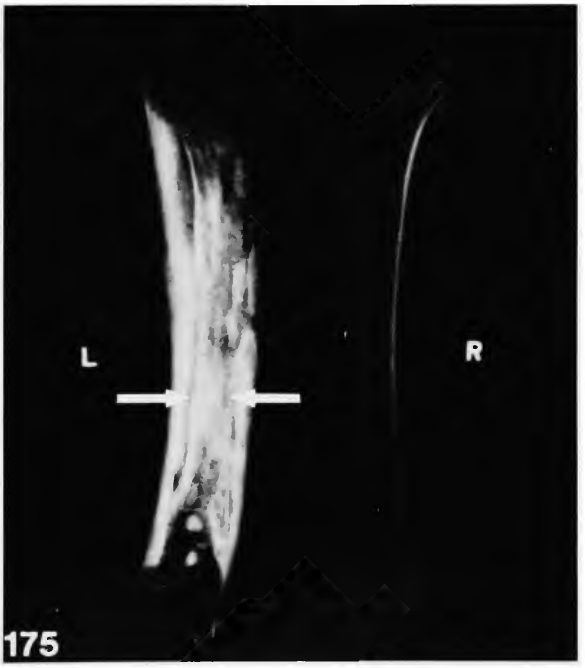
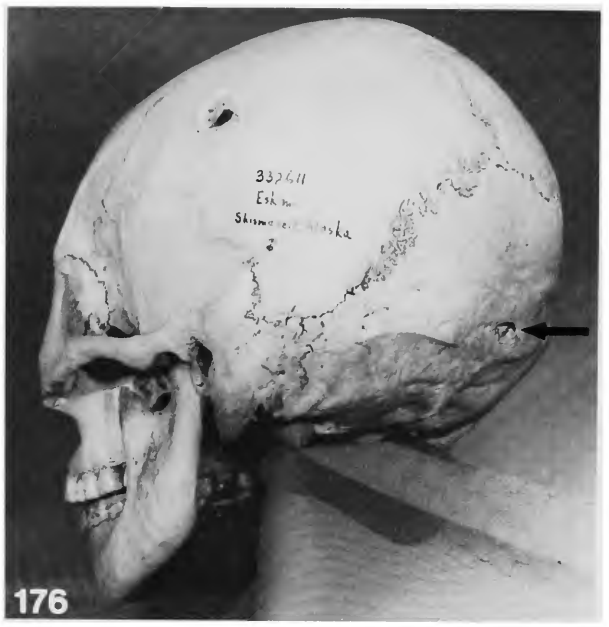






FIGURES 169–175.—Osteomyelitis of the left tibia compared with the normal (right) tibia of a child about 9 years of age from an archeological site near La Oroya, Peru: 169, Anterior view; note the sequestrum (arrow) and cloacae. 170, Detail of sequestrum and cloaca (arrow) in left tibia. 171, Medial view of pathological and normal tibia. Large channel penetrates the distal growth plate (arrow). 172, Detailed view of channel penetrating the distal growth plate of the pathological tibia. 173, Posterior view of pathological and normal tibia. 174, Anteroposterior X-ray film of pathological and normal tibia; note the presence of Harris' lines in the normal tibia (arrows). 175, Medio-lateral X-ray film of pathological and normal tibia. The cortex, which was sequestrated at the onset of the disease, is apparent (arrows) but is completely surrounded by involucrum, which forms the externally visible cortex. (NMNH 378243; scales in cm.)



FIGURES 176–178.—Possible osteomyelitis in a young adult male Eskimo skeleton with a bullet wound in the skull and destructive lesions of the lumbar vertebrae: 176, Posterolateral view of skull, showing entry (arrow) and exit wounds from a bullet. 177, Destructive lesions primarily affecting the third and fourth vertebral bodies in an anterior view of the lumbar vertebrae and first two segments of the sacrum. 178, Right, lateral view of lumbar vertebrae and first segment of the sacrum showing virtually complete destruction of the third and fourth vertebral bodies. (NMNH 332611.)

the disease suggesting a somewhat chronic condition. In the third lumbar vertebra, the entire vertebral body, the left transverse process, and the left superior articular process have been entirely destroyed. There is a slight amount of reactive bone adjacent to the lytic areas. The spinous process is normal. Like the third lumbar, the vertebral body of the fourth lumbar vertebra has been completely destroyed. However, all articular processes are present, although the left articular facets have been partially destroyed. There are long spurs coming off the transverse processes with the most prominent one found on the right side. The anterosuperior plate of the body of the fifth lumbar vertebra is destroyed. However, the articular surface of the inferior body is largely intact. There is a large depression on each of the sides of the vertebral body and evidence of cavitation, mostly destroyed postmortem, on the anterior vertebral body. There has been some osteoblastic activity, which had begun to fill in spongy bone forming a new cortex. The vertebral processes are generally intact although the left superior articular facet is porous with fine reactive bone around the porous joint area. There is some periosteal reaction on the left dorsal lamina. In the first segment of the sacrum the anterosuperior portion of the body is destroyed as is the inferior body of the same segment. There are large, well-circumscribed cavities in the body. Both alae of the first segment have a lytic cavity extending into them. However, the articular surface with the innominate bone is normal. The articular facets, sacral canal, as well as spinous and articular tubercles, are unaffected. The second segment is affected only on the superior portion of the body. There are lytic lesions 1 to 2 centimeters in diameter, some of which appear to be continuous with lesions in the body of the first segment.

In this case the lesions are predominately lytic, but with some time for an osteoblastic response to the disease process before the individual died. The skeletal remains suggest a disease process with an initial, acute, destructive phase followed by a short but more chronic phase with an exclusive involvement in the lumbosacral vertebrae. Both osteomyelitis and tuberculosis are strong possibilities and neither can be ruled out. There are several factors, however, that more strongly indicate osteomyelitis.

It has been noted that vertebral arches, joints, and spinous processes are almost never destroyed in tuberculosis (p. 145). In this specimen there is destruction of transverse processes, diarthrodial intervertebral joints and portions of the arch. Furthermore, the extent of osteoblastic response at this stage in the lytic process would be less likely in tuberculosis than in osteomyelitis.

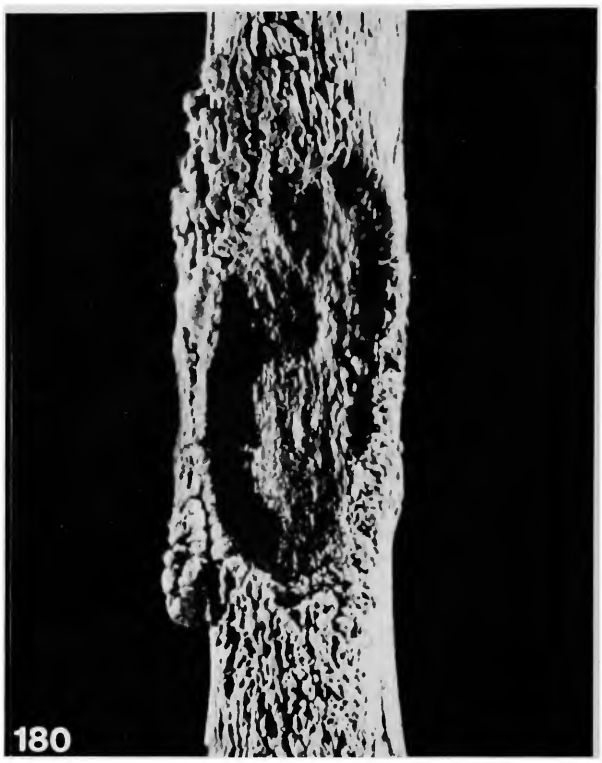
Periostitis

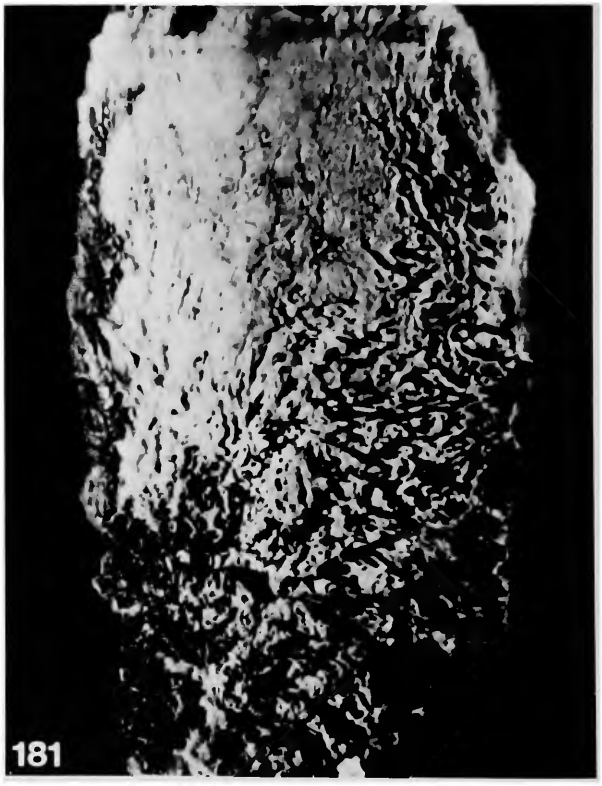
Pathology

Periostitis, as a disease by itself, is uncommon. It usually represents part of, or a reaction to, pathologic changes of the underlying bone. The inner layer of the periosteum retains osteoblastic capacity throughout life, even after termination of growth. It is, therefore, not surprising that the periosteum reacts to many different insults with formation of woven bone, which later may become incorporated into the underlying cortex and remodeled into lamellar bone. This periosteal bone formation is not always an expression of inflammation and certainly not restricted to infection. In most of the infections discussed in this chapter, periosteal bone formation is one of the significant changes. Of these changes, first and foremost, inflammatory periostitis plays a role in the infections of bone, specific and nonspecific. Obviously, transitory periosteal reaction, which neither leaves permanent bone deposition nor resorptive pitting of the underlying cortex, cannot be recognized on dry bone. This brings up the question: What are the characteristics of inflammatory periosteal bone? There are no unequivocal characteristics. Generally, inflammatory periosteal bone deposited over a long period of time tends to be unevenly distributed, not involving the entire bone. The surface tends to be irregular and the thickness often variable. The marked,

FIGURE 179.—Periostitis and superficial osteomyelitis of right tibia beneath skin ulcers. Notice shield-like periosteal buildup under major ulcer. (WM S 47.2 from 1831.)

uneven hypervascularity visible on dry bone in the form of smaller and larger pores in periosteal bone is often striking. In most cases, changes in the underlying bone will answer the question, but a purely subperiosteal bone deposit may be difficult or impossible to classify pathogenetically. In areas where the periosteum is close to the skin surface, as on the anterior surface of the tibia, localized ossifying periostitis can be observed secondary to trauma. Such longstanding periosteal bone deposition can be completely incorporated into the cortex in form of remodeled lamellar





FIGURES 180, 181.—Periostitis and superficial osteomyelitis of tibia beneath leg ulcer. 180, Superficial cortical defect surrounded by reactive periosteal bone. 181, Sequestrum of reactive bone over cortical defect, beneath leg ulcer. (HM P673 and 674.)

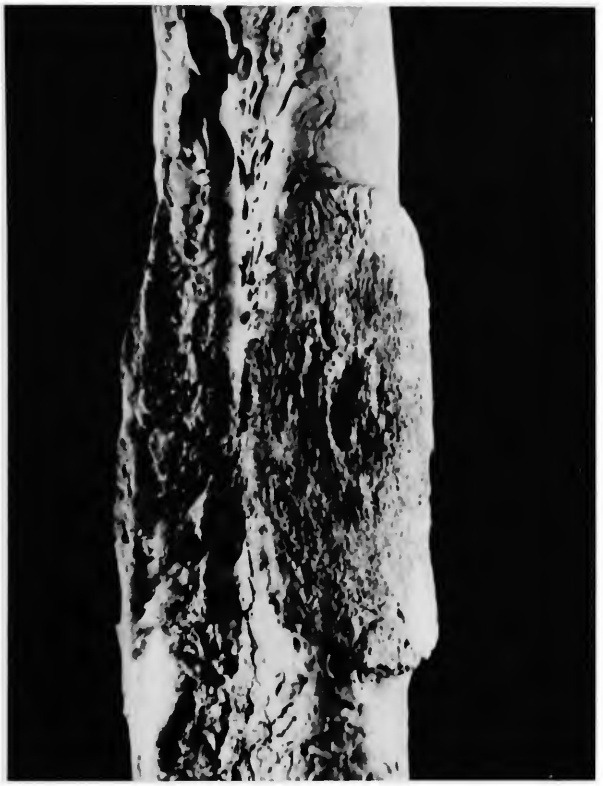


FIGURE 182.—Ossifying periostitis of right tibia beneath leg ulcer, synostosis of tibia and fibula. (Adult, WM S 47.1 before 1831.)

bone. Large chronic ulcers on the skin, especially those due to venous stasis, not uncommonly produce a reactive, local, ossifying periostitis on the tibia (Figure 179). This may represent a plaque-like deposition of periosteal bone of considerable thickness, roughly copying the outline of the ulcer. Occasionally such deposits may separate as shallow sequestra (Figures 180, 181). In other cases, rough exostosis-like osteophytes, which flatten out below the ulcer, may occur. This is observed particularly in tropical ulcer, a skin ulcer over the shin, characterized by presence of fusiform bacilli and saprophytic spirochetes (Brown and Middlemiss, 1956). Occasionally, such reactive bone deposits may bridge the gap between tibia and fibula (Figure 182). Massive reactive periosteal bone formation of the mandible used to be observed in the nineteenth century in persons exposed to vapors of yellow phosphorus for a long period (Figure 183).

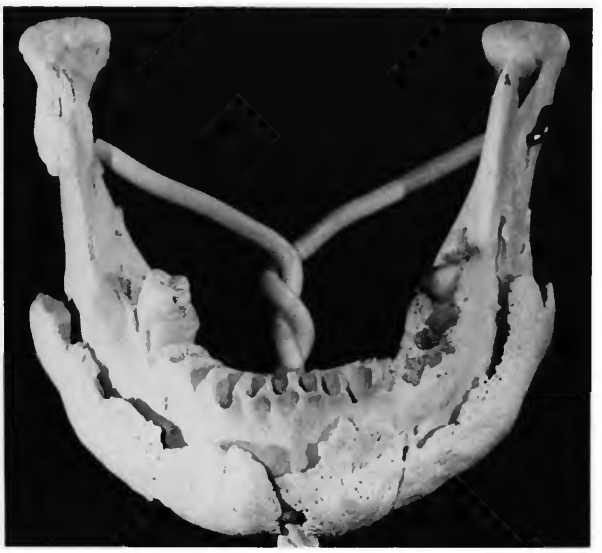


FIGURE 183.—Phosphorus necrosis of mandible with involucrum. (IPAZ 1758, no date.)

PALEOPATHOLOGY

While periostitis, as an isolated disease process, is uncommon in modern medical literature and clinical practice, undifferentiated, nonspecific, periosteal lesions of long bones are rather common in archeological skeletons. The main reason for this difference between clinical and paleopathology is that many of the periosteal reactions may be part of the expression of a specific disease process, which can be identified in a living patient, whereas in archeological specimens the pathological characteristics necessary to make a specific diagnosis are not available. This would have the effect of increasing the frequency of nonspecific periostitis in archeological skeletal series.

It is important to emphasize that periostitis (as is also the case with osteomyelitis) has both a general descriptive usage as a term and a discrete usage. Thus, periostitis is part of a disease syndrome such as syphilis, but it is also a specific disease itself. This varied usage of the term is well illustrated in Brothwell and Sandison's book, *Diseases in Antiquity* (1967). The index to their work

lists six references to periostitis. Of these six, the term is used five times in the context of a syndrome associated with a specific disease, such as syphilis. Only once is the term applied to primary periostitis. Since we discuss secondary periostitis in the context of specific disease entities, the following discussion on the paleopathology of periostitis will focus on the primary type. However, the designation of primary periostitis in paleopathological skeletons may mean only that a more specific diagnosis is impossible and does not eliminate the possibility that the periosteal reaction is secondary to a specific disease process.

Primary periostitis is most often the result of two pathological conditions, trauma and infection (Senn, 1886:5–6). Putschar (1966:60) notes that it is often impossible to determine which of these two conditions gave rise to a given lesion in an archeological skeleton. While the periosteum will always be activated in fracture, periosteal reactive bone can also be stimulated by injury that does not produce fracture. The latter may resemble periosteal reactions stimulated by localized infectious foci.

Traumatic periostitis is brought on by sudden or chronic insult to bone. Wood-Jones (1910b: 285–286) has described a periostitis of the skull common in Nubian archeological specimens. The initial lesion is characterized by hypervascularity either on or near the vertex of the skull. However, there are also small spicules of bone formed, and later stages may be characterized by hypervascularity and, in extreme cases, exfoliation of portions of the skull. Wood-Jones attributes this periostitis to chronic insult produced by women carrying water jars on their heads. This observation is supported by the fact that the lesion occurs much more often in female skulls. Periosteal reactions of the skull are occasionally associated with trephination; however, here the periostitis is probably due to complication of the surgical procedure by infection.

Periostitis of long bones is often seen in the tibiae of archeological skeletons. The reasons for this localization remain obscure. It is, however, instructive to note that periosteal reaction in syphilis forms on bones that tend to be near the skin surface, such as the tibia and skull vault. Such bones may be somewhat cooler than bones like the femur, which are enclosed in a heavy mass of muscle and fat. It is also true that bones near the skin are more exposed to direct trauma than bones protected by overlying muscle. Perhaps both these factors are significant in localization of periostitis.

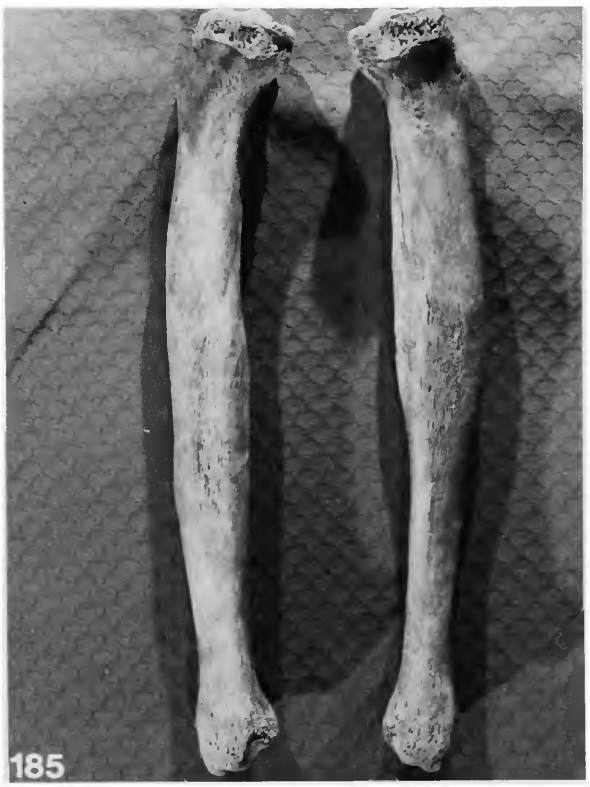
The distinction between osteomyelitis and periostitis cannot always be made in dry bone specimens. However, in periostitis there will be no cloacae, involucrum, or changes in the marrow cavity. Furthermore, periosteal bone tends to be superficial to the normal cortex, at least in early stages of the disease condition. The superficial nature of periosteal lesions can be seen in a skeletal specimen from Ossuary II at the Juhle site near Nanjemoy, Maryland, USA (NMNH 384380). The specimen is dated to the sixteenth century A.D. There is no evidence of European contact. The secondary nature of the burials makes it virtually impossible to obtain complete skeletons. However, the two long bones in question, the left ulna and tibia, exhibit similar periosteal lesions and both are from an individual between 14 and 18 years of age. Thus, the probability is high that the two bones came from the same individual. The periosteal, reactive bone is superficial to the intact cortex (Figure 184) and is porous fiber bone rather than compact bone. There is no evidence of a cloaca or marrow involvement. Thus, a diagnosis of primary periostitis is justified.

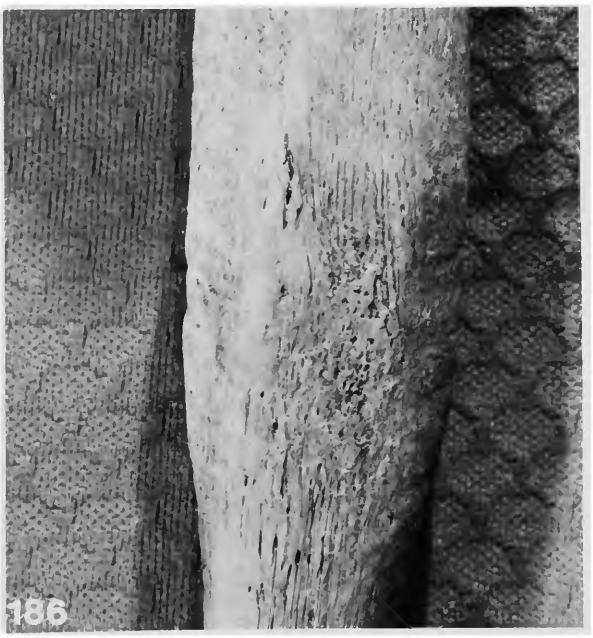
In his study of pre- and post-Columbian Pecos Pueblo skeletal material, Hooton (1930:308–309) distinguished between periostitis and osteomyelitis. Although he does not indicate the criteria by which this distinction is made, Hooton diagnosed periostitis in 13 of 503 skeletons, an incidence of 2.6 percent. There were four cases of osteomyelitis found in 503 skeletons indicating that periosteal reactions were more common. Morse (1969:17, 106–111) presents four prehistoric cases of periostitis in his monograph on paleopathology in archeological skeletons from the midwestern United

FIGURE 184.—Periosteal reactive bone on the left ulna of an individual between 14 and 18 years of age. Some of the periosteal reactive bone was broken away postmortem revealing the underlying cortex (arrow), which exhibits minimal changes. Specimen from an archeological site in Maryland, USA. (NMNH 384380; scale in cm.)

States. With the possible exception of Burial 42 from the Klunk site in Illinois, these cases would appear to be secondary reactions to infection. In the following cases from the collections of the National Museum of Natural History, Washington, D. C., USA, I illustrate a few examples of periostitis ranging from what would appear to be primary periostitis to those in which a distinction between primary and secondary periostitis cannot be made.

One example of a nonspecific periosteal lesion is found in a skeleton (NMNH 308679) from the Hawikuh archeological site in New Mexico. The skeletons associated with the site are dated archeologically between A.D. 1200 and 1670. The individual was female and between 25 and 35 years of age. The preservation of the skeleton is excellent, with virtually all bones present except some





FIGURES 185, 186.—Bilateral midshaft thickening: 185, Left and right tibiae of an archeological skeleton from the site of Hawikuh, New Mexico, USA. 186, A detailed view of the thickened midshaft of the left tibia; note the lack of any foci of active reaction. This lesion represents a healed periosteal lesion. (NMNH 308679.)

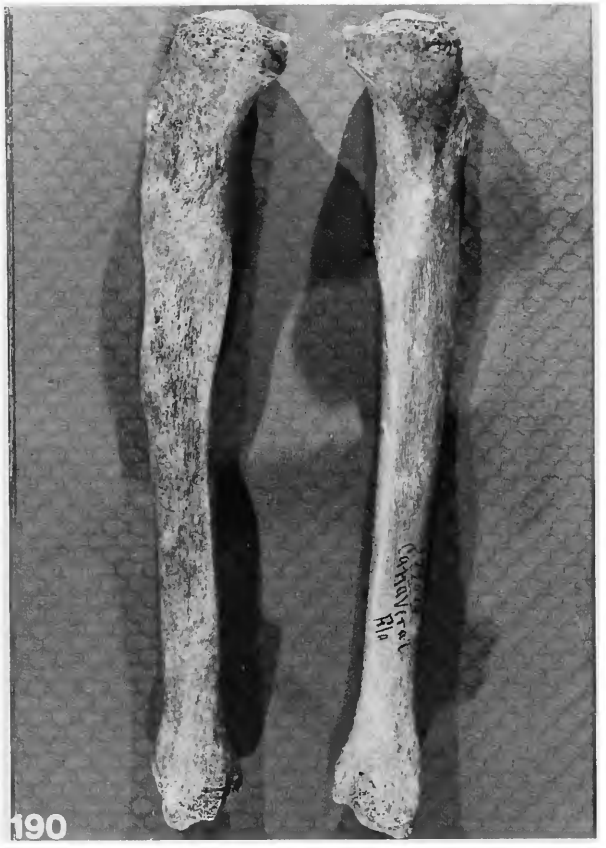
FIGURES 187–189.—The right and left tibiae of a post-Columbian adult male skeleton from an archeological site in Florida, USA: 187, Lateral view; note the presence of a porous, thickened lesion below midshaft on the right tibia. The left tibia is normal. 188, A detailed view of the lateral aspect of the lesion on the right tibia. 189, The medial view of the right and left tibiae. The thickened area is less porous than the lateral aspect of the lesion. (NMNH 377457.)

of the hands and feet. The skull is normal except for a slight amount of occipital flattening, dental caries, and the loss of the lower left second molar antemortem. There is a supernumerary premolar in the left mandible. The postcranial skeleton is also normal except for a bilateral, diaphysial, cortical thickening of the tibiae (Figures 185, 186). Grossly, the thickening takes place circumferentially. The lesions of both tibiae are remarkably similar. The bone surface of both lesions is smooth except for a medial focus, which has a slightly raised plaque adjacent to a region of longitudinal striations. The overall appearance of the lesions is one of a well-contained chronic process. There are no cloacae and no evidence of fracture.

The original cortex of both tibiae has been partially remodeled near the main focus of the lesion. A somewhat greater amount of periosteal bone has been added on the lateral side, and it is on this side that the maximum cortical remodeling has taken place. The predominant lesion is periosteal and thus it is appropriately classified as periostitis. However, there is evidence of encroachment by bone reacting to the disease in the marrow cavity. This means that the focus of the lesion could have been the marrow cavity with secondary activation of the periosteum, as does occur in osteomyelitis. However, the superficial nature of the lesion and minimal medullary change make a periosteal origin somewhat more probable.

Another example of periostitis is seen in an archeological skeleton from the Canaveral Peninsula of Florida, USA (NMNH 377457). The skeleton is an adult male approximately 30 years of age as estimated by pubic symphysis morphology. Arthritic degeneration elsewhere in the skeleton is suggestive of a somewhat older age. The presence of trade beads indicates a post-Columbian date for the site. The skull vault is normal, although the temporal portion of the temporomandibular joint exhibits considerable arthritic degeneration. The mandibular portion of the joint is much less severely affected. The dentition is considerably worn, as is typical of this population, and there is evidence of at least two abscessed teeth. The postcranial skeleton is incomplete. With the exception of severe arthritic degeneration of the right elbow, there is no evidence of pathology except for a lesion on the shaft of the right tibia (Figures 187–189). This lesion begins at midshaft and extends distally 10 centimeters. It involves both the medial and lateral sides of the shaft and encompasses most, but not all, of the circumference. On the medial side, the lesion consists of fine porous bone with a longitudinally striated appearance at the boundary with normal bone. Laterally the lesion has a more irregular surface but is still composed of fine porous bone. Posteriorly there are two small isolated plaques of fine periosteal bone located distally to the main lesion. The overall appearance of the lesion suggests an active, but chronic, inflammation limited to a single bone of the skeleton. The location, morphology, and circumscribed nature of the lesion is suggestive of the bony reaction to an overlying skin ulcer. The X-ray picture of the two tibiae is of poor quality due to soil infiltration into the cortex and medullary cavity. There has been a slight degree of remodeling of the original cortex beneath the lesion, in which the lytic process predominates. There may have been slight encroachment into the medullary cavity by reactive bone, although the predominant disease process is periosteal.

Another case from the same site in Canaveral, Florida (NMNH 377516) illustrates a periosteal reaction apparently resulting from a penetrating wound into the medial tibia just proximal to the midshaft. The main focus of the pathology on the tibia is a central, elongated, sharply outlined, traumatic defect (Figures 190–191), surrounded by a thickened periosteal lesion. There is a second periosteal lesion just inferior to the tuberosity. This lesion does not contain a central cavity. The skeleton is an adult male with an age estimated to be 24 to 30 years on the basis of pubic symphysis morphology. There is a hint of reactive periostitis on the skull near vertex and a small depression on the right frontal bone. The right humerus has a thickened periosteal lesion in the distal medial region. Adjacent to this lesion is a lytic focus. The left ulna has a slight swelling of

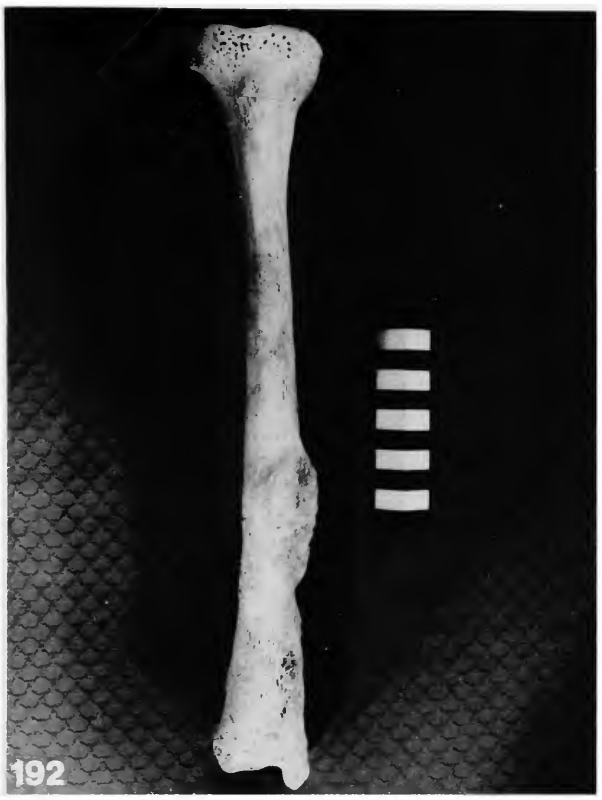


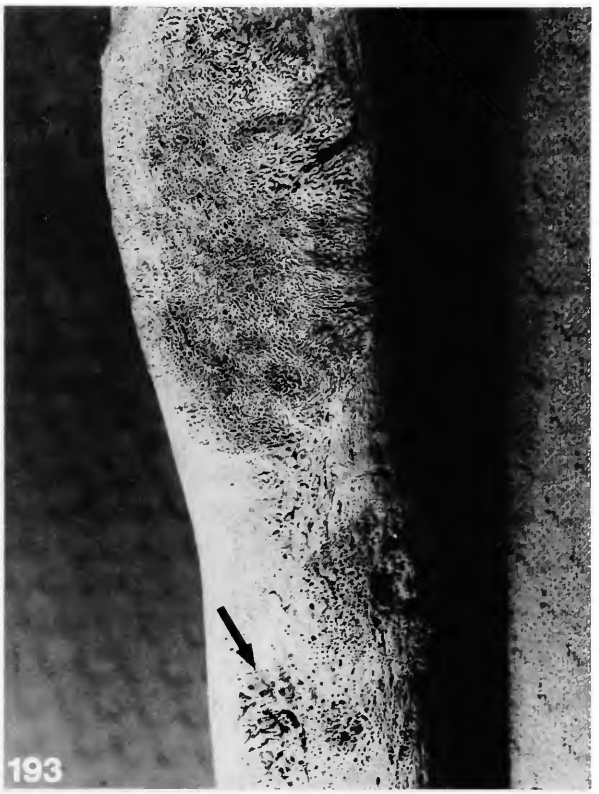
the distal diaphysis with an irregular surface. There is no corresponding pathological involvement on the radius. A comparison of the lateral roentgen images of the two tibiae suggests a slight reactive remodeling in the marrow cavity of the pathological right bone. However, the predominant lesion is periosteal. There is no apparent sclerotic reaction surrounding the central cavity in the most distal lesion.

If the tibial lesion was the only lesion in the skeleton, a periosteal reaction to a local trauma (perhaps a sharp penetrating object embedded in the bone) would be the most probable diagnosis and, indeed, this remains a strong probability. However, the complications introduced because of the other bone lesions raises the possibility of a systemic disease affecting many parts of the skeleton, but producing a more pronounced lesion in a bone traumatized by a penetrating wound. In any case, we do have a lesion on the tibia, which is not bilateral and seems to be associated with a localized trauma. As such, it provides an example of what one might expect to find in periostitis resulting from injury to bone.

There are several examples of periosteal reactions to skin ulcers in the human skeletal collections of the National Museum of Natural History, Washington, D. C. One of these illustrates the distinctive external morphology associated with this condition. This specimen is a right tibia excavated from Ossuary II at the Juhle site near Nanjemoy, Maryland, USA (NMNH 384299). This site is dated to the sixteenth century A.D. There is no evidence of European trade contact. There is a large area of reactive thickening on the medial part of the bone just below the midshaft. This lesion measures 65 by 40 millimeters (Figure

FIGURES 190, 191.—Right and left tibiae of a post-Columbian adult male skeleton from an archeological site in Florida, USA: 190, Medial view; note the periosteal reactive bone proximal to midshaft. There is a central lytic focus that might have been traumatically induced. 191, Detailed view of the central lytic focus (arrow) on the right tibia. There is evidence of active periostitis surrounding the lytic area. The lytic depression does not penetrate to the marrow. (NMNH 377516.)

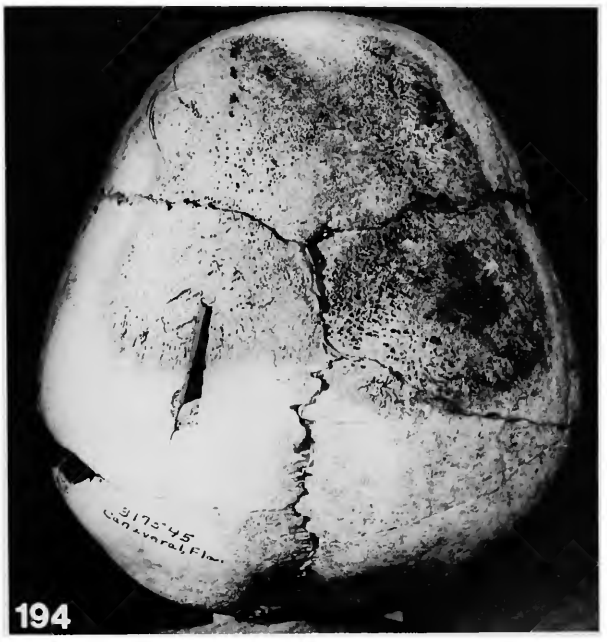


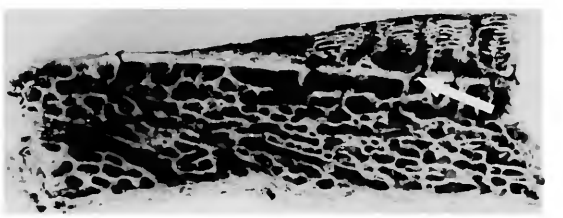


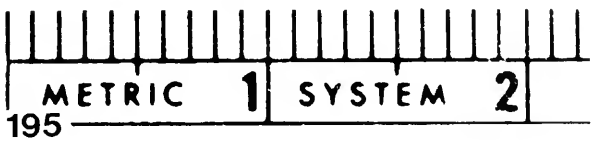
FIGURES 192, 193.—Right adult tibia from the Juhle site in Maryland, USA. This site is dated to the sixteenth century. There is no evidence of European contact. 192, Anterior view; note the large area of reactive bone on the medial side just distal to midshaft (scale in cm). 193, A detailed view of the bony reaction to an overlying skin ulcer. Distal to this lesion is an additional periosteal reaction, which is not as well organized (arrow). (NMNH 384299.)

192). There is a nodular thickening superior to this lesion and another area of reactive bone on the distal metaphysis (Figure 193). The sharply demarcated, thickened nature of the central lesion is characteristic of a bony response to a skin ulcer. This case appears to have been complicated by disseminated infection giving rise to the two other foci of periostitis.

Periosteal skull lesions are also seen in archeological materials and present a challenge to the paleopathologist because of their resemblance to the expansive, porous bone seen in the anemias. An example of skull periostitis is seen in a skull of a young child (NMNH 377545). The dental age of the specimen is about 3 years. The presence of trade beads indicates a post-Columbian date for the site, located on the Canaveral Peninsula in Florida, USA. The major lesion on the skull involves the external surface of the skull and includes the contiguous portions of the frontal and both parietal bones (Figure 194). Less extensive lesions can be seen on the supraorbital region of the left frontal, lateral left parietal, and left temporal bones. These last two lesions may have been bilateral, although postmortem damage prevents confirmation on the right side. Porous lesions are also seen in the left orbital roof. A diffuse, porous lesion appears on the posterior part of the skull, primarily on the parietals near the lambdoid suture. The maxilla, both facial and palatine portions, is also affected, as is the anterior portion of the mandible. In the temporal bone, the maxilla, and the mandible, the lesions appear to involve the spongiosa. The other lesions, including the large lesion on the vault, are superficial. A cut section from the left parietal







FIGURES 194, 195.—An extensive periosteal lesion concentrated in the anterior region of the skull vault: 194, Top view of skull. 195, Detailed view of cut section of bone including areas of the lesion and normal bone. Note that the periosteal bone has been layed down on the intact cortex (arrow) and that there is no expansion of the diploë. (3-year-old child from a post-Columbian site on the Canaveral Peninsula, Florida, USA, NMNH 377545.)

shows the outer cortex to be intact with abnormal bone, added in layers, superficially over the outer cortex (Figure 195). Unfortunately there are no postcranial bones associated with the skull.

Brucellosis

PATHOLOGY

Brucella is a genus of gram negative rods (bacilli) containing three species pathogenic to domestic animals and through them to man: *Brucella abortus*, which causes miscarriage in cattle and horses; *Brucella melitensis*, mainly affecting goats in the Mediterranean area, transmitted to man through milk; and *Brucella suis* in domestic pigs, transmitted through infected meat. The human disease is a chronic infection of lungs and other organs, characterized by recurring bouts of fever (undulant fever) (Spink, 1956). In a number of cases the skeleton is involved by the hematogenous route. Skeletal involvement varies from 2–70 percent of the cases and is especially high in *B. melitensis* infections (Jaffe, 1972:1048). Adult males are affected much more frequently than females; children are usually spared (Glasgow, 1976). The most common skeletal lesion is on the spine or pelvis. Ganado and Craig (1958) observed 130 instances of spondylitis in 6300 patients with brucellosis. Long bones are rarely involved. Kelly, Martin, Schirger, and Weed (1960) observed in 36 cases the following localizations: spine 17, humerus 3, femur 2, ilium 1, hand 1, foot 1. The spinal lesion locates in the vertebral bodies, especially of the lower dorsal, lumbar, and lumbosacral areas, often involving more than one vertebra (Lowbeer, 1948, 1949). The lesion is a lytic cavitation. Grossly and on X-ray, it frequently can be seen penetrating the vertebral end-plate and extending through the nucleus pulposus of the disc into the next vertebral body (Figure 196). The cancellous bone within the focus is destroyed without formation of significant sequestra. The cortex also may be perforated, leading to parosteal abscesses. There is usually very little, if any, reactive bone formation except in the healing phase (Glasgow, 1976). In contrast to tuberculosis, which it resembles in several ways, complete collapse of vertebrae with gibbus formation is usually not observed (Keenan and Metz, 1972) and paravertebral abscess is rare (Kelly, Martin, Schirger, and Weed, 1960; Glasgow, 1976). Lesions in the pelvis likewise show,

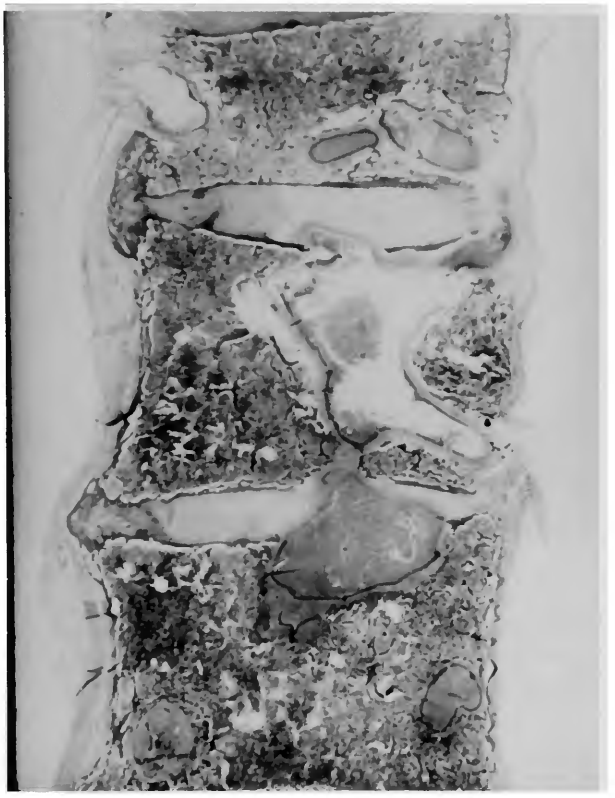


FIGURE 196.—Brucellosis of vertebral bodies, multifocal cavitating abscesses with perforation of intervertebral disc. (Histologic section. Courtesy Dr. L. Lowbeer, Tulsa, Oklahoma, USA).

usually multifocal, cavitation of the spongiosa, occasionally with perforation of the overlying cortex. Monoarticular or polyarticular involvement in large joints occurs, but usually without significant destruction.

PALEOPATHOLOGY

Since the spread of brucellosis between humans is very rare (Glasgow, 1976:283), the presence of the disease depends on the presence of domestic vectors including cattle, horses, goats, sheep, pigs, and dogs. Clearly, if the disease was endemic in ancient times, it should be seen in archeological skeletons. However, brucellosis receives little attention in the literature on paleopathology. Brothwell (1965b:690, 692–693) reports a case of bone inflammation in Early Bronze Age remains from Jericho in the Near East, which he tentatively attributes to brucellosis. However, Brothwell, initially attributed the diseased lower lumbar vertebrae to infectious arthritis. Although the description of the Jericho specimen is incomplete, the disease involves the two fibulae and the lumbar spine. One fibula is irregular and has a somewhat thickened shaft, the other is also enlarged but has no evidence of active infection.

One of the pathological skeletons in the National Museum of Natural History, Washington, D. C., has lesions that possibly can be attributed to brucellosis. This case (NMNH 227474) is a female Lapp skeleton from Norway accessioned by the museum in 1904. The estimated age at death on the basis of epiphysial fusion and pubic symphysis morphology is 20 to 25 years. The archeological date is unknown. The skull of this specimen is normal. Unfortunately, there is some mixture of bones from other skeletons. This problem is particularly apparent with the hand and foot bones, in which there are representatives of at least four individuals. However, there is no gross evidence of disease in any of these bones, so in all likelihood the hands and feet were not affected by disease in this case. Of the remaining bones of the upper extremity, only the left humerus is abnormal. Grossly only the head of the humerus is affected. Indeed, on superficial inspection the affected bone could easily be confused with postmortem damage due to burial conditions. However, the presence of periosteal, reactive bone peripheral to the destroyed area on the humeral head provides evidence of a pathological process. The humeral head itself has been eroded away, leaving exposed somewhat cavitated spongy bone, in which there has been no osteoblastic response to the disease process. However, the picture of the lytic process is obscured by what is undoubtedly some postmortem damage to the pathological bone.

The glenoid cavity of the left scapula exhibits a similar destructive, pathological process obscured by postmortem damage. Here, again, there is a slight perifocal bony reaction. The left radius and ulna are normal.

The X-ray film of the left humerus indicates

much more extensive involvement than is seen grossly. The cortex is much thinner than in the right humerus and the appearance of the bone resembles a fairly coarse, net-like structure, suggesting multiple foci for the disease process. Virtually the entire left humerus is affected.

The cervical vertebrae are all normal as are the first three thoracic vertebrae. An initial X-ray film of the thoracic and lumbar vertebrae indicated that the fourth through the sixth thoracic vertebrae were markedly osteoporotic. However, comparison of the corresponding articular facets and shape of the vertebral bodies revealed that these vertebrae were from another individual. Initially, this fact was not obvious since both the size and color of these vertebrae were appropriate. This problem serves to highlight the need to insure that all bones associated with a paleopathological specimen actually belong to a single individual and that great care is taken to prevent mixing bones during excavation and subsequent processing.

The seventh and eighth thoracic vertebrae are normal. However, in T9 there is a large lytic process that has created a cavity in the body. The inferior plate of the body is largely destroyed, and there is a hole one centimeter in diameter into the neural canal. There is no marked gross evidence of bony reaction to the disease process. The tenth thoracic vertebra contains a similar lesion except that the superior plate of the body is involved. Thus the vertebral bodies adjacent to a single intervertebral disk are affected. The anterior cortex of the bodies of the tenth thoracic through the second lumbar vertebrae have enlarged vascular foramina but are otherwise normal. There is a suggestion of a lytic cavity on the anterior body of L3. On the anterior cortex of the body of L4 there are four depressed lytic lesions, which are not confluent. In the largest of these there is slight scalloping, suggesting coalescence of two or more lytic foci. The fifth lumbar vertebra exhibits slight reactive bone on the lateral cortex of the body.

In the sacrum, the anterior surface of the first and second bodies shows evidence of slight erosion and reactive bone. However, the left and right articular surfaces of the sacrum show a massive lytic process with multiple foci (Figure 197). The large cavities created by the disease process subsequently have been well circumscribed. There is corresponding circumscribed, lytic destruction on the articular portion of the innominate and adjacent bone as well. However, the cavities are somewhat larger than those in the sacrum. The major lesion on the left innominate measures 25 × 40 millimeters, on the right 20 × 30 millimeters. Both cavities are at least 10 millimeters deep.

FIGURE 197.—Large circumscribed, lytic lesion in the sacroiliac joint surface of the innominate (arrow) with multiple lytic foci on the corresponding surface of the sacrum, possibly due to brucellosis. (Adult, female skeleton from Norway, NMNH 227474.)

The long bones of the lower extremities are all normal with the exception of the left femur. In this bone there is a well-circumscribed lytic lesion 15 × 18 millimeters and approximately 5 millimeters deep, which is lateral to the lesser tubercle on the posterior diaphysis. Some of the edges of this lesion have been broken postmortem, suggesting that the lesion was a cyst rather than a shallow depression during life.

Disease entities in differential diagnosis for this case must include, in addition to brucellosis, tuberculosis, osteomyelitis, mycotic infections, and cancer. The lesions of greatest significance in this case are the lytic cavities found in corresponding

endplates of the ninth and tenth thoracic vertebrae. The total bilobed appearance of the two cavities suggests an initial focus for the disease in the intervertebral disk. This is an unlikely focus for tuberculosis, osteomyelitis, or mycotic infections, but is typical for brucellosis (Glasgow, 1976: 286). The fact that the vertebral bodies have not collapsed argues against tuberculosis. The roentgen appearance of the humerus has at least a superficial resemblance to patterns seen in multiple myeloma (plasmacytoma), although the irregular distribution of the lytic process argues against this disease.

Glanders

Pathology

Glanders is an infectious disease of horses and donkeys, caused by the gram negative bacterium *Malleomyces mallei*. The disease is transmissible from animal to man and from man to man. In animals and man the nasal mucosa is frequently involved and often the portal of entry. Bone lesions in glanders are rare. The organism may affect the periosteum from adjacent soft tissue abscesses and skin ulcers. There can be marked periosteal reactive bone formation. Hematogenous osteomyelitis in glanders is very rare and inconspicuous (Beitzke, 1934c). The part of the skeleton most often affected is the skull, secondary to nasal and oral mucosal lesions. Involvement with defects of nasal bones, nasal septum, ethmoid, and sphenoid have been observed. Destruction of turbinates with perforation into the maxillary sinus and perforations of the hard palate occur. Cranial vault lesions, secondary to ulcerating glanders of the scalp, have been seen. Other bone lesions in glanders are almost exclusively limited to the lower extremities, particularly the tibia. The lesion may present as periostitis or osteomyelitis, occasionally with secondary infection by staphylococci, which modify the picture towards the appearance of ordinary osteomyelitis.

The bone lesions in glanders are not diagnostic in dry bone. The main reason to mention them is to point out the similarity to tertiary syphilis and to some lesions seen in leprosy. Joint involvement in glanders is not rare, occurring mostly by extension from adjacent soft tissue lesions, and only occasionally secondary to an epiphysial bone focus. In 27 cases gathered from the literature, Beitzke (1934c) found distribution as follows: Knee 16, elbow 4, ankle 4, toes 2, fingers 1, tarsus 1.

Tuberculosis of Bones and Joints

Pathology

Tuberculosis is a chronic infectious disease caused by *Mycobacterium tuberculosis* of the human or bovine type. The route of infection is usually through the respiratory tract, leading to the formation of a primary focus in the lung, followed by single or multiple foci in the regional hilar lymph nodes. Both lesions together form the primary complex. Much less common is the intestinal pathway with formation of a primary complex in the intestinal wall and mesenteric lymph nodes. The later course of the disease depends on the size of the inoculum, the virulence of the organisms, and the resistance of the host. In most instances (at least in Western populations of the nineteenth century), the primary complex heals without leading to a progressive disease. If the primary complex fails to heal, the lung lesion progresses and tubercle bacilli may be early disseminated through the bloodstream to other organs and tissues. Again, the number of organisms and the immunological capacity of the patient determine whether this early hematogenous dissemination will lead to fatal miliary tuberculosis and/or tuberculous meningitis or to isolated organ tuberculosis. Organ tuberculosis may not make its appearance until years after the early dissemination of organisms, and is precipitated by lowered host resistence due to malnutrition, other disease, or possibly local trauma. Since dormant primary pulmonary foci may harbor viable organisms for many years, late hematogenous dissemination also may become the source of organ tuberculosis. Skeletal tuberculosis is, with rare exceptions, the result of limited hematogenous dissemination.

Statistical Data

Although, obviously, statistical information is not available for earlier historic and prehistoric populations, data concerning the frequency and mortality of tuberculosis in the late nineteenth and early twentieth centuries, before chemotherapy and antibiotics, might serve as a guideline of limited value. The tuberculosis mortality in Germany in 1892 was 260 per 100,000 living inhabitants. For the beginning of the twentieth century (1901) statistics are only available for part of Europe and the United States and cover mortality of pulmonary tuberculosis only, ranging from 111 to 289 per 100,000 population. The age and sex distribution of all tuberculosis deaths for Western Europe, the United States, and Canada for 1949 were published by the International Union Against Tuberculosis in 1964. All statistics show a predominance of male over female deaths at a rate of almost 2:1. One must also appreciate that the figures for 1892 and 1901 are already modified by successful surgery and the figures for 1949 by the availability of effective antibiotics and chemotherapy. Therefore, these data represent less than a minimum of incidence which one should expect in earlier periods in areas involved by tuberculosis. The overwhelming majority of tuberculous infections affect the lungs. Skeletal tuberculosis comprises only about 3 percent of the total and about 30 percent of extrapulmonary tuberculosis (Kastert and Uehlinger, 1964:444, 445). Even so, the official yearbook of tuberculosis for West Germany still reports 20,342 cases for 1956, 3358 of them new. It is particularly noteworthy that not only the total tuberculosis mortality has steadily declined but, even more so, that in infants and children.

Among 560 cases of fatal hematogenous tuberculosis autopsied in the 10-year period 1923–1932 were 115 incidents of skeletal tuberculosis. Of these, 58 were limited to the skeleton while 21 showed tuberculous foci in other organs, 12 showed active pulmonary tuberculosis and 24 organ and lung tuberculosis (Kastert and Uehlinger, 1964:447).

The distribution and frequency of tuberculosis of bones and joints in a large clinical series (1752 cases) before 1892 has been published by Alfer (1892) (Table 5). In addition to the tabulated cases, 91 patients presented with multiple tuberculous bone and joint lesions. The age distribution of the same series of cases is shown in

TABLE 5.—Localization of skeletal tuberculosis listed in order of decreasing frequency (after Alfer, 1892)

Location	*No. of cases*
BONES	
Spine	239
Tarsals and metatarsals	184
Carpals and metacarpals	109
Ribs	67
Tibia and fibula	49
Radius and ulna	48
Phalanges of fingers	38
Temporal bone	33
Phalanges of toes	31
Pelvis	27
Sternum	21
Femur	14
Humerus	10
Mandible	9
Scapula	8
Orbital margin	7
Parietal bone	5
Frontal bone	5
Maxilla	5
Sacrum	3
Zygoma	2
Patella	2
Clavicle	2
Occipital bone	1
Coccyx	1
JOINTS	
Knee	281
Hip	241
Elbow	113
Ankle	43
Shoulder	28
Wrist	20
Metacarpal phalangeal joints	5
Metatarsal phalangeal joints	4
Sternoclavicular joint	4
Acromioclavicular joint	1

TABLE 6.—Age distribution of skeletal tuberculosis in bones and joints listed in order of decreasing frequency (after Alfer, 1892)

Location	*Years old**															
	0–5	5–10	10–15	15–20	20–25	25–30	30–35	35–40	40–45	45–50	50–55	55–60	60–65	65–70	70–75	75–80
BONES																
Spine	89	59	32	23	9	10	3	6	3	1	4	–		–		–
Tarsals and metatarsals	9	20	26	38	14	10	10	7	6	8	11	9	9	5	2	1
Carpals and metacarpals	16	12	16	23	12	5	1	5	3	6	2	2	4	2	–	
Ribs	–	4	9	8	5	9	5	5	10	–	7	2	2	1	–	
Tibia and fibula	12	5	7	8	3		1	7	–	1	1	–	1	2	1	
Radius and ulna	6	9	6	8	4	2	5	1	2	–	2	1	2	–	–	
Phalanges of fingers	15	7	4	4	1	2	2	–	–	–	1	1	1	–	–	
Temporal bone	6	4	2	3	7	5	2	3	1	–		–	–		–	
Phalanges of toes	2	6	5	7	1	1	2	1	2	–	1	1	–	2	–	–
Pelvis	1	1	3	7	5		2	3	3	–		–	–	1	1	
Sternum	–	1	–	3	1	1	1	3	2	3	2	3	1		–	
Femur	2	2	1	1	3	2	1	2		–		–	–		–	–
Humerus	1		1	2	1	1	–	1	1	1	1	–	–		–	–
Scapula	–		1	2	2	3	–	–		–	–	–	–		–	–
Orbital margin	1	2	1		–	1		2		–	–		–	–	–	–
Frontal bone	–		1	2	1	1	–	–		–	–	–	–		–	–
Maxilla	–		–	1	1	1		–		–	2	–	–		–	–
Mandible	2	–	–	1	3	–		1	1	–	–		1	–	–	–
Parietal bone	–	2	1	1	–	1		–	–	–	–		–	–	–	–
Sacrum	–	–	–	1	–	1		–	1	–	–		–	–		–
Patella	–	–	–	–	–	–		–	–	1	–	1	–	–		–
Clavicle	–		–	–	–	–	1	–	–	–	1		–		–	–
Zygoma	–	1	–	–	–	1		–	–	–	–		–	–		–
Occipital bone	–	–	–	–		–		–	1		–		–	–		–
Coccyx		–	–	–		–		–	–		–	1	–	–		–
JOINTS																
Knee	47	52	47	37	20	11	23	11	11	3	2	8	6	3		–
Hip	58	59	43	46	9	11	6	–	4	1	1	3		–		–
Elbow	7	14	14	21	12	9	6	5	9	8	5	2	2	–		
Ankle	5	9	10	5	2	1	1	3	2	–	3	–	2	–	–	
Shoulder	–	2	2	6	3	5	3	1	1	2	2	1		–	–	
Wrist	1		–	1	5		–	3	1	3	2	1	3	–	–	
Metacarpal phalangeal joints	–		–	1	1		–		1	1	1	–		–		
Metatarsal phalangeal joints	–		1	3	–		–	–		–		–		–	–	
Sternoclavicular joint	–	–	1	–	1	1	–	1		–		–	–		–	
Acromioclavicular joint		–		–		–		1		–		–	–	–	–	
Multiple foci	19	13	14	11	6	5	4	3	2	1	5	4	3	1	–	

* Overlap in age categories is from Alfer and is not resolved here.

Table 6. It indicates the great preponderance of skeletal tuberculosis affecting infants and children.

General Pattern of Bone and Joint Tuberculosis

Tubercle bacilli circulating in the bloodstream locate within the skeleton, particularly in areas of hemopoietic (red) marrow, which has a high circulatory and metabolic rate. These are, essentially, the areas of cancellous bone rather than the cortex or medullary cavity. As far as long bones are concerned, this especially means metaphyses and epiphyses in adults. In infants and young children, the distribution of hemopoietic marrow is much wider so that tuberculous foci often occur in tubular bones of hands and feet and in ossification centers of tarsal and carpal bones in addition to occasional diaphysial lesions in long bones. At all ages, vertebrae, ribs, and sternum have hemopoietic marrow, which explains the frequent tuberculosis of the spine at all ages. The flat bones, particularly the cranial vault, are more frequently involved in infants and children than in adults. Joint tuberculosis is intimately linked to involvement of the adjacent bones and for that reason discussed here rather than separately. The process may begin in the synovial membrane or in the bone or simultaneously, in both. This is explained by the joint blood supply to the epiphysis and the joint capsule. In an advanced stage, the original focus often cannot be ascertained.

The morphology of tuberculous lesions in dry bones is not specific and overlaps considerably in appearance with manifestations of other bone infections. However, there are some general characteristics of diagnostic value, in addition to the age-modified distribution of the lesions in the skeleton.

The tuberculous process, in its exudative phase, permeates the marrow spaces, devitalizing areas of cancellous bone, thus leading to formation of centrally located sequestra of cancellous bone (caries). In its proliferative granulomatous phase, the process leads to local destruction and cavitation in the cancellous bone. In either case very little, if any, perifocal reactive bone formation is elicited, and often the involved bone shows perifocal or general osteoporosis. The process in long bones tends to remain localized, mostly to the metaphysial or epiphysial portion. In contrast to purulent osteomyelitis, massive sequestra, especially of cortical bone, are very uncommon. Periosteal reactive bone is very limited or absent with the exception of tuberculosis of small tubular bones in infants and children, characterized by destruction or sequestration of the cortex and formation of an expanded shell of periosteal reactive bone (spina ventosa). Similar changes affecting part of an involved long bone are observed also in the young age group. Perforation of the cortex with formation of an extraosseous abscess, with or without fistulous perforation of the skin, is common. Traces of such an abscess can sometimes be seen in the presence of reactive periosteal bone in the vicinity of the opening and, occasionally, ossification of portions of the abscess wall.

In joints, destruction of the articular surface may be minimal if the process is limited to the synovium. Undermining and resorptive grooving of the articulating bones frequently occurs along the line of synovial or ligamentous attachments. If the process started in the bone or involves bone extensively, destruction of the articular surface and of the epiphyses with formation of cancellous sequestra and/or cavitation often occurs. Skeletal tuberculosis can heal without specific or surgical therapy. Small foci, particularly in infants and small children, may leave no trace, because the area is removed in the process of subsequent growth and remodeling. Foci destroying a growth plate will leave a growth deficit and/or deformity of the involved bone in the young age group. Foci in the vicinity of a growth plate may lead to excessive growth. This is not different from the effect of osteomyelitis. Joint tuberculosis may heal with obliteration of the joint cavity, often terminating in bony ankylosis with varying degrees of volume loss of the constituent bones. After healing, the spongiosa undergoes remodeling along altered stress lines but never reaches the original density of cancellous bone (Kastert and Uehlinger, 1964:467).

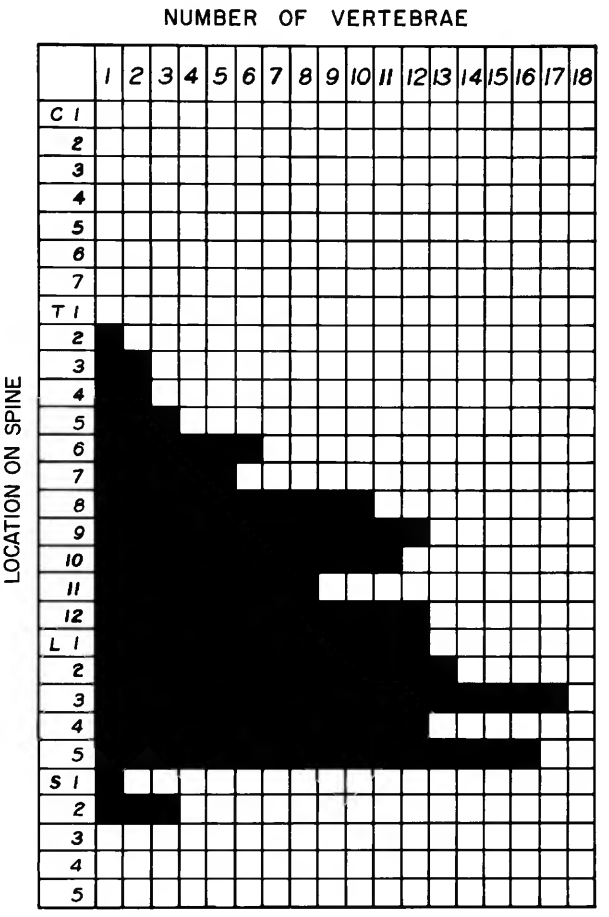
The Spine

Vertebral tuberculosis is, in practically all of the clinical and autopsy series, the most common and most characteristic lesion. The disease begins usually in early childhood. A well-documented series of 1490 cases after 1920 has been published by Sorrel and Sorrel-Dejerine (1932). It shows the rapid decline in the number of new cases after 7 years of age. The fourth, fifth, sixth, and seventh decades showed only 100, 50, 18, and 2 new cases, respectively. Since the disease takes a very chronic course, active and/or healed cases may be observed at any age. All studies indicate the predominant frequency of involvement of the lumbar vertebrae at all ages. Autopsy data concerning the frequency of involvement of individual vertebrae have been published by Uehlinger (in Kastert and Uehlinger, 1964:486) (Table 7). As far as the number of vertebrae affected is concerned, different statistics reported by Kastert and Uehlinger (1964:486) show that in about 80 percent of the clinical and autopsy cases at least two adjacent vertebrae are involved, while three or more are affected in about 10 percent of the total. Multiple foci separated by intact vertebrae are observed in about 4 percent.

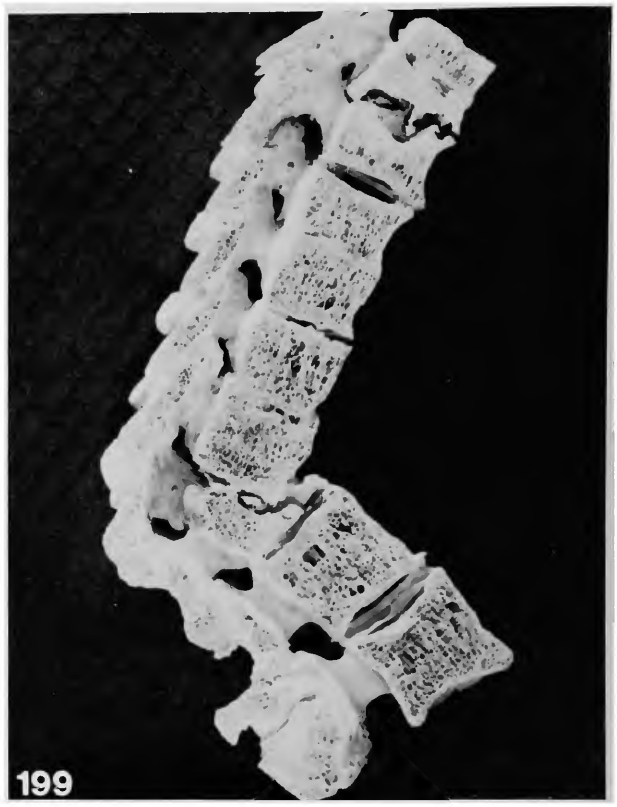
The part of the vertebra involved is almost exclusively the vertebral body. Even after extensive destruction of one or several adjacent vertebral bodies, extension into the vertebral arches is uncommon and the true intervertebral joints and spinous processes are almost never destroyed. An exception from this rule is suboccipital tuberculosis involving the atlas and the axis.

Because of the rudimentary bodies of these vertebrae, their intervertebral joints are not uncommonly involved (Oehlecker, 1924:242). Isolated tuberculous foci in posterior elements of vertebrae are extremely rare. Sorrel and Sorrel-Dejerine (1932:500) observed only three cases of isolated spinous process and one case of lumbar transverse process tuberculosis in their large series. The destruction of the vertebral body is usually purely lytic, leading to cavitation. Central spongiosa sequestra can occur (Figures 198, 199). The vertebral collapse can be combined with a pathologic fracture. Small wedge-shaped remnants of the affected vertebra often remain in contact with the end-plate and are displaced anteriorly or posteriorly by the collapse. Extension to adjacent vertebrae mostly occurs through the area of the nucleus pulposus of the intervertebral disc.

TABLE 7.—Distribution of lesions in 62 autopsied cases of vertebral tuberculosis (after Uehlinger, in Kastert and Uehlinger, 1964) (C = cervical, T = thoracic, L = lumbar, S = sacrum)

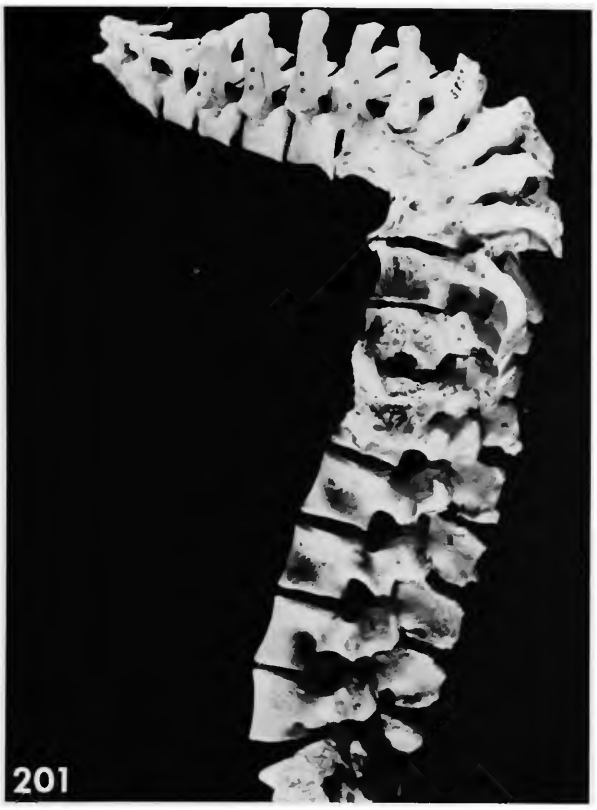


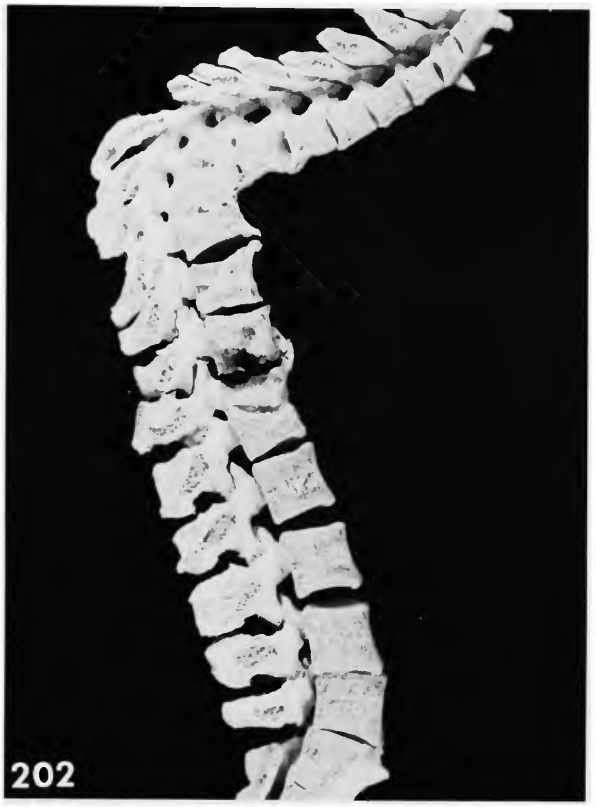
A common complication of vertebral tuberculosis is the formation of uni- or bilateral paravertebral abscess or fistula (Figure 200). The presence of a paravertebral abscess on dry bone is characterized by flaring, shelf-like bony extensions from the affected vertebra (Figures 201, 202). In Johansson's (1926:143) clinical material of 86 cases, 32 developed only abscess, 22 abscess



FIGURES 198, 199.—Tuberculosis of spine, partly healed, involving thoracic (Th) vertebrae 7 and 8 and first lumbar (L) vertebra, with gibbus: 198, Lateral view, showing fusion of vertebrae and periosteal bone. 199, Cut surface, showing perforation of disc Th7–8 and sequestrum in L1. (52-year-old female, IPAZ autopsy S901 from 1948.)

FIGURE 200.—Tuberculosis of the dorsolumbar spine with gibbus and large right psoas abscess. (Adult, PMUG, no number.)





FIGURES 201, 202.—Tuberculous spondylitis, healed with gibbus (thoracic vertebrae 6 to 9) and active with paravertebral abscess (thoracic 10 to lumbar 1): 201, Lateral view. 202, Cut surface. (48-year-old female with visceral and miliary tuberculosis, IPAZ S 793 from 1936.)

FIGURE 203.—Ossification in wall of right tuberculous psoas abscess extending to the femur, secondary to tuberculosis of the spine. (55-year-old male, FPAM 2894.)

and fistula, and 2 fistula only. The abscess extends, usually downward, following the line of gravity, beneath the anterior longitudinal ligament and along the fascial plane of the psoas muscle, occasionally showing ossification of the abscess wall (Figure 203). The abscess may become an important source of contact infection of additional vertebrae, especially below the original focus. The tuberculous process erodes the cortical surface and slowly extends into the anterior portion of the vertebral bodies (Figures 204, 205). Such secondary extension into adjacent ribs is occasionally observed.

Collapse of one or several vertebral bodies with remaining vertebral arches and spinous processes leads to a sharply angular kyphosis (gibbus). This deformity was observed in about 60 percent of the cases of spinal tuberculosis in the preantibiotic

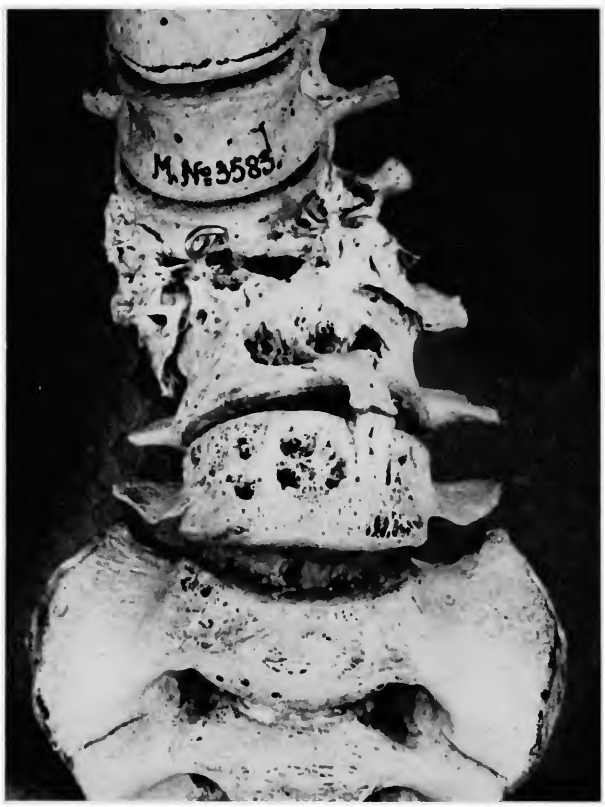


FIGURE 204.—Tuberculosis of lumbar vertebrae 2 and 3 and anterior excavation of lumbar vertebrae 4 and 5 by a prevertebral cold abscess; marked kyphotic angulation. (20-year-old male, FPAM 3583.)

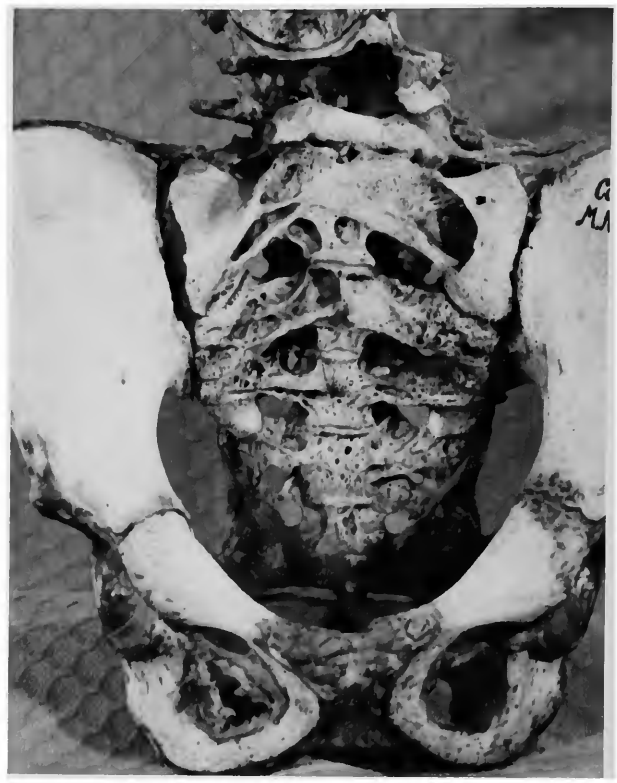


FIGURE 205.—Lumbosacral tuberculosis with gibbus and presacral tuberculous abscess. Notice the almost complete destruction of the lumbar vertebral bodies and the excavation of the sacrum from without. (About 10 years old, FPAM 1493.)

era (Reinhart, 1932). The kyphosis is most marked in the thoracic spine (Figures 201, 202), while lumbar lesions may terminate with telescoping of the defect rather than severe angulation (Girdlestone, 1965:81–83). Healing may occur with permanent preservation of the deformity by means of bony fusion of the remnants of the involved vertebral bodies. New formation of spongiosa and of cortex is rather meager. Residual cavitary defects may remain. There is usually secondary bony ankylosis between the true intervertebral joints of the involved segment and often also ossification of the interspinous ligaments. If a sharp, angular gibbus develops in childhood, increased height of the vertebrae below, due to compensatory growth, is often observed.

The differential diagnosis is mostly against osteomyelitis and healed fracture. In osteomyelitis,

the massive destruction of several vertebral bodies leading to the sharply angulated gibbus is uncommon. Paravertebral abscesses are also less frequently observed and, if present, may extend above the lesion as well as below because they form rapidly. In healed fracture with angular deformity, usually only one vertebra is involved with a diagonal break, and there is much less extensive destruction of the vertebral body. Besides, the predilected areas for fracture are the cervical and upper dorsal spine, a less common location for vertebral tuberculosis.

Isolated tuberculosis of the sacrum and of the coccyx are rare (David, 1924). Abscess formation is a frequent complication, and in the case of the coccyx, total sequestration is not unusual (Konschegg, 1934:409).

The Pelvis

The acetabulum is most frequently involved by tuberculosis but, being part of the hip joint, it is discussed there.

Sacroiliac Joint—Of the pelvis in the stricter sense, the sacroiliac joint is most commonly involved (Figure 206), usually by extension of the lumbosacral focus unilaterally or bilaterally. This is observed more often in young adults than in children. Isolated sacroiliac tuberculosis is very rare. In their large series, Sorrel and Sorrel-Dejerine (1932:501) observed only two cases as compared with 114 cases secondary to lumbosacral involvement. There may be considerable destruction of the sacral wing with some reactive osteosclerosis (Kremer and Wiese, 1930:198). Healing with bony fusion may lead to asymmetrical pelvic deformity.

Ilium.—Isolated tuberculous foci in the ilium are rare, usually consisting of round or oval cav-

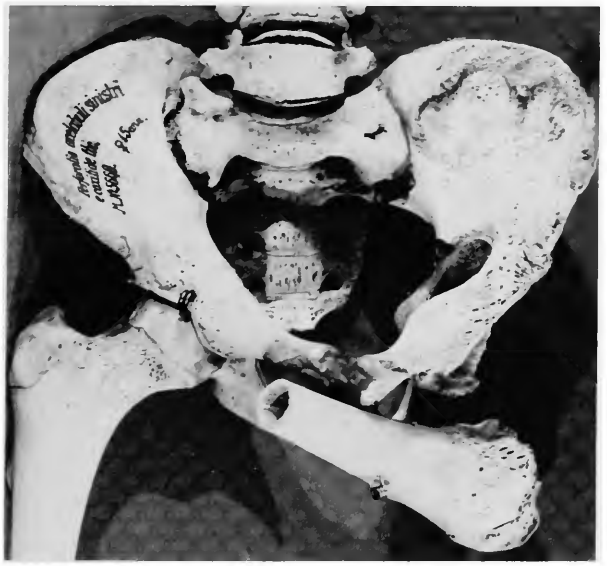


Figure 206.—Tuberculosis of the left hip with destruction of the femoral head and perforation of the acetabulum, tuberculosis of left ilium and of both sacroiliac joints. Notice minimal reactive bone in all affected areas. (15 years, FPAM 5669 from 1895.)

Figure 207.—Tuberculosis of spine and left ilium; reactive bone in wall of cold abscess in left inguinal area; notice round defect in ilium, erosion and bony buildup in inguinal area. (28-year-old male, FPAM 2488, autopsy 33582 from 1853/54.)

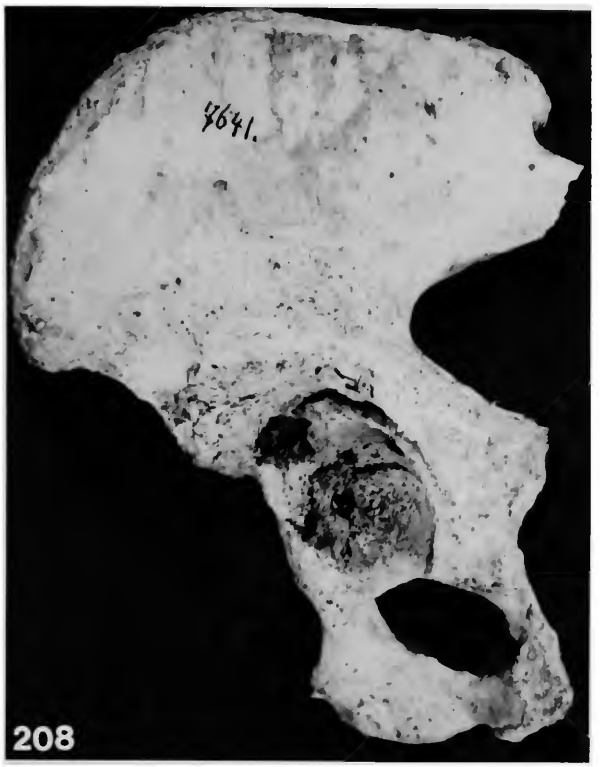
ities with or without a central sequestrum, perforation of the cortex, or fistula (Konschegg, 1934: 409–410). Extension of a psoas abscess into the ilium does occur (Figure 207).

PUBIS AND SYMPHYSIS.—The pubis is rarely affected by tuberculosis. Both sexes participate about equally and most instances concern older children and young adults. The largest series published are 27 cases of Fares and Pagani (1966), of which about half showed other adjacent or remote tuberculous skeletal foci. There is apparently no relationship to trauma or parturition. The lesion in adults is usually close to the symphysis, which may be involved. In that case, both pubic bones may be affected (Kremer and Wiese, 1930:195). The lesion is lytic and destructive, occasionally with formation of small sequestra. Abscess formation and fistula are common. In children, the lesion may be medial to the hip joint because of the incomplete ossification of the pubic area.

ISCHIUM.—Similar lesions occur in the ischial ramus (14 ischial compared to 20 pubic lesions in the series of Sorrel and Sorrel-Dejerine, 1932:76). In addition, occasionally an isolated lesion can occur in the tuber ischii, similar to the more common lesion in the greater trochanter (Blankoff, 1927).

The Hip

Tuberculosis of the hip joint is the second most frequent skeletal lesion after tuberculous spondylitis. All statistical series agree that the majority of cases start in childhood and that onset of the disease after 25 years of age is rare (Johansson, 1926:159; Sorrel and Sorrel-Dejerine, 1932:309). In the series of Sorrel and Sorrel-Dejerine, comprising 995 cases, the maximal incidence of onset is at 4 to 6 years of age with a second smaller peak around puberty. There were only 28 cases in the fourth, 11 cases in the fifth decade, and 4 cases beginning after 50 years of age. In the majority, the lesion starts with an osseous focus (Konschegg, 1934:461–462). The anatomy of the hip joint allows early access to the joint space, not only to acetabular and femoral epiphysial foci but also to metaphysial foci of the femoral neck. In the Kastert series, one-third of the cases started from an extra-articular focus (Kastert and Uehlinger, 1964:508). In addition to the usual hematogenous route, direct extension to the hip joint can also occur by contact with long-standing abscesses from vertebral or pelvic tuberculosis. In advanced stages of the disease, the point of origin can no longer be determined. A series of 416 cases of tuberculous coxitis reported by Vacchelli in 1922 (in Kremer and Wiese, 1930:202) gives the following breakdown: total destruction of femoral head and acetabulum 22, diffuse involvement of femoral head and acetabulum 220, diffuse synovial tuberculosis 52, isolated osseous tuberculous foci 122. The isolated foci were distributed as follows: femoral head 60, femoral neck 24, acetabulum 30, greater trochanter 8. The foci in the femoral head or neck may be small cavitating lesions or larger triangular foci with a spongiosa sequestrum in the center. These foci may represent territories of terminal arteries. Acetabular foci predilect the posterior superior rim and the cartilage-free center around the origin of the round ligament (Figures 208, 209). Foci in the neck of the femur are often adjacent to the medial inferior cortex (Girdlestone, 1965:44,45). Extension of primarily synovial tuberculosis into the bone occurs along the synovial attachment on the neck of the femur. The weight-bearing articular surfaces are longest preserved. Ultimately, destruction of the bones may be very extensive with sloping upward extension of the acetabulum, leading to upward partial or complete dislocation of the remnant of the femoral head and/or neck (Figures 210, 211). If the dislocation is complete, a neo-acetabulum is formed on the lateral surface of the iliac wing. In contrast to the appearance in congenital dislocation of the hip, the head of the femur is more eroded and there is no groove for the round ligament, which is destroyed by the infection before the dislocation occurs. The old acetabulum is not rudimentary and the neo-acetabulum shows evidence of infection. If there is extensive necrosis and destruction of the acetab-



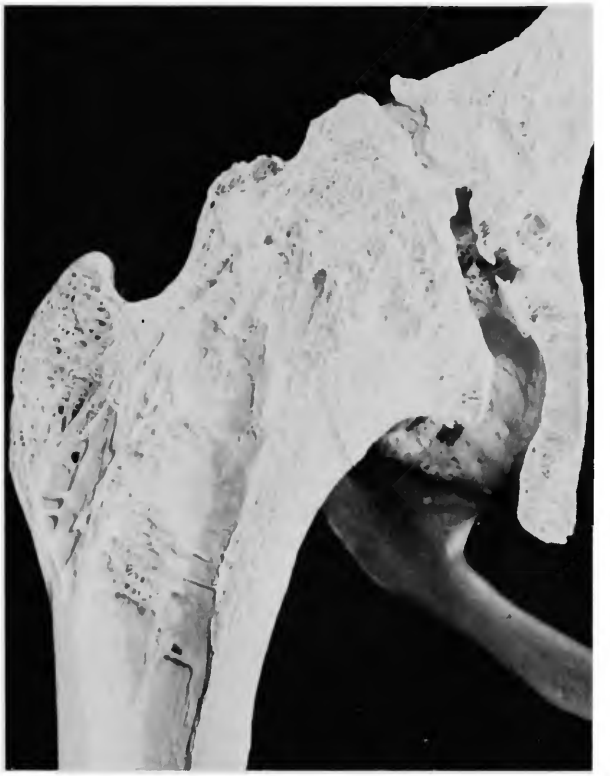
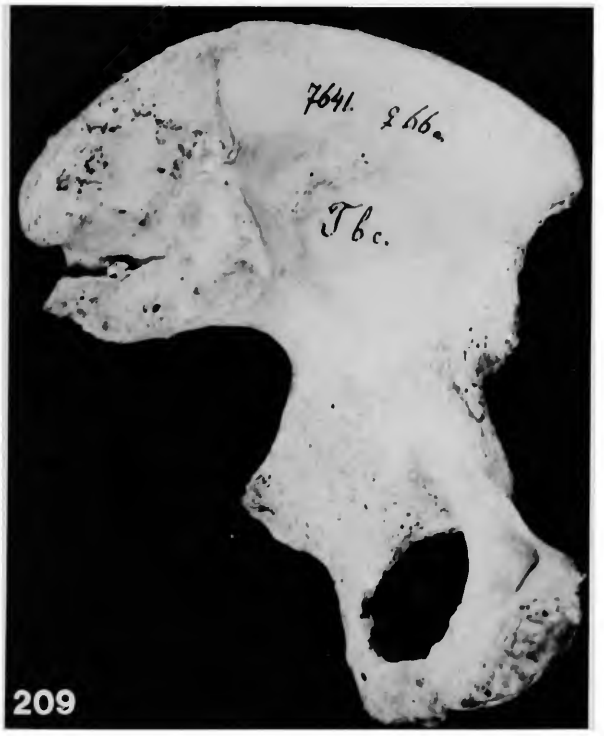


FIGURE 210.—Tuberculous arthritis of right hip with upward subluxation, partly healed. Notice restoration of subchondral bone plate on the partly destroyed femoral head. (16-year-old male, died of tuberculous meningitis, IPAZ S325 from 1936.)



FIGURES 208, 209.—Tuberculous arthritis of left hip, three months after resection of femur; evidence of upward subluxation: 208, Lateral view. Periosteal buildup on ischium probably secondary to cold abscess. 209, Medial view, hypervascularity of acetabular base. (66-year-old female with chronic pulmonary tuberculosis; DPUS 7641, autopsy 880 of 1912.)

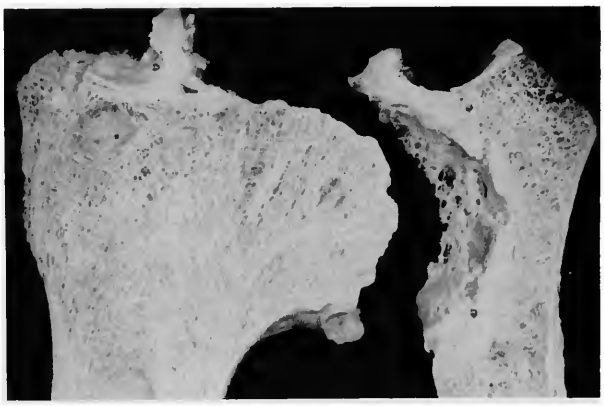
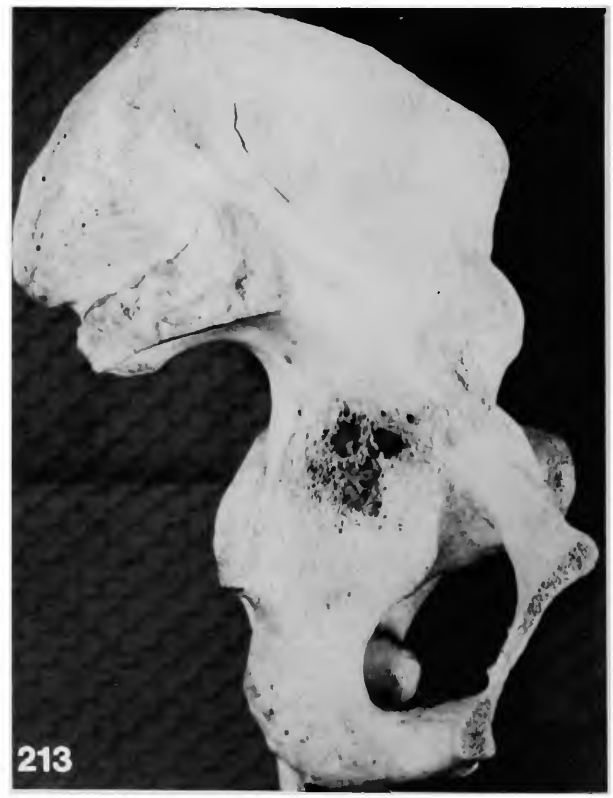


FIGURE 211.—Destructive arthritis of right hip with upward subluxation, probably tuberculous. Note severe osteoporosis of femur and ilium and iliac shelf secondary to subluxation. (88-year-old female with pulmonary and lymph node tuberculosis, IPAZ autopsy 1445 from 1954, museum no. 5947.)

ulum, its pelvic aspect shows lytic defects (Figures 212, 213); perforation of its floor with central dislocation of the remnants of the proximal femur can occur (Figures 214, 215). Ménard (1900) observed 105 acetabular perforations in the course of 268 hip resections. Tregubow (1929) reported 12 acetabular perforations in a series of 500 radiologically studied cases of tuberculous coxitis. In healed cases, bony ankylosis usually occurs. Growth deficit also may be observed. Differential diagnosis is mainly between tuberculous and septic arthritis. The septic process is rapid and bone destruction is much more limited. Dislocation upward or centrally is not observed. Bony ankylosis as final outcome usually reveals little, if any, bone loss of the joint constituents. Only in infants with septic arthritis is complete



FIGURES 212, 213.—Tuberculosis of the left hip: 212, Anterior view. 213, Medial view. Notice destruction of femoral head with exposure of porotic spongiosa and perforations of the involved acetabulum. (Adult, IPMI KM 352.)

destruction of the femoral head seen. But this condition is usually accompanied by osteomyelitis of the shaft. Statistically tuberculous coxitis was much more common than septic arthritis.

FIGURES 214, 215.—Tuberculous arthritis of right hip with complete destruction of the acetabulum and central dislocation of the remnant of the femoral head; notice sparsity of reactive bone: 214, Lateral view. 215, Medial view. (Male about 30 years old, IPAZ 1940, old no. 2167.)

The Trochanter

Chronic tuberculosis of the greater trochanter of the femur is an uncommon but characteristic lesion. Sorrel and Sorrel-Dejerine (1932:70) observed, in a series of 6578 cases of skeletal tuberculosis, 32 instances of tuberculosis of the trochanter (11 in children and 21 in adults). McNeur and Pritchard (1955) reported 38 clinical cases treated at the Royal National Orthopaedic Hospital (London, England) in a period of 35 years. The age distribution was as follows: 30 of the cases were between 10 and 40 years, only 3 cases concerned children below 10, and 5 patients were above 40 years. The infection may start in the trochanteric bursa or in the bone. It takes a very chronic course and has a great tendency to recur

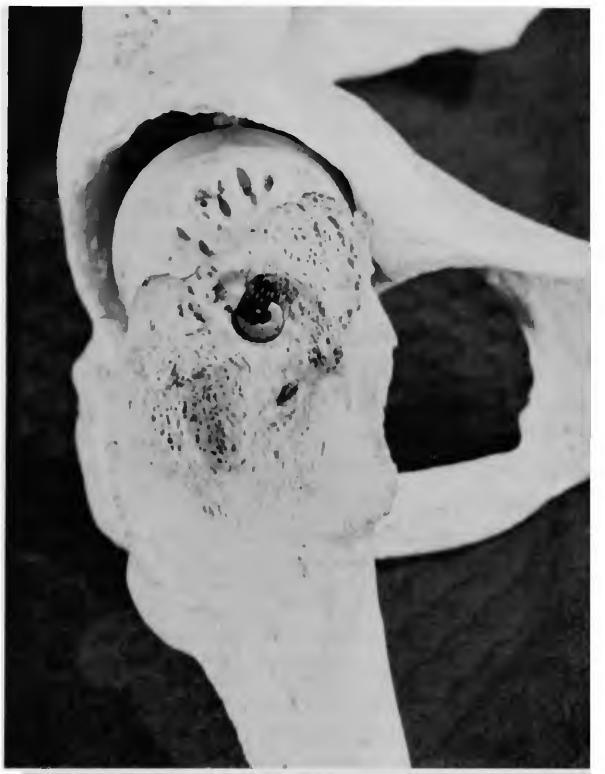


FIGURE 216.—Chronic tuberculosis of right greater trochanter with fistulating abscess, hip joint free. Notice scalloped destruction of greater trochanter exposing porotic cancellous bone. (54-year-old female with pulmonary tuberculous focus, FPAM, Jubiläumspital 857, autopsy from 1924.)

over many years. This has also been the experience with three cases followed at the Massachusetts General Hospital in Boston. The lesion tends to remain localized to the trochanter area, which is progressively destroyed (Figure 216). The cavity often contains a sequestrum of cancellous bone (Sorrel and Sorrel-Dejerine, 1932:70). Abscess and fistula formation are common. This actually represents the most identifiable tuberculous bone lesion of the adult with exception of tuberculous spondylitis. This entity is relatively little known except to orthopedists, but a number of similar series have been published (Meyerding and Mroz, 1933; Wassersug, 1940; Alvik, 1949; Ahern, 1958).

The Knee

Tuberculosis of the knee joint occurs about as often or even more frequently than tuberculous coxitis. Again, the majority of cases begin in infancy, childhood, and adolescence. Johansson (1926:177) observed 50 percent before the age of 5 years, with about an equal distribution between the sexes. The series of Sorrel and Sorrel-Dejerine (1932:247) comprises 558 cases, of which 34 occurred in the first year of life and only 51 cases began after age 20. The majority of cases start as synovial tuberculosis and may remain so. Extension of the synovial infection occurs along the capsular insertions of the femur and tibia and along the attachments of the cruciate ligaments. Linear cortical erosion and undermining destruction of the adjacent portion of the articular surface occurs. Significant amounts of localized destruction of femoral condyles or of the tibial plateau are observed only if a primary or simultaneous hematogenous osseous focus, with or without sequestrum, is present (Figure 217). Such foci are more often found in the femoral condyles or in the tibial epiphysis, rarely in the patella or fibula. König (1906:112) observed, in a series of 720 cases, 50 with involvement of the patella. In 33 of these the patella was the only focus, with or without secondary extension to the knee joint. In healing, fibrous or bony ankylosis results. If bone destruction was absent or limited, differentiation

FIGURE 217.—Tuberculosis of knee, recurrent after previous surgery. Notice the destruction of subarticular bone and the minimal reactive bone formation. (13-year-old female, DPUS 3921 from 1899.)

from the end-stage of rheumatoid or septic arthritis may be impossible. Both rheumatoid and tuberculous gonitis are usually accompanied by osteoporosis of the involved limb. Tuberculous and septic arthritis are more often unilateral than rheumatoid arthritis. In severe cases, particularly in children, dislocation and valgus or varus deformity of the knee is observed, depending on the relationship of the osseous focus to the growth plate. Generally, the adult cases are less destructive than the infantile ones (Sorrel and Sorrel-Dejerine, 1932:267).

The Ankle and Tarsal Bones

Tuberculosis of the ankle most commonly involves the tibiotalar joint (Figure 218), much less

FIGURE 218.—Tuberculous arthritis of right ankle with extensive destruction of distal tibial epiphysis and partial ankylosis with remnants of talus; histologically proven, 9-years duration. Initiated by crushing trauma. (25-year-old male, PMES 2 FT 16(1) from 1930.)

commonly the talocalcaneal joint. The lesion is most common in children. In the large series of Sorrel and Sorrel-Dejerine (1932:210), the tibiotalar joint was involved in 185 cases of which, 121 cases were below 13 years of age and only 64 were adolescents or adults. The maximum incidence is at 3 years of age. In most cases the process seems to start with an hematogenous osseous focus. In the majority, the primary bony focus is in the talus, less commonly in the distal tibia and rarest in the fibula. The ossification of the talus begins at birth and essentially fills the cartilage model at 2 years of age, leaving only the articular cartilage between the ossification center and the adjacent joints. This explains why isolated tuberculosis of the talus without involvement of the adjacent joints is not observed (Sorrel and Sorrel-Dejerine, 1932:210), but early extension into the tibiotalar, and, less often, into the talocalcaneal joint as well, is the rule. In tibiotalar tuberculosis of talar origin, the talus is cavitated and often ultimately destroyed. In the cases of tibial origin, there is significant destruction of the distal tibial epiphysis and sometimes also of the metaphysis. Healing always leads to tibiotalar bony ankylosis. In advanced cases of tuberculosis of the ankle, the talocalcaneal joint frequently becomes involved at any age. If the talus was completely destroyed, tibiocalcaneal bony ankylosis develops with uptilted position of the calcaneus. This would not be the case in ankylosis following juvenile rheumatoid arthritis. Since the ankle is a weight-bearing joint, limited perifocal osteosclerosis does occur.

Isolated involvement of the talocalcaneal joint usually is due to secondary extension of a calcaneal focus. This tuberculosis, limited to the lower ankle joint, is observed only in older children between ages of 7 and 16 (Sorrel and Sorrel-Dejerine, 1932:207). Healing terminates with broad bony fusion of talus and calcaneus.

Of the tarsal bones, after the talus, the calcaneus is most frequently affected by tuberculosis. Sorel and Sorrel-Dejerine (1932:207) observed 131 cases of calcaneal tuberculosis but only 29 cases in all other tarsal bones. The explanation for the frequent and often isolated involvement of the calcaneus rests with its development. An ossification center usually appears as early as the last trimester of intrauterine life and ossification is not completed until 17 to 18 years of age. This makes a highly vascular area available for tuberculous seeding in infancy and early childhood, while thick layers of the cartilage still separate the focus from the adjacent joint cavity (Sorrel and Sorrel-Dejerine, 1932:268). In early childhood, central tuberculosis of the calcaneus is fairly frequent and may heal without permanent traces, because of the effect of growth and remodeling. At age 7 to 9 years an apophysial ossification center appears on the posterior portion of the calcaneus, and during later childhood tuberculous foci adjacent to the anterior surface of the posterior growth plate appear. The lesion usually

shows a cavity with a central spongiosa sequestrum and often some perifocal osteosclerosis. After termination of growth in the adult, tuberculous foci of the calcaneus readily breaks through or around the articular cartilage into the talocalcaneal joint and from there spread to the tibiotalar joint (Sorrel and Sorrel-Dejerine, 1932:59–60).

The cuboid is much less often involved and most of the patients are children (11 children, 2 adults). The ossification of the cuboid begins at 3 months of age and terminates at 9 to 10 years. Occasionally the navicular also is involved in older children (6 cases in children, 1 adult). The ossification of the navicular begins at 2½ to 5 years of age and terminates between 10 and 12 years. The cuneiformes are rarely involved; if destroyed, medial deflection of the foot may occur. Foci like these may remain isolated in children and may heal without joint involvement. All these data are taken from the discussions of Sorrel and Sorrel-Dejerine (1932:63–65). In adults, tarsal bones not uncommonly participate in extensive tuberculosis of the ankle. The differential diagnosis between tuberculosis and subacute osteomyelitis on isolated tarsal lesions may be impossible on dry bone.

The Tubular Bones of the Hands and Feet (Spina Ventosa)

The most frequent localization of skeletal tuberculosis in infancy and early childhood is the often multiple involvement of phalanges, metacarpals, and metatarsals (spina ventosa). Sorrel and Sorrel-Dejerine (1932:2) reported that among 4660 children with skeletal tuberculosis, there were 649 cases of spina ventosa with 1 to 10 foci in individual patients. Bailleul (1911:4) reported that in 274 patients, there were 495 lesions of spina ventosa, of which 381 were located in the hands and 114 in the feet. The age distribution in Johansson's (1926:144) material is as follows: 15 percent in the first year, 62 percent below 3 years, and 77 percent below 5 years. The lesion rarely occurs after 10 years of age. Of his patients 50 percent showed solitary lesions. The localizations in his cases were fingers 108 times, metacarpals 68 times, metatarsals 32 times, and toes 11 times.

In infancy and early childhood these short tubular bones still have hemopoietic marrow throughout the shaft. In these small bones a focus will readily occupy the whole diaphysis, leading to ischemic necrosis and/or penetration of the thin cortex. The cortex may be rapidly resorbed or form a sequestrum. The elevated periosteum forms a new bony shell, which accounts for the ballooned appearance of the involved bone (Figure 219). These lesions often accompany other skeletal manifestations of tuberculosis. If the child does not die from tuberculosis located elsewhere, the lesion usually heals. Destruction of the growth plate in metacarpals and metatarsals, and, less commonly, of phalanges, may lead to marked shortening of the digit after healing. If this is not

FIGURE 219.—Tuberculous dactylitis (spina ventosa) of the basal phalanx of the finger of a child. Notice expanded involucrum surrounding remnant of diseased phalanx; epiphysis spared. (PMES 1 FT 12(1).)

the case, evidence of the healed lesion will disappear in remodeling. Very similar lesions are produced by osteomyelitis and by congenital syphilis. However, those lesions are usually singular, and expansion of the involved bone is usually much less marked.

In children, tuberculous dactylitis spares the interphalangeal joints. In adults, rarely, phalanges may be involved, the lesion extending into the joint and not giving the distended appearance of the bone (Girdlestone, 1965:183).

The Shoulder

Tuberculosis of the shoulder is much less common than that of the hip or knee. Kastert observed 77 cases in a series of 2457 skeletal tuberculosis patients (in Kastert and Uehlinger, 1964: 517). It is seen at any age, but in adults more often than in children. There is prevalence of male cases and in both sexes the right side is three times more affected than the left (Kremer and Wiese, 1930:291). The complicated relationships

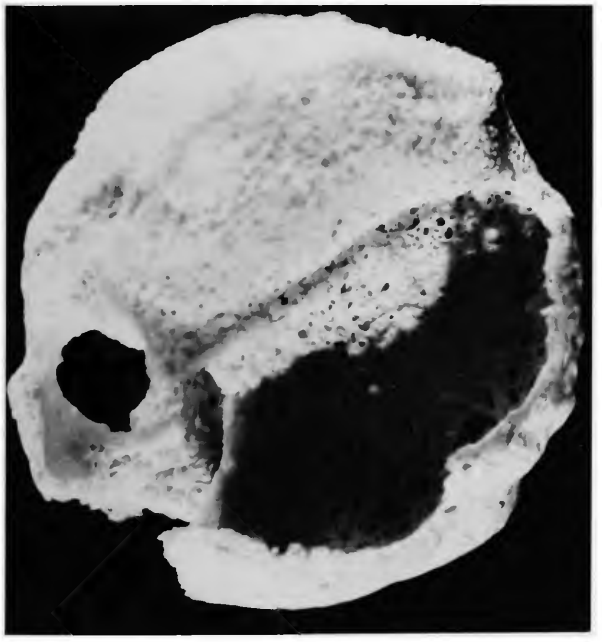


FIGURE 220.—Tuberculosis of humeral head with cavitation, sequestrum removed; resection specimen. Note exposed porotic hypervascular bone with little reactivity. (17-year-old female, PMWH W0709.)

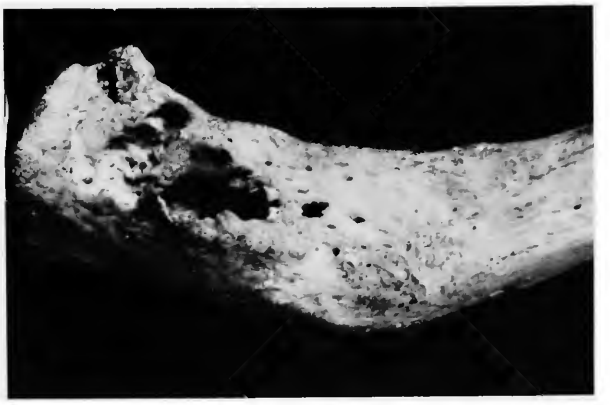


FIGURE 221.—Cavitating tuberculosis of the lateral portion of a right clavicle. Notice the sparsity of reactive bone. (From an adult with tuberculous arthritis of the shoulder, PMES 1 FT 5(2) from before 1920.)

of the shoulder joint to the synovial sheath of the long biceps tendon and to the subdeltoid bursa favor extensive synovial involvement. If osseous foci are present, they are more frequently found in the head or proximal metaphysis of the humerus than in the scapula (Figure 220). In children, cavities with sequestra occur in the epiphysis and metaphysis of the proximal humerus (Sorrel and Sorrel-Dejerine, 1932:165–168). In the synovial form, the extension to the humerus is along the capsular attachment, creating a resorption groove on the lateral aspect of the humeral head. In children, shoulder tuberculosis may heal. In adults, ultimate extensive destruction of the humeral head and of the glenoid fossa are common. Occasionally the acromion and clavicle may also be involved (Figure 221). Abscess formation and fistula are less frequent than in other large joints. In the differential diagnosis, septic arthritis is the main consideration. In that condition bone destruction is usually much less extensive and the lateral grooving and undermining defect on the humeral head is not observed.

The Elbow

Tuberculosis of the elbow is in many series the most frequent joint tuberculosis of the upper extremity (50 percent, Kremer and Wiese, 1930: 304). The majority of the lesions develop between

FIGURE 222.—Tuberculosis of right elbow with bony ankylosis between radius, ulna, and humerus. Notice marked involvement and enlargement of the distal humerus. The defect of the olecranon process is artificial. (Amputate from a child, PMES 2 FT 7 (4).)

1 and 20 years of age (König, 1906:141, Cheyne, 1911:328). Sorrel and Sorrel-Dejerine (1932:121) observed, in the 10 years preceding 1928, tuberculosis of the elbow in 164 children and 72 adults. Osseous foci, if present, are most common in the distal humerus, second in the proximal ulna, and least common in the proximal radius. König (1906:141–142) found bony foci in 91 of 128 patients. The distribution was as follows: distal humerus 43 (mostly in the lateral condyle), ulna (olecranon) 36, proximal radius 2. In advanced cases, several of the adjacent bones may be involved. In very young children a central tuberculous focus in the olecranon is not uncommon as part of multiple skeletal foci (fingers, toes, calcaneus, zygoma). This is a cavitary lesion with central sequestrum and reactive periostitis resembling spina ventosa. The joint may not be involved (Sorell and Sorrel-Dejerine, 1932:121). After 6 years of age, the joint is often involved by extenstion of the ulnar focus through the joint cartilage. This leads to deeper excavation of the semilunar incisure of the ulna with elongation of its coronoid process. In children, the process may heal with fibrous ankylosis. In adults, destruction of the adjacent bones may be extensive, usually least and last of the head of the radius. Healing usually terminates in bony ankylosis (Figure 222). Ankylosis without major bone loss may be impossible to differentiate from rheumatoid or septic arthritic sequelae. Periarticular osteophytosis can occur in extensive capsular involvement (Figure 223).

FIGURE 223.—Tuberculosis of right elbow. Notice exposure of subchondral porotic spongiosa on all joint surfaces and partial destruction of the radial head. The periarticular osteophytosis suggests massive capsular involvement. (68 years, IPAZ 1969, old no. 253.)

The Wrist and Carpal Bones

The wrist consists of three partly separated joints: the radiocarpal joint, the intercarpal joints, and the carpometacarpal joint. Any one or all of them may be involved in tuberculosis. The following discussion is mainly based on the detailed study of Sorrel and Sorrel-Dejerine (1932:91–120). They observed 63 instances in children and 51 in adults. The location and manifestation of the lesions vary greatly in different age groups. In children, the carpometacarpal joint is mainly involved and the radiocarpal joint is spared. In adults, the process usually begins in the radiocarpal joint and spreads rapidly throughout the joint compartments of the wrist (Figure 224). The difference is explained by the anatomy and maturation of the constituent bones. In children below 4 years of age, the carpus is mainly a block of cartilage with minimal focal ossification. At this stage carpal tuberculosis is not observed. From 4 to 12 years of age, carpal ossification centers become larger and more numerous. At this time, localized carpometacarpal involvement is observed, since adjacent bones are still protected by thick layers of unossified cartilage. The carpometacarpal joint is uncommonly involved by extension of a spina ventosa of an adjacent metacarpal. In general, an active growth plate serves as a barrier for spread to the adjacent joint. The metacarpals two to five are devoid of a proximal growth plate. These localized joint lesions may heal along with the healing of the accompanying spina ventosa, leading to bony fusion between individual carpals and metacarpals. With increasing age, the cartilage cover diminishes and extensive joint involvement becomes the rule. In children and adults, the joint lesion may originate directly in the synovium or by contact with tuberculous tenosynovitis. In the adult, osseous destructive foci are not infrequently present in the distal radial epiphysis and/or metaphysis. The disease rapidly spreads through the entire wrist and, in contrast to the infantile type, the proximal row of carpal bones is more severely involved. In healing, with various degrees of bone loss, the entire carpus becomes a solid bone block, fused to the radius and often to the base of the metacarpals as well.

FIGURE 224.—Tuberculosis of the left wrist. Wet preparation with soft tissue attached, showing extensive destruction of distal radius, ulna, carpal bones, and carpometacarpal joints of 1-year duration. (56-year-old male with pulmonary tuberculosis, WM S82.1 from 1876.)

The Shaft of Long Bones

Tuberculosis of the shaft of long bones is uncommon. It is almost exclusively observed in children and frequently as a manifestation of multiple skeletal foci, particularly spina ventosa. Sorrel and Sorrel-Dejerine (1932:2 and 21) observed about 100 cases in infants and children compared with 649 cases of spina ventosa. The lesion consists of eccentric cavitation, usually in the metaphysis, with a small sequestrum (Figure 225), and often marked reactive periosteal bone formation over the overlying cortex (Figure 226). Thus, in children, there is considerable resem-

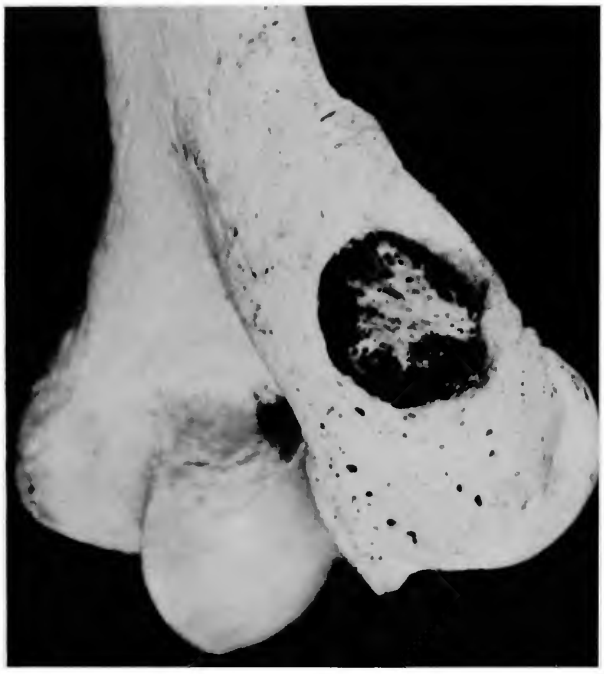


FIGURE 225.—Tuberculosis of lateral epicondyle of right humerus. Notice smooth cavity with porotic sequestrum and minimal reactive bone in vicinity. (Adult, DPUS 5989, French catalog no. 834.)

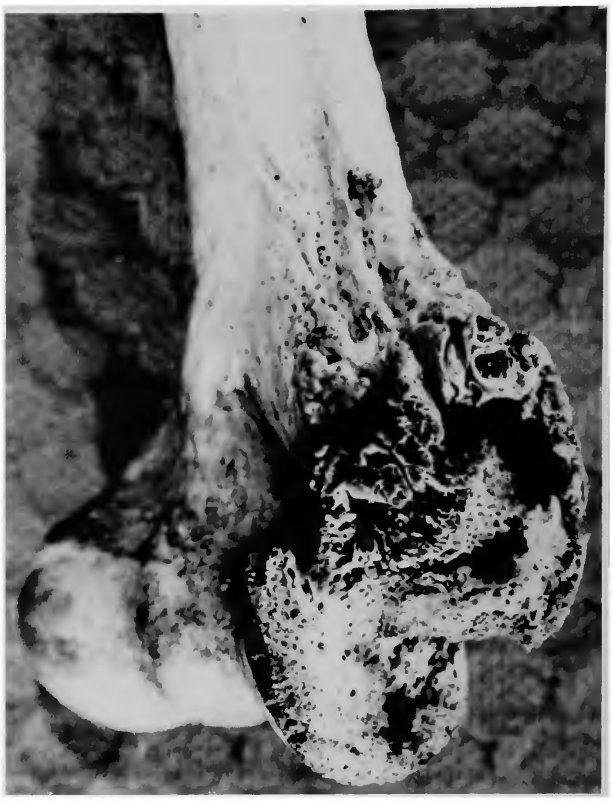


FIGURE 226.—Tuberculosis of medial epicondyle of right humerus. Notice porotic destruction with moderate periosteal reactive bone. The joint is not involved. (Adult, DPUS 5982, French catalog no. 827.)

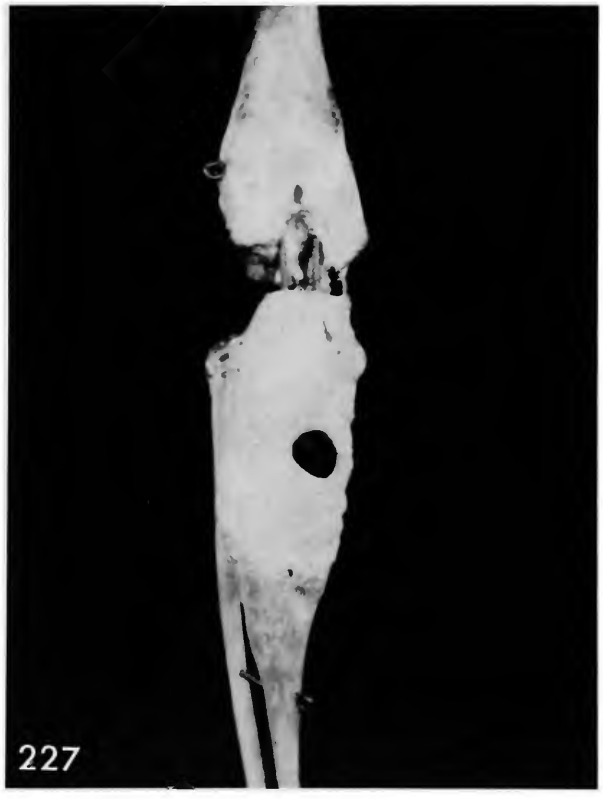
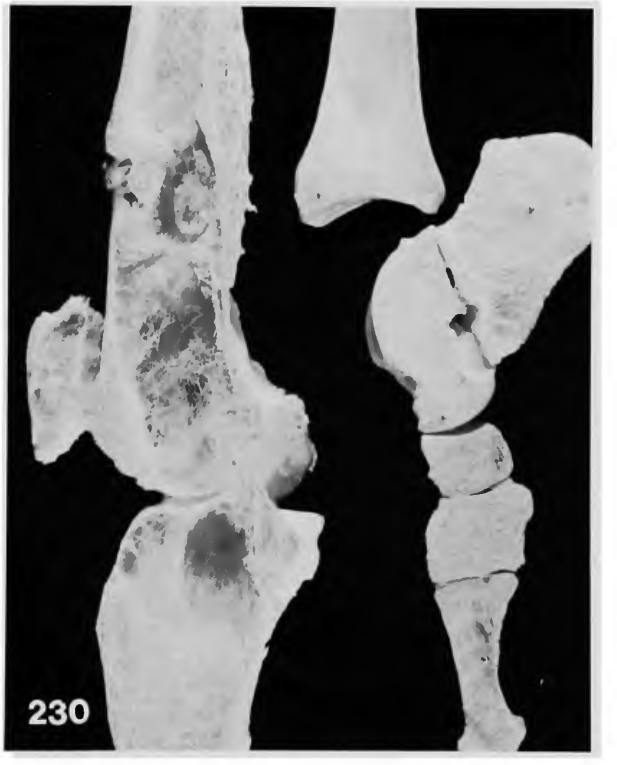


FIGURE 227.—Tuberculosis of proximal left ulna (spina ventosa) with extension to the elbow joint and humerus, posterior view. Notice the bulbous expansion of the new ulnar cortex with single cloaca, destruction of olecranon, and erosion of humerus. (From a child, PMES 2 FT 7(6).)

blance to the appearance of spina ventosa (Figures 227, 228). In adults, the lesion is extremely rare; periosteal bone formation is meager (Figure 229, 230). Perifocal osteoporosis may be followed in long-standing cases by perifocal osteosclerosis.

FIGURE 228.—Tuberculosis of proximal radius (spina ventosa) with bulbous enlargement of new cortex and multiple cloacal perforations; epiphysis and articular surface spared. (6-year-old female with tuberculosis of cervical lymph-nodes, PMES 1 FT 10 (2) from before 1900.)



FIGURES 229, 230.—Healed tuberculosis of distal femur with partial ankylosis of knee and severe osteoporosis due to disuse: 229, External view; note periosteal hyperostosis. 230, Cut surface; note cavitation in femur metaphysis and severe osteoporosis. (68-year-old male, IPAZ 6052, surgical specimen MB 6936 of 1955.)

A certain differentiation from osteomyelitis and Brodie's abscess may be impossible on the dry bone.

The frequency of involvement is: tibia, ulna, radius, humerus, femur, and fibula, respectively (Konschegg, 1934:422).

The Thoracic Cage

RIBS.—Tuberculous involvement of one or several ribs is not rare. Sorrel and Sorrel-Dejerine (1932:84–88) observed 93 instances, of which 56 concerned children and 37 adults. The infection is usually hematogenous, but direct extension from paravertebral abscesses and other adjacent tuberculous foci can occur (Kremer and Wiese, 1930:283). The hematogenous foci predilect the area near the osteocartilaginous border and may involve the cartilage secondarily (Kastert and Uehlinger, 1964:515). The process creates a lytic lesion with fusiform enlargement of the involved area and often perforations of the cortex lead to chest wall abscesses. Periosteal reactive bone formation is usually rather meager. The middle ribs are more often affected than the upper or lower ones and secondary involvement of adjacent ribs can occur (Konschegg, 1934:407). Differentiation from osteomyelitis, fibrous dysplasia, and eosinophilic granuloma may be very difficult on dry bone.

FIGURE 231.—Tuberculosis of sternum (posterior view) probably by direct extension from visceral tuberculosis. Note excavation of manubrium and corpus sterni from without with moderate amount of reactive bone. (PMWH WO 706.)

STERNUM.—The sternum is much less frequently involved than the ribs. König (1906:155) observed that ribs are five times as often affected as the sternum. The most frequent location is in the manubrium (Figure 231). These lesions may extend into the sternoclavicular joint and involve the medial portion of the clavicle. The sternal lesion is mostly lytic and may perforate the anterior or posterior cortex or both. In the differential diagnosis, erosion of the manubrium by an aortic aneurysm must be considered.

SCAPULA.—The scapula is very rarely involved except by extension of tuberculosis of the shoulder joint into the glenoid fossa or the acromion. In hematogenous tuberculosis of the scapula, the vertebral margin is predilected (Kremer and Wiese, 1930:301).

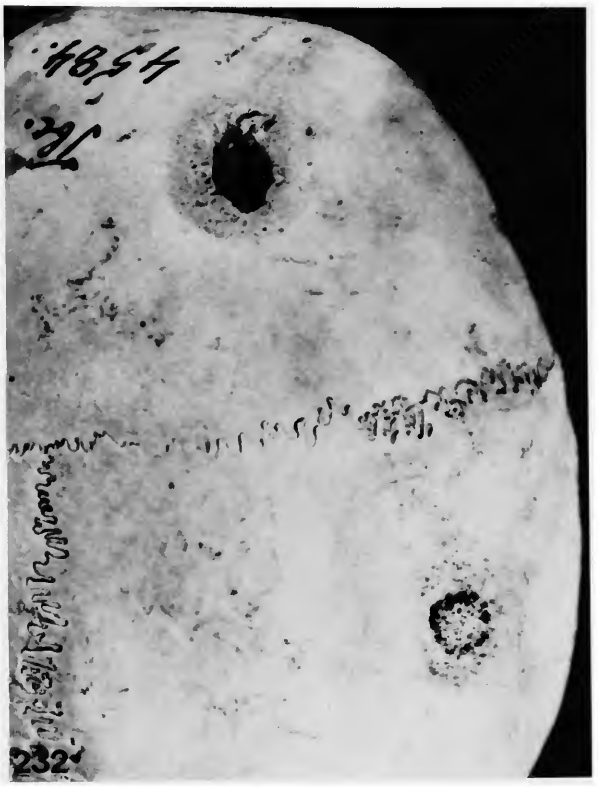
The Skull

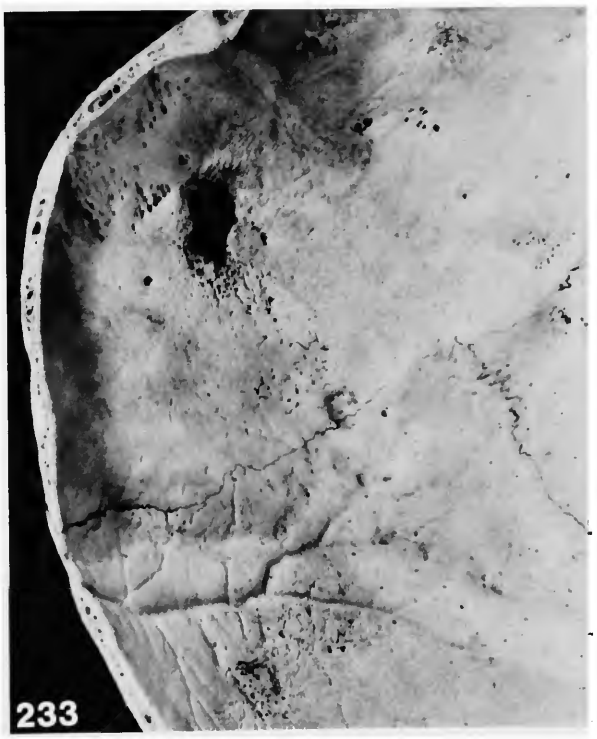
The skull is a rare area of involvement except in young children. Tuberculosis of the skull has to be separated into three areas: cranial vault, cranial base, and face.

CRANIAL VAULT.—This is the most common location of cranial tuberculosis. Sorrel and Sorrel-Dejerine (1932:78) saw 21 cases, of which 16 were children and 5 adults. The localization in the different bones, in a statistical survey by Straus (1933), is as follows: frontal 86, parietal 86, occipital 18, temporal 16. The majority of the cases are infants and children below 10 years of age. The infection usually is through the hematogenous route. In children, the lesions are often multiple and secondary to or coexistent with other active tuberculous skeletal foci (Sorrel and Sorrel-Dejerine, 1932:79–80). Apparently the presence of hemopoietic marrow and the growth activity of the cranium at this age determine the frequency of involvement. The most characteristic lesion is a round lytic focus of not more than 2 centimeters

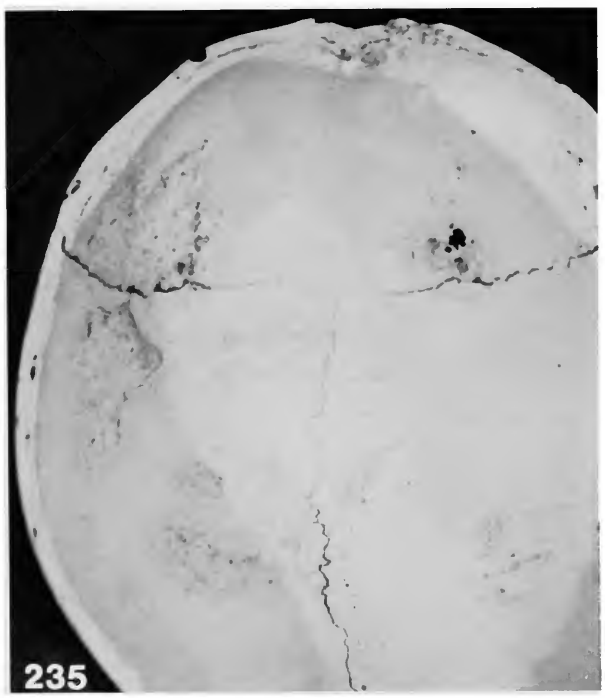
in diameter, with or without a "moth-eaten" central sequestrum, terminating in complete perforation of the inner and outer tables (Figures 232, 233). There is often abscess formation and fistula with transcutaneous elimination of the sequestrum. The lesion does, not uncommonly, cross suture lines. The margin of the lesion shows active resorption, but no reactive bony wall. Spread along the internal periosteum, with perifocal bone resorption and hypervascularity, is often observed. In this age group, the lesions have to be differentiated from eosinophilic granuloma and metastatic neuroblastoma. Solitary lesions of eosinophilic granuloma usually do not contain a central sequestrum and do not cross suture lines. Multiple lesions of reticulosis usually do not spare the skull base. Metastatic neuroblastoma often shows marked osteoblastic reaction. In adults, the cranial vault lesion is almost always solitary and often much larger than in infants and children. In addition to the hematogenous route, extension from tuberculoma of the brain or dura does occur (Figures 234–236). The process is characterized by a chronic progressive destruction of the cranial vault with irregular margins. Major sequestration is uncommon, in contrast to osteomyelitis, and bony reaction is very limited or absent, in contrast to tertiary acquired syphilis. The defect of the inner table is usually larger than that of the outer (Figures 237, 238), while in tertiary syphilis the greater defect is usually outside, and the inner table may be completely intact (Erdheim, 1932: 355).

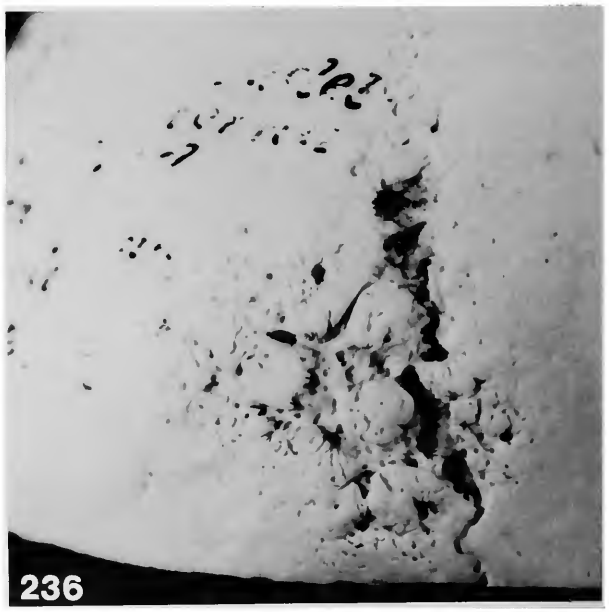
Cranial Base.—The base of the skull is rarely involved in tuberculosis. Tuberculous otitis media is not rare in infants, representing 50 percent of middle ear infections in the first year of life and about 3 percent at all ages (Fraser and Stewart, 1936; 402). There is occasional destruction and sequestration of the petrous bone and mastoid process by secondary extension of mucosal middle





Figures 232, 233.—Tuberculosis of cranial vault: 232, External view; notice little reactive bone around frontal defect and porous sequestrum in parietal lesion. 233, Endocranial view; the frontal lesion is broader based on the inner table. (Adult, DPUS 4584.)



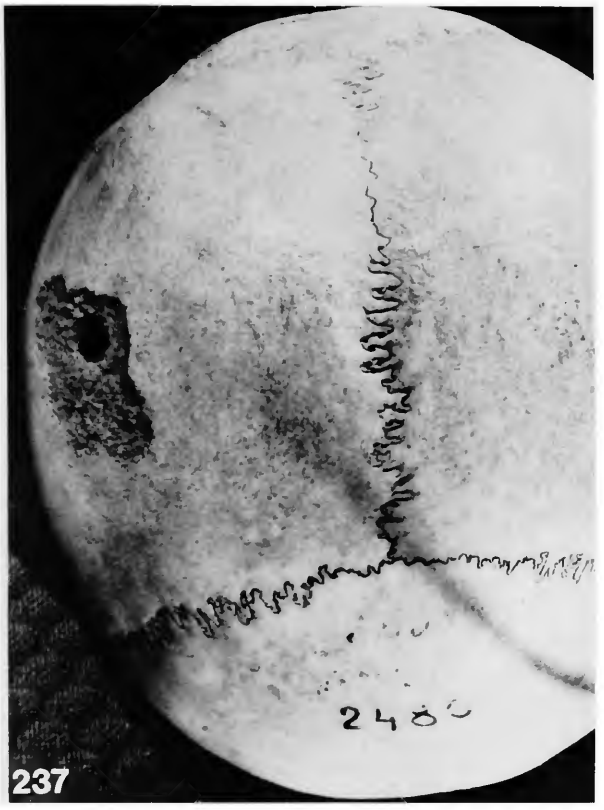


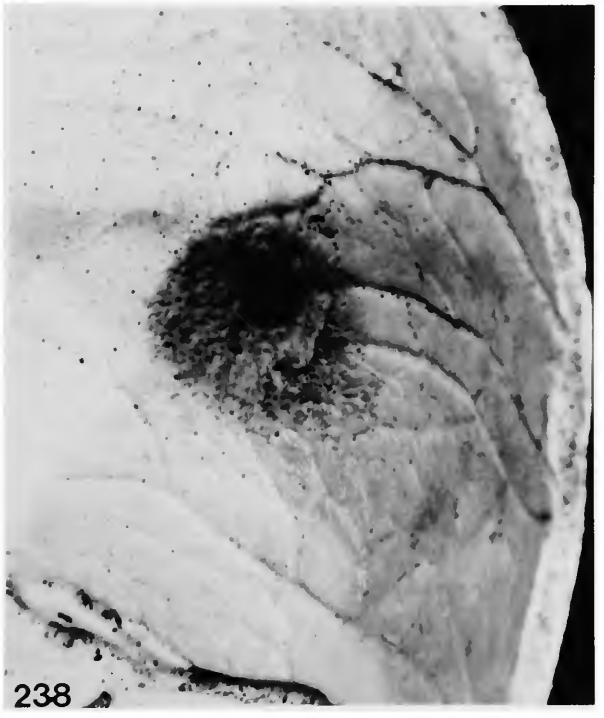
FIGURES 234–236.—Tuberculosis of the cranial vault secondary to tuberculous external pachymeningitis: 234, External view showing small perforation of the outer table. 235, Endocranial view; notice the lesions are much larger on the inside. 236, Left lateral view, showing plaque-like healing. (12-year-old female, FPAM, Jubiläumspital 40, from 1914.)

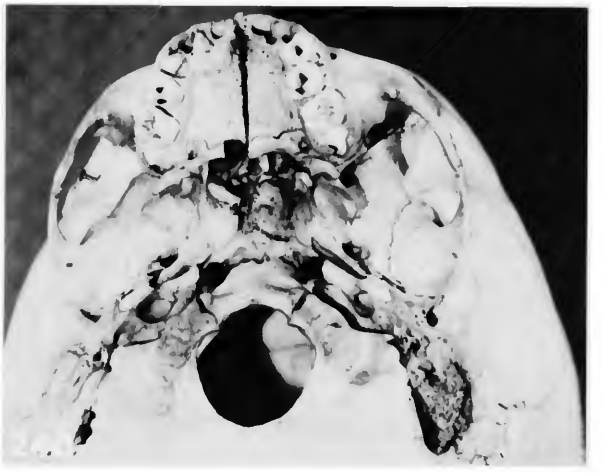
ear tuberculosis (Krause, 1899:54) (Figures 239, 240). Occasionally the base of the occipital bone in the vicinity of the foramen magnum may participate in suboccipital tuberculosis of the atlas and axis (malum suboccipitale) (Figure 241).

FACIAL BONES.—In small children focal tuberculosis of the inferior lateral orbital margin is not uncommon, involving the maxilla, especially at the junction with the zygoma. Involvement of the zygoma itself also is seen frequently. Chronic tuberculosis of the zygomatic arch may lead to an abscess, which typically ascends on the temporal squama along the temporal muscle (Figure 242). Most of these cases show multiple skeletal tuberculosis elsewhere. The lesions are superficial, leading to small sequester formation eliminated through a fistula (Krause, 1899:54–55; Kremer and Wiese, 1930:153–155).

The bony walls of the nasal cavity may be secondarily affected by extension of mucosal tuberculosis (Figures 243–246). The facial bones also can be secondarily involved by long-standing tuberculosis of the facial skin and soft tissues (lupus vulgaris), which often leads to destruction of the nasal bones, as in leprosy (Figures 247–249).







FIGURES 237, 238.—Tuberculosis of right parietal bone: 237, External view; notice small perforation in exposed diplöe. 238, Endocranial view, showing larger defect on inner table sloping to smaller perforation on outer table. (55-year-old female died of tuberculosis, ANM 2480, autopsy 658-(16) from 1896.)

FIGURES 239, 240.—Cranial tuberculosis with lesion in right frontal bone and left middle ear: 239, Lateral view, showing exposed diploë and posterior penetration of the inner table. 240, Basal exterior view, showing involvement of the left middle ear, necrosis of mastoid process and extension to base of sphenoid. (About 8 years old, DPUS 5266, French catalog 779 from 1837.)

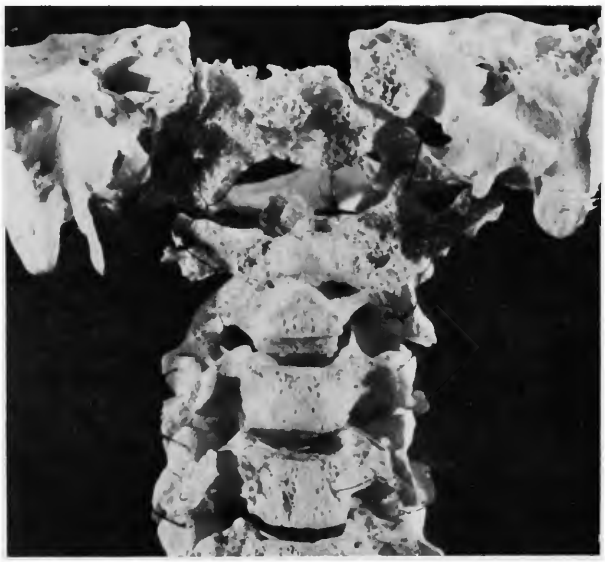


FIGURE 241.—Tuberculosis of cranial base and atlas with superficial erosion of anterior surface of cervical vertebrae suggestive of prevertebral abscess. Sudden death by compression of the medulla. (55-year-old male with chronic pulmonary tuberculosis, PMWH WO 702.)

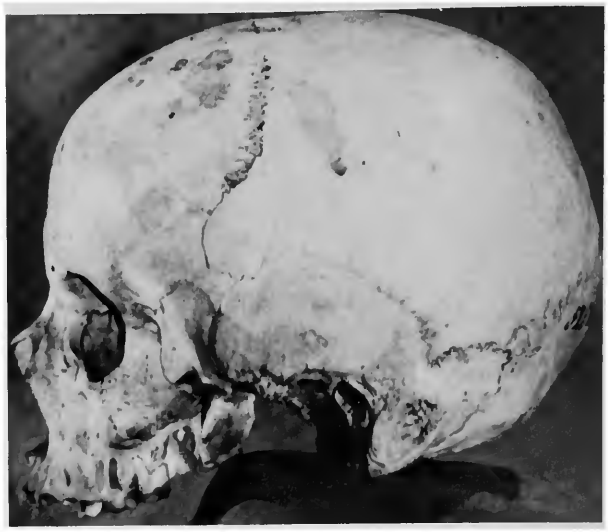


FIGURE 242.—Cranial tuberculosis with destruction of left zygomatic arch and involvement of temporal bone and mastoid process with periosteal reaction to subtemporal cold abscess. Separate foci in frontal and left parietal bone. (Adolescent, DPUS 5268, French catalog 778a from 1837.)

In infants the mandible occasionally also shows hematogenous foci, near the angle. These lesions may be very similar to foci of eosinophilic granuloma. In adults, in advanced stages of open pulmonary tuberculosis, extension of oral mucosal lesions into the alveolar process of the mandible uncommonly and, even less often, of the maxilla has been described (Zandy, 1896).

PALEOPATHOLOGY

The earliest report on presumptive tuberculosis in ancient Old World human remains is an article by Bartels (1907) on a neolithic skeleton found near Heidelberg, Germany. The fourth and fifth thoracic vertebrae have collapsed and fused with the somewhat abnormal sixth vertebra creating an angulation often, but not exclusively, seen in spinal tuberculosis. There are several reports of lesions attributed to tuberculosis in Egyptian skeletal material (Derry and Elliot-Smith, 1909; Elliot-Smith and Dawson, 1924; Derry, 1938b). Elliot-Smith and Ruffer (1910) reported a case of probable tuberculosis in an Egyptian mummy dating from the Twenty-first Dynasty (ca. 1000 B.C.). This specimen shows extensive destruction of the last four thoracic and first lumbar vertebrae. There was some bony reaction to the disease process on the first lumbar vertebra. Inferior to the lesion on the lumbar is a swelling in the soft tissue, which Elliot-Smith and Ruffer judged to be a psoas abscess. Although no tubercle bacilli were found in any of the soft tissue lesions, the morphological evidence for tuberculosis is strong. Indeed, while discounting much of the purported evidence for tuberculosis in the ancient Egyptian material, Williams (1929: 869–873) concluded that the evidence for tuberculosis in the Egyptian skeletal and mummy remains was convincing. Morse, Brothwell, and Ucko (1964) reached a similar conclusion. Although Hrdlička (1909:1) claimed that there were no pre-Columbian skeletal lesions attributable to tuberculosis, there have been several authors who subsequently have reported cases. Hooton (1930:319) reported a case of possible vertebral tuberculosis among the pre- and post-Columbian Pecos Pueblo skeletal series. Ritchie (1952) coordinated a study of three pre-

Columbian kyphotic Indian spines from New York state. In this study he submitted photographs and roentgen films of the three specimens to 17 specialists. The majority opinion of the medical specialists was that in all three abnormal cases the lesions were the result of tuberculosis, although there were some dissenting opinions.

Lichtor and Lichtor (1957) published their observations on a pre-Columbian kyphotic spine from Tennessee. The tenth thoracic through the fourth lumbar were involved with vertebral body destruction followed by collapse, angulation, and fusion. The authors conclude that the lesion is characteristic of spinal tuberculosis. Roney (1966: 103) reports a case of thoracic vertebrae destruction followed by kyphosis in a spine from an archeological site in California.

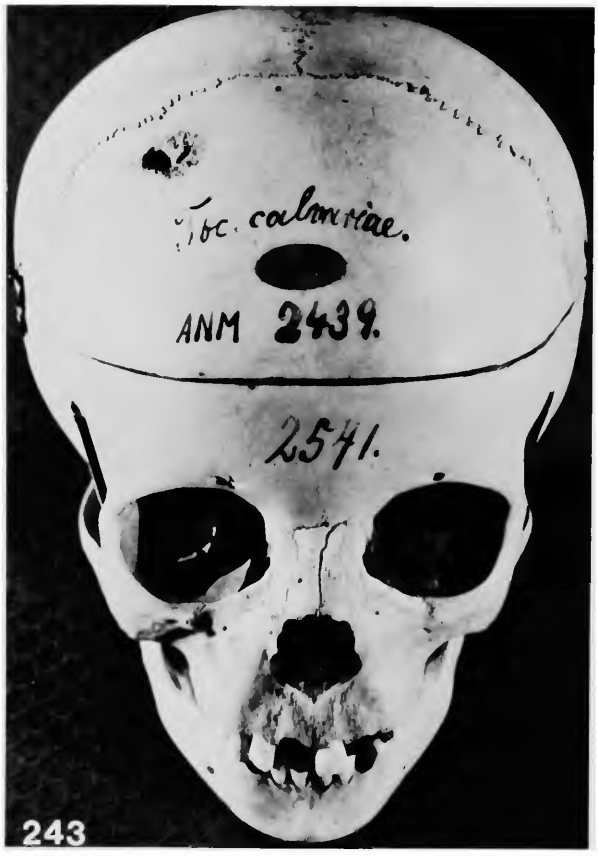
Morse (1961, 1967) reviews the evidence for prehistoric tuberculosis in America. He bases his review on artistic representations of abnormal human figures suggestive of tuberculosis and on a critical evaluation of 15 cases of Indian skeletons that have vertebral lesions, possibly attributable to tuberculosis. Nine cases were from published reports and six were based on original observations by Morse. Of these 15 cases, Morse considered only four to be sufficiently typical to allow a strong presumption of spinal tuberculosis. One of these four cases could not be dated to the pre-Columbian period with certainty. Despite the strong evidence of pre-Columbian tuberculosis in the other three, Morse concluded that the existence of the disease among the prehistoric American Indians had not been demonstrated.

It would seem appropriate to review critically the criteria on which Morse bases his judgment. The following are the characteristics that he attributes to classic vertebral tuberculosis: (1) One to four vertebrae are usually involved, rarely more. (2) Bone destruction occurs with little or no bone regeneration. (3) Angular kyphosis of the affected spine is a frequent sequela. (4) Neural arches, transverse and spinous processes are rarely involved. (5) Cold abscesses drain mostly along the anterior surface of the vertebrae; they almost never drain posteriorly through the skin. (6) Massive regeneration is a great rarity; even spontaneous fusion of affected vertebrae is uncommon.

In addition to these criteria, Morse argues that the rarity of lesions in pre-Columbian skeletal material argues against the disease being present. He bases this conclusion on the following argument. In his survey of American Indian groups living in the American Southwest around the turn of the twentieth century, Hrdlička (1909) found the incidence of bone tuberculosis to be 7 cases in every 100 cases of tuberculosis. Since one of every 3 deaths among the Indians was due to this disease, Morse (1961:493) suggests that about 22 of every 1000 burial specimens should show some evidence of tuberculosis. Morse (1961:493) concludes that if tuberculosis was endemic among the prehistoric Indians, we should see more evidence of it than we do. Since lesions that might be attributed to tuberculosis are rare, paleopathological cases of such lesions are more likely to be one of the several other pathological conditions that can produce similar lesions. One problem with this argument is that social and environmental conditions in the post-Columbian period were very much worse for the American Indians. Both these factors adversely affect the incidence and course of tuberculosis. Thus the incidence reported by Hrdlička (1909) may not be indicative of the incidence in pre-Columbian times.

In his review of the 15 purported cases from the Western Hemisphere, Morse (1961) most often argues against the diagnosis of tuberculosis on the basis of two criteria: (1) destructive lesions extending to more than four vertebrae and (2) significant amounts of bone regeneration. However, Morse (1961:489) himself, notes that the skeletal response to the tubercle bacilli is dependent on the virulence of the organism and the host response. If the host had little ability to defend himself, then tuberculosis could cause a rapidly progressive type of acute osteomyelitis and early death. "If the host's resistance was considerable, it is presumed that the disease then would be extra-chronic with the possibility of extensive healing and bone regeneration."

It is significant to note that in only one of the



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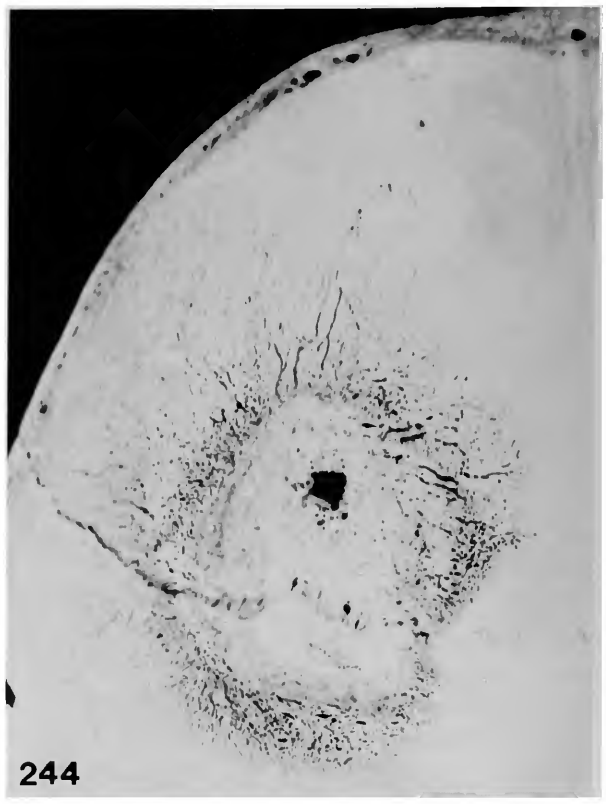
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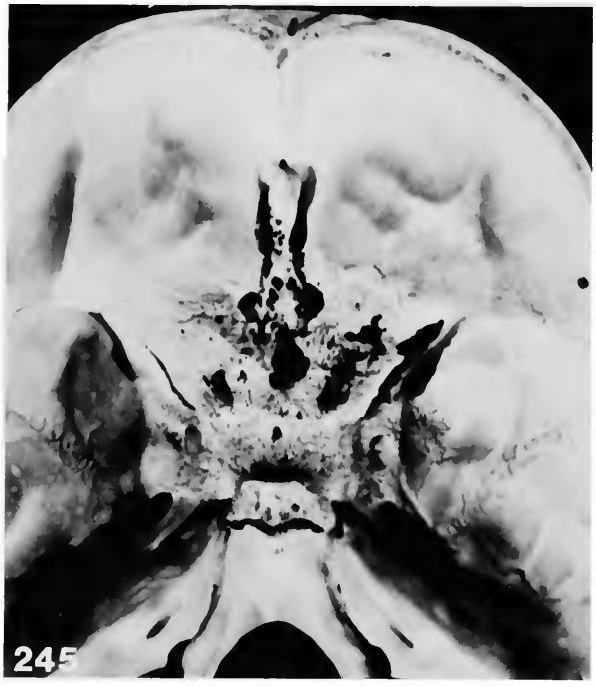
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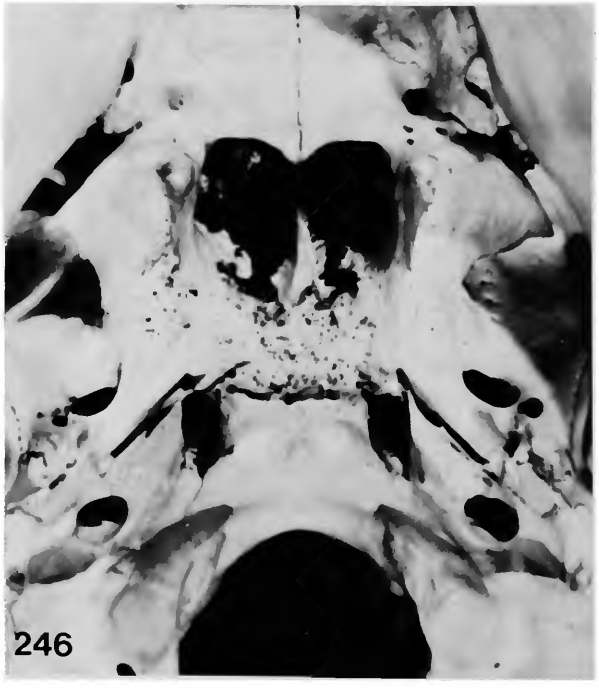
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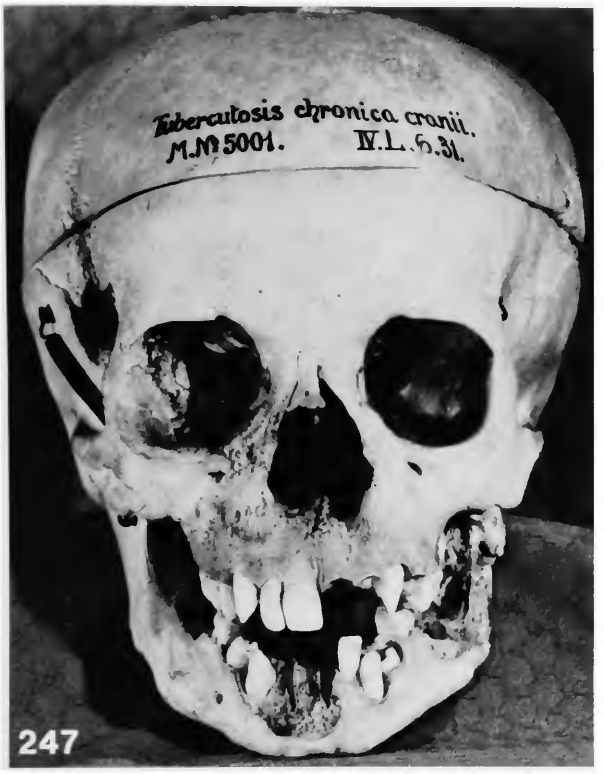
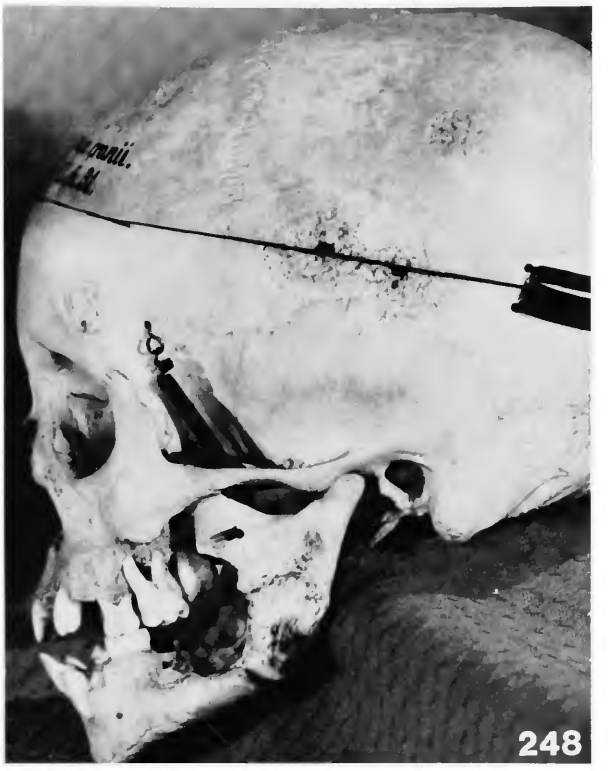




FIGURES 243–246.—Tuberculosis of cranium with right frontal and basal lesion: 243, External view showing small sieve-like perforation of outer table. 244, Endocranial view showing large dura-based lesion crossing the coronal suture. 245, Destructive lesion of sphenoid-ethmoid area, endocranial view. 246, Exterior view of skull base. (10-year-old female, ANM 2439 from 1870.)

FIGURE 247, 248.—Craniofacial tuberculosis in lupus vulgaris with destruction of nose: 247, Anterior view, showing porotic erosion around nasal aperature and maxilla; extensive destruction of mandible. 248, Left lateral view, showing multiple superficial erosive lesions of cranial vault and extensive destruction of mandibular angle. (12-year-old male with visceral tuberculosis and meningitis, FPAM 5001, autopsy 87916 from 1888.)

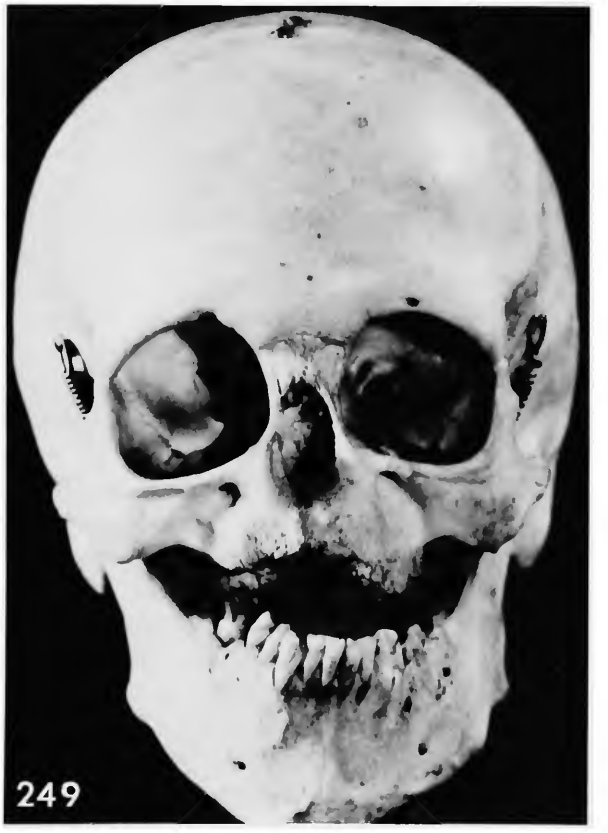


FIGURE 249.—Cranial tuberculosis in lupus vulgaris, with partial destruction of nasal bones, conchae, nasal septum, maxilla, and palate; periostitis of mandible. 10 years duration. (15-year-old male, PMES 1 FT 2(1) from before 1920.)

nine cases does Morse indicate age and it is that of a child. Furthermore, it is this case that provides the most typical expression of vertebral tuberculosis, implying that the acute or subacute cases do not survive to adulthood. As a result acute or subacute cases show the primarily lytic, bony response typical of spinal tuberculosis. As has been stated, vertebral tuberculosis begins most often in early childhood (p. 145). This means that the vertebral tuberculosis occurring in the adult probably reflects a chronic condition in which the host response is at least sufficiently adequate to insure survival through childhood and adolescence. In such long-term, chronic, adult cases the criteria of no more than four vertebrae being involved with little bone regeneration may be less applicable.

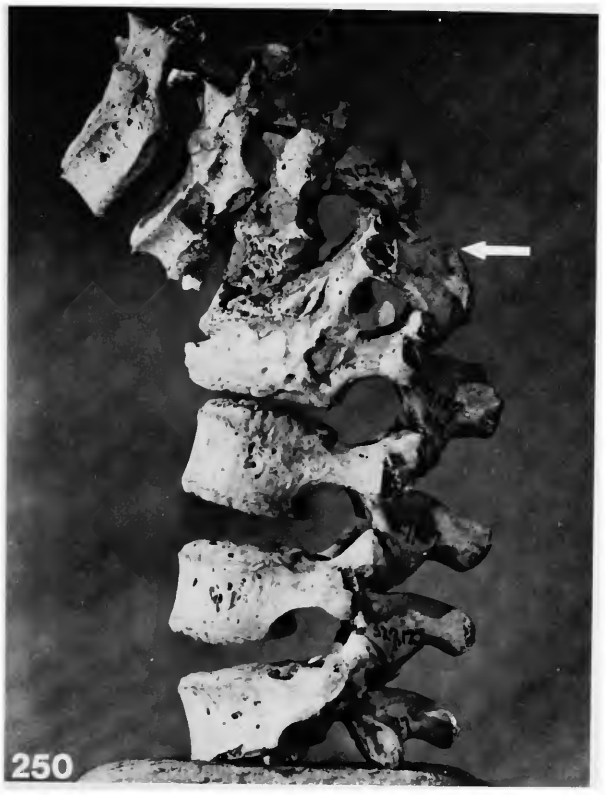
Morse's (1961:489) caution about attributing a specific bone lesion in an archeological specimen to tuberculosis is understandable and, indeed, commendable, particularly at a time when unequivocal evidence for tuberculosis in the Western Hemisphere during pre-Columbian times does not exist. Allison, Mendoza, and Pezzia (1973) have published a report of a case of tuberculosis in a naturally preserved body of a child from southern Peru. The case appears to be well-dated to about A.D. 700, clearly long before European contact. The child was a male between 8 and 10 years of age. The roentgen and gross appearance of the lumbar spine indicates partial destruction of the first three lumbar vertebrae. The authors indicate that the burial position was unusual and suggest that the position of the legs of the burial is commonly seen in living individuals with paralysis of the lower limbs.

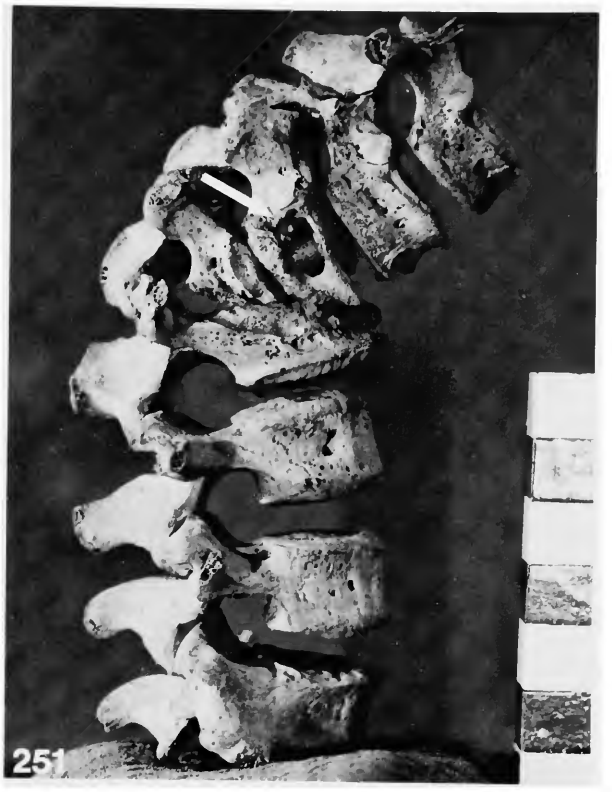
At autopsy, the mummy had a psoas abscess overlying the lumbar vertebrae and nodules resembling tubercles in the right lung. By rehydrating some of the recovered tissues it was possible to prepare stained histological sections, which revealed acid-fast bacilli in the tubercles. The evidence from the roentgen, gross, and histological sources leave little doubt that the child suffered and probably died from tuberculosis. This study would seem to provide unambiguous evidence regarding the presence of tuberculosis in the Western Hemisphere during pre-Columbian times.

The existence of valid evidence for tuberculosis in the New World in the prehistoric period has considerable significance for the diagnosis of lesions in skeletal remains that could be attributed to tuberculosis. Morse (1961:491) lists 13 diseases that produce lesions that can be confused with tuberculosis. The most serious problems in differential diagnosis arises in distinguishing between pyogenic osteomyelitis, perhaps the mycotic infections, and tuberculosis. Brucellosis should be added to this list. Assuming that all of these infectious conditions are endemic, tuberculosis, because it is the most frequent morbid condition, becomes the most likely diagnosis on the basis of probability. Thus, the unequivocal demonstration of tuberculosis in the pre-Columbian New World means that the disease was endemic and acceptance of this diagnosis in more questionable cases becomes a more acceptable conclusion.

Three cases of possible bone tuberculosis in archeological specimens are presented to illustrate the morphology of bone lesions associated with this disease. The first of these cases is a child's skeleton from Pueblo Bonito, New Mexico (NMNH 327127). The archeological date for cultural materials from this site is A.D. 828–1130. The right femur of this specimen measures 265 millimeters (without the epiphyses) which indicates an age at death of about 9 years. This estimate is in good agreement with age based on dental eruption. Determination of sex in a child's skeleton of this age is not reliable.

While some of the bones are damaged or missing, the majority are present including some of the hands and feet. There is no grossly observable evidence of disease in any of the bones except the vertebrae. The first through the fifth cervical vertebrae are missing, although the sixth and seventh are normal. The first through the tenth thoracic vertebrae are normal. The pathological process begins with the eleventh thoracic (Figures 250, 251). The superior surface of the body of this vertebra is normal, as are the transverse, spinous,





FIGURES 250, 251.—Slight to pronounced destruction of the vertebral bodies of the eleventh thoracic through first lumbar vertebrae probably due to tuberculosis: 250, Destruction of the thoracic and lumbar (arrow) vertebrae has resulted in kyphosis; there has been a limited amount of healing. 251, The right lateral view of the lower vertebrae; note the large cavity (arrow) on the remnant of the body of T12. (9-year-old child from the pre-Columbian site of Pueblo Bonito in New Mexico, USA, NMNH 327127; scale in cm.)

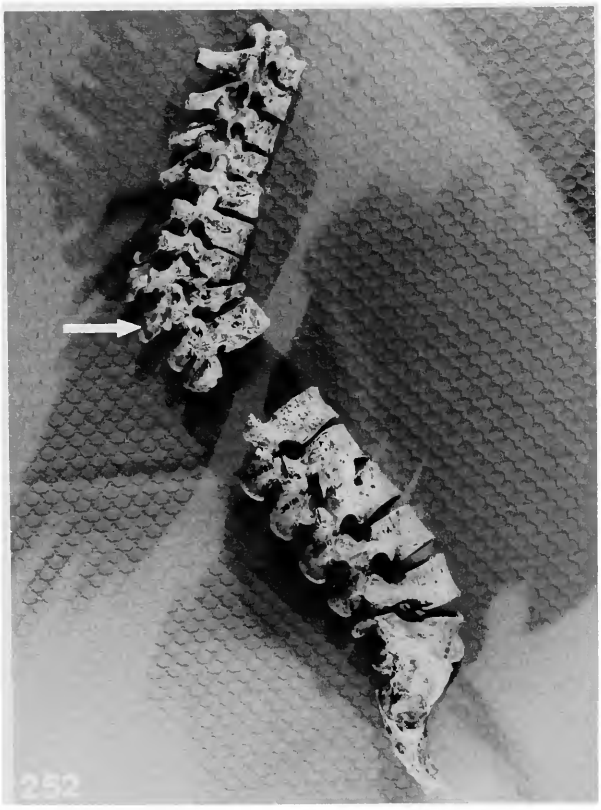
and articular processes. Cavitation is seen on the anterior and left portion of the vertebral body. An enlarged canal resembling a cloaca occurs in the left sector of the body. The inferior surface of the body is eroded from the central to the left side. All vertebral processes are normal.

The anterior body of the twelfth thoracic vertebra is destroyed leaving a scalloped spongy surface in the remaining bone. There is no bony fusion and the spinous, transverse, and articular processes are normal.

The first lumbar vertebra is the most extensively affected bone with almost complete destruction of the body. The left pedicle is fused with the body of L2. All vertebral processes are normal; however, there is slight deformation apparent in the lamina. This suggests the possibility of compensatory remodeling after the destruction of the vertebral body.

The superior surface of the body of L2 is eroded and fused with the remnant of L1. The inferior body is intact but has slight exostoses on the left side. All processes are normal. The third through the fifth lumbar vertebrae are all normal except for slight evidence of deformity on L5, which, like the deformation seen in the lamina of L1, may be a reaction to the kyphosis produced by the collapse and fusion of L1 and L2.

X-ray films of the vertebrae demonstrate the anterior angulation (kyphosis) of the normal axis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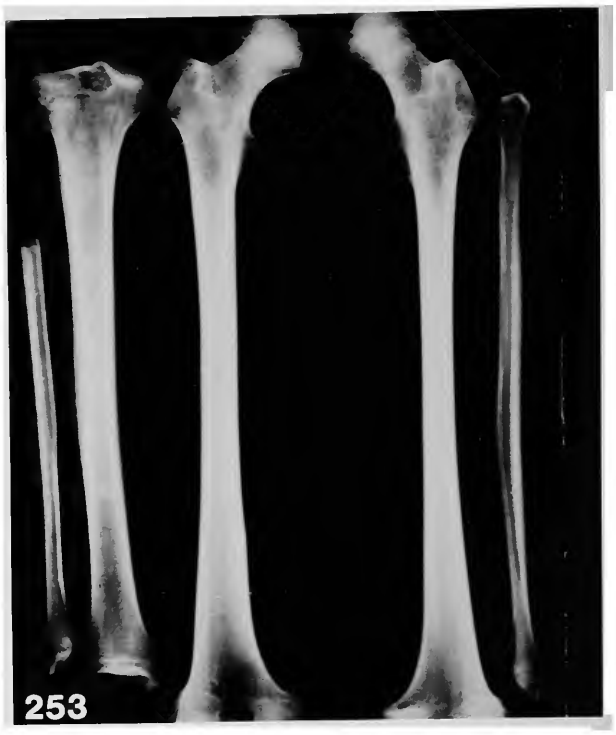
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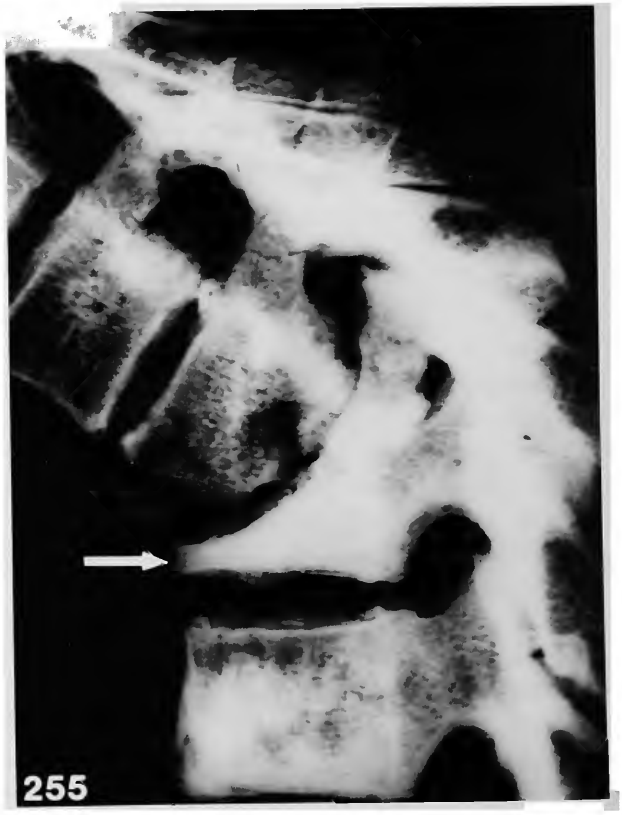
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of the spine. The collapse of the vertebrae produced an angle of about 110 degrees (170°–180° normal). The film also shows evidence of bony response to the deformity in the form of reinforced trabecular bone in the affected vertebrae. X-ray films of the femur suggest two, and possibly three, Harris' lines indicative of acute disease episodes at various times before death. The skull is badly broken and deformed by actions subsequent to death and burial. However, there is no evidence of disease on the skull. The erupted teeth are normal. The age of the individual and the morphology of the lesion are compatible with a diagnosis of tuberculosis, although other diseases are possible. Morse (1961:500) provides a brief descriptive summary of this case (his No. 10) and concludes that it is "quite typical of tuberculosis with destruction of vertebral bodies, angular deformity, and very little bone regeneration."

The second case (Figures 252–255) is a skeleton from Illinois, USA (NMNH 381853), whose archeological provenience cannot be accurately determined. In his brief report of this specimen, however, Morse (1961:502) indicates that it was probably late Hopewell or post-Hopewell Woodland (A.D. 200–1000). The age at death of this individual was about 22 years and the sex is male.

Only a few of the bones of the hands and feet are present. None of these bones show any evidence of disease. The bones of the upper extremity are normal. In the pelvis the sacrum is missing. There are no gross lesions of the innominates, although the left ilium is visibly less flared than the right.

In the lower extremities, the left tibia is missing, although the right is normal, as are both fibulae. Comparative measurements of the two femora are presented in Table 8 and suggest significant differences.

TABLE 8.—Comparative measurements of the left and right femur in possible case of skeletal tuberculosis of archeological specimen from Illinois, USA (NMNH 381853)

Measurement	*Left* (mm)	*Right* (mm)
Maximum length	428	421
Maximum midshaft diameter	27	29
Minimum midshaft diameter	18	21
Neck-shaft angle	152°	136°

The left femur is grossly more gracile than the right. The neck and head of the left femur are more nearly in line with the long axis of the shaft than the right (Figure 253). This condition (coxa valga) is typical when partial to complete paralysis of the limb occurs during the growth phase. The abnormally large neck-shaft angle of the left femur, the reduced size of the shaft diameters, and the diminished flare of the left ilium suggest partial paralysis of the left limb perhaps caused by the collapse of the vertebrae.

In the vertebral column (Figure 252) the first cervical is missing. The remaining cervical vertebrae are normal, as are the first three thoracic vertebrae. On T4 there is a lytic lesion on the anterior inferior surface of the vertebral body, with a similar but smaller lesion on the right portion of the body. T5–T7 have a slight periosteal bony reaction on both sides of the posterior portion of the body. The inferior articular surface of the body of T7 is eroded but shows evidence of circumscription, although there is no exuberant bone formation. The body of T8 is destroyed with the posterior remnant fused with T9 (Figures 254, 255). Both inferior articular facets are fused with their corresponding facets on T9 producing an anterior angulation. The superior plate of the body of T9 is destroyed. There is some peripheral

FIGURES 252–255.—Probable spinal tuberculosis in an archeological specimen from Illinois, USA: 252, The right lateral view of the vertebral column. The entire vertebral body of T8 is destroyed and the posterior remnant is fused with T9 (arrow). 253, Anteroposterior X-ray film of the right and left femora, the fibulae, and the right tibia. Compare the left femur with the right, noticing the diminished diameter of the midshaft and the increased neck-shaft angle of the left femur. 254, Detailed view of the major lytic and kyphotic focus in the vertebrae. The body of T8 is completely missing and the partially eroded body of T7 is resting on the partially destroyed body of T9 (arrow). 255, Lateral X-ray film of the area of angulation of the spine. Osteosclerosis is apparent in the partially destroyed remains of the T9 body (arrow) and in the body of T10. (NMNH 381853.)

reactive bone formation with a large, bony spur on the right lateral portion of the vertebral body. In T10 the articular surface of the vertebral body is intact. There are bilateral bony spurs on the vertebral body with slight cavitation on the anterior surface. T11 and T12 are missing.

The first lumbar vertebra has slight erosion of the superior articular surface of the body and lateral bony spurs with anterior cavitation. The inferior articular surface of the vertebral body is normal. Both L2 and L3 show evidence of destructive lesions on the anterior part of the body. There is a reactive spur joining L2 and L3 on the right portion of their respective bodies with a draining sinus affecting primarily the body of L3. L4, L5, and the sacrum are normal. The skull and mandible were badly crushed and not available for study.

In his review of this case Morse (1961:502) expresses the opinion that such an extensive, destructive process would have resulted in rapid death and that the bony fusion of T8 and T9 must have occurred early in the disease process. Initially he suggests a type of acute osteomyelitis of short duration as the cause of the lesion. However, he did not rule out tuberculosis and later (1967:257–258) included this case as one of four cases "considered typical enough to give a strong presumption of spinal tuberculosis." An alternative opinion is that the destruction is suggestive of an acute initial condition. However, the bony spurs, fusion, and draining sinuses are all suggestive of limited recovery and a more chronic response with life prolonged for some time (a year or two) beyond the acute stage. The abnormality of the left femur suggests a longer time.

The third case (Figures 256–258) is a skeleton of an Eskimo from the Yukon River in Alaska (NMNH 345394). The archeological age of the specimen is uncertain. The estimated age on the basis of epiphysial union and pubic symphysis morphology is 20 years. The sex appears to be male, although the small size for the long bones and the abnormal shape of the pelvis make sex determination difficult. The skull, which does not exhibit any gross pathology, is male in appearance.

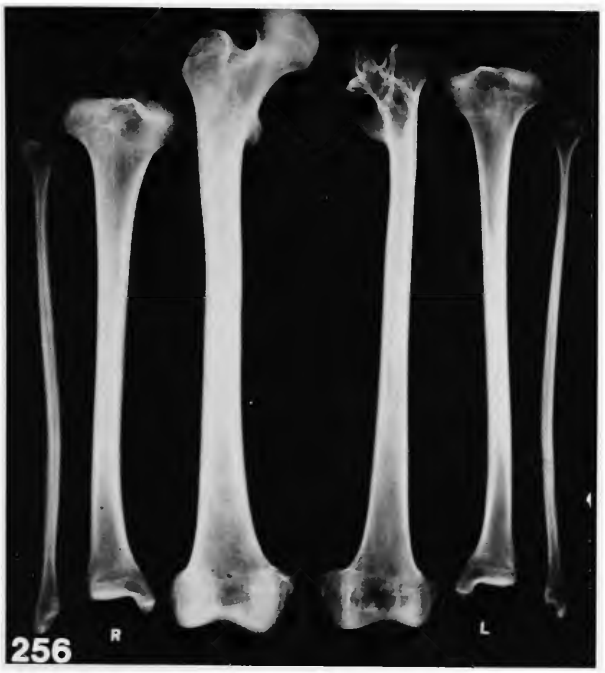
The upper extremities are normal although small and suggest robust muscle activity. The bones of the hands are small but appear normal. There is no evidence of disease in the vertebrae either grossly or on the X-ray film.

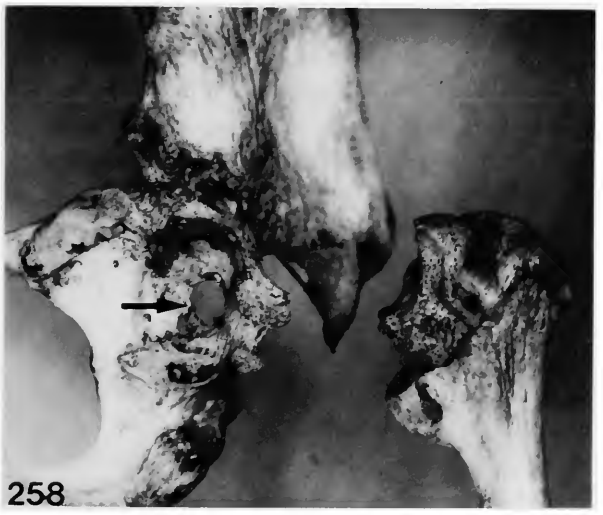
The sacrum is normal, although small. The right innominate is small and somewhat deformed in response to the pathological condition in the left hip. The left innominate is grossly deformed, but the major focus of the infection is the acetabulum. The acetabulum is very shallow, an effect created by the destruction of the acetabular rim and articular surface with subsequent remodeling and healing. There are large, well-organized osteophytes in the posterior rim area and a pronounced cavity in the inferior rim area. A well-developed bony ridge on the anterior-superior margin of the acetabulum is suggestive of upward migration and subluxation of the joint. There is a large hole through the acetabulum in the region of the fovea. The overall gracility of the innominate is exemplified by the very delicate ischial ramus.

The left femoral head is destroyed with subsequent cavitation. Apparently this destruction took place in late childhood with diminished growth of the bone following the acute phase of the disease. The measurements of the two femora highlight the differences in growth occurring after the onset of the disease (Table 9).

TABLE 9.—Comparative measurements of the left and right femur and tibia in a possible case of tuberculosis of the hip (NMNH 345394)

Measurement	*Left* (mm)	*Right* (mm)
FEMUR		
Length from medial condyle to greater trochanter	332	340
Maximum midshaft diameter	20	22
Minimum midshaft diameter	15	20
TIBIA		
Length from lateral condyle to medial maleolus	299	290
Maximum midshaft diameter	18	20
Minimum midshaft diameter	13	15





FIGURES 256–258.—Possible tuberculosis of the left hip in an archeological skeleton: 256, X-ray film of the major long bones of the lower extremity, anteroposterior view; note that the bones of the left leg are more gracile. 257, The anterior view of the right and left femora. The femoral head of the left femur has been destroyed. The lesser trochanter is elongated and the shaft diameter of the left femur is much smaller. 258, A detailed view of the left hip joint; note that the lytic process has produced a hole (arrow) completely through the wall of the acetabulum. (NMNH 345394.)

The measurements of femora length do not include the diseased femoral heads and thus reflect the diminished growth of the pathological left femur. Notice that the left tibia has undergone accelerated growth, perhaps in an attempt to compensate for the shortened left femur. Indeed, the total lengths of the femur plus the tibia for the two sides are virtually identical. This, however, would have left the left leg somewhat shorter due to the destroyed femoral head and the pathological superior subluxation of the hip joint. The resulting abnormal gait undoubtedly contributed to the deformity of both innominates. In addition to diminished growth in length (endochondral ossification) there is reduced appositional growth (intramembranous ossification) of the shaft diameter. While the left tibia has undergone compensating growth in length exceed-

ing that of the right side, its development in midshaft diameter is relatively deficient, suggesting only limited use of the limb in locomotion. The powerful development of the arms may be the result of using some type of crutch to assist in locomotion.

The developmental difference between the legs is also seen in the bones of the feet. The left metatarsals have attained the same length as the right, but have visibly narrower shafts. Curiously, there is a lytic lesion in the right talocalcaneal joint, which has affected the medial articular surface of both bones. The lesion is somewhat circumscribed, indicating containment of the disease process. The relationship of this lesion to the disease process in the left leg remains problematical.

It has been noted that differential diagnosis of distinctive lesions in major joints is difficult (p. 144). Morse (1961:489) indicates that bone lesions from tuberculosis in locations other than the spine are indistinguishable from many other diseases. In view of the obvious problems in differential diagnosis it is, of course, appropriate to avoid dogmatic assertions. However, with this caution in mind, let us review the evidence presented thus far and seen in the illustrations of this case.

In addition to tuberculosis, osteomyelitis and septic arthritis are important entities in the differential diagnosis. In tuberculosis, it has been noted that the active phase leads to destruction and cavitation in the cancellous bone with little perifocal reactive bone. This description certainly applies to the destructive process in the femoral head of this case. There is no evidence of sequestration or involucrum as would tend to occur in osteomyelitis, nor is there fusion of the joint, which is a common sequela in both septic arthritis and tuberculosis of the hip.

Furthermore, tuberculosis of the hip is second in frequency to the involvement of the spine in bone tuberculosis and is the most common destructive lesion of the hip. Dislocation and perforation of the acetabulum in the region of the fovea are also associated with hip tuberculosis. Although ankylosis is a common sequela if healing occurs, the early death of this individual would have precluded this effect. Furthermore, subluxation is not typical in septic arthritis. While other diseases obviously cannot be ruled out, this case does provide an example of what tuberculosis of the hip could look like in an archeological specimen.

Leprosy

Pathology

Leprosy is a very chronic granulomatous infection of man caused by *Mycobacterium leprae*, which is acid-fast and Gram-positive. There is no known animal reservoir for the organism. Leprosy used to be worldwide in distribution with the arctic regions the only exception. Presently, the most affected areas are the tropical and subtropical regions of Asia (India, Thailand, Indonesia, Philippines), Africa, and of the Americas. There is a predominance of male over female in a ratio 2:1 (Faget and Mayoral, 1944). The infection mainly involves skin, mucous membranes, soft tissues, and nerves. The disease manifests itself either as lepromatous, with abundant bacteria in the lesions, or as tuberculoid, with scanty organisms, or an intermediate type. Peripheral, especially sensory, nerves of the extremities are often involved (neural leprosy) leading to loss of sensory perception in affected areas. The development and progression of the disease extends over decades. Various skeletal changes can be observed, based on three different pathogenetic mechanisms: (1) lepromatous osteomyelitis and periostitis, (2) neurotrophic bone and joint lesions, in neural leprosy, and (3) ordinary osteomyelitis and septic arthritis due to secondary infection. In a series of 483 lepers, 306 showed bone lesions radiologically (Esguerra-Gómez and Acosta, 1948).

Lepromatous osteomyelitis is an uncommon and frequently insignificant lesion, which may take decades to develop and involves only a small portion of a bone. Only 5.6 percent of the lepromatous type showed specific lepromatous bone disease (Faget and Mayoral, 1944). Small bones of the extremities (fingers and metatarsals) are

predilected, mostly the epiphysis (Beitzke, 1934d: 595). These lesions appear as lytic destructive foci, which may extend into the adjacent joint. There is no periosteal reaction on the phalanges, and no sclerosis is observed in the healing phase (Paterson and Job, 1964:430). The bone may be infected by the hematogenous route or, more commonly, by direct extension from adjacent skin or mucosal lepromas. On the skull, destruction of nasal bones, nasal septum, turbinates, and of the hard palate does occur (Job, Karat, and Karat, 1966). Erosions of the cranial vault, secondary to lepromatous scalp lesions, can be observed. Møller-Christensen (1953) has described the so-called facies leprosa, which he observed on skeletons with neurotrophic lesions from a medieval Danish cemetery of a leprosarium. The characteristic features frequently found were the disappearance of the anterior nasal spine, the rounding and widening of the nasal aperture, and the partial resorption of the premaxillary alveolar process with or without loss of the upper incisors. He usually found the bony nasal septum and, in most cases, the hard palate intact. Similar lesions occur in tertiary syphilis and in lupus vulgaris.

Periostitis with subperiosteal new bone deposition is not uncommon (de la Camp, 1900; Riordan, 1960). Møller-Christensen (1953) frequently found finely, longitudinally striated, subperiosteal bone deposits on the tibia and/or fibula on skeletons of medieval Danish lepers.

Neurotrophic changes in advanced leprosy are the most common bone and joint lesions observed in this disease. Faget and Mayoral (1944) found such lesions in 66 percent of neural and 33 percent of mixed cases. Destruction of the sensory nerves with ensuing anesthesia, circulatory alterations, and local pressure (Coony and Crosby, 1944) lead to slowly progressive concentric atrophy of terminal phalanges of fingers, metatarsals, and toes. Anesthesia often begins in the distribution of the ulnar nerve and, therefore, the terminal phalanx of the fifth finger is usually involved first (Hopkins, 1928). In the hands, the resorption begins in the distal phalanges and progresses proximally, occasionally including the metacarpals. In the foot, the concentric resorption starts in the proximal phalanges and distal metatarsals, often sparing the subluxed distal toes. The difference is probably due to the effect of local trauma (Faget and Mayoral, 1944). At all times, one observes a complete cortex on these tapered pointed remnants of involved bones. Similar changes, with a similar pathogenic mechanism, are seen in advanced cases of rheumatoid arthritis, but in leprosy no primary arthritic changes are seen. Severe degenerative arthritis and even neuropathic arthropathy, similar to Charcot's joint, can be seen in weight-bearing joints of ankles and feet in advanced cases of leprosy. In addition, the existing anesthesia facilitates traumatic mutilations and secondary infections.

Osteomyelitis and septic arthritis may modify the appearance of the skeletal lesions of leprosy in the terminal phase. In mutilating leprosy, hands or feet may be almost completely destroyed. Even secondary tuberculous arthritis is not uncommon (Beitzke, 1934d:611). In seven of eight autopsied lepers, active pulmonary tuberculosis was found (Brutzer, 1898). The specific granulomatous bone lesions, especially in the phalanges of hands and feet, resemble very closely those observed in sarcoidosis (Paterson and Job, 1964:432). Mutilations also occur in both diseases. Differentiation of these lesions on dry bone alone may be impossible (see also page 233).

Paleopathology

Leprosy is one of the diseases in which features identified in archeological specimens have been of value in modern diagnosis (Møller-Christensen, 1965:603). It has been noted in the discussion of the clinical aspects of leprosy that its distribution was virtually worldwide until recently. However, while the evidence of leprosy in the ancient Old World is convincing, its presence in pre-Columbian New World populations is doubtful (Cochrane, 1964:10).

The earliest purported evidence of leprosy is from a Canaanite jar from Beth-Shan in Palestine (Yoeli, 1955:331). The vessel is dated between

1411 and 1314 B.C. and shows features of the face that resemble those associated with leprosy. Cochrane (1964:2) questions Yoeli's interpretation but indicates that there are references to a disease like leprosy in China dating to 600 B.C. The disease is first reported in Europe around A.D. 150, increasing to its most serious intensity between A.D. 1000 and 1400. By 1798, the disease had died out in Great Britain, although it continued to be a minor problem in continental Europe (Cochrane, 1964:7). It was present in Norway through the nineteenth century. Andersen (1969:123) suggests that the disease may have been introduced into the Western World by the soldiers of Alexander the Great on their return from the campaign in India in 327–326 B.C.

One of the problems in using historical documents in studying the history of leprosy is the vagueness of the descriptions and imprecision of terms. MacArthur (1953:8) suggests that the term "leprosy" was used for nonspecific skin diseases, elephantiasis, small pox, bubonic plague, mange, etc. He further suggests that many of the people relegated to lazar houses in medieval times may have been suffering from diseases other than leprosy. However, Chapman Binford (1965), responding to a scientific paper read by Møller-Christensen (1965), emphasized that the work by Møller-Christensen on the cemeteries of leper houses or hospitals in Denmark showed that most if not all of the people buried there could have had leprosy. This suggests greater precision in the diagnosis of leprosy than MacArthur (1953) proposes.

Møller-Christensen's research (1952, 1953, 1961, 1965, 1967, 1978) has provided empirical evidence for the presence of leprosy in Denmark during the Middle Ages. Wells (1962a) reports a probable case of leprosy in a sixth-century A.D. Saxon burial in Great Britain. Brothwell (1958: 287) found lesions in a post-Conquest skull from Scarborough in England, which he attributes to leprosy. Wood-Jones (1910b:283) describes an ancient Nubian skull in which the nasal bones, including the turbinates and nasal septum, were destroyed. In the same specimen the major portions of the hard palate were also destroyed. Møller-Christensen and Hughes (1966:243) concluded that this destructive process was probably the result of leprosy. They also found an additional Nubian skull from the same series, in which leprosy was a probable diagnosis. Gladykowska-Rzeczycka (1976) attributes to leprosy the lesions seen in an adult male skeleton (her No. 20) from a medieval cemetery located in Suraż, Lapy County, Poland. There is atrophy of the anterior nasal spine, evidence of truncated metatarsals, and extensive periostitis of the long bones.

If, as seems probable, leprosy was introduced into the New World during the colonial period, archeological examples of leprosy should be rare and limited to the post-contact period. I know of no reports suggesting the presence of leprosy in the New World before Columbus. Indeed, leprosy remains a condition rarely mentioned in the New World paleopathology in the post-contact period as well. This is not too surprising, since leprosy is one of the least contagious of transmissible infectious diseases (Browne, 1970:640). Thus, its introduction into the New World would not have had nearly the impact of other infectious diseases, such as small pox, malaria, and measles. Møller-Christensen and Inkster (1965:12) have a reference to an Indian skull from Vancouver Island, off the coast of Canada, found in 1866, which they suggest had the facies leprosa syndrome associated with leprosy.

Møller-Christensen's experience with many skeletons from leper cemeteries having lesions presumably produced by leprosy provides significant insight regarding the bone changes to be expected from this disease (Møller-Christensen, 1961). The major foci for the disease are the face and the small bones of the hands and feet, although other bones may be affected. The skull lesions include the destruction of the nasal bones, particularly the anterior nasal spine and hard palate. He also found a high incidence of cribra orbitalia (usura orbitae) in leprous specimens (1953:134–135; 1961:29). Henschen (1961:729) observed that cribra orbitalia may result from insufficient nutrition and chronic inflammation. This suggests that cribra orbitalia may not be a primary lesion of leprosy but arise from the poor

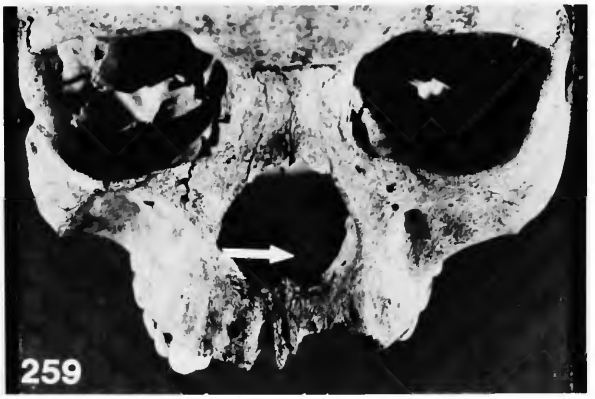
nutrition and other problems that may attend the disease.

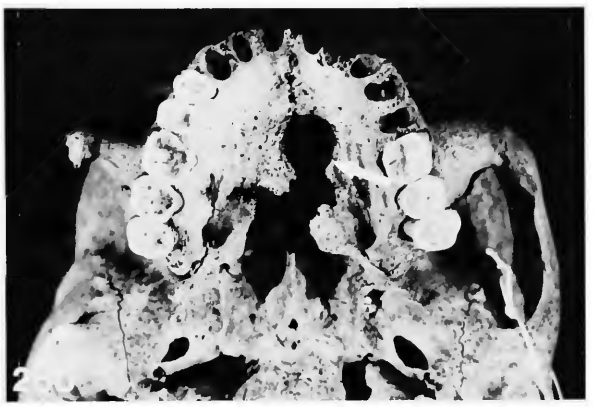
The postcranial lesions are most pronounced in the extremities, particularly the hands and feet, with concentric atrophy and truncation of the phalanges (Møller-Christensen, 1961:15). The bony destruction appears to affect the subarticular, more highly vascularized, bone in the epiphysis adjacent to the articular surface. This process compromises the joint leading to collapse and a cupping deformity of the joint.

Møller-Christensen (1961:13) states that atrophy of the anterior spine and destruction of maxillary alveolar bone are pathognomonic of leprosy. However, my experience with anatomical specimens of syphilis indicate that very similar conditions are found in syphilis. Møller-Christensen (1965:604) indicates that in leprosy, pathological changes are not found on the skull vault. This contrasts with syphilis where changes on the vault predominate. This is certainly a helpful criterion but should not be considered absolute. I have studied pathological cases of purported syphilis in recent materials in which there was nasal destruction with no vault involvement (see Figures 274, 275, 277, 278, 279). The facies leprosa syndrome, in combination with atrophy and truncation of the fingers and toes, would appear to be almost pathognomonic for leprosy, although it is conceivable that combinations of other diseases (e.g., syphilis and frostbite) might cause this condition.

Probably the best documented, archeological examples of leprosy are those skeletons excavated by Møller-Christensen at Naestved, Denmark. This town was the site of a medieval leper hospital and was part of a complex called the St. Jorgen's Hospitals, which were built throughout Denmark (Møller-Christensen, 1953:14–15). Most of the 200 individuals excavated showed bone changes attributable to leprosy and formed the basis for the classic work by Møller-Christensen on such changes.

One of the specimens recovered during these excavations was presented by Møller-Christensen to the British Museum (Natural History). This specimen (BMNH 1962.1.1.2) is the cranium of





FIGURES 259, 260.—Female leper from the medieval leper cemetery at Naestved, Denmark: 259, The nasal aperature is enlarged with evidence of an inflammatory response in the bone adjacent to the aperture. Note the lytic lesion in the palate seen through the aperature (arrow). 260, The palate of the female leper showing a possible lytic focus (arrow) and the porous bone adjacent to the lytic lesion indicative of an inflammatory condition. (BMNH 1962.1.1.2.)

an adult female leper dated to between A.D. 1250–1550. The destructive lesions focus on the nose, whose lateral margins have been eroded, producing an oval rather than the normal elongated opening (Figure 259). The anterior nasal spine is destroyed, and there is a suggestion of alveolar resorption, although postmortem destruction complicates the picture (Figure 260). There is a rounded, penetrating defect in the left portion of the hard palate, which extends slightly across the midline. The central, lytic lesion is surrounded by hypervascular, partly resorbed bone. Neither the mandible nor the postcranial skeleton was avail-

able for study. However, Møller-Christensen (1953) does provide descriptions of bone lesions in the extremities of other skeletons from the Naestved cemetery, which resemble those discussed earlier (p. 176). One possibly significant diagnostic feature is that the metacarpals are not as severely affected in leprosy as the metatarsals (Møller-Christensen, 1953:141).

Treponemal Infections

Pathology

Treponemal infections occur in four clinically different diseases having a worldwide distribution through the tropical and temperate zones of all continents. They are venereal syphilis, endemic syphilis (treponarid), yaws, and pinta, of which only the first three may affect the skeleton. Whether they are caused by four different species of treponema or by variants of the same species is still unresolved and does not concern us here. The important feature is that bone lesions of venereal syphilis, endemic syphilis, and yaws are so similar that diagnostic differences of individual lesions cannot be elicited with certainty (Hackett, 1976). However, there is no evidence that yaws produces the "worm-eaten" appearance of the outer table of the skull that is seen in syphilis (Williams 1935:613). The geographic distribution before antibiotic eradication was very characteristic. Yaws was a worldwide disease of tropical indigenous populations. Pinta was restricted to tropical America. Endemic syphilis was widespread among indigenous populations of subtropical North Africa, the Near East, and temperate Asia, but not found in the Americas. Venereal syphilis is a sporadic disease following the habitations and colonizations of urbanized white races. In all treponematoses, the infecting organisms enter the body through the skin or mucous membrane near the skin surfaces. In venereal syphilis, *Treponema pallidum* enters mostly in areas of sexual contact, especially the genitals. In endemic syphilis and yaws, the infection may locate anywhere on the body surface by intimate contact, mostly already occurring in childhood. In all three treponematoses affecting bone, the organisms are disseminated throughout the body and reach the skeleton by the bloodstream.

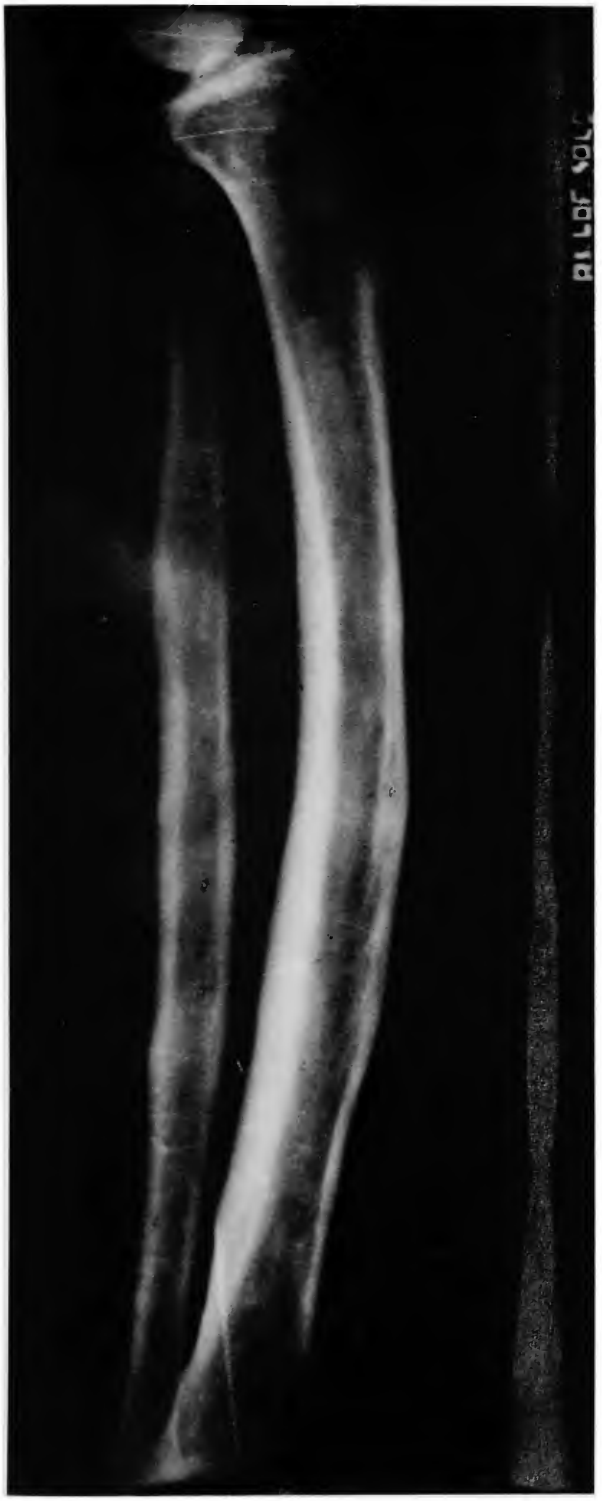
Yaws

Bone lesions in yaws occur in about one percent of the patients (Wilson and Mathis, 1930). The location of such lesions in 101 cases observed by Goldmann and Smith (1943) was as follows: tibia 46, fibula 20 (19 times with tibia), femur 13, ulna 10, humerus 9, radius 7 (6 times with ulna), spine 5, clavicle 4, hand 4, foot 4, skull 3, ribs 3, pelvis 2.

Since yaws is mostly acquired in childhood, the most active lesions are seen in children and in adolescents. Many are similar to those seen in congenital syphilis. This is particularly true of the often symmetrical dactylitis of the spina ventosa type and of the periostitis (Hackett, 1957:14–15). However, since it is not a disease of the newborn, osteochondritis is not seen in yaws. The early bone lesions of yaws may completely heal without leaving permanent bone changes. Another finding frequently observed in yaws is bending of the tibia (boomerang leg)(Figure 261), which is very similar to the saber tibia of congenital syphilis and usually begins before 15 years of age (Wilson and Mathis, 1930). Hackett (1936:54–56) describes disseminated radiologic lucencies in the anterior cortex in the early stages followed by anterior cortical thickening and bending. In the late stage, the posterior concave cortex is thickened and the anterior cortex thinned, as in late rachitic deformities. Rarely, the fibula participates in the deformity and occasionally radius and ulna show similar bending (Hackett, 1936: 52–54). These changes are indistinguishable from postrachitic deformities and may not be due to yaws alone.

Late yaws show destructive dactylitis of single phalanges. The long bones, especially the tibia and bones of the forearm, show gummatous periostitis and osteomyelitis, very similar to tertiary syphilis. In contrast to early yaws, the overlying soft tissues frequently ulcerate and thus open the way for secondary pyogenic infections (Hackett, 1957:16).

Skull changes mainly consist of destruction



around the nasal cavity, particularly extensive in gangosa (nasopharyngitis mutilans), a rare complication of yaws in Africa and South America. Destruction of the nasal bones, the nasal septum and the hard palate are much less frequent in ordinary tertiary yaws than in syphilis. The same is true of the involvement of the cranial vault. If the frontal bone is involved in tertiary yaws, it shows shallow, pitted defects rather than the characteristic appearance of syphilitic caries sicca.

Endemic Syphilis (Treponarid)

This treponemal infection occurred in Syria, Bosnia, and certain areas of Africa. In a survey of 25,000 persons in Bechuanaland, 26 percent showed latent and 1.4 percent active treponemal infections (Murray, Merriweather, and Freedman, 1956:991). In this study the predilected skeletal locations were tibia and ulna. Bone lesions were not seen in children less than 2 years old (Murray, Merriweather, and Freedman, 1956:1000). In a study from Syria, the changes observed resemble those of acquired or late congenital syphilis. They consisted mostly of periosteal bone deposits causing fusiform swelling but little, if any, medullary changes. Intracortical, sharply lytic, rounded gummas are occasionally found. Saber tibia does occur (Rost, 1942:321–323). Periostitis and gummas in short bones of hands and feet have been observed in Bosnia by Grin (1935:482). Charcot's joint is not observed in endemic syphilis (Rost, 1942:323; Murray, Merriweather, and Freedman, 1956:1001). Destructive nasal lesions leading to perforation of the hard palate are not uncommon (Grin, 1935: 482; Murray, Merriweather, and Freedman, 1956:1000).

The detailed discussion of the bone lesions is

FIGURE 261.—X-ray of saber tibia in yaws. Notice the bowed and elongated tibia with anterior cortical osteoporosis and posterior cortical thickening. Compare with the normal fibula. (48-year-old Javanese male with yaws since childhood; studied by W. Putschar at Orthopedic Rehabilitation Center, Solo, Java, Indonesia.)

essentially based on cases of venereal syphilis, with the understanding that similar or identical lesions can occur in yaws and in endemic syphilis (treponarid).

Venereal Syphilis

Venereal syphilis is transmitted through sexual contact. This type is called "acquired syphilis." However, the actively infected female can pass the organisms transplacentally to a fetus, so that the infant is born with the disease. This type is called "congenital syphilis" and will be discussed separately (p. 198). In view of the mode of transmission, it is obvious that, as a rule, acquired syphilis commences beyond the age of puberty. After an incubation period of several weeks following the infective contact, a distinct primary lesion (chancre) usually develops at the point of entry. The ensuing long lasting disease is customarily divided into three stages.

The primary stage begins with the appearance of the chancre and ends with the involvement of the regional lymph nodes, to which the organisms migrate. The secondary stage begins with dissemination of the organisms through the blood stream, characterized by transitory skin rash and mucous membrane lesions. The borderline between the secondary and the tertiary stage is not as clearly defined. However, the tertiary stage is mostly characterized by progressive syphilitic involvement of different organs, as, for instance, the skeleton. It is in this stage that the tissue reaction may assume a distinct granulomatous appearance of nodular foci with central liquifying necrosis (gumma). The character of the inflammation at all stages is more chronic, with lymphocytes and plasma cells predominating. Neutrophilic leucocytes are not prominent and there is no significant formation of pus. Hypervascularity is usually marked. In the secondary stage periostitis is not uncommon, producing some periosteal bone. These changes are usually transitory and leave no characteristic permanent alterations on the bone.

Our main interest centers on the bone lesions of the tertiary period. For the study of these one has to go back to specimens collected before the advent of effective chemotherapy and antibiotics, which means, approximately before 1910. It is estimated that at that period about 10 percent of inhabitants of European cities gave positive serological reactions for syphilis, which is not identical with active disease (McElligott, 1960). In a survey of 2000 cases of syphilis in Norway covering the three decades from 1890 to 1920, Gjestland (1955) found that approximately one percent of the patients showed bone lesions. The combination of these figures would result in the frequency of syphilitic bone lesions in about one in 1000 Europeans in the time period before effective treatment was discovered (Hackett, 1976:114).

The tertiary syphilitic bone lesions usually develop between 2 and 10 years after the infection, but may occasionally occur earlier or much later. Often more than one bone is affected. Although any bone can be occasionally the seat of a syphilitic lesion, there are a few areas that are greatly predilected: the tibia, the bones surrounding the nasal cavity and the cranial vault. These three locations combined represent about 70 percent of all tertiary syphilitic bone lesions. These are followed in decreasing frequency by the cancellous bone, rich in hemopoietic marrow (ribs, sternum), and the long bones of the extremities. A survey of frequency from the greatest series of Fournier (1906, as cited in Beitzke 1934a:471) is given in Table 10. The spine is an exception to the behavior of other cancellous bones, rarely being involved by syphilis.

Tertiary syphilitic bone changes are the result of either chronic nongranulomatous inflammation or granulomatous (gummatous) processes. In many cases a combination of the two is present. Either one of these changes can affect only a localized area of the bone or the entire bone. The inflammation may begin on the periosteum or in the bone. Ultimately, however, periosteum and cortex, more rarely the medullary cavity, are involved. All tertiary syphilitic bone lesions are characterized by an excessive osteosclerotic response to the infection. In many instances adjacent mucosal surfaces, as in the nasopharyngeal area, and overlying skin, as on the scalp or shin, or soft tissue are involved and ulcerated. The

FIGURE 262.—Tertiary syphilis of cranial vault, outside view. These are relatively early lesions with central destruction and peripheral reactive bone. The inner table shows only minimal diffuse reactive bone deposition. (Adult, ANM 2232.)

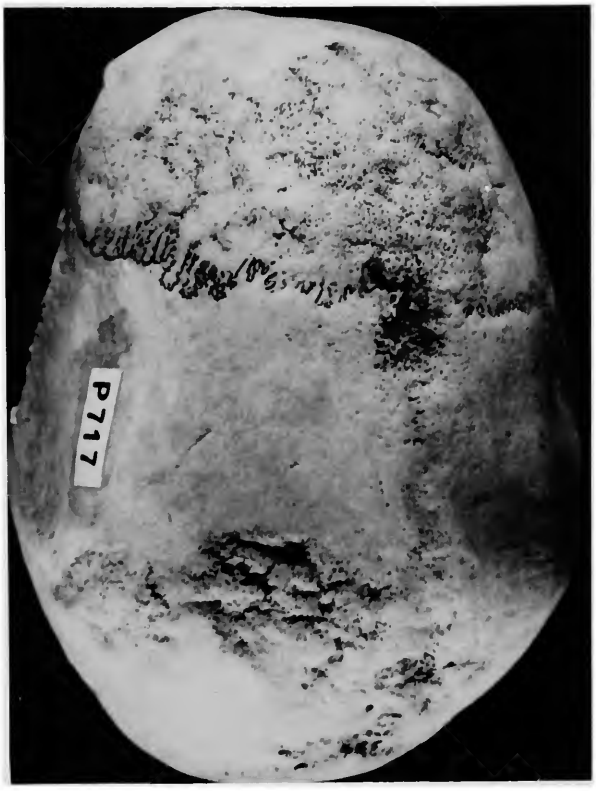
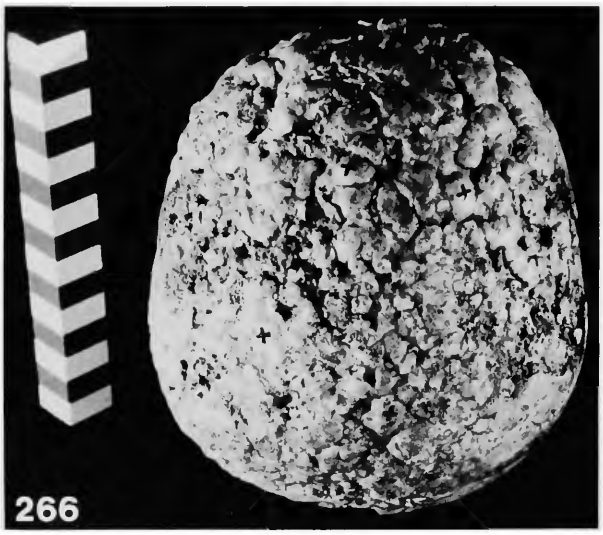


FIGURE 264.—Syphilis of cranial vault with considerable healing; top view. (HM P717.)

FIGURE 263.—Tertiary syphilis of cranial vault, external view, multiple small cavitating lesions with advanced and partly complete healing; inner table unchanged. (Adult, ANM 2468.)

FIGURE 265.—Extensive syphilis of cranial vault with advanced sclerotic healing. (Adult, WM S50a.2 from 1828.)



FIGURES 266–268.—Syphilis of cranial vault (caries sicca): 266, Top view, showing advanced healing with nodular sclerotic bone (scale in cm). 267, Endocranial view, showing only slight bony buildup with hypervascularity. 268, Cut surface, showing marked thickening of vault with sclerotic and porotic areas alternating. (Adult, PMSG 9/1.372E.)

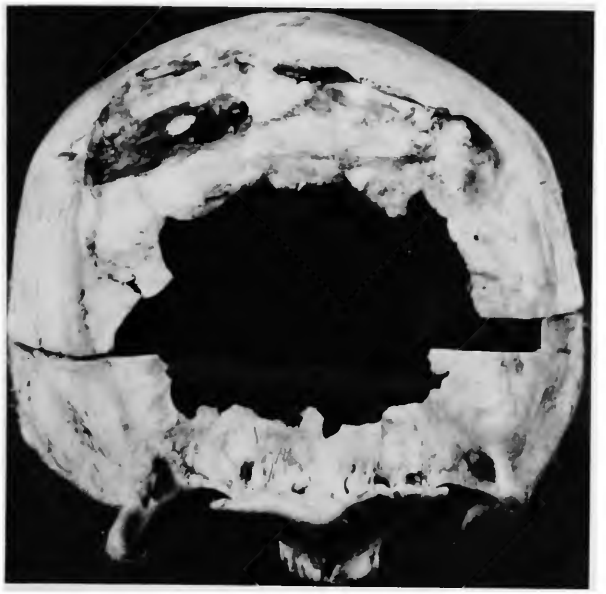
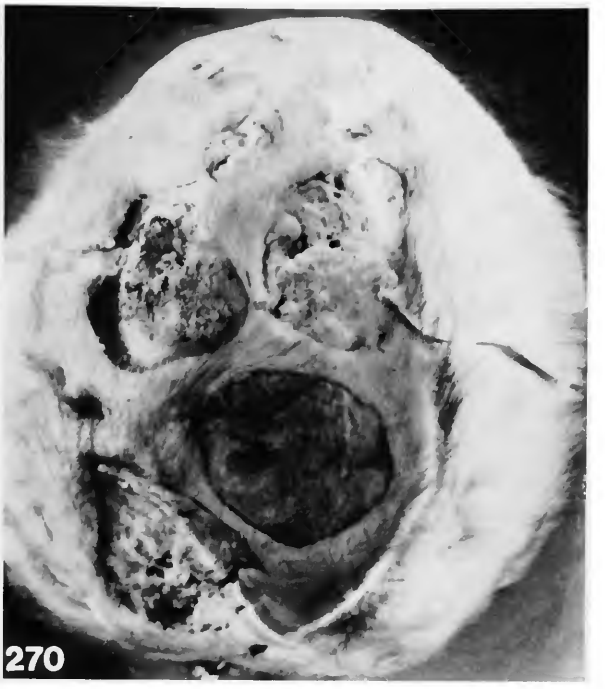
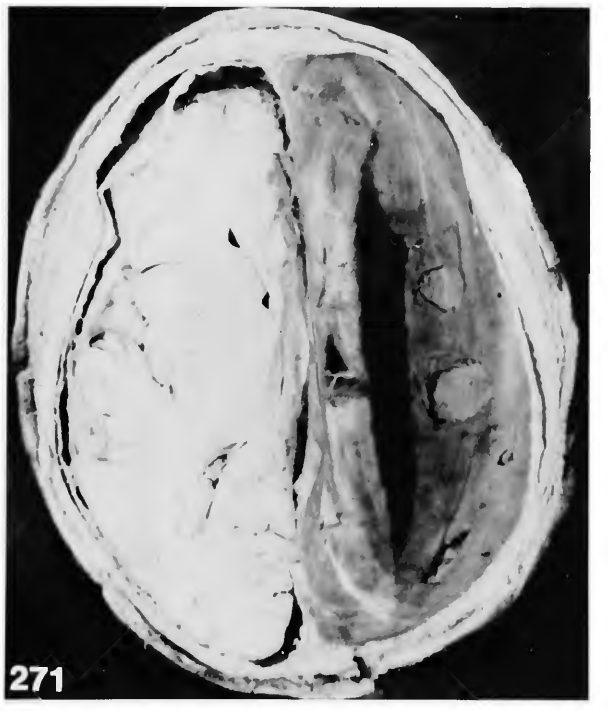


FIGURE 269.—Infectious defect of frontal bone covered by scarred soft tissue, probably syphilis with superimposed osteomyelitis, 25 years duration. (58-year-old male, PMES 1E.B.1(4) from 1899.)

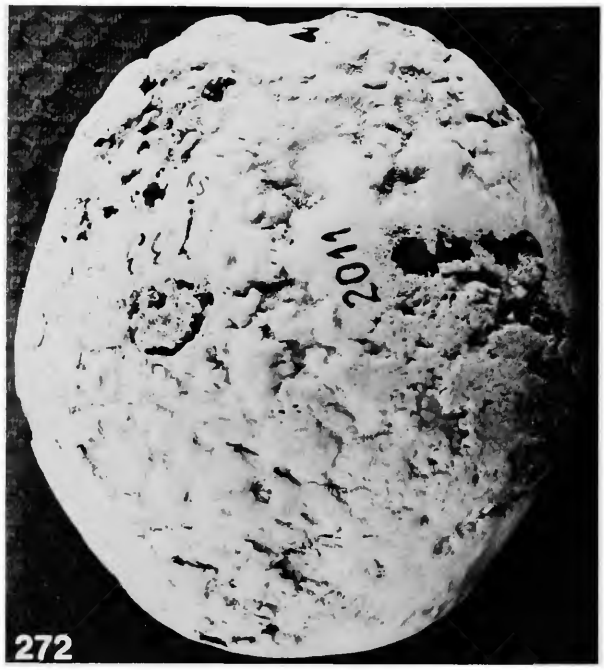
TABLE 10.—Localization of 945 tertiary syphilitic bone lesions in order of decreasing frequency (after Fournier, 1906)

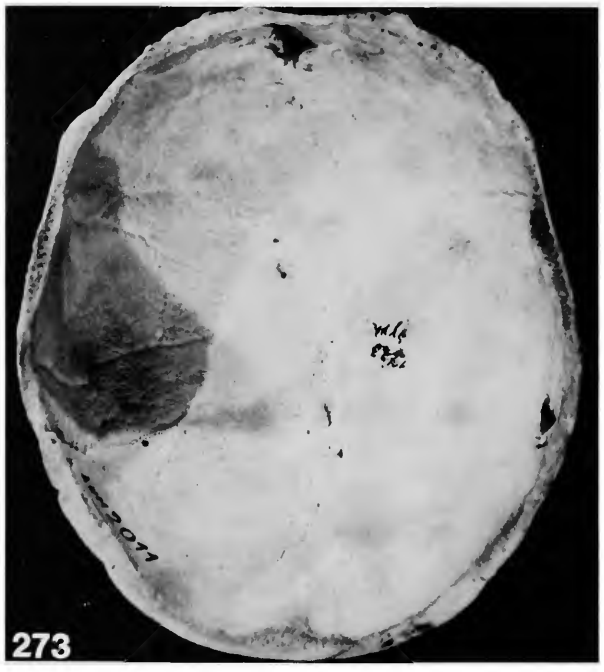
Bone	*No.*
Tibia	248
Nose and palate	238
Skull	179
Ulna	37
Ribs	35
Sternum	29
Clavicle	27
Metacarpals	21
Humerus	20
Radius	17
Femur	16
Mandible	14
Fibula	12
Spine	9
Nasal bone	9
Fingers	7
Pelvis	5
Metatarsals	4
Scapula	4
Ribs and sternum	3
Tarsals	3
Toes	3
Maxilla	3
Patella	1
Carpals	1



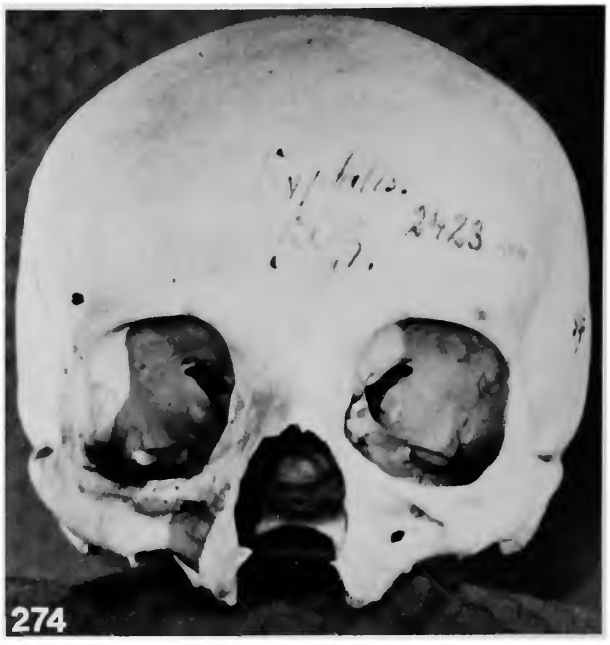


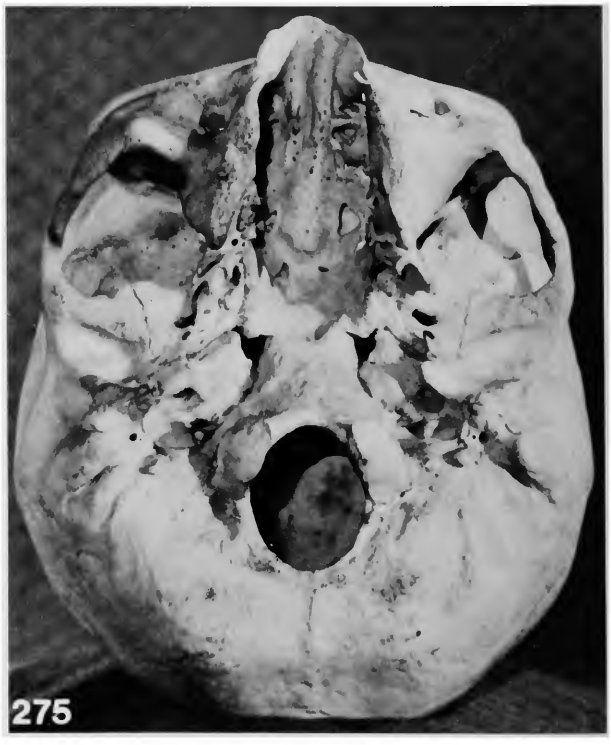
FIGURES 270, 271.—Tertiary syphilis of cranial vault: 270, Surface view, wet preparation, showing multiple scalp ulcerations exposing the affected bone. 271, Endocranial view, wet preparation, showing multiple perforations of the dura over the affected bone. (GHPM 3914 from 1914.)





FIGURES 272, 273.—Tertiary syphilis of cranial vault (caries sicca): 272, Surface view, showing extensive sclerotic healing between areas of active destruction. 273, Endocranial view, showing only small perforations and slight deposition of reactive bone on the inner table. (Adult, ANM 2011.)





FIGURES 274, 275.—Syphilis of nasal cavity: 274, Frontal view, showing widening of the nasal cavity, complete destruction of the nasal septum and large perforation of the hard palate. 275, Exterior basal view, showing eroded hypervascular bone at margin of defect. (Old female, ANM 2423.)

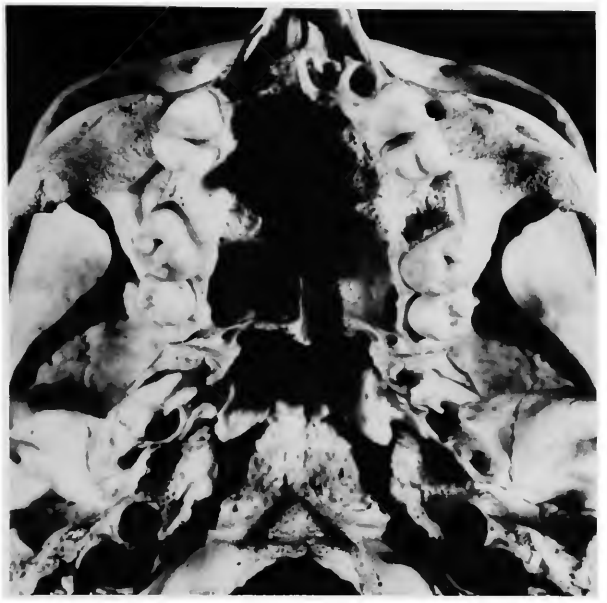


FIGURE 276.—Syphilis of nasal cavity with destruction of hard palate and perforations of ossa palatina; exterior basal view. (24-year-old male syphilitic, WM HR 4.1 from 1848.)

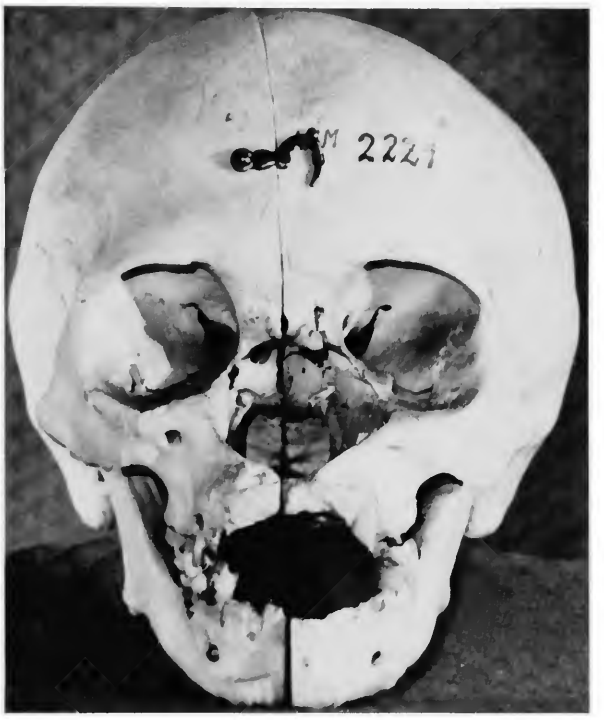


FIGURE 277.—Nasomaxillary syphilis with extensive endonasal destruction and some sclerotic healing, destruction of the palate and involvement of the maxilla. (43-year-old female, ANM 2221 from 1889.)

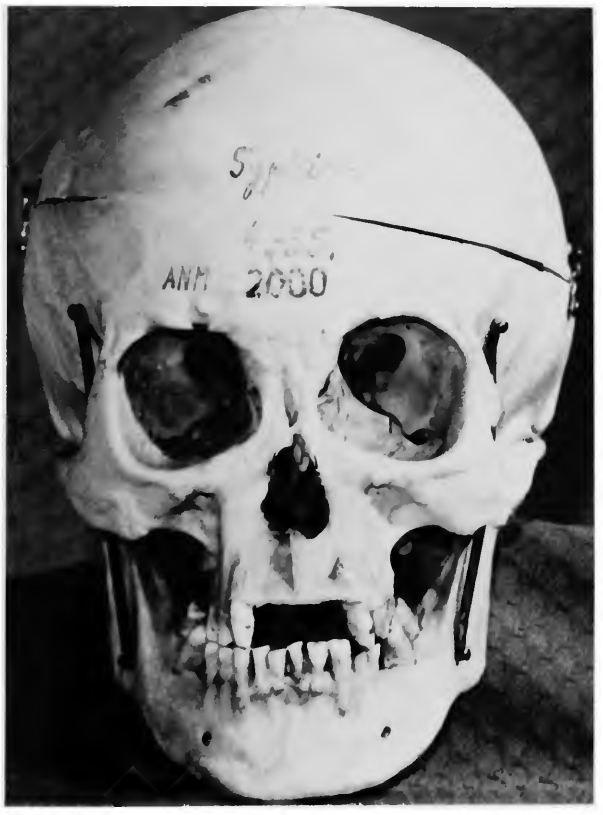


FIGURE 278.—Cranionasal syphilis, mostly healed. Notice scarring of nasal and frontal bones; also preservation of the inferior nasal spine. (43-year-old male, ANM 2000 from 1893.)

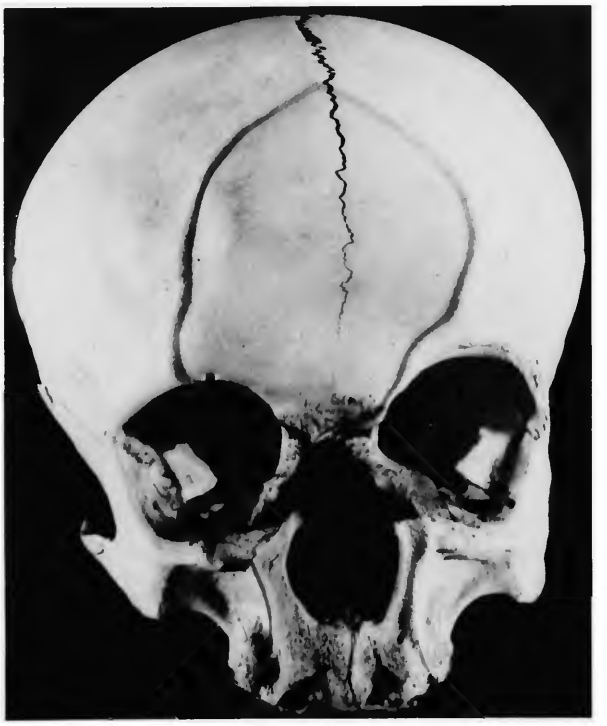


FIGURE 279.—Frontonasal syphilis with active destruction of the nasal bones and medial orbital walls, healed margin of enlarged nasal aperture and completely healed lesions of frontal bone. (Adult, WM HS 50a.2 from 1841; lines on forehead are shadows from plastic box containing skull.)

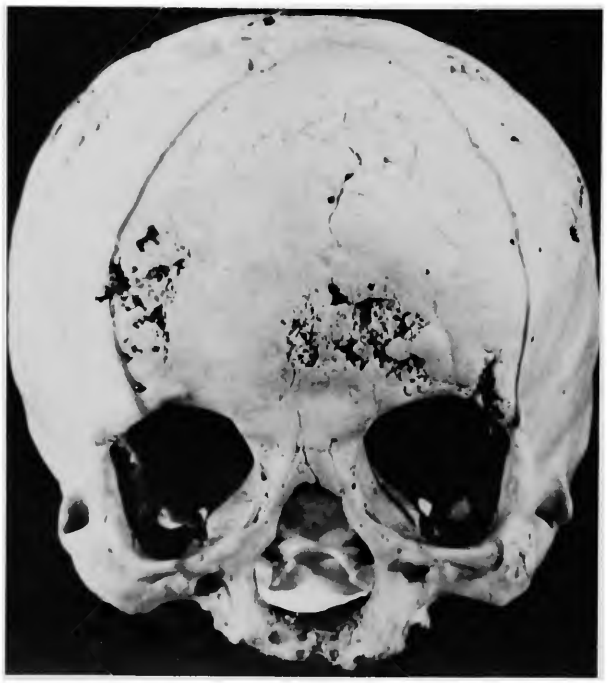


FIGURE 280.—Syphilis of calvarium and nose. Notice extensive sclerotic healing combined with active destruction, especially frontal, sequestration of the nasal septum and perforation of the palate. (30-year-old female syphilitic, WM S 50a.3 from 1869.)

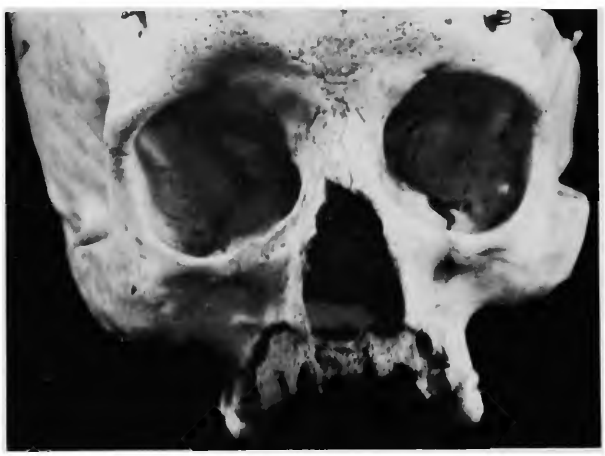


FIGURE 281.—Craniofacial syphilis, showing endonasal destruction with perforation into both orbits, perforation of palate, and sequestration of anterior portion of maxilla. (Adult male, four years after syphilitic primary lesion, PMES 1.GD.1(136) from before 1824.)

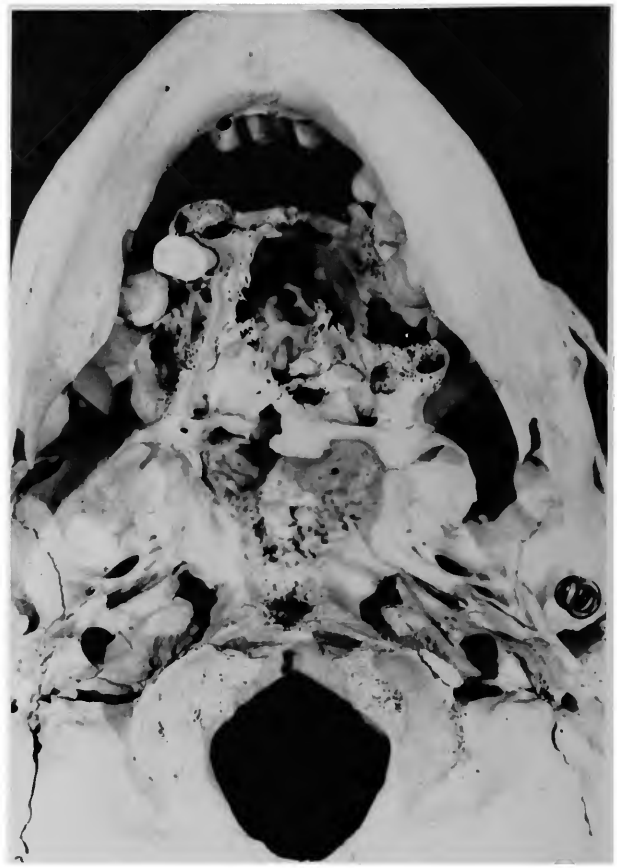


FIGURE 282.—Cranionasal syphilis, external basal view, showing active destruction of palate, sclerotic scarring of nasal roof with extension to sphenoid; frontal exterior lesions almost healed. (30-year-old male syphilitic, FPAM 1552, autopsy 8826 from 1834.)

most characteristic lesions are those with a gummatous destruction and perifocal osteosclerotic reaction involving the periosteum and the underlying bone. (For a detailed analysis of morphologic features and their relative diagnostic specificity, see Hackett, 1976.)

THE SKULL.—The most common location of tertiary syphilitic lesions is in the skull, particularly in the perinasal area and in the cranial vault. Furthermore, calvarial lesions represent the most specific diagnostic features.

Transient cranial periostitis is common in the earlier stages of syphilis, and chronic syphilitic nongummatous periostitis probably contributes to the cranial hyperostoses designated as leontias-

FIGURES 283, 284.—Syphilis of left tibia: 283, Medial external view with marked periostitis and two small round defects. 284, Cut surface, showing sclerotic obliteration of medullary cavity. Autopsy showed also gummatous periostitis of right tibia and cranial syphilis. (40-year-old male, IPMI KM229, autopsy 2922 from 1892, annual no. 2; scales in cm.)

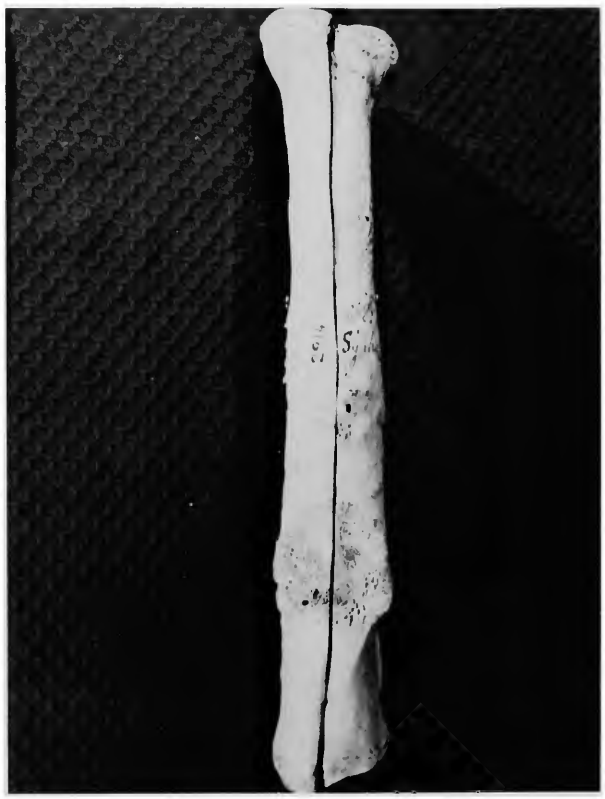


FIGURE 285.—Tertiary syphilis of left tibia complicated by skin ulcer. Notice the mid-diaphysial involvement and the erosion at the ulcer base below. The medullary cavity was blocked by reactive bone. (27-year-old female, ANM 2914 from 1872.)

sis ossea. However, these are not diagnostic etiologically. The main interest concentrates on the tertiary gummatous, osteoperiostitic lesions of the cranial vault, the majority of which begin in the frontal bone from where they may spread across sutures to the adjacent parietal and facial bones. Less commonly, the first lesion in the cranial vault begins in a parietal bone. The occipital bone is usually spared, even if the process extends to the lambdoid suture. The characteristic lesion has been classically described by Virchow (1858, 1896) under the term of "caries sicca." Hackett (1976:30–49) has revived this term, added more detail to the sequential events of this lesion, and designated it as the most diagnostic feature in dry bone. The lesion begins at or near the osteoperiosteal border, usually of the outer table, eliciting hypervascularity which, on inspection of the outer table of a dry skull, reveals itself in the form of grouped, enlarged vascular foramina. This initial change is shared with the reaction to tuberculous foci and to metastatic cancerous deposits. However, in the latter two the inner table is predilected and ultimately shows the larger defect. In osteolytic metastatic cancer, due to its frequently rapid growth rate, full thickness destruction of both tables and the intervening diploë is common, resulting in a hole with crenelated nonreactive border. In tuberculosis, in addition to the broad-based involvement of the inner table, there is usually little reactive bone formation perifocally.

The syphilitic lesion leads to a destruction of the outer table and part of the diploë by the syphilitic granulation tissue but often spares the inner table almost completely. It elicits strong sclerotic response of the surrounding bone, forming a sclerotic base and an elevated sclerotic margin around the defect in the table (Figure 262). Microscopically, focal bone necrosis is common and may well be a major stimulant to reactive sclerosis in the process of remodeling (Axhausen, 1913). However, pus formation is not significant and large sequestra usually do not form. In the chronic course of syphilis, even if untreated, individual foci will heal, but new foci will form in the vicinity. The healed individual focus of caries sicca of the skull leaves a depressed, slerotic, radially grooved stellate scar (Figure 263). This is somewhat less obvious in confluent healed areas (Figure 264). The continued process leads to confluent pitting in circinate arrangement surrounded by ivory-hard reactive bone, partly with smooth, partly with hypervascular surfaces (Figure 265). In advanced confluent caries sicca, the diploë may be markedly thickened and sclerotic while the inner table exhibits only minor reactive

FIGURES 286–289.—Tertiary syphilis involving multiple bones: 286, Distal left femur (P374) with periostitis and sclerotic placquing. 287, Right ulna (P372) with periostitis and slight pitting. 288, Left clavicle (P369), with diffuse periosteal bone and focal pitting. 289, Left scapula (P370) with enlargement and erosion of the acromion and spine. (HM 372.)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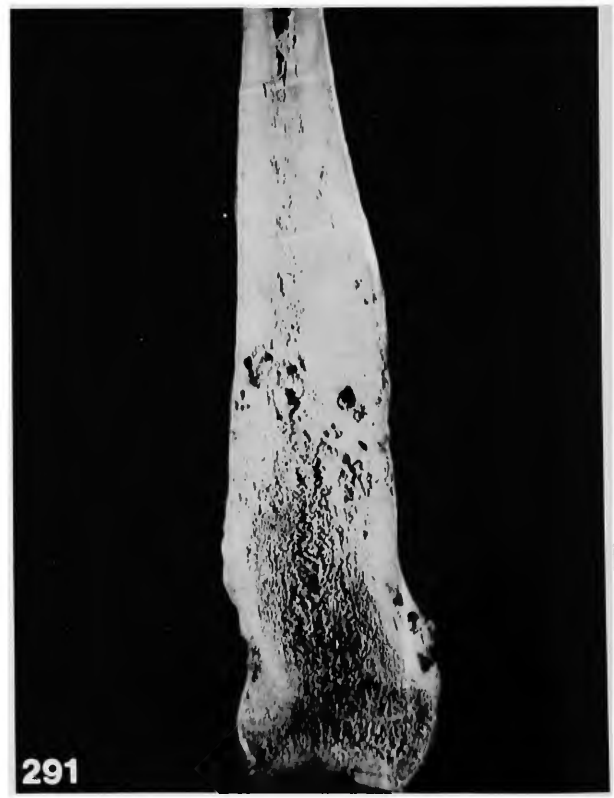
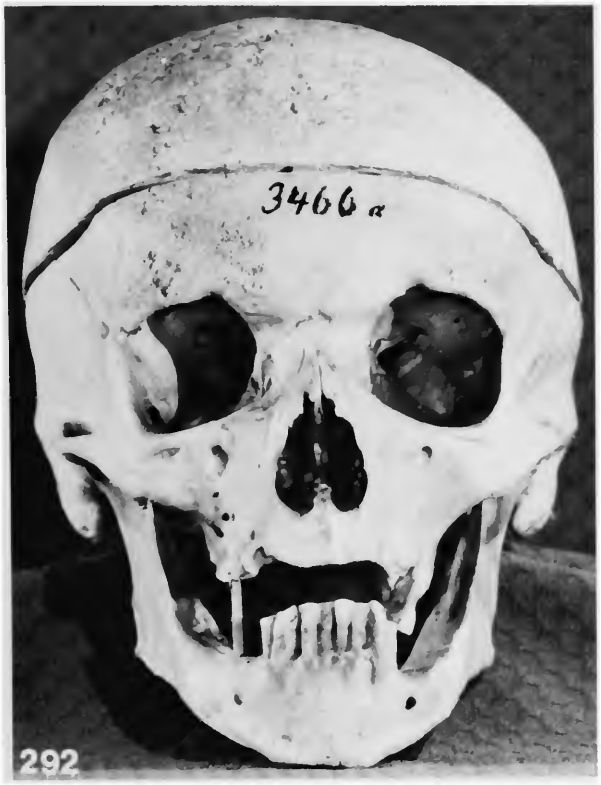
bone (Figures 266–268). Full thickness destruction and perforation of the cranial vault does occur (especially in cases with secondary pyogenic osteomyelitis), but even then the changes on the inner table are less pronounced (Figure 269). Major sequestra are seen in European syphilitic skulls, often showing darker discoloration of the necrotic bone. This is due to exposure of the affected bone in scalp ulcerations and probably largely represents the result of secondary pyogenic infection, presumably from without (Figures 270, 271). In contrast to the sequestra in pyogenic osteomyelitis, these sequestra show a "worm-eaten" appearance, indicating their involvement in the disease process before becoming necrotic (Figures 272, 273). Hackett (1976:57) did not find sequestra in skulls of Australian aborigines affected by endemic syphilis or yaws. He attributes this to the presumed absence of pyogenic infections in these populations.

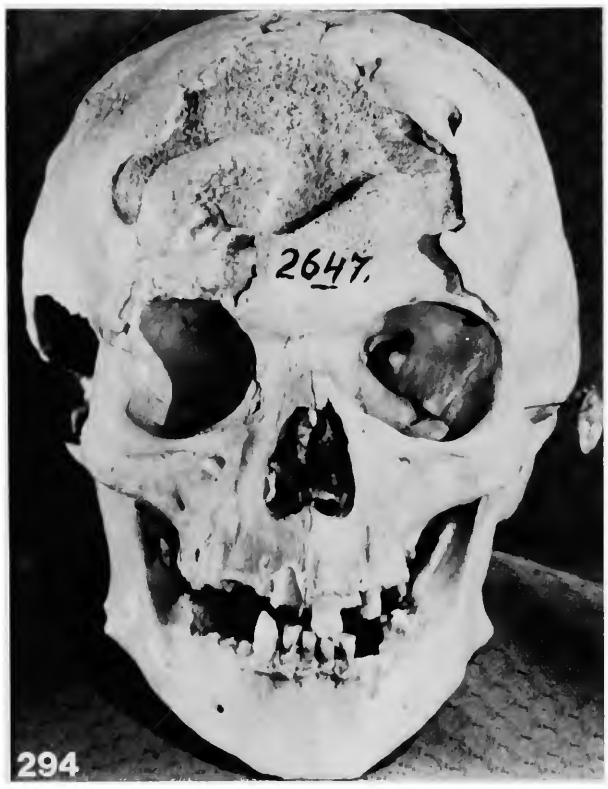
The facial bones most often affected by tertiary syphilis are the nasal bones, the bony nasal septum, the hard palate, the turbinates, and the medial walls of the maxillary antrum. These bones are involved secondary to common syphilitic lesions of the nasal mucosa. The thin bones are often destroyed, resulting in perforation of the nasal septum, the hard palate, and of the medial walls of the maxillary sinuses (Figures 274–277). The nasal cavity appears enlarged and empty in the dry skull. However, in contrast to neoplastic destruction in this area, the margins of the defect are bordered by smooth sclerotic bone. In contrast to leprosy, the frontal bone is usually involved, the perforation of the nasal septum and hard palate is common, the sclerotic response is marked and the inferior nasal spine may be spared (Figure 278). The zygoma, the ossa nasalia, and medial orbital walls may be affected by direct extension of the process in the frontal bone (Figure 279). Destruction of the bony support of the bridge of



FIGURES 290, 291.—Syphilis of left femur with fusiform hyperostosis and medullary osteosclerosis: 290, Anterior view, showing snail-track pitting, especially on the metaphysis. 291, Cut surface, showing extensive sclerotic obliteration of medullary cavity. (Adult, WM S 506.1 from 1831.)

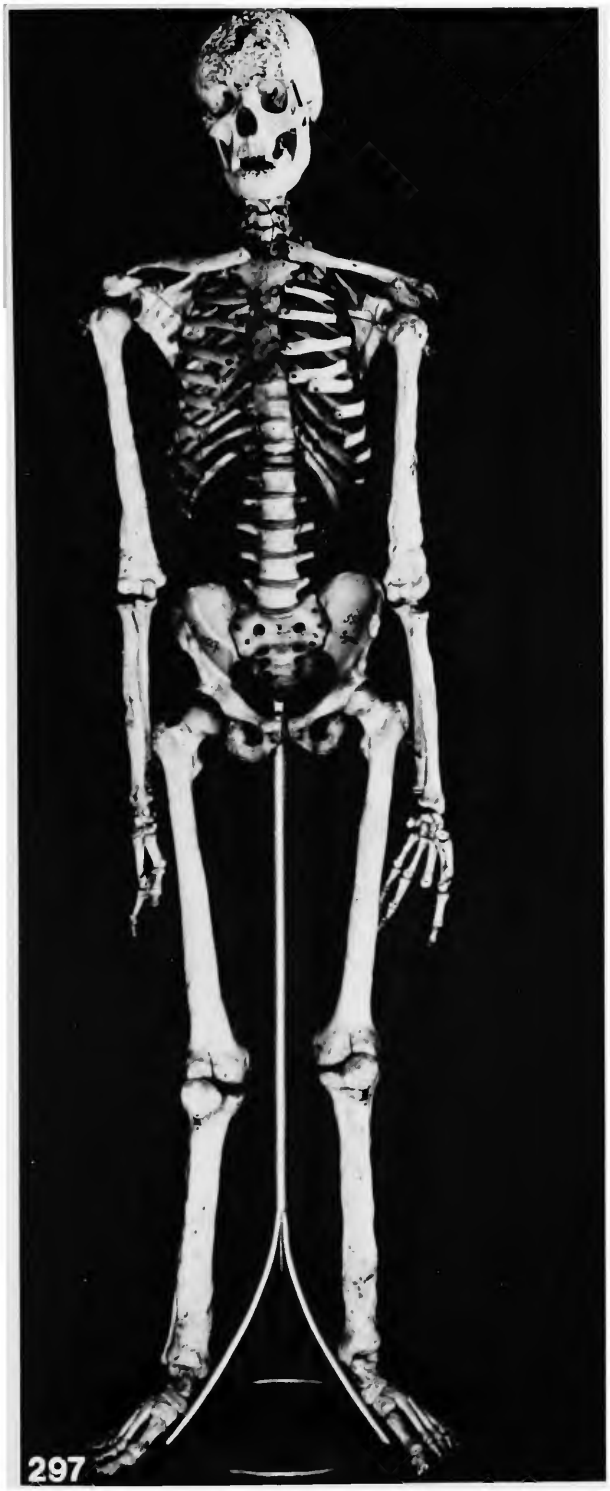


FIGURES 292, 293.—Tertiary syphilis of cranium and right femur: 292, Skull, frontal view; active superficial lesions on frontal bone with hyperostosis. 293, Right femur, frontal view; massive gummatous periostitis with typical snail-track pattern. (55-year-old male, PMUG 3466.)



FIGURES 294–296.—Syphilis of cranial vault and of long bones: 294, Frontal view, showing quiescent external defect with sclerotic healing. 295, Top view, showing more active destructive lesions in both parietal bones. 296, Left arm bones; notice absence of sequestra and cloacas (scale in cm). (45-year-old female, PMUG 2647, autopsy 6262 from 1874.)

FIGURES 297–299.—Tertiary syphilis of skull and extremities: 297, Total skeleton, frontal view. 298, Both arms, posterior view, showing especially massive symmetrical involvement of both humeri. 299, Both legs, anterior view, showing especially marked symmetrical involvement of both tibiae. (Adult male, DPUS 5527 from before 1820.)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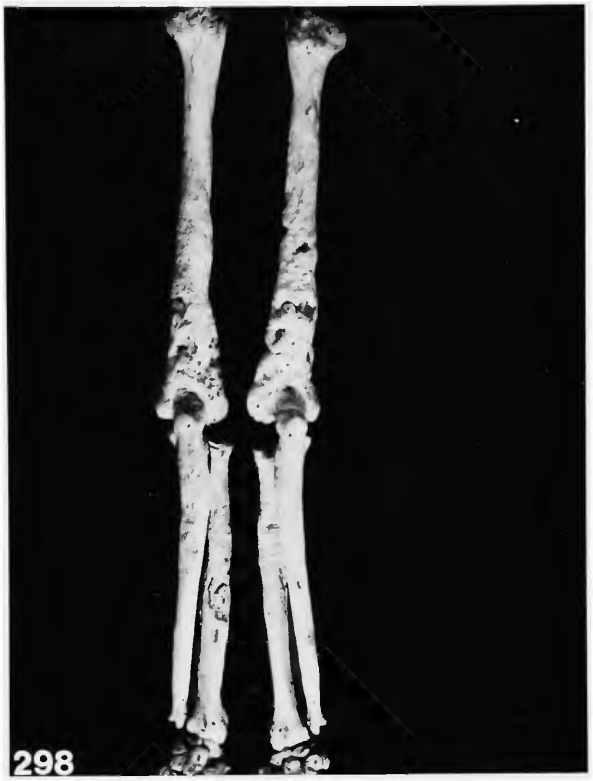
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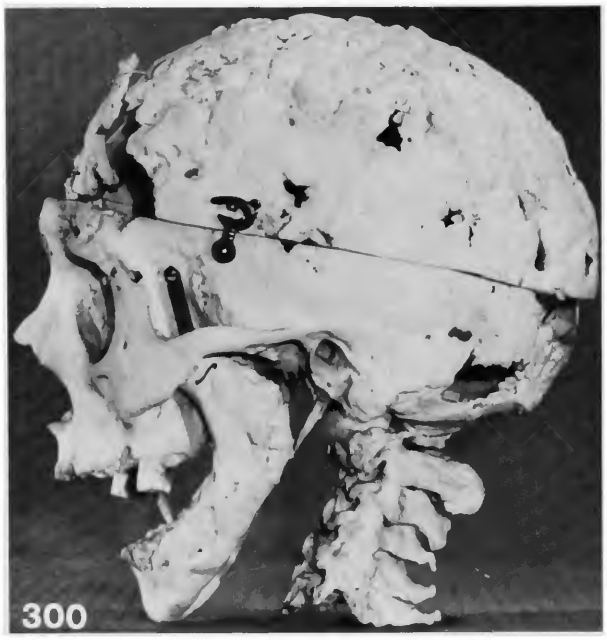
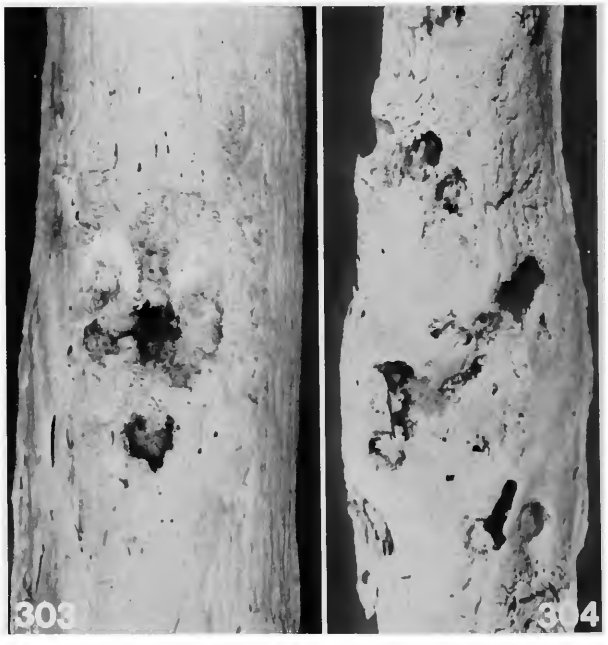




FIGURES 300–304.—Details from a total skeleton with numerous syphilitic bone lesions: 300, Lateral view of skull showing three unusual features: involvement of the mandible, lesions on the inferior portion of the occipital bone, and fusion of the diarthrodial joints including the occipital condyles through the sixth cervical. 301, Severe involvement of clavicles and sternum. 302, Extensive destruction right scapula and ribs. 303, Lateral view, right femoral diaphysis, showing penetrating defect surrounded by periosteal buildup. 304, Posterior view, right tibial diaphysis, showing fusiform swelling and multiple defects with isolated bone fragments. (51-year-old male with visceral and cerebral syphilis, DPUS 4479, autopsy 697 from 1907.)

the nose results in the syphilitic saddle nose (Figure 280). Major sequestration may be the result of secondary infection (Figure 281).

The skull base is rarely involved, but extension of the nasopharyngeal process into the sphenoid does occur, resulting in marked sclerosis of the area (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952:627)(Figure 282).

THE LONG BONES.—The tibia is approximately 10 times more often the site of syphilitic lesions than any other long bone of the extremities (Figures 283, 284). The syphilitic lesions of long bones can be separated into nongummatous and gummatous osteoperiostitis. The nongummatous lesions are suggestive but probably not diagnostic of treponemal infection (Figure 285). Hackett (1976:87–90) puts them into his "on trial" category of diagnoses. The localized form of nongummatous syphilitic periostitis may leave elevated, placque-like exostoses on the cortex of bones with a large muscle mantle (as the femur or humerus)(Figures 286–289), or circumscribed buildup of surface parallel lamellar bone of varying thickness and density on bones immediately beneath the subcutis (as the anterior surface of the tibia). Extensive periosteal thickening is often combined with cortical thickening, including the endosteal surface. Diffuse nongummatous osteoperiostitis tends to leave the bone thick and heavy. The entire surface, with exception of the cartilage-covered articular facets, may be involved. The periosteal bony buildup may be thick and becomes firmly fused with the old cortex. The outer surface is rough and markedly hypervascular. In late stages of nongummatous osteoperiostitis, the medullary canal may be completely obliterated by sclerotic trabeculae, while the old cortex may be loosened up by Haversian resorption so that, on cross-section, the bone appears uniformly coarsely cancellous with loss of the distinction of cortex and medulla. Differential diagnoses from Paget's disease can be made microscopically by the absence of the typical Paget mosaic patterns in syphilis, in which bone deposition is not interrupted by many phases of active resorption.

Gummatous osteoperiostitis is a much more characteristic lesion. In its localized form it may result in tumor-like enlargement of the affected bone area (Axhausen, 1913). In dry bone, the marked hypervascular periosteal bony buildup surrounds a scooped-out defect, extending into the cortex. This corresponds to the location of a destructive gumma in the fresh specimen. At times the scooped-out lesions are smaller and grouped, resembling the picture of cranial caries sicca, but individual gummatous defects tend to be larger than on the skull. The underlying cortex is hyperostotic and endosteal bone formation may encroach upon the medullary canal (Figures 290, 291). Diffuse gummatous osteoperiostitis resembles closely the surface appearance of cranial caries sicca (Figures 292, 293). In some cases larger defects are seen in the mantle of hypervascular periosteal bone, exposing to view deep cortex (Figures 294–296) and sometimes small sequestra. These lesions must not be confused with cloacal openings of hematogenous osteomyelitis. In contrast to that condition, major sequestra are missing, and the margins of the defects are rough and thin, and not smooth and sclerotic as on the cloacas of pyogenic osteomyelitis. Hackett (1976: 82, 93–97) accepts this form of osteoperiostitis as diagnostic of treponemal infection. Central gummas of the medullary cavity occur in the form of larger lytic lesions surrounded by a marked perifocal reactive sclerosis. This lesion may not be differentiated with assurance from a Brodie's abscess, although the sclerosis is more marked than in the latter. Pathologic fractures in bones weakened by syphilitic osteoperiostitis are not uncommon.

THE SPINE.—Syphilitic osteomyelitis of the spine is rare (Whitney and Baldwin, 1915) and the changes in dry bone would not be diagnostic in themselves. The outcome may be gibbus, after destruction of adjacent vertebrae, similar to tuberculosis. In contrast to tuberculosis, a paravertebral abscess is missing; however, this may not be obvious on dry bone. Also, in contrast to tuberculous spondylitis, the cervical spine is affected three times as often as any other segment

(Beitzke, 1934a:510). This may suggest extension from nasopharyngeal mucosal lesions rather than hematogenous origin.

THE JOINTS.—Syphilitic arthritis occasionally occurs in the large joints (knee, shoulder, elbow) during the secondary stage (Clutton's joint). This affection is transitory and leaves no changes visible on dry bone. Occasionally an epiphysial gumma may perforate into a joint and lead to gummatous arthritis (Axhausen, 1913). The perforation into the joint predominantly occurs near the margin of the articular cartilage (Freund, 1933). Subsequent bone changes, if present, are not significantly different from other forms of arthritis, particularly degenerative arthritis. Only the presence of the gummatous bone lesion may be a clue to the treponemal etiology.

Indirect suspicion of syphilitic infection may be aroused by observation of pressure erosions of sternum, ribs, or thoracic vertebrae or from findings of a Charcot joint. However, it should be realized that not all aneurysms of the thoracic aorta are syphilitic and not all neuropathic arthropathies are the result of tabes dorsalis (nerve degeneration associated with syphilis).

The multiplicity and complexity of tertiary syphilitic bone lesions is illustrated in selected views from two complete skeletons of the Pathology Museum of the University of Strasbourg, France (Figures 297–304).

Congenital Syphilis

Congenital syphilis was a common disease with high mortality before the possibility of effective treatment of the infected mother during pregnancy. Among 4500 consecutive autopsies at Johns Hopkins Hospital before 1933, there were 189 fatal cases of congenital syphilis, only two of which were older than 4 years of age (Smith, 1933). In congenital syphilis the treponemal infection is transmitted transplacentally from the actively infected mother to the fetus. The result is fetal death followed by abortion in the first half of pregnancy, fetal death with delivery of a premature or mature diseased stillborn fetus, or delivery of a living infected infant. If the infection is mild it may not manifest itself for several years (syphilis congenita tarda). In massive infections, all organs and tissues of the fetus are permeated with numerous treponemal organisms. Since the immune capacity to mount an inflammatory response is not yet developed in the first half of pregnancy, such aborted fetuses show no tissue changes. Furthermore, such fetuses are obviously subject to rapid disintegration and, therefore, do not concern us here. In premature and full-term stillborns, as well as in actively infected newborn living infants, characteristic skeletal changes are almost always present in form of syphilitic osteochondritis. This is the result of hematogenous dissemination of *Treponema pallidum* throughout the fetus in utero. It affects all areas of enchondral growth in the entire skeleton but is most marked in the fastest growing metaphyses (distal femur and proximal tibia). The lesions are symmetrical. They consist of accumulation of calcified cartilage adjacent to an area of lucency due to poor bone formation (Figure 305). This may represent merely a toxic effect on the enchondral ossification ("passive osteochondritis" of Schneider, 1923–1924:205) or be the result of formation of syphilitic granulation tissue in this area ("active osteochondritis" of Schneider, 1923–1924:205). Similar disturbances of enchondral growth are seen in a variety of conditions and are not diagnostic of syphilis (Caffey, 1939). It is appreciated that these fetal and neonatal alterations will have very little chance of recognition in archeological material. In the surviving infants, transverse metaphysial pathologic fractures through this weakened area of the metaphysis not uncommonly occur (Parrot's pseudoparalysis). Since, in the infant, before walking, the arms are more mechanically stressed than the legs, the distal humerus is the predilected site of such fractures. This osteochondritis heals in the infant even if untreated. Syphilitic periostitis often develops during infancy, mostly following the osteochondritis. Occasionally it has already begun in intrauterine life and is present at birth. It consists of multiosseous, usually symmetrical, circumferen-

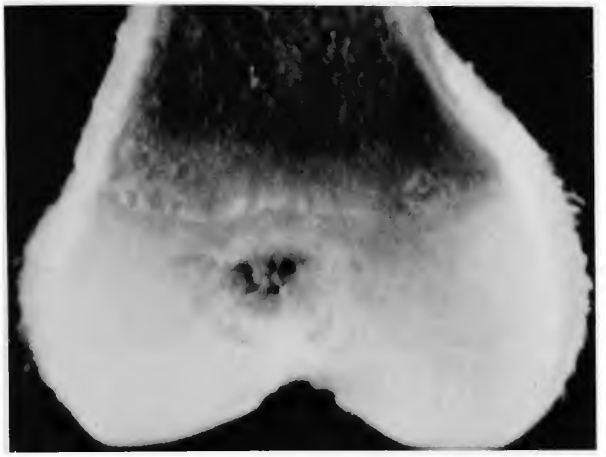


FIGURE 305.—Syphilitic osteochondritis of distal femur, wet preparation. Notice the broadened and irregular area of provisional calcification at the meta-epiphysial junction. (2-month-old male, WM S 51.2 from 1910.)

FIGURE 306.—Humerus with subperiosteal bone formation on the shaft in congenital syphilis. Most long bones showed the same changes. (3½-month-old infant, WM S 51.1 from 1880.)

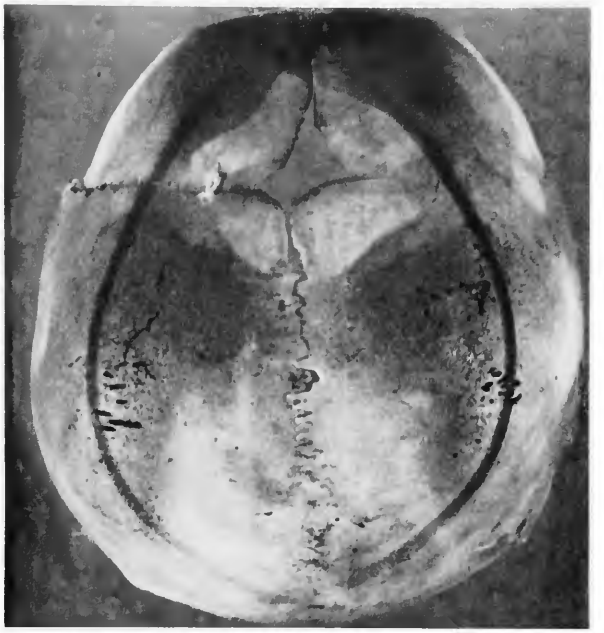


FIGURE 307.—Congenital syphilis of cranial vault with periosteal bony buildup on both frontal and parietal bones. The mother had extensive ulcerated nasofacial syphilis. (1 year, PMWH WO 734 from 1886; lines on skull vault are shadows from box containing specimen.)

tial deposition of subperiosteal bone on the shafts of long bones. The trabeculae of this bone deposit often shows a radial arrangement. This change is frequently transitory but may be recognizable on infant bones under conditions of ideal preservation (Figure 306). Similar periosteal bone deposits may occur on the cranial vault (Figure 307).

Gummatous periostitis and osteomyelitis occasionally occur, especially in older children who have passed through mild, unrecognized, and untreated manifestations in infancy (syphilis congenita tarda)(Wimberger, 1925:307–370). In this age group the bone involvement is neither so frequent nor so generalized as in the infant period but comes closer to the distribution and appearance of acquired syphilis (Figures 308–310)(Pendergrass, Gilman, and Castleton, 1930). Only 32 of 462 patients of over 13 years of age with late congenital syphilis showed active syphilitic bone changes (Smith, 1933). Such lesions in the long bones predilect the tibia, ulna, and

FIGURES 308–310.—Tertiary congenital syphilis involving multiple bones of about a 12 year old: 308, Left femur, external view, showing periosteal buildup with scooped-out defects and snail-track pattern. 309, Bisected left femur with cortical resorption and medullary reactive bone; notice absence of medullary penetration and sequestrum (AMN 2999). 310, Fibula, ulna, and radius (bisected), showing similar lesions (ANM 3163, 3166, 3185).

FIGURE 311.—Tertiary congenital syphilis of radius and ulna, bilateral (all bones bisected longitudinally); notice pitting destruction and marked hyperostosis without cloacas or sequestra. (About 6 years, ANM 3386.)

radius (Figure 311). Skull lesions occur but usually appear as multiple, rounded, destructive foci without the characteristic features of the caries sicca sequence (Figures 312, 313). Syphilitic saddle-nose in congenital syphilis may be mainly the result of disturbed enchondral ossification of the base of the skull. In adolescents, tertiary syphilitic bone lesions may more closely simulate those observed in adults, and differentiation of a late stage of congenital from venereal syphilis may be impossible without clinical data (Figures 314, 315). Occasionally extensive involvement of facial bones can occur in congenital syphilis (Figures 316, 317).

Nongummatous periostitis results in osteosclerosis with ultimate fusion of the subperiosteal bone deposits and the underlying cortex. The most characteristic lesion of this type is the so-called saber tibia of Fournier (1886:265–269). This apparently represents the combination of stimulated growth and layered bone deposition on the anterior surface. The tibia is longer than normal and shows a true forward curve, while the fibula remains of normal length and configuration. As previously mentioned, similar bony buildup on the anterior tibial surface occurs in nongummatous periostitis of acquired syphilis (Figures 318, 319). In that case, however, the tibia is not truly elongated and curved, and the posterior contour remains straight.

Syphilitic dactylitis used to be more frequently observed in congenital than in acquired syphilis. It concerned the fingers more often than the toes and predilected the basal phalanges. The condition often affected more than one finger, not infrequently bilateral but not symmetrical. The appearance, especially in small children, may closely simulate that of tuberculous spina ventosa with widening of the diameter and formation of a thin, bony shell. In adults there is less expansion and more reactive osteosclerosis.

Hutchinson's and Moon's teeth, although present in a variable number of individuals with congenital syphilis, also have been questioned in their specificity (Kranz, 1927:263).

Although there are a variety of skeletal changes occurring in different phases of congenital syphilis, the findings are more indicative than diagnostic and must be critically evaluated in the entire context of the skeletal findings. Individual lesions may not be distinguishable from tuberculous or other infectious changes.

PALEOPATHOLOGY

In the history of disease, probably no single pathological condition has generated more scholarly debate than treponemal infections and, in particular, syphilis. The debate has focused on the origin of syphilis, with some scholars arguing that the disease was introduced into the Old World by Columbus and his crew after their discovery of the New World. Another possibility is that syphilis existed in the Old World before

FIGURES 312, 313.—Congenital syphilis with destructive lesion of right frontal bone involving both tables: 312, Outside view. 313, Endocranial view. (8-month-old female, ANM 2233 from 1865.)

Columbus and may have been introduced into the New World by Columbus. A third point of view is that treponemal diseases existed in both the Old and New World before Columbus.

These different and conflicting opinions are based on three distinct, but not necessarily mutually exclusive, sources of information: (1) historical documents, (2) pathological remains, and (3) speculation based on evolutionary theory. The relative dependence of a scholar on one of these sources appears to have considerable influence on his subsequent conclusions. The problem, of course, is that so far there is no source that can provide overwhelmingly convincing evidence for any of the three hypotheses.

While a comprehensive review of the basic literature on the subject of the history of treponemal diseases is beyond the scope of this work, a brief summary of the major concepts is essential to our subsequent presentation of paleopathological specimens. It also serves to highlight the pitfalls of uncritical reliance on published sources.

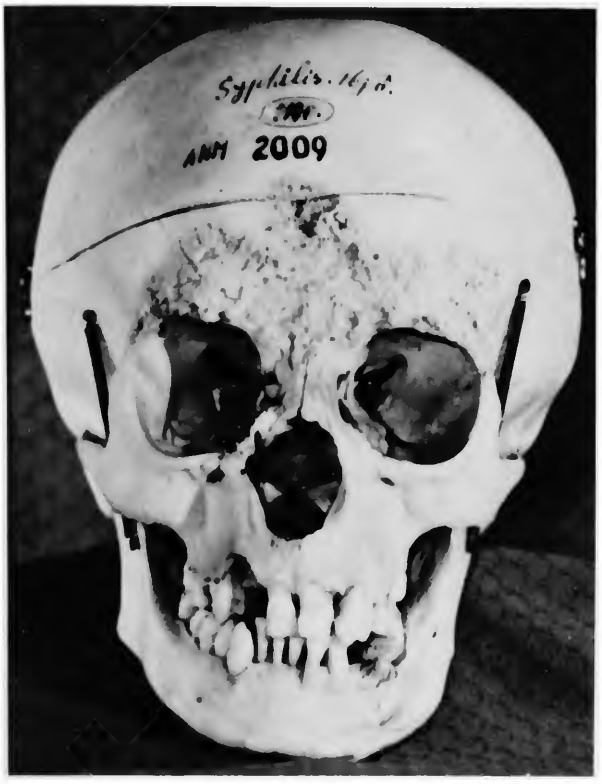


FIGURE 314.—Tertiary cranial syphilis (congenital?). Notice active and healing lesions in frontonasal area, widened nasal aperture, loss of nasal septum and inferior nasal spine, and perforation of palate. (16-year-old male, ANM 2009 from 1870.)

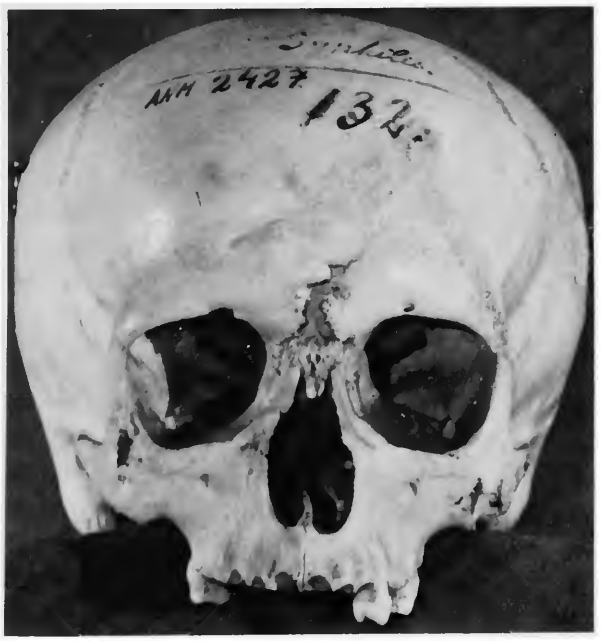
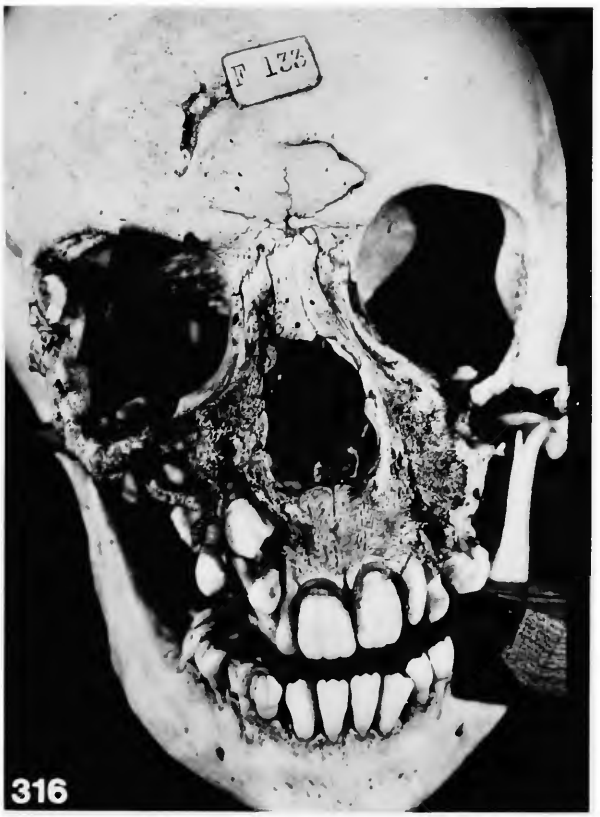
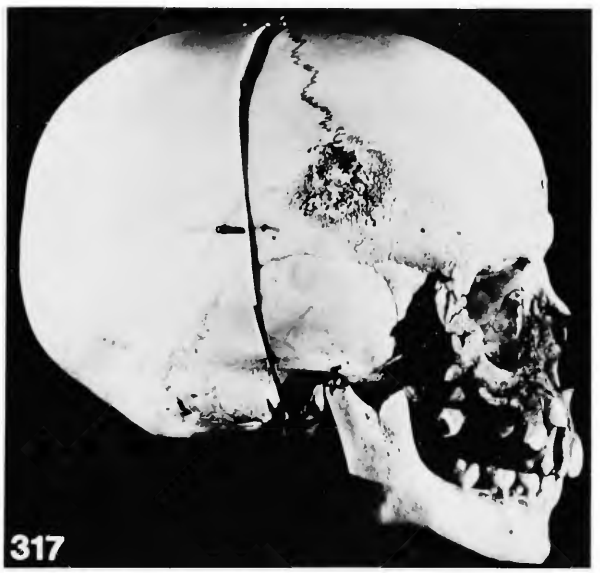
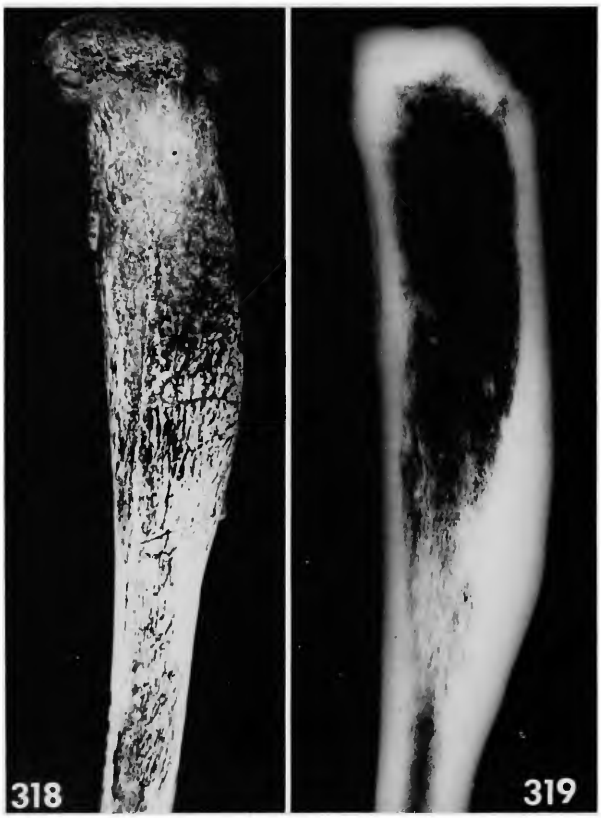


FIGURE 315.—Frontofacial syphilis with destruction of nasal bones and involvement of zygomas, the frontal lesion partly healed. The young age raises the question of tertiary congenital syphilis. (17-year-old female, ANM 2427.)





FIGURES 316, 317.—Congenital syphilis of maxilla, right orbit, and right frontoparietal area: 316, Frontal view, showing sclerotic healing of frontal and nasal bones; teeth normal. 317, Lateral view, showing active frontoparietal lesion. (8 year old, with history of syphilis, OM F 133.)



FIGURES 318, 319.—Syphilitic saber tibia: 318, Right tibia, lateral view, showing massive anterior periosteal bone deposition. 319, X-ray of lateral view, showing anterior cortical thickening and preservation of the medullary cavity. (Adult Caucasian male; Pathology Department of the University of Otago, New Zealand, courtesy of Dr. B. Ragsdale, Armed Forces Institute of Pathology, Washington, D.C., USA.)

Bruhl (1880) cites many primary historical sources, including early chroniclers of colonial life in the Americas, such as Oviedo and Las Casas, who supported the view that syphilis existed in the New World at the time of its discovery by Europeans. He also cites Diaz de Isla, the physician who treated Columbus' men for presumed syphilis. Bloch (1901, 1911) provides a review of older, relevant source material on the subject. In another report (1908:4), he concludes that "all available statements and facts point to the last decade of the fifteenth century—particularly the years 1493–1500—as the time when syphilis first appeared in the Old World. There is not a particle of evidence to show that the disease existed in Europe before that time." Dennie (1962:61–63) notes that syphilis was particularly virulent after its initial introduction from the New World and subsequently became milder. Dennie argues that this reflects the normal progress of a newly introduced disease and thus supports the theory of New World origin for syphilis.

In contrast with the above opinions, Holcomb (1941) cites early descriptive medical reports of a disease process attributed by the early writers to leprosy. However, Holcomb (1941:151–152) indicates that many of these purported cases of leprosy were clearly syphilis, both in terms of appearance and the venereal mode of transmission. He concludes (1941:167) that "syphilis in a congenital and acquired form certainly prevailed in Europe long before the discovery of America." This conclusion is supported by a description reported by Thorndike (1942:474) on a disease process resembling syphilis dated 1412.

Elsewhere in the Old World, Hyde (1891:117) states that the ancient medical literature of China, India, Greece, and Italy contains unmistakable proof that early in the world's history genital lesions were known to occur from sexual contact. However, Crosby (1969:219) offers an opposing opinion that there is no unequivocal description of syphilis in the ancient Old World medical literature. In particular, Crosby cites Wong and Wu (1936:218), who state that no Chinese writer has ever described a case that could be attributed to syphilis.

There is one, final, historical argument pertaining to the origin of syphilis that bears noting. Bloch (1908:8) suggests that the presence of syphilis would have led to its widespread prevalence had it existed in the ancient Old World. In view of the "unbridled licentiousness of Imperial Rome and the excesses of the Middle Ages," he argues, there should be considerable evidence for syphilis in ancient skeletons. Bloch found no such evidence in European skeletal material of pre-Columbian age. There is, unfortunately, no indication regarding the sample size of pre-Columbian

European skeletons on which this conclusion was based. Undoubtedly, Bloch's reference to unnumbered thousands of skeletons of pre-Columbian age refers primarily to the work of Elliot-Smith and Wood-Jones on Egyptian material, where other epidemiological considerations need to be considered before one eliminates the possibility of syphilis in that population.

In discussing evidence for pre-Columbian syphilis in the New World we are primarily limited to human remains and most of the evidence is from the skeleton. With skeletal material there are two fundamental requirements: (1) unambiguous identification of treponemal disease and (2) unambiguous determination of pre-Columbian archeological age. Several purported cases of pre-Columbian syphilis have foundered on one or both of these factors.

The earliest reference to syphilitic lesions in New World skeletons is Jones' (1876:66) report on archeological skeletons from Tennessee. Indeed, this early report appears to have been the basis for subsequent reports in which the authors concluded that syphilis was in the New World before Columbus (e.g., Lamb 1898). Morgan (1894) challenges Jones' conclusions calling into question both the diagnosis of syphilis and the pre-Columbian date.

Many other purported cases of skeletal syphilis have been reported in New World specimens. One of the most respected scholars studying this material was H. U. Williams, a pathologist, who concluded on the basis of a critical review of published material and his own observations that the case for pre-Columbian syphilis was probable beyond reasonable doubt (Williams, 1932:978). He found the evidence for Old World skeletal syphilis before 1490 to be much less convincing. After a careful study of Eskimo and Aleut skulls, Holcomb (1940:189) concluded that syphilis probably did not exist in these northern regions of the New World until after contact with Russian sailors and traders.

The third source of speculation on the history of treponemal diseases is based on theoretical reconstructions with evolutionary theory as the major integrating concept. In essence this concept of treponemal diseases suggests that they have evolved with man and migrated with him throughout the world and thus were endemic in both the Old and New World long before Columbus (Hudson, 1968; Cockburn, 1961; Hackett, 1963). Although not phrased in evolutionary terms, the concept of treponemal diseases being endemic in both Europe and the Americas goes back to an early report by Krumbhaar. He states (1936:232): "Personally, I regard as most convincing the theory of the existence of syphilis in both continents as far back as prehistoric times." Similarly, Stewart and Spoehr (1952) speculate that treponemal diseases existed in the Old and New World but had developed different strains while isolated from each other. When contact between the European explorers and indigenous Americans occurred, they traded strains of treponemal organisms to which neither had developed any immunity. This they suggest is the reason for the syphilis epidemic in Europe after Columbus and the increase in bony lesions possibly attributable to syphilis in post-Columbian Indian skeletons in the New World. Stewart and Spoehr's (1952) observations on the increase in syphilitic type lesions in post-Columbian indigenous American skeletons is significant, because of Stewart's extensive experience with New World skeletal material.

Hudson, in several papers, has attempted to reconstruct the evolutionary history of the treponemal organism. In one of his earlier papers (1958:23) he suggests that the treponeme evolved from a saprophyte (a microorganism that is closely related to the treponemal organism) early in man's evolutionary history through the introduction of such an organism into a break in the skin. He further speculates that this event may have taken place in Central Africa in an environment similar to the rain forest of today (Hudson, 1965:890–891). Such an early disease would probably have been similar to yaws of today, in which the organism survives on the moist skin.

Crucial to an understanding of Hudson's evolutionary history of treponemal diseases is the

concept that treponemal organisms represent a "biological gradient" rather than separate species. In this scheme the various treponemal diseases reflect adaptations by the same microorganism to varying epidemiological conditions. Thus, when man moved to more temperate regions the organism migrated to the more moist regions of the body (the mouth, axillae, and the crotch), creating a new disease syndrome called endemic syphilis by Hudson (1965:891).

Hudson (1965:895) argues that the development of venereal syphilis was the result of development of the cities with the improved hygiene associated with city life. Also the increased use of clothing prevented the frequent skin contact in children necessary for transmission of endemic syphilis. After man's migration to the New World, the development of the South American treponemal disease, pinta, was, in Hudson's reconstruction, the result of a local adaptation, in which the treponeme was again introduced to environmental conditions similar to those found in Central Africa (Hudson, 1965:892).

Hudson recognizes the inherent question in his theory which is, why did venereal syphilis not develop sooner in the evolutionary process if all treponemal diseases are the result of adaptations in a single species? In response to this question he notes that immunity to the treponeme would be acquired during childhood as the result of exposure to nonvenereal syphilis so that venereal transmission after sexual maturity was not possible until children no longer contracted the disease (Hudson, 1965:892).

Hackett (1963) takes a somewhat different approach in reviewing the origin and history of treponemal diseases. While recognizing that the organisms associated with the four contemporary treponemal diseases are indistinguishable microscopically, he continues to distinguish the diseases as clinical entities (Hackett, 1963:9). In Hacketts's evolutionary scheme, treponemal diseases began in the Afro-Asian landmass about 15,000 B.C. The syndrome was pinta, which spread throughout the world, subsequently becoming isolated in America. Yaws, also, developed in the Afro-Asian landmass as a mutant of the pinta treponeme and spread throughout the Old World. Endemic syphilis evolved from yaws during the drying trend following the last Pleistocene glacial period. As in Hudson's reconstruction, Hackett associates the development of venereal syphilis with the emergence of cities. However, he suggests that the disease was mild until a virulent mutation toward the end of the fifteenth century A.D. gave rise to the European epidemic attributed to the return of Columbus from the New World. Subsequent to this event venereal syphilis was spread throughout the world during the European colonial expansion in the sixteenth and seventeenth centuries (Hackett, 1963:38). Like Hudson, Hackett does find a likely association between the evolution of treponemal diseases and environment, particularly climate. However, these two scholars differ in the evolutionary mechanism. Hudson supports the concept of a pluripotential organism; Hackett evokes multiple mutations and different organisms.

While these arguments may seem rather arcane, scholarly debates that are probably unresolvable, it is important to stress that these speculations do have a bearing on the interpretation of skeletal lesions or the lack of skeletal lesions. The historical and contemporary expression of treponemal diseases suggests some variation in the organ systems affected (Hackett, 1963:8–9) and the virulence of the disease. These basic principles apply to the effect of treponemal disease on the skeleton. Thus, the absence of skeletal evidence of treponemal disease is necessary but not sufficient support for the absence of the disease in an archeological population. Indeed, the arguments presented, particularly by Hudson and Hackett, should make one cautious about assumptions of the absence of treponemal diseases on any continent at any point in man's history of the last 10,000 years.

Crosby's observation (1969:219) that the typical evolutionary development of some infectious diseases is characterized by decreasing virulence is a well-known epidemiological principle and is based on the fact that those organisms that kill

the host generally die as a result, while organisms that are less virulent or infect individuals with a more effective immune response tend to survive. Where a disease has been endemic for centuries, as Hudson and Hackett suggest was the case for treponemal diseases, milder forms of the disease and improved host-response could have developed in which the skeleton was not affected, as is the case with pinta today.

Congenital Syphilis

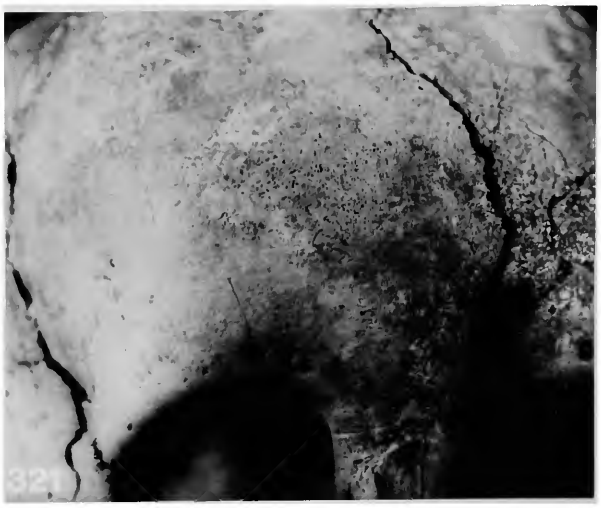
Because of the problems in differentiating various expressions of treponemal disease in paleopathological specimens, most of the following case descriptions will not be attributed to a specific syndrome. An exception to this is an archeological example of probable treponemal disease in a 6- to 7-year-old American Indian child from Virginia (NMNH 379177). The sex of the child is unknown. There were no European trade goods found at the site. Both the pottery types and the presence of stone pipes suggest a date before 1400. While a later date cannot be ruled out, the evidence points to the pre-Columbian period.

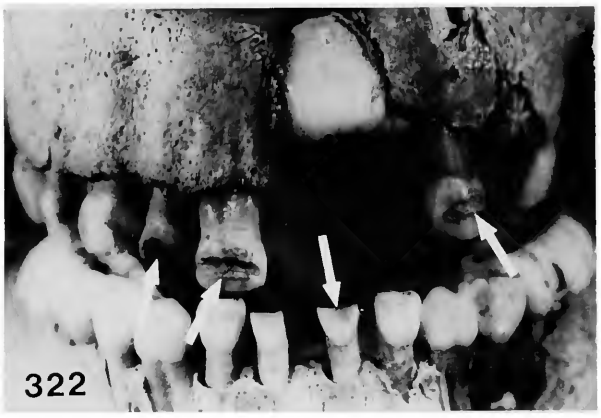
Lesions on the skull appear to be confined to the face and forehead (Figure 320). There is some postmortem damage to the skull, which complicates the picture somewhat. The lesion on the forehead is located near the midline and measures about 35 millimeters by 45 millimeters. It may extend to the left of the frontal bone, but postmortem damage precludes confirmation. The lesion itself consists of periosteal bone, which shows only slight evidence of porosity (Figure 321). The most markedly abnormal bone tissue occurs around the nose. The nasal bones and the maxillary bone adjacent to the nasal bones and nasal aperture have thickened, porous, periosteal bone on their external surfaces (Figure 323). The remaining portions of the maxilla appear to be normal. There is a slight degree of porosity of the orbital roof (cribra orbitalia). There are plaques of porous periosteal bone on the inferior and anterior portions of the mandibular body. This is accompanied by a slight expansion in the thickness of the body on the left side. The maxillary deciduous incisors are missing on the left. However, both right maxillary incisors have marked hypoplastic enamel defects (Figure 322). Indeed, the defect was so severe that the lower portion of the right lateral incisor appears to have broken off antemortem. There is a less severe hypoplastic defect on the left deciduous maxillary canine. The enamel of the right canine has been damaged postmortem, although there is a slight vestige of a defect on this tooth as well. The deciduous and first permanent molars are normal, as is the emerging permanent left central incisor.

The dentition of the mandible is less severely affected. The central incisors are hypoplastic with antemortem loss of the superior portions of the crown. The lateral incisors have a very slight enamel defect. The remaining teeth appear to be normal, although the severe caries of both deciduous second molars suggests weakened enamel. Caries of the lower left first deciduous molar has resulted in an abscess on the left side.

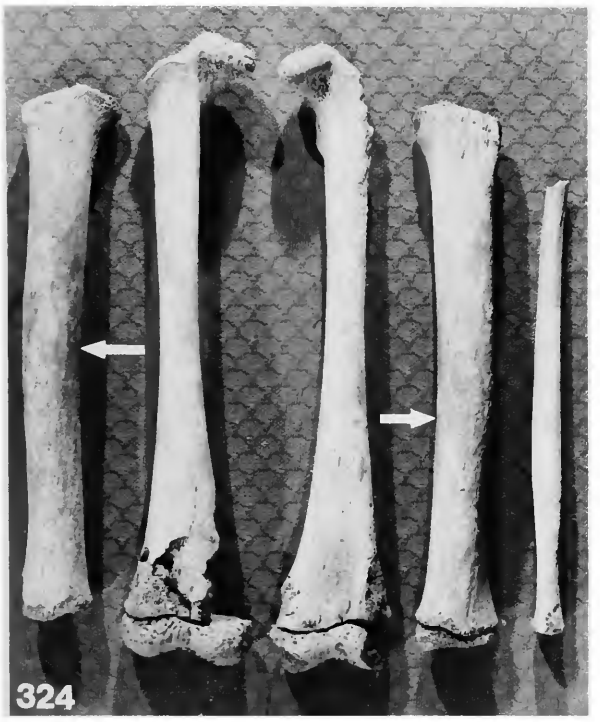
All of the major bones of the postcranial skeleton exhibit porous periosteal lesions. The severity and thickness of these lesions varies and is significant in reaching a preferred diagnosis. Both scapulae are normal, as is the manubrium. Both clavicles have periosteal bone deposits on the anterior medial aspect of the diaphysis; the metaphyses are normal. Both humeri have patchy, periosteal bone deposits, which are limited to the diaphysis. Involvement is slight and there is no appreciable expansion of the cortex. The radii are somewhat damaged but clearly indicate a general deposition of periosteal bone on the diaphysis, which is more severe distally. There is a slight expansion of the diaphysis, and the metaphysis is normal. A similar distribution of periosteal bone occurs on the ulnae, but the diaphysial expansion is more marked. This expansion occurs through the apposition of periosteal bone, but this is also associated with an expansion of the marrow space, although there appears to be a net increase in the thickness of the cortex.

Many of the smaller bones of the hands were not recovered during excavation, although all the





FIGURES 320–327.—Probable congenital syphilis in a 6 to 7-year-old child from an archeological site in Virginia, USA (artifacts associated with the burial suggest a pre-Columbian date): 320, Facial view showing abnormal periosteal, reactive bone (arrows) on the frontal, nasal, and maxillary bones. 321, Detailed view of the frontal lesion. 322, Detailed view of the hypoplastic development of some of the deciduous dentition. Note the hypoplastic enamel lines and the loss of a portion of the crowns of some of the defective incisors (arrows). 323, Detailed view of the periosteal reactive bone on the nasal and maxillary bones. 324, The anterior view of the femora, tibiae, and left fibula. Note that the diaphyses of the tibiae (arrows) are much thicker than the diaphyses of the femora. 325, A detailed view of the expansive periosteal bone on the left tibia. 326, The laterosuperior view of the right fifth metatarsal; note the thickened porous nature of the periosteal reactive bone. 327, The mediosuperior view of the right fifth metatarsal; note the diminished amount of reactive bone on the medial aspect (arrow). (NMNH 379177.)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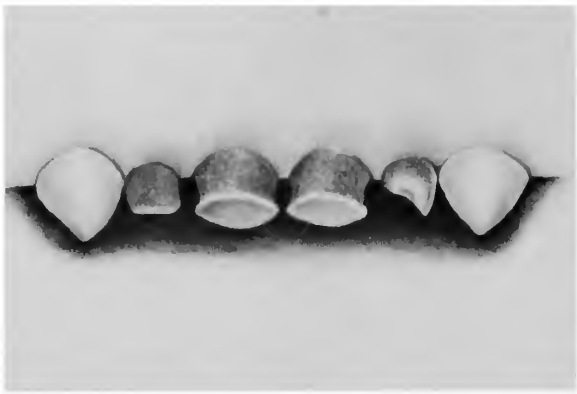


FIGURE 328.—Upper incisors and canines of a 3-year-old child with congenital syphilis. Note that the crowns are partly to completely missing from the incisors. (Redrawn from Hutchinson, 1909, plate 31, figure 3, facing page 460).

metacarpals are present. In general they exhibit a low-grade, diaphysial periostitis, which is somewhat expansive. All bones appear to be affected to approximately the same degree except the fourth metacarpal, which is only slightly affected on the right and normal on the left.

The vertebrae and pelvis are normal, as are the existing fragments of ribs. There is slight periostitis on the femora, which is limited to the anterior distal diaphysis and metaphysis with involvement extending almost to the growth plate. Both tibiae exhibit a marked, expansive, porous periostitis with the major focus being the anterior proximal portion of the diaphysis (Figures 324, 325). However, the entire shaft is abnormally thickened. The fibulae are less severely affected with a similar lesion. The major focus is the mid- to distal portion of the shaft with the right fibula somewhat more affected than the left.

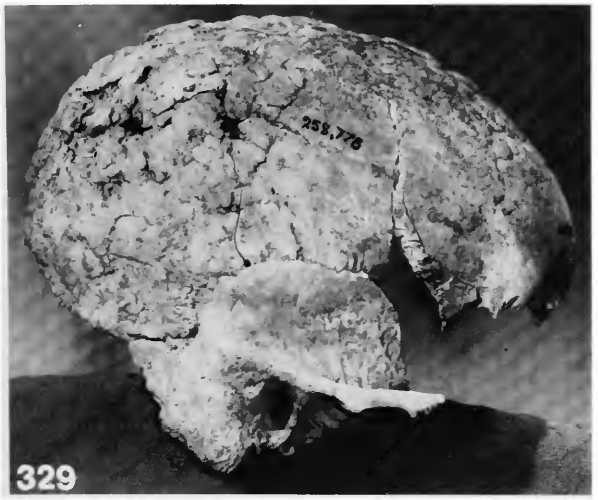
Many of the smaller bones of the feet are missing. Both tali are normal, although the calcanei exhibit periostitis. The involvement is more severe on the right calcaneus. The metatarsals show a bilateral pattern of involvement, in which the first and fifth metatarsals are most severely affected. The fifth metatarsal, in particular, exhibits a much more severe condition than that seen on the other metatarsals (Figures 326, 327), and both the first and fifth metatarsals are more severely affected than any of the metacarpals.

The overall pattern in this case is that the bones, which have minimal overlying or adjacent tissue, are most severely affected. These bones include the frontal, nasal and adjacent maxillary bones, the ulnae, tibiae, and first and fifth metatarsals. The position of the marked hypoplastic defect on the incisors could only have occurred in utero, since that portion of the tooth was developing at about the seventh fetal month. This indicates a congenital disease and, in combination with the pattern of relatively severe lesions in the skeleton, suggests congenital syphilis. Hutchinson (1909, pl. 31: fig. 3) illustrates the upper incisors and canines of a 3-year-old child with congenital syphilis. The canines were normal but the incisors have hypoplastic defects, which resulted in part, or all, of the crown breaking off (Figure 328).

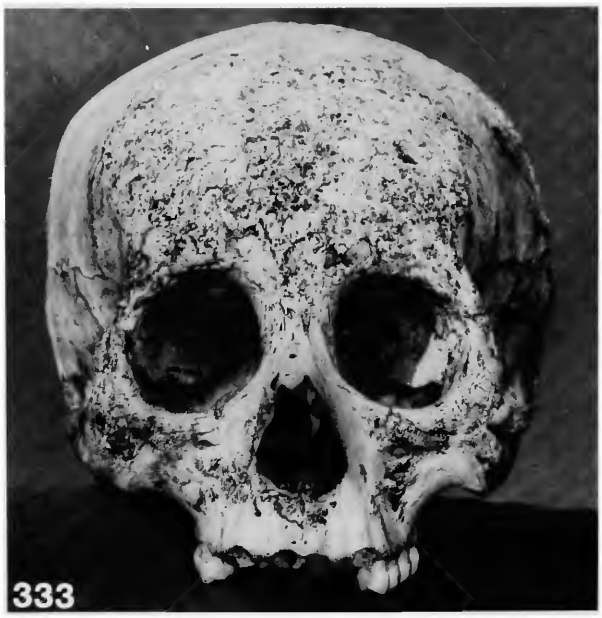
Adult Treponemal Disease

The second case of probable treponemal disease is the skeleton of an adult female Indian from Arkansas, USA (NMNH 258778). This skeleton was excavated by Clarence Moore during field work conducted in 1909 and 1910. Moore (1910: 258) believed that the burial was pre-Columbian. However, the date remains uncertain since documentation is poor. The bones that were recovered include a fragmentary skull, the right clavicle, both humeri, the left radius and proximal ulna, the distal right radius, the proximal right tibia, the shaft of the right fibula, and the right talus and calcaneus. Of these bones, the skull, left ulna, both femora, and the left tibia have obvious lesions. The right calcaneus has a slight periostitis of the superior and lateral cortex. The other bones are normal.

The lesions of the skull (Figures 329, 330) are most pronounced on the vault and are much less developed on bone underlying the temporal muscle mass. However, both temporal bones show a thickened, irregular, and slightly porous surface. The major focus for the lesions is the external table, although some of the lesions penetrate to the inner table and others seem to originate there. However, inner table involvement is much less



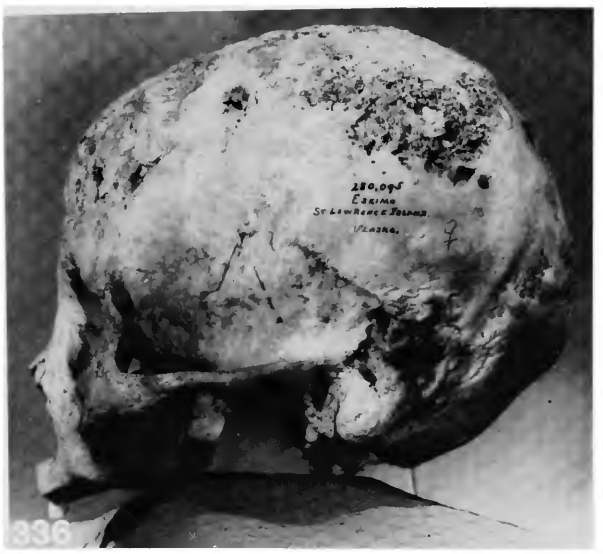
FIGURES 329–332.—Probable acquired syphilis in an adult female Indian from Arkansas, USA: 329, Right lateral view of skull; note that the reactive bone is minimal below the area of the temporalis muscle. 330, Detailed view of the irregular, lumpy outer table of the right parietal. This is a typical bony reaction to gummatous lesions. 331, Anterior view of the femora and tibiae. 332, Detailed view of medial side of right femur near midshaft; note the bony bridging that took place over blood vessels (white arrows) and the undercut periosteal bone (black arrow). Such lesions are typical of bony reactions in treponemal disease. (NMNH 258778.)

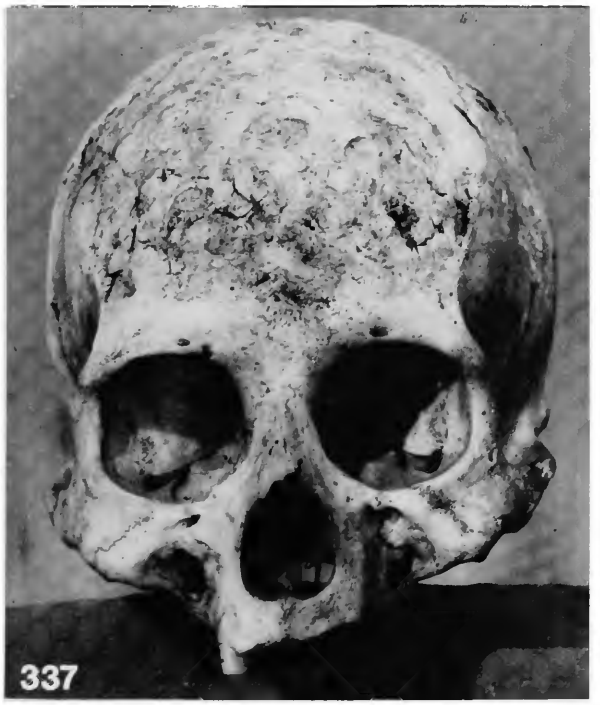


FIGURES 333–335.—Probable acquired syphilis in a young adult female skull from Alaska, USA: 333, Facial view, exhibits an active, bony reaction typical of a gummatous condition. 334, Left lateral view. 335, Detailed view of the reactive bone on the left parietal showing mixed active and healing condition. (NMNH 372956.)

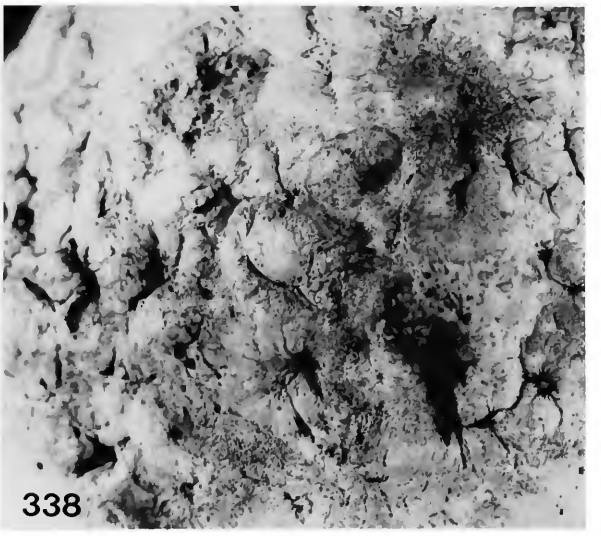
extensive. Broken sections of the frontal and the parietals reveal a thickening of the skull with almost complete fill-in of the diploë by compact bone. The lesions of the external table are typical gummatous lesions characterized by a mixture of bone formation and destruction, creating an irregular lumpy appearance. In this case, the smoothed surfaces of the lesions indicate a long-term chronic condition. The left orbital roof was not recovered; however, the right orbital roof exhibits a slight labyrinthian periostitis, which may or may not be associated with the disease process affecting the rest of the skeleton.

The proximal left ulna is the only long bone of the upper extremity that exhibits a morbid condition. The lesion consists of an expansion of the proximal metaphysial cortex with slight porosity. Because of the rotation of the biceps tendon and the biceps tubercle of the radius, the superior lateral portion of the metaphysis could not par-



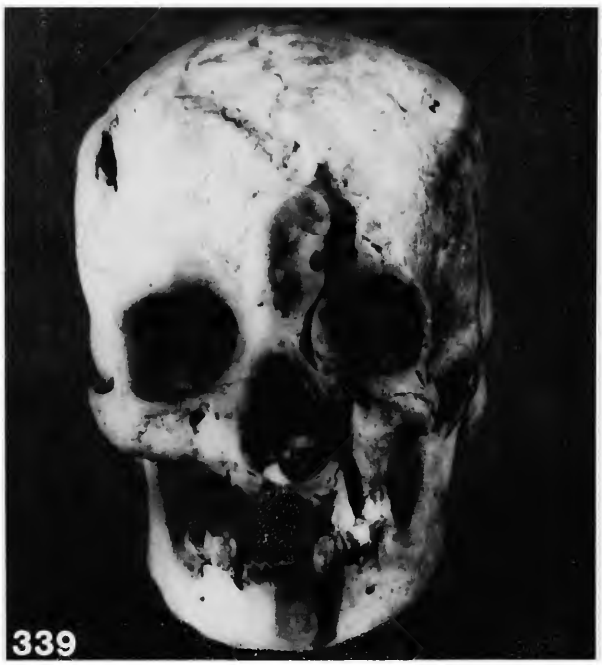


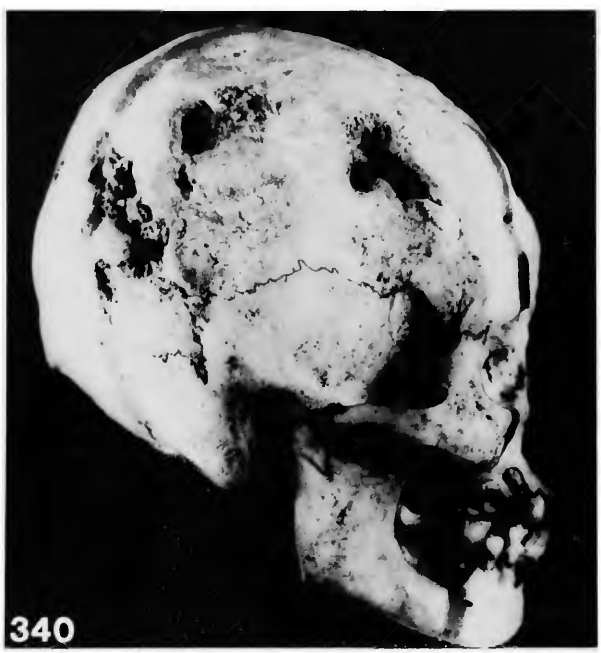
FIGURES 336–338.—Probable acquired syphilis in an adult female skull from Alaska, USA: 336, Left, lateral view; note that the more active lesions are on the posterior part of the skull. 337, Facial view showing mostly healed lesions on the frontal bone. 338, Detailed view of the lesions on the left frontal bone. Healing was taking place in the central portions of the lesions with porous reactive bone on the periphery of many lesions. (NMNH 280095.)



ticipate in this expansion, creating a depression in this region of the ulna. The insertion of the brachialis muscle is markedly rough with spicules occurring on the periphery of the insertion area. The joint surface itself shows a slight degree of breakdown on the coronoid portion, which may reflect an expansion of the disease process into the joint. However, the joints of the affected bones of the lower extremity are normal.

The abnormal bone of the left femur is restricted to the proximal shaft and metaphysis (Figure 331). The lesion does not extend to the trochanters or the femoral neck. The disease process has resulted in concentric thickening of the





FIGURES 339, 340.—Probable treponemal disease in a skull from the Cook Islands: 339, Facial view; note the extensive but largely healed lesions of the frontal bone and resorption of the alveolar bone. 340, Right lateral view, showing active lesions on the parietal and occipital bones. (WM S 50 A5.)

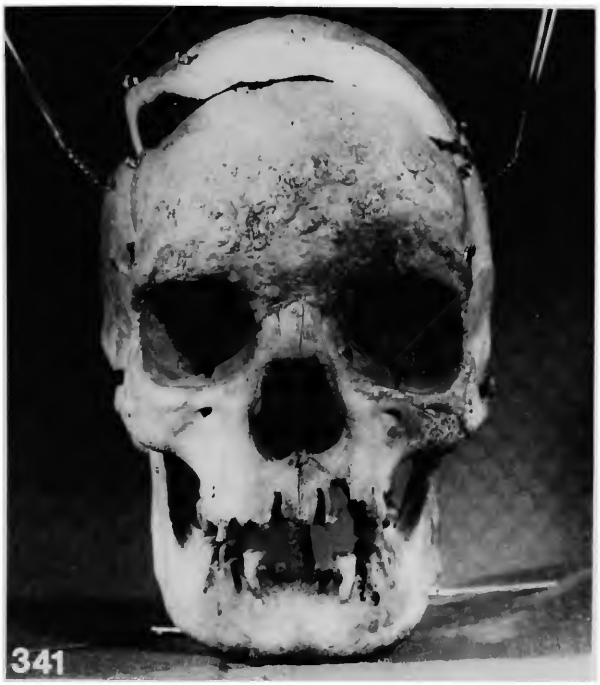
cortex, which is most pronounced on the posterior aspect. The surface is generally smooth with isolated regions of slight porosity. Muscle attachments in the affected area are noticeably rugose. The entire shaft of the right femur is abnormal, somewhat more so distally than proximally (Figures 331, 332). The gross lesion consists of thickening with isolated patches of porosity. Proximal muscle attachments appear normal. However, there has been considerable thickening of bone with a very irregular surface in the distal region of the linea aspera. The anterior surface shows raised plaques and bony spicules, which appear to be bridging over superficial blood vessels. There are no cloacae. The last three features are very typical of conditions occurring in long bones of known cases of syphilis, and in my experience are not found together in nontreponemal infectious diseases.

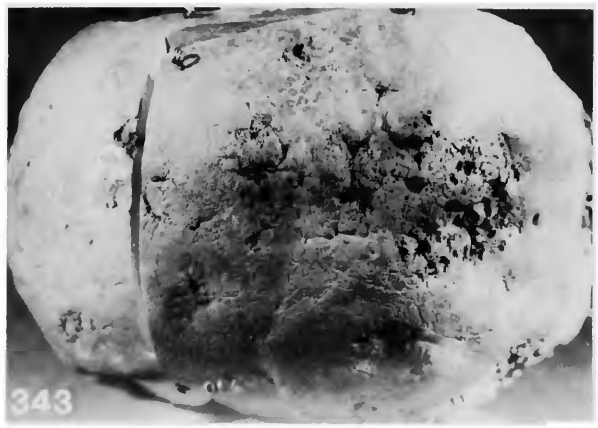
A very similar pattern of abnormal bone is seen on the left proximal tibia. The abnormal bone is expanded and has isolated foci of porosity. There are occasional, slightly raised plaques and bony bridges over superficial blood vessels. The attachments of the proximal posterior muscles and ligaments, including the soleus, popliteus, and the tibial collateral ligament, are markedly rugose. The surface appearance suggests that the periosteal bone grew around the ligament attachments but did not affect the attachment area.

The roentgen picture on both femora and the left tibia reveals the thickening of the cortex. The original cortex is still apparent but has been significantly cancellized in focal areas of disease in the three bones. Endosteal surfaces of the cortex do not encroach on the marrow cavity as often happens in osteomyelitis and occasionally in periostitis.

An isolated Eskimo skull from Mud Bay in the Alaska Peninsula (NMNH 372956) demonstrates a more active phase of probable treponemal disease. The skull is adult and is labeled female. The age is unknown, but minimal wear of the teeth suggests young adult. The archeological age is uncertain.

The lesions are distributed extensively over the



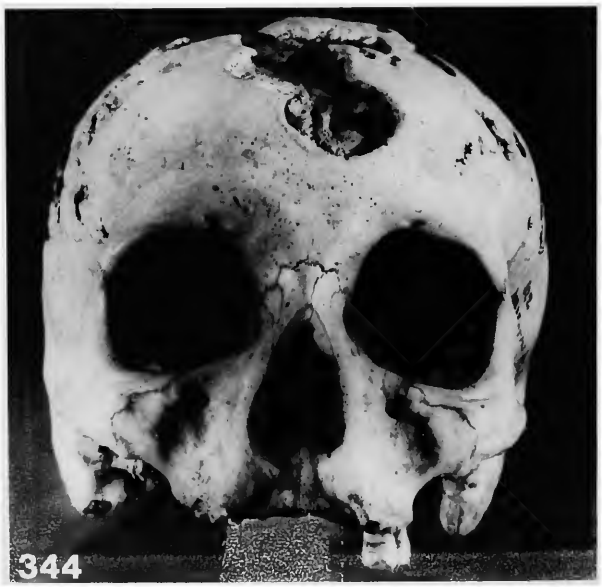


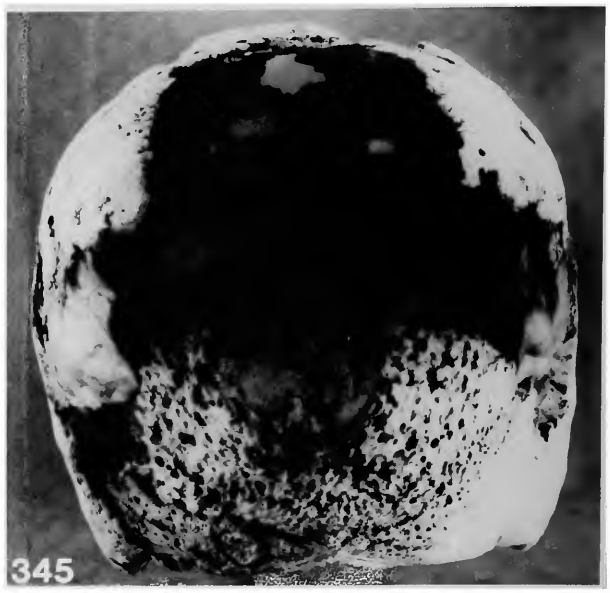
FIGURES 341–343.—Probable treponemal disease in an adult aboriginal Australian: 341, Facial view of the skull; note the healed nature of the frontal lesions and the sclerotic diploë in the sectioned portion of the frontal bone. 342, Lateroposterior view, showing active lesions on the occipital and posterior parietal bones. 343, Top view of the skull showing the more active nature of the lesions toward the posterior portion of the skull. (PMES 1GD1(116).)

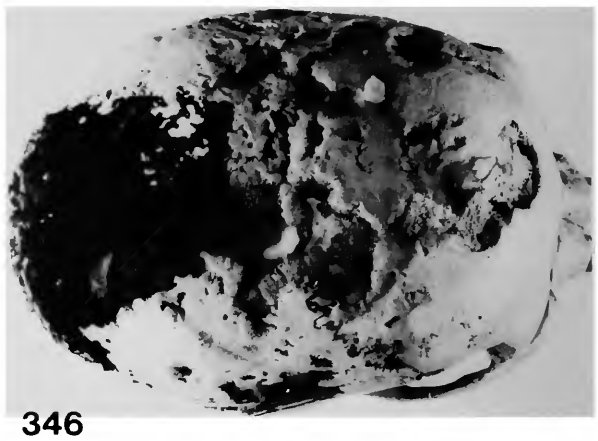
skull, sparing only part of the outer table underlying the temporalis muscle, the alveolar portion of the maxilla, the base of the skull, and most of the occipital bone (Figures 333, 334). In general, the lesions are almost purely lytic, coalescing foci, which leave more or less intact projections of bone between the foci. This creates the very irregular surface seen on the frontal bone and on the posterior portion of both parietals. The destructive process was sufficiently severe to expose the frontal sinuses. The bones of the face are affected as well, including the nasal spine, which has been destroyed. At the edges of the large coalescing lytic lesions one sees fine, porous, periosteal, reactive bone (Figure 335). The lesions are largely confined to the outer table, although slight porosity is apparent on the inner table, particularly in association with the most extensive lesions on the outer table. There is very little evidence of healing and this is largely restricted to a slight remodeling (smoothing) of remaining bone projections. The penetrating damage on both parietals and the frontal appears to be postmortem.

The lesions in this specimen have a distribution and morphology compatible with treponemal disease, although the predominantly active nature of the lesions is less commonly seen in such skeletons. The young age of the specimen, along with the active nature of the lesions, suggests that the individual died during the active phase of the disease.

The above specimen contrasts with another Eskimo skull (Figures 336–338), St. Lawrence Island, Alaska (NMNH 280095). The mandible







FIGURES 344–346.—Probable treponemal disease in an adult aboriginal Australian: 344, Facial view of skull; note the large, confluent, lytic lesions of the frontal bone. 345, Posterior view of the skull showing an active, lytic morbid process. 346, Top view of the skull showing a large area of bone loss. This is the result of extensive necrosis followed by sequestration and possible exfoliation of the sequestrated bone. (WM HS 50a.4.)

and postcranial bones were not recovered. Regrettably, the archeological age is unknown, but other specimens collected at the same time were thought to be post-Russian contact. The skull is female and the teeth in the maxilla, except the right canine, have been lost before death. The canine is badly worn which, with the loss of teeth, suggests an older age than the previous case. The lesions are largely confined to the skull vault, although there is a focus on the left parietal bone superior to the mastoid process, which also involves the adjacent temporal bone (Figure 336). While the most obvious lesions are confined to the outer table, the external lesions penetrate to the inner surface in the region of the left mastoid, the right parietal, and the posterior sagittal suture. There is considerable fine porosity on the inner surface of the vault with some lytic depressions about 0.5 centimeter in diameter, particularly on the right parietal.

The lesions in the frontal region of the outer table reveal a fairly typical, coalescing, lytic-blastic response associated with the chronic, less active phase of treponemal disease. In the posterior portion of the skull, the lesions appear to be more active and are largely lytic in nature. However,

FIGURES 347, 348.—Tibial lesions attributed to yaws or treponarid (endemic syphilis) in a specimen from Australia prior to European contact: 347, Posterior view of tibiae. 348, Medial view of tibial lesions. (AIAC SF 19:27, in Hackett, 1978; photograph courtesy of Dr. C. J. Hackett and the South Australian Museum.)

there is a slight development of reactive bone at the margins of the more active lesions.

The general picture in this case is that of a more chronic course of the disease process with considerable healing in the frontal region but with active lesions elsewhere. The more active lesions on the younger specimen from Mud Bay, Alaska, may reflect a more aggressive course of the disease, an earlier infection by the disease, or death occurring during the active phase of the disease and before any healing could occur. The specimen from St. Lawrence Island may have acquired the disease later in life or the disease may have taken a less aggressive course.

The pattern of healing lesions on the frontal bone with more active lesions in the posterior portion of the skull is also seen in three specimens located in the European pathology collections. The first of these is a specimen now located in the Wellcome Museum in London, but found on Atui in the Cook Islands (WM S 50 A5). The skull probably dates to before 1850. Age and sex are unknown, but the condition of the teeth suggests fully adult age and the skull morphology is female. There is considerable destruction of the nasal bones and the hard palate (Figure 339). The frontal bone shows extensive confluent lytic foci, which have been remodeled and healed. More active lesions are seen in the right parietal bone (Figure 340).

The second specimen is the skull of an adult aboriginal Australian. The skull appears to be female and judging from toothwear was over 30 years of age. It dates to before 1920 and probably is much older. Currently the specimen is in the Pathology Museum of The Royal College of Sugeons of Edinburgh (PMES 1GD1(116)). The frontal bone exhibits extensive, but mostly healed, lesions (Figure 341). However, the lesions become more active in appearance toward the occipital region (Figures 342, 343). The most active lesions are seen in the occipital bone but extend up into the left parietal (Figure 342). The skull has been sectioned and reveals a sclerotic diploë similar to that seen in the archeological specimen from Arkansas described earlier.

The third case of probable treponemal disease is also the skull of an Australian aborigine (Figures 344–346) currently located in The Royal College of Surgeons of England Wellcome Museum (WM HS 50a.4). The date in the catalog is 1811; thus the skull dates to before that year. In contrast to previously described specimens, this shows much more extensive destruction, particularly in the posterior portion of the parietals and the superior occipital bone (Figure 346). On the frontal bone there are large carious defects with porous reactive bone on the boundary with normal bone and evidence of some healing (Figure 344). However, the occipital bone exhibits an active destructive process, which could be active syphilis or possible secondary pyogenic osteomyelitis (Figure 345). Again, the lesions exhibit a pattern of increasing activity toward the posterior portion of the skull.

Hackett (1978) describes several cases of bone lesions in pre-European skeletons from Australia. He attributes these lesions to yaws or treponarid (endemic syphilis), since venereal syphilis is not thought to have been present until European contact. The long bone lesions of one of these cases is shown in Figures 347 and 348. The lytic lesions in this case are quite different from bony reaction seen in the femur of the specimen from Arkansas, USA (Figures 331, 332).

Actinomycosis and Nocardiosis

Pathology

Actinomyces israelii is a higher bacterium, rather than a true fungus, and is a common saprophyte of the human oral cavity. Infections with this organism are uncommon and occur mainly in three areas of the body: cervicofacial, thoracic, and abdominal. The organism enters through the oral mucosa in the first area, by aspiration into the lungs in the second, and by the gastrointestinal route in the third. The cervicofacial location is the most common (Nathan, Radman, and Barton, 1962). Of 60 clinical cases reported by Baracz (1902), 55 involved head and neck, 3 thoracic,

and 2 abdominal lesions. The distribution of the disease is worldwide. The great majority of the human infections is restricted to the soft tissue and organs (lungs, liver, intestines). It consists of a protracted purulent infection with great tendency for local invasion and extension and frequent fistula formation. Bone involvement is uncommon. In a study including statistics of 486 cases of human actinomycosis, Grässner (1929) reported 73 incidents of skeletal involvement (15 percent). The bone or bones are usually affected by direct extension of the infection from an adjacent soft tissue focus. This means that, in most instances, the bone infection starts on the periosteal surface and frequently remains limited to it. Hematogenous dissemination, especially to the long bones of the extremities, rarely occurs. The frequency of actinomycotic lesions in the different

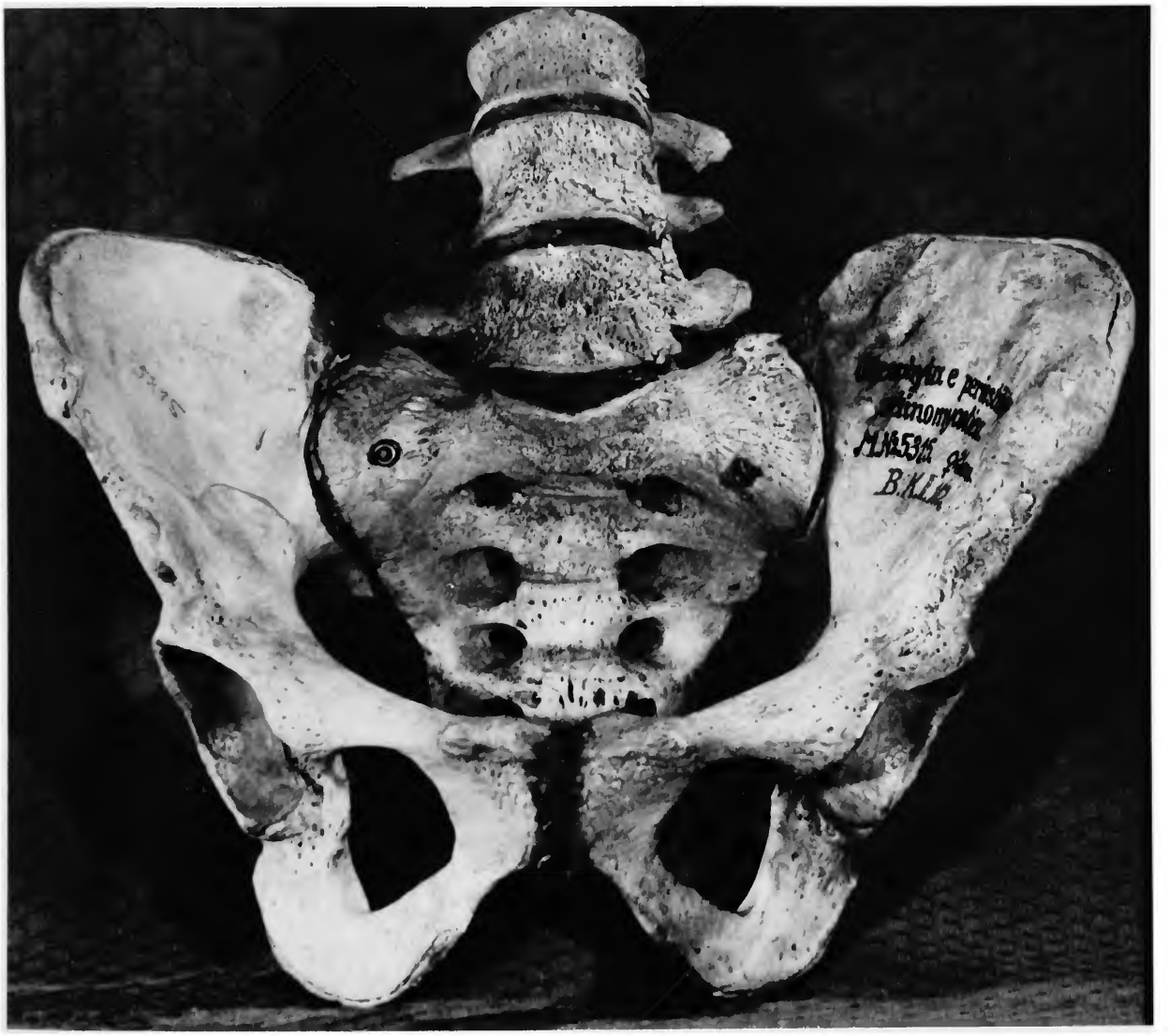


FIGURE 349.—Actinomycotic periostitis of lumbosacral spine and left ilium, secondary to actinomycosis of sigmoid colon, left ovary, and presacral soft tissue. (21-year-old female, FPAM 5315, autopsy 93144 from 1891.)

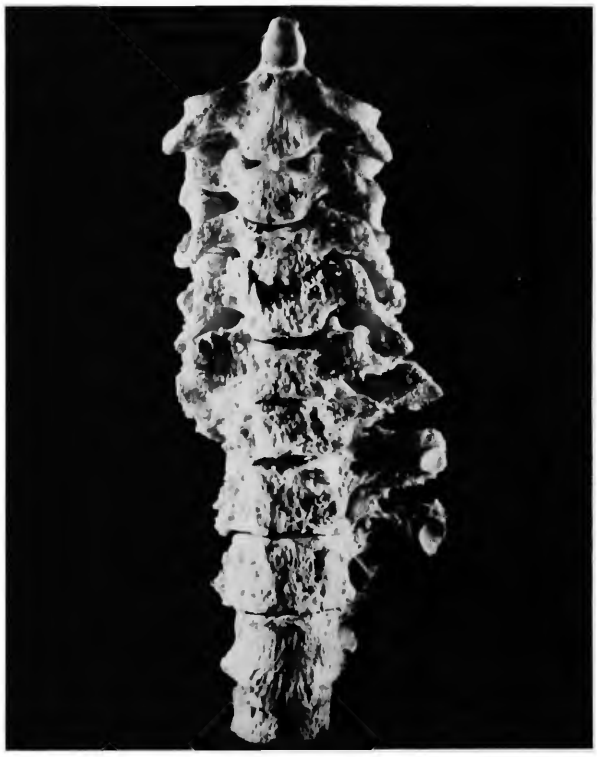


FIGURE 350.—Actinomycosis of cervical spine. Notice periosteal reactive bone including transverse processes and superficial erosion of vertebral bodies from without. (53-year-old male with actinomycosis of cervical soft tissues and both lung apices, UGPM autopsy 203 from 1907.)

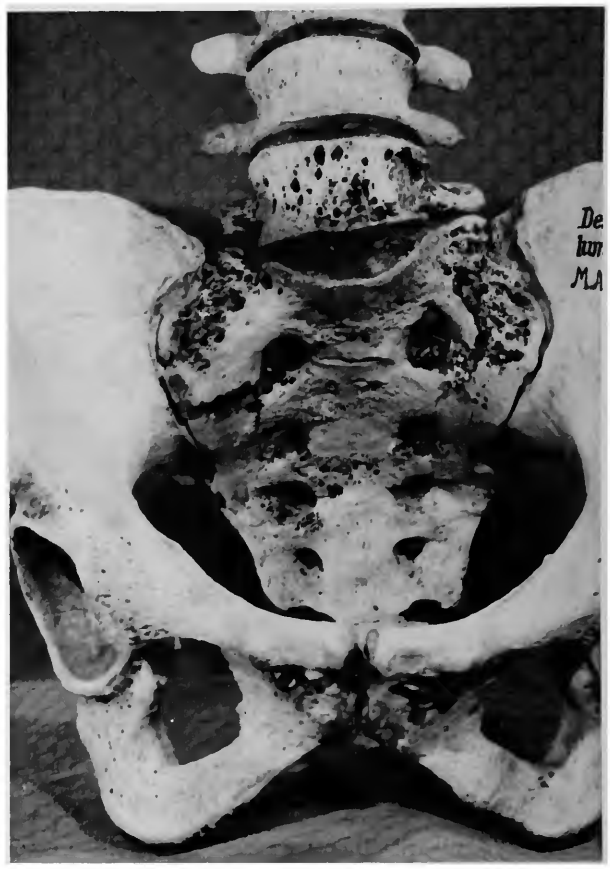


FIGURE 351.—Actinomycosis of fifth lumbar vertebra and sacrum. Notice pitted transcortical erosions based on periosteum. (46-year-old female, FPAM 5649 from 1897.)

bones in Grässner's study is as follows (quoted from Beitzke, 1934b:540): vertebrae 37 percent, mandible 25 percent, ribs 10 percent, maxilla 8 percent, extremities 5 percent, skull base 4 percent, pelvis 4 percent, sternum 3 percent, zygoma 3 percent, and clavicle 1 percent.

The periosteal involvement usually leads to hypervascularity, which is noticeable on the dry bone by the increased number and size of vascular channels and foramina. In addition, there is usually a varying degree of reactive subperiosteal bone formation of porous or solid character. There is superficial erosion of the cortex and a varying degree of destruction of the adjacent cancellous bone, with little or no endosteal sclerotic response, in contrast to bacterial osteomyelitis. Sequester formation also is uncommon.

The most characteristic lesion is that of the spine (Young, 1960). The segmental location, in decreasing frequency, is as follows: Thoracic, lumbar, cervical, and sacral (Beitzke, 1934b:554). Since the spine is usually infected from spreading pleural, abdominal, or cervical soft tissue lesions in broad contact with its anterior surface, the anterior periosteum of several or many vertebral bodies is involved (Figure 349). Transverse processes and, in the thoracic portion, attached ribs often participate in the lesions (Figure 350). The involvement of the vertebral bodies, if present, starts anteriorly and seldom extends very deeply (Figure 351). Vertebral collapse and gibbus formation hardly ever occur. The neural arches and

FIGURES 352, 353.—Actinomycosis of lumbar spine and left pelvis, showing mainly periosteal reactive bone deposition and hypervascularity with focal erosion of cortex: 352, Interior view. 353, Exterior view. (18-year-old male, FPAM 5686, autopsy 130155-1353 from 1909.)

spinous processes are usually spared. The intervertebral discs are usually preserved. Lumbosacral actinomycosis may spread to the pelvic bones (Figures 352, 353).

The mandibular lesions are mostly limited to periosteal reactive bone formation of moderate extent with occasional destruction and focal necrosis of the underlying bone, especially of the alveolar process. A central involvement of the mandible, possibly through a dental alveolus, leading to cavitation and expansion of the bone, is very rare in the human. By contrast, this is the most frequent type observed in bovines and equines infected with *Actinomyces bovis*, which is usually not pathogenic for humans. In the rare actinomycosis of the maxilla, adjacent paranasal sinuses, facial bones (zygoma), and middle and inner ear may become involved. Destruction of mastoid process and petrous bone have been observed.

In small and flat bones (ribs, sternum, pelvis), the destruction may be more extensive, creating a "worm-eaten" appearance and large cortical perforations. Actinomycosis of long bones is rather rare and probably mostly the result of hematogenous dissemination from a pulmonary focus. The area most frequently affected is the metaphysis. Extensive destruction of the cortex with reactive periosteal bone formation may be observed, but large cortical sequestra, as in coccal osteomyelitis, do not occur. Focal cavitation may closely resemble a Brodies's abscess, but perifocal osteosclerosis is usually slight or wanting.

Joint involvement may occur, secondarily, to lesions in adjacent bones. This is most often the case in costovertebral and less commonly in intervertebral joints (Beitzke, 1934b:563). Pelvic actinomycosis can extend to the hip joint and proximal femur. Periosteal reactive bone is seen in such areas. If actinomycotic arthritis heals, bony ankylosis results.

Nocardia asteroides is a bacterium of the actinomycetales group which, in contrast to actinomyces, grows aerobically and stains acid-fast. Infections occur through the pulmonary route and mostly are confined to the lung and pleural cavity (Pizzolato, 1971:1064–1066). In rare instances hematogenous spread to bones occurs. The lesions predilect cancellous areas and mainly consist of lytic cavitation, often complicated by fistula (Figure 354). There is little, if any, reactive bone formed.

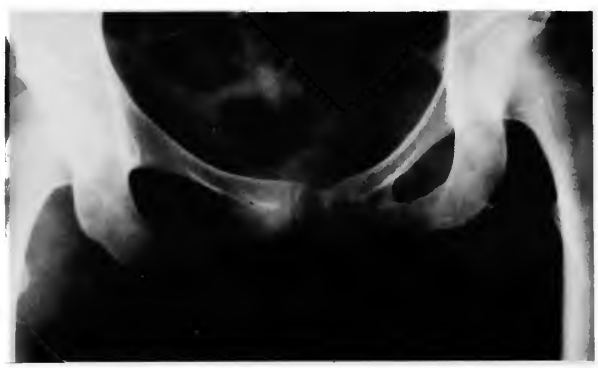
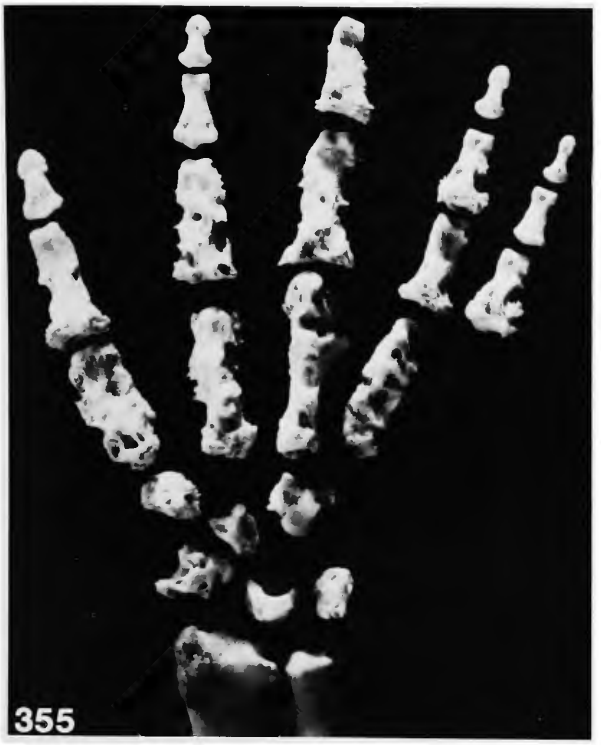


FIGURE 354.—X-ray of nocardia osteomyelitis of pubis with fistula, proven by culture. (27-year-old female with pulmonary cavity, MGH 1293348.)

Mycetoma (Maduromycosis)

PATHOLOGY

Mycetoma or maduromycosis is a clinicopathological entity, caused by a variety of different higher bacteria (*Actinomyces*, *Nocardia*, *Streptomyces*) and different true fungi of various genera that occur in the soil (Winslow, 1971:591). The infection almost always is located in the foot (Madura foot), much less commonly in the upper extremity (Oyston, 1961)(Figures 355, 356). The disease is practically limited to tropical and subtropical areas. The geographic distribution illustrates this point. In Asia, the disease is common in India, particularly in the south, in Ceylon, and in the Philippines. In Africa, the affected areas are Madagascar, Zanzibar, East Africa, Senegal, Tunis, Algiers, Morocco, and southwestern Africa. In the Americas, the disease is not uncommon in Cuba, Nicaragua, Costa Rica, Guyana, Brazil, Argentina, and Chile. In the United States, 30 cases had been observed before 1921, mostly in the southwestern states. In Oceania, cases were reported from Yap (Carolines). In Europe, mycetoma was observed in Sardinia, southern Italy, and Greece (Plehn, 1928). The infection occurs through skin lacerations in barefoot-going populations, producing long-lasting, progressive, soft tissue lesions with formation of multiple fistulae. The bones become secondarily infected through the periosteum. The tarsal and metatarsal bones usually show multiple lytic foci and progressive osteoporosis (Figure 357). The tarsal and ankle joints frequently participate in the process. The distal tibia and fibula as well as the metatarsals are involved in advanced cases, showing multiple cortical perforations and destructions with very little reactive bone formation. If the process heals, bony ankylosis of the affected joints is to be expected (Figure 358).



FIGURES 355, 356.—Actinomycosis (maduromycosis) of left hand: 355, Volar view; notice extensive periosteal involvement of carpals, metacarpals and phalanges. 356, X-ray; notice subcortical involvement and cloacal openings on some bones. (Adult Iranian male, AFIP 859604.)

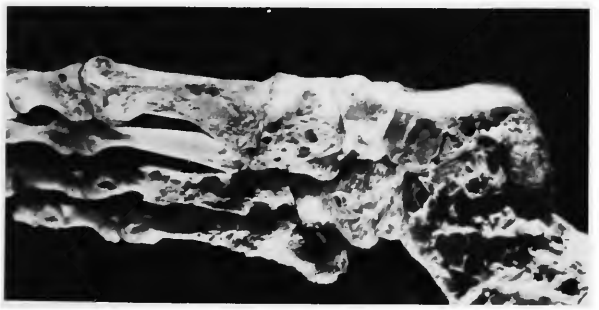


FIGURE 357.—Maduromycosis of right foot, plantar view, showing extensive lytic destruction of most bones. (Adult Asian Indian from India, WM S 52.1 from 1875.)

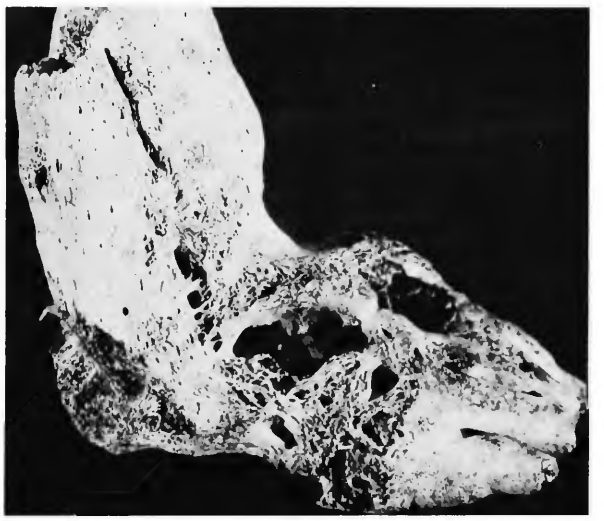


FIGURE 358.—Infection of right ankle and foot, with ankylosis of ankle, tarsal and tarsometatarsal joints. Notice the extensive destruction of the tarsal bones and the large cloacas, suggestive of actinomycosis or fungal infection, but ordinary osteomyelitis cannot be ruled out. (HM P948.)

Fungal Infections

Pathology

Of the numerous fungal infections of man, only those capable of involving bones concern us here. There is a variety of yeast-like, budding fungi with different geographic distributions causing human infections. This group of fungi all enter through the respiratory tract with the principal internal lesion usually in the lung. Occasionally, in systemic cases, hematogenous dissemination to the skeleton occurs. The bone lesions produced are similar to and not distinguishable from each other on X-ray or dry bone specimens. Therefore, the geographic location becomes a key factor in attempted identification.

North American Blastomycosis

North American blastomycosis is caused by *Blastomyces dermatitidis*, which usually enters the body through the respiratory tract. It can manifest itself as a skin infection, but frequently presents as a systemic disease (Witorsch and Utz, 1968:169–171). Its main distribution is in North America, especially in the Mississippi and Ohio valleys and North Carolina, but presently it has worldwide distribution to some extent. In Canada, Ontario and Quebec have a significant number of cases. The male:female ratio in disseminated cases is at least 5:1. There is no distinct racial predilection. The highest incidence is between 30 and 50 years of age (Chick, 1971:466–468). Two-thirds of the disseminated cases show bone involvement. The predilected areas are vertebrae and ribs (often multiple), skull, tibia, and tarsus. The lesions are usually purely lytic with sharp borders (Boswell, 1959). In contrast to tuberculosis, vertebral collapse is rare. Paravertebral abscesses occur. In long bones the focus can mimic an infarct. The foci in long bones predilect subarticular epiphysial and metaphysial areas. Periosteal reactive bone can be found (data from Chick, 1971:485–486).

Cryptococcosis

Cryptococcosis is the result of infection by *Cryptococcus neoformans*, mainly in the form of a systemic disease. The main distribution of cases is through Europe, with occasional cases occurring in all continents. The disease often develops in debilitated patients, especially in leukemia and lymphoma. There is no age, sex, or race predilection (Salfelder, 1971:385–390). About 10 percent of the disseminated infections show bone involvement. The predilected areas are bony prominences, cranial bones, and vertebrae, but any bone may be affected occasionally (data from Salfelder, 1971:420–421). The lesions usually appear lytic and well circumscribed on X-ray (Collins, 1950).

Paracoccidioidomycosis

Paracoccidioidomycosis is due to infection with *Paracoccidioides brasiliensis*. This American fungus disease mainly occurs in Brazil, Venezuela, and Colombia but is found from Mexico to Argentina with the exception of Chile, San Salvador, and Panama. The disease mainly affects middle-aged persons with a male:female ratio of at least 12:1. About half of the patients are farmers. Whites are most often involved and Negroes least (Angulo and Pollak, 1971:511–513). Bone involvement is not rare in disseminated cases. The lesions are round and lytic, single or multiple, or may mimic ordinary osteomyelitis (Emmons, Binford, and Utz, 1970:336). The predilected locations are clavicles, ribs, vertebrae, and extremities (Angulo and Pollak, 1971:555–556).

Coccidioidomycosis

Coccidioidomycosis is the result of inhalation of spores of *Coccidioides immitis*. This disease is heavily concentrated in parts of Southern California (San Joaquin Valley) but occurs in the dry areas of adjacent southwestern states and Mexico with additional foci in Central America, Venezuela, Bolivia, and Argentina (Huntington, 1971:

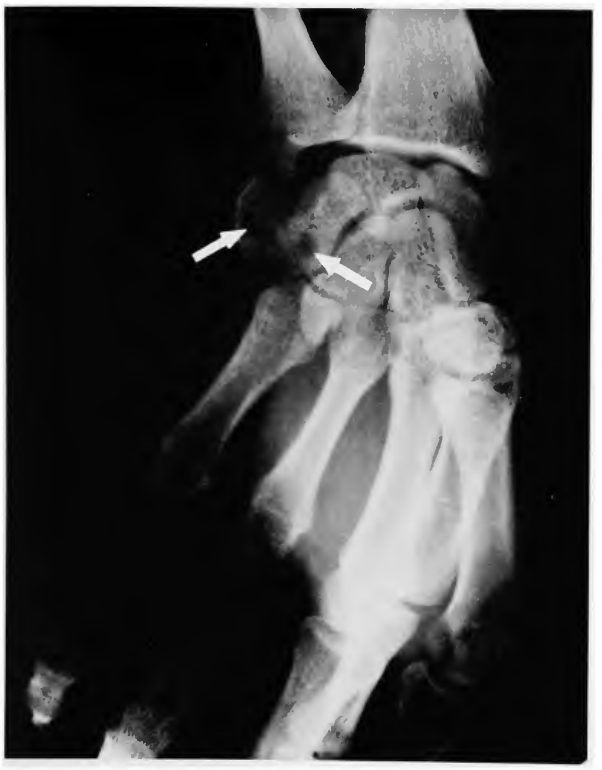


FIGURE 359.—X-ray of coccidioidomycosis of right carpal bones. Notice round lytic focus in os triquetrum and os pisiforme (arrows). (38-year-old male from California with pulmonary lesion, MGH admission no. 1187633.)

172–174). About 25 percent of the fatal disseminated cases show bone lesions (Huntington, 1971: 184). The mortality is considerably higher in Negroes and Filipinos than in Whites and Mexican Indians in California (Huntington, 1959). (For clinical and radiological reviews, see Miller and Birsner, 1949; Dykes, Segesman, and Birsner, 1953; and Birsner and Smart, 1956.)

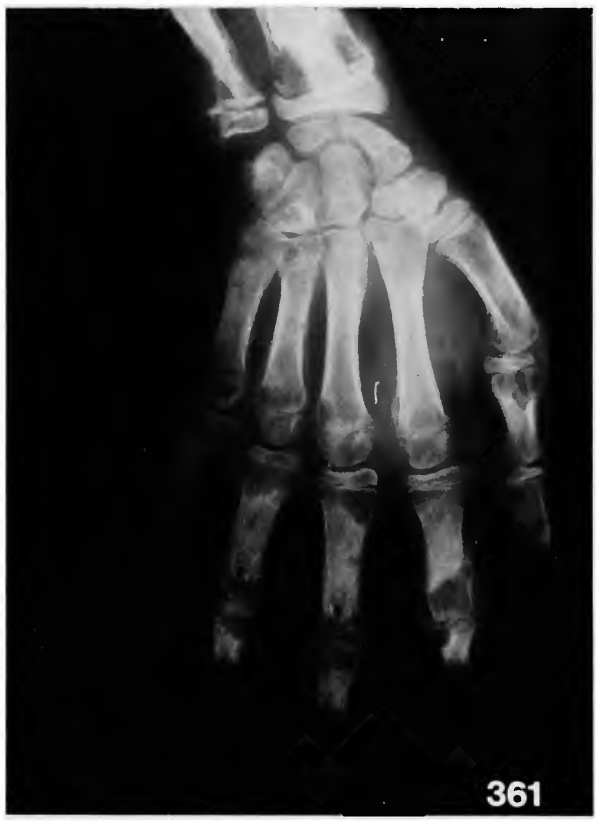
The bone lesions of all these fungus infections are very similar. Cancellous bone areas are predilected and multiple lesions are not uncommon. In the spine, the lesion may be indistinguishable from tuberculosis, although in these conditions the posterior elements are not spared and vertebral collapse with gibbus formation usually does not occur. In general, the bone lesions tend to be lytic on X-ray and show central cavitation, usually combined with osteoporosis, in the gross. Perifocal reactive sclerosis can be found, making the differentiation from Brodie's abscess difficult or impossible. Fistula formation is common. The most characteristic aspect of these systemic fungus lesions is that they predilect areas of the skeleton usually spared by other infections. Such areas are bony prominences such as the acromion, coracoid process, styloid processes of radius and ulna, condyles of the humerus, ends of clavicles, málleoli and tibial tuberosity. There is also involvement of small bones: carpals (Figure 359), tarsal bones, and patella. Marginal destructive lesions of ribs and destructive lesions of the skull, limited to the outer table, do occur (Carter, 1934). In carpal and tarsal lesions, adjacent joints are often implicated.

Histoplasmosis

Histoplasmosis is a common infection of the respiratory tract, caused by *Histoplasma capsulatum.* Infections are usually slight and self-healing and are most common in the Mississippi and Ohio valleys. A few cases occur, sporadically, worldwide. Bone lesions are uncommon even in the systemic cases but, if present, resemble those in other systemic fungal diseases (Schwarz, 1971:95–97). There is, however, a different species (*Histoplasma duboisii*), which causes severe systemic infections in young Negroes in tropical West Africa (Nigeria). In this disease, multiple round lytic lesions usually are present in the cranial vault (Figure 360) and in long and small bones of the extremities (Figure 361)(Cockshott and Lucas, 1964). Such lesions in long bones may lead to expansion of the shaft (Edington, 1971:131–138).

Sporotrichosis

Sporotrichosis mainly manifests itself as a skin and soft tissue infection, caused by *Sporotrichum schenkii* entering the body through skin lacerations. The disease has a worldwide distribution between latitudes of 50° north and south. There is a male:female ratio of 3:1. The majority of cases are children and young adults (Lurie, 1971:



616–621). In about 10 percent of the cases, mostly hematogenous osseous lesions occur (DeBeurmann and Gougerot, 1912:329). The majority of cases described are in the French literature. Bone lesions represent the most frequent extracutaneous foci in sporotrichosis. The bones affected, in decreasing order of frequency, are tibia, hands and feet, ulna and radius, skull and facial bones, ribs, clavicles and vertebrae. The process may extend into joints, terminating in severe destruction (Altner and Turner, 1970). The joints involved, in decreasing frequency are knee, elbow, wrist, and sternoclavicular joints (data from Lurie, 1971:629–630). Bone lesions may be mild, healing without residual alterations. Localized bone abscesses and ossifying focal or extensive periostitis have been observed. Spontaneous fractures can occur (Zeiliger, 1911:13–28). The lesions are probably too rare and too uncharacteristic to be identified without recovery of the causative organism. The bone lesions most frequently affect the tibia and the skull. Hands and feet, especially metacarpals and metatarsals, are more often involved than long bones of the extremities. The spine and bones of the thorax seem to be spared. The prevailing lesion is periosteal rather than a deep lytic bone abscess or osteomyelitis (The above data are from Beitzke, 1934e:616–619).

Aspergillosis

Several species of *Aspergillus*, especially *Aspergillus fumigatus*, may act as opportunistic invaders in debilitated individuals. Aspergillosis occurs worldwide, without predilection of sex or race (Peña, 1971:766–767). Destruction and necrosis of bone between affected paranasal sinuses and

FIGURES 360, 361.—Bone lesions in *Histoplasma duboisii* infection: 360, X-ray of lateral view of skull; notice multiple round lytic lesions of cranial vault. 361, X-ray of right hand; notice multiple lytic lesions in bones of forearm and hand, partly with sclerotic border. (Adolescent negro, courtesy Dr. Stanley Bohrer, Ibadan, Nigeria.)

orbit or anterior cranial fossa can be observed. Hematogenous dissemination is rare but may involve ribs or vertebrae. In vertebral aspergillosis, collapse and gibbus occurs, which may mimic tuberculous lesions (Peña, 1971:807–812).

Mucormycosis (Phycomycosis)

Several genera of Mucoraceae (*Rhizopus*, *Mucor*, and *Absidia*) can occasionally become invasive opportunistic pathogens. Most of the patients affected are diabetics in the acidotic state (Straatsma, Zimmerman, and Gass, 1962). Extension into bone is seen in the cranial form of mucormycosis, starting from the nasal cavity with involvement of the paranasal sinuses and their walls. The main reason for discussing this lesion here is that perforation of the hard palate, with or without sequestrum, occurs, which may be confused with a tertiary syphilitic lesion. Since only one maxillary sinus is usually affected by mucormycosis, the perforation of the hard palate is more often unilateral (Baker, 1971:835–860).

Paleopathology

Fungal infections and their effect on human bone have received little attention in the literature on paleopathology. Poswall (1976) has called attention to the presence of both coccidioidomycosis and blastomycosis in North America. She suggests that skeletal lesions from these fungal diseases may be distinguishable from other infectious diseases of bone and may be a more probable diagnosis for bone lesions seen in skeletal remains of some American Indian groups. Morse (1969:45) had included both blastomycosis and coccidioidomycosis as diseases that must be considered in the diagnosis of destructive lesions of the spine. Similarly, Buikstra (1976) has called attention to the need to differentiate fungal infections from other infectious conditions that affect bone. She indicates the importance of epidemiological factors, as well as the morphology and location of the lesions in the skeleton, as helpful diagnostic features.

There are, however, no published cases of bone lesions in paleopathological specimens attributed to fungal infections. Clearly in geographical regions where such diseases are known to have been endemic in the historic period, it is probable that they existed in ancient times as well. Lesions in skeletal material recovered from such areas, such as the American Southwest, should be studied with an awareness that fungal diseases represent a significant cause of conditions that can produce skeletal lesions.

Viral Infections

Pathology

Of the viral infections involving the skeleton, only smallpox and rubella need to be discussed. In chickenpox, bone and joint participation is extremely rare, and vaccinia osteomyelitis, representing a rare complication of prophylactic vaccination, does not fall within the scope of this book.

Smallpox

Smallpox was a worldwide common viral infection, especially in infants and children, before the use of vaccination. Until recently, large areas of Africa had a high incidence of smallpox. As late as 1958, Cockshott and MacGregor reported a series of 2500 cases observed in 18 months in Nigeria, and Davidson and Palmer (1963) report 400 cases from southern Rhodesia. During my work in India and Indonesia in the 1960's I had ample opportunity to study the radiological changes of smallpox osteomyelitis and arthritis. The skeletal involvement varies in different series from 2 to 5 percent (Cockshott and MacGregor, 1958:377, 1959:57; Middlemiss, 1962:11–13) to about 20 percent (Davidson and Palmer, 1963: 687). The skeletal infection starts between one and four weeks after onset of the disease; 80 percent of the patients in the series of Davidson and Palmer (1963) were less than 5 years of age. Skeletal involvement is not seen in the adult. Variola osteomyelitis, in contrast to most other

FIGURE 362.—X-ray of smallpox osteomyelitis of both arms and hands; notice the involvement of humerus, radius, and ulna and of most metacarpals and phalanges. (2-year-old female Indonesian; courtesy Dr. L. A. Tamaela, Djakarta, Indonesia.)

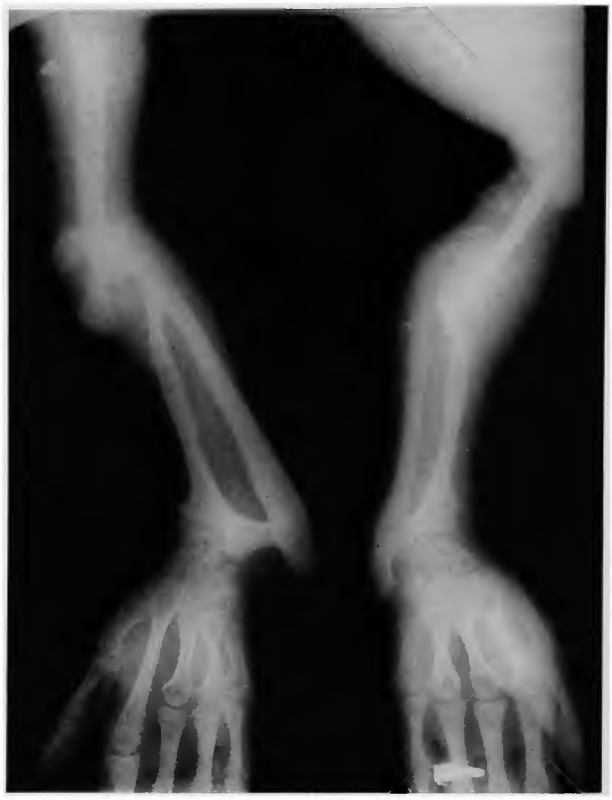


FIGURE 363.—X-ray of growth deformity of forearms 7 years after onset of smallpox osteomyelitis. (13-year-old male Asian Indian; courtesy X-ray Department, All India Institute of Medical Sciences, New Delhi, India.)

skeletal infections, predilects the upper extremities, especially the elbow areas. The lesion usually starts in the metaphysial area near the growth plates and often spreads to the adjacent joint. There is destruction of metaphysial bone, leading to separation of the epiphysis and, in severe cases, to pathological fracture. There is no massive sequestration, in contrast to bacterial osteomyelitis, but pronounced formation of reactive periosteal bone attached to the cortical surface is usually present. In about 80 percent of the 81 patients of Davidson and Palmer (1963), the elbow was affected and very frequently bilaterally, which is uncommon in any other infection. Often all three bones (humerus, ulna, and radius) participated, while in most other infections, including tuberculosis, the radius is usually spared. Second to the elbows, the wrists, knees, and ankles are most often affected, but any joint may be involved. In a review of 124 published cases of variola arthritis, Cockshott and McGregor (1958:376) report the following locations: elbows 97, wrist and hands 25, knees 17, ankle and foot 16. In more than half of the cases more than one area was affected. In carpal and tarsal bones, which are often involved, uneven, patchy, lytic destruction is seen on X-ray (Figure 362). Lesions in the calcaneus are common (Davidson and Palmer, 1963:692). In fingers and toes dactylitis occurs. Ribs, spine, pelvis, and cranium are usually spared in smallpox. During the active disease, the bones may be sufficiently weakened to permit bending deformities to arise in weight-bearing areas. Permanent changes are mainly deformities due to arrested growth, sec-

ondary to destruction or slipping of the growth plate (Figure 363). Dactylitis may lead to short stubby digits for the same reason. Joint involvement may terminate in bony ankylosis.

Rubella

Rubella is a relatively harmless viral infection of children and adults which, however, often has serious effects on the fetus, if acquired by the mother during the first trimester of pregnancy. In addition to the triad of congenital cataract, heart defects, and mental retardation, frequent skeletal changes have been appreciated only recently. In a series of 81 infants with a history of maternal rubella during pregnancy, 34 showed skeletal involvement (Singleton, Rudolph, Rosenberg, and Singer, 1966). The most characteristic lesions are seen on X-ray in the metaphysial areas of the long bones of upper and lower extremities of the newborn. The changes affect all metaphysial areas but are most obvious on the distal femoral and proximal tibial metaphysis, reflecting the greatest growth rate in these areas. These changes consist of poor mineralization adjacent to the growth plate and coarsening of the reduced trabecular pattern. The diaphyses are not involved. The lesion is similar to that of congenital syphilis, but in contrast to syphilis, ossifying periostitis is not observed in rubella (Highman, 1967). There is general retardation of growth, and the skull may show enlargement of the anterior fontanel and poor mineralization. If the infant survives, the bone changes disappear at about 3 months of age without leaving residual irregularities of architecture; but the mortality rate in this group with skeletal involvement was 32 percent (Singleton, Rudolph, Rosenberg, and Singer, 1966).

Parasitic Infections (Echinococcosis)

Pathology

The only parasitic infestation of the human causing significant bone changes is the *Echinococcus. Echinococcus granulosus* is a tapeworm, which inhabits the small intestine of canines. The larval stage causes echinococcosis in humans and domestic animals (sheep, cattle, and pigs), which serve as intermediate hosts. This circumstance gives this disease a special paleopathological interest, since human involvement would depend on the domestication of the dog and the herding of domestic animals. The present and recently past geographic distribution of the disease is closely tied to the use of dogs in cattle and sheep herding by people living under primitive hygienic conditions. The disease occurs in two different manifestations, probably caused by two different but closely related parasites. The common manifestation is the hydatid cyst, which has worldwide distribution. The European areas presently or previously involved are Iceland (with the greatest frequency in the past), Mecklenburg, Pomerania, Friesland, Spain, Sardinia, Sicily, Yugoslavia, Hungary, Bulgaria, Rumania, Greece, Russia, parts of France and Switzerland, and England. Outside of Europe, the areas of highest incidence are Australia and Argentina, followed by New Zealand, South Africa, Uruguay and Chile. In my own experience, even presently, cystic echinococcosis is a common disease in Iran. The alveolar type of echinococcosis is much rarer and occurs in a more limited area: in Europe, particularly in the Alps and their forelands (Bavaria, Austria, eastern France, Switzerland, northern Italy) and, to some extent, the Balkans and southern Russia. I have seen one case in northern Iran. In the areas where the alveolar form occurs, the hydatid form is rare (Posselt, 1900:238, 314).

In the heavily infested areas, echinococcosis is a major health problem and a significant cause of death. As recently as 1935, Račić reported that in Split (Yugoslavia) 70 to 100 percent of the animals coming to the slaughterhouse were infected with hydatid cysts and that he, alone, in 20 years operated on 269 cases of human echinococcosis, including five involving the skeleton. Ivanissevich (1934) surveyed the problem in Argentina and found that 1 percent of all hospital patients and 3 percent of all operated patients in a large hospital in Buenos Aires suffered from hydatid cysts. Similar figures are reported from

Australia. In Concepción (Chile), hydatid cysts were found in about 1 percent of all autopsies and over 0.5 percent of all surgicals (Behn, 1938). Slightly lower autopsy figures were reported from Virchow's Institute in Berlin (Boecker, 1868). In this series of 33 cases, none were found in individuals below 15 or above 70 years of age.

The infection occurs either by direct contact with dogs or by contamination of pasture, crops and drinking water with dog feces containing the ova of the parasite. Domestic animals form the reservoir, reinfecting dogs through their eating of or contact with the offal of slaughter. The embryonated ova, ingested, lose their shells by digestive action of the stomach and penetrate with their six hooks the intestinal blood capillaries of the portal system. Through the portal blood they most commonly infect the liver but may, with varying frequency, settle in any part of the body. In the organ or tissue involved, the larva forms a cyst, which consists of laminated chitin and is usually surrounded, by the host, by a fibrous capsule. The inner layer of a fertile cyst forms many little tapeworm heads (scolices), each with a crown of hooks. If such a cyst ruptures, each scolex can form a new cyst. Hydatid cysts are usually well tolerated by the affected tissue. In contrast, the rare alveolar *Echinococcus* continues to invade and destroy the infected tissue in the form of a microcystic invasion, producing extensive necrosis of the tissue and giving the gross appearance of a malignant neoplasm. It was Virchow (1856) who first recognized the parasitic origin of this lesion.

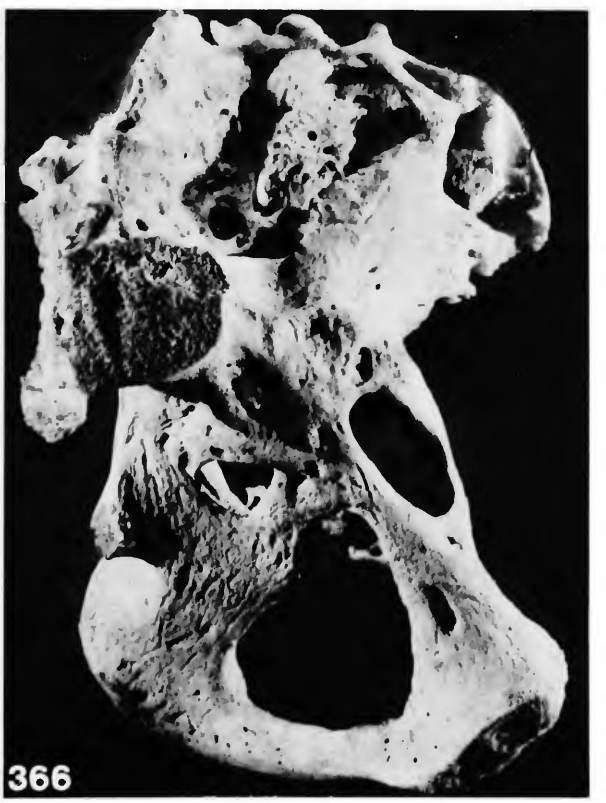
The behavior of the hydatid type of echinococcosis in the skeleton is different from that in organs and soft tissues. Usually, no major cysts are formed, but small cysts rupture and disseminate between the bone trabeculae, causing osteoclastic resorption. There is often foreign body giant cell reaction around ruptured cysts but no inflammatory reaction or fibrous encapsulation (Ivanissevich, 1934). Extensive permeation of the marrow spaces may cause local ischemic necrosis of the bone with formation of sequestra. By contrast, in the few instances of alveolar echinococcosis involving bone, the microcysts may not be recognizable grossly but, as in other tissue, the parasite shows a more aggressive growth and produces extensive necrosis and inflammatory response (Klages, 1930:141). The main difference between the two types is that the hydatid form resumes its macrocystic patterns when it escapes from the confines of the bone into the soft tissue, while the alveolar form always maintains its microcystic destructive growth. There are very few cases of alveolar echinococcosis of bone on record and this type of bone lesion has never been found in Africa, Australia, or America.

The frequency of skeletal involvement in echinococcosis of the hydatid type is about 2 percent in the statistics of Ivanissevich (1934:17) from

FIGURE 364.—Echinococcus granulosus cyst of left scapula showing ballooning of the inferior angle and the lateral margin with sparse reticulated new cortex. (Adult, specimen provided by Lord Joseph Lister's collection, before 1912, WM S 54.1.)

Argentina and other series give similar data. According to collective worldwide statistics of 406 cases of skeletal echinococcosis published by Pasquali (1930:369), the distribution through the skeleton is as follows: vertebrae 41.6 percent, hip bone 22.1 percent, humerus 9.4 percent, tibia and fibula 8.9 percent, femur 8.1 percent, skull 5.2 percent, sternum and ribs 2.2 percent, scapula 1.5 percent, phalanges 0.7 percent. All bones are occasionally involved, with the exception of the carpal and tarsal bones and the ulna (Ivanissevich, 1934:21). The vertebral lesions form close to one-half of all cases, followed in frequency by the pelvic bones. The segments of the spine most often affected are the middorsal and sacral region (Ivanissevich, 1934:21).

The parasites most often settle in areas of cancellous bone with hemopoietic marrow. This means predilection, in the long bones, of the metaphysis and, in the spine, of the vertebral bodies. The infection progresses very slowly over decades and may remain asymptomatic for a long time. The changes in different bones are different. In the spine, progressive destruction of one or several vertebral bodies may lead to angular kyphosis, similar to a tuberculous gibbus. Transverse processes and posterior elements are more often involved in echinococcosis than in tuberculosis. In flat bones, the outer contour is often expanded by slow destruction of the old cortex and formation of a new bony shell (Figure 364). The progressive destruction, especially of pelvic bones (Doebbelin, 1898), may be extreme, leading to multiple large cystic cavities separated by residual bony septa (Figures 365, 366). The large cavities do not contain large parasitic cysts but



FIGURES 365, 366.—Echinococcus granulosus cysts of left pelvis with secondary infection and destruction of acetabulum, 18 years duration: 365, Lateral view. 366, Medial view. (Adult male, GHPM 4196.)

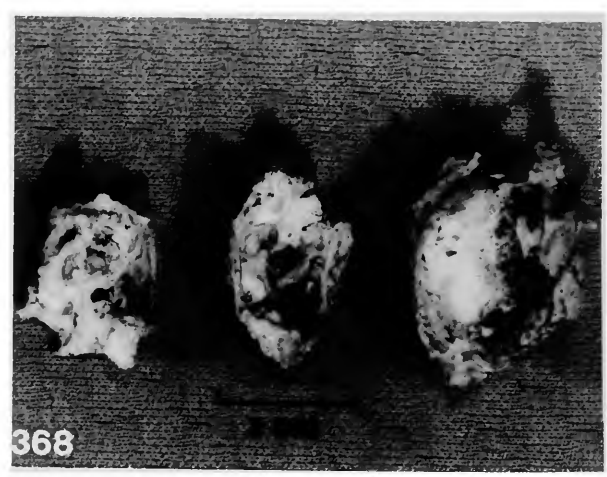
numerous small cysts and tube-like membranes. Destruction of the floor of the acetabulum with central dislocation of the head of the femur has been observed (Račić, 1935). On X-ray, the polycystic transformation and great reduction of bony density is apparent. In the differential diagnosis on dry bone, fibrous dysplasia has to be excluded. In this latter condition, the destruction of the bony structure between the fibrous foci is much less pronounced. The cystic form of hyperparathyroidism can give a similar picture but would not be limited to one area. The rest of the skeleton would show other evidence of hyperparathyroidism.

In long bones, the parasitic small cysts starting in the cancellous bone of the metaphysis may ultimately occupy the entire marrow cavity. Articular or epiphysial cartilage forms a barrier, which is not penetrated by the parasite. This explains the absence of joint involvement. The outer contour of the involved bone may not be altered, but scalloping of the endosteal cortical surface may be apparent on X-ray. Pathological fractures occur frequently in the late stage of the disease. Gangolphe (1894:675) observed in 25 cases of long bone involvement 14 pathological fractures (humerus 5, femur 5, tibia 4). Ultimately, even long bones may be extensively destroyed if the cortex is transgressed and the parasites extend into the soft tissue. Reactive osteosclerosis and periosteal new bone formation is uncommon except in cases complicated by superimposed bacterial osteomyelitis.

Paleopathology

Paleopathological evidence of echinococcosis may occur in the skeleton, but it may also be inferred from the presence of mineralized outer shells of hydatid cysts occurring in the thoracic or abdominal cavity. Wells and Dallas (1976) found a probable example of such a cyst in the thoracic cavity of a woman about 45 years of age. The skeleton of this woman was excavated from a site at Orton Longueville, Cambridgeshire, England. The site was dated to the late first or early second century A.D. and is part of a Belgic and early Romano-British farmstead. In the left side of the thoracic cavity, the excavators found a mineralized shell approximately ellipsoid in shape measuring 47 × 35 millimeters. The object was not attached to bone. Wells and Dallas conclude that the specimen is the mineralized shell of a hydatid cyst produced in response to infestation by the tapeworm *Echinococcus granulosus.*

Similar cyst-like structures are associated with an adult female skeleton from Jones Point, Kodiak Island, Alaska, USA (NMNH 374623). The skeleton is associated with what Hrdlička called



Figures 367, 368.—Three cyst-like structures associated with an adult female skeleton from Kodiak Island, Alaska, USA. These structures may be the mineralized outer shells of a hydatid cyst resulting from infestation by Echinococcus: 367, External view. 368, Internal view. (NMNH 374623.)

the intermediate, pre-Koniag period, which would have been before Russian contact. The skeleton is incomplete and fragmentary, but the skull and postcranial bones exhibit no external evidence of disease. X-ray films of the major long bones, vertebrae, and innominates do not reveal any cystic changes with the exception of a partial cyst-like structure in the cancellous bone of the right iliac crest. The postmortem damage of this bone makes firm identification impossible. There are, however, three fragmentary, ovoid structures associated with the skeleton but not attached to any bones (Figures 367, 368). These fragmentary structures appear to represent three different mineralized cysts. The external surfaces of the cysts are irregular with calcified spurs and holes in the surface, indicating that the external shell of the cyst was not completely mineralized. Internally there is no evidence of bony divisions in the cyst. Wells and Dallas (1976) note that hydatid cysts of *Echinococcus* typically are unilocular and may range in size from a pigeon's egg to as large as a human skull. The mineralized cysts from this specimen are very similar to the cyst illustrated by Wells and Dallas (1976, pl. 6). I would agree that the most likely diagnosis is that they represent the mineralized partial outer shell of a hydatid cyst resulting from *Echinococcus* infestation.

Sarcoidosis

Pathology

Sarcoidosis is a granulomatous disease of unknown etiology affecting mainly lymph nodes, lungs, and skin. The disease is more often observed in adolescents and young adults than in children and is even rarer in infants. There is a marked racial predilection for Negroes. About 14 percent of the cases show bone involvement (Mayock, Bertrand, Morrison, and Scott, 1963: 76). The bone lesions are usually multiple and often bilateral.

The bone lesions are most commonly located in the fingers and toes, less often in metacarpals and metatarsals. The second and third phalanges are most frequently involved. Within the phalanges, the lesions predilect the distal epiphysis but do not break through into the interphalangeal joints. The granulomas cause purely lytic, round, or slightly lobulated defects in the spongiosa, varying from one millimeter to one centimeter or more. If the granuloma is located in the diaphysis, the cortex is markedly thinned and the shaft may be slightly widened, with loss of the dumbbell shape of the bone. There is usually no significant perifocal sclerosis. There is no sequestra formation. Perforations of the thin diaphysial cortex by the granulomas are not uncommon and create round defects. There is usually only minimal or no reactive periosteal bone formation. Occasionally phalanges, especially terminal, may be completely destroyed. Destruction of nasal bones occurs secondary to granulomas of the skin of the bridge of the nose (Holt and Owens, 1949).

Lesions of similar lytic character have occasionally been described in other bones, such as skull, pelvis, long tubular bones, especially in the metaepiphysial spongiosa, and vertebrae. The vertebral lesions are located in the vertebral bodies and usually spare the intervertebral discs and the pedicles. Vertebral collapse may occur. Lesions usually involve multiple, sometimes widely separated, vertebrae and may show sclerotic rimming.

In differential diagnosis, the hand and foot lesions have to be differentiated from leprosy, spina ventosa, and from osteochondromatosis (Ollier's disease). In tuberculous and syphilitic spina ventosa the widening of the affected bone is more pronounced; there is marked periosteal new bone formation and a central sequestrum is often present. In Ollier's disease the enlargement and distension of the involved phalanges is much more marked. There is also usually similar involvement of large tubular bones of the same extremity while bilaterality is uncommon. The greatest similarity exists between the bone lesions of sarcoidosis and those of lepromatous leprosy (Paterson and Job, 1964:432). Both show the same predilection of the phalanges of fingers and toes, the lytic character of the lesions, and absence of reactive bone formation. A differentiation of individual lesions may be impossible on dry bone alone. The cranial lesion may be more character-

istic, since sarcoidosis mainly affects the nasal bones but not particularly the inferior nasal spine and maxilla. The mutilating lesions have to be differentiated from leprosy, which would not usually show simultaneously lytic lesions in other phalanges. The vertebral lesions have to be differentiated from those of tuberculosis and of osteomyelitis. Osteomyelitis usually does not spare the intervertebral space, and, therefore, often involves two adjacent vertebrae, while discontinuous involvement of vertebral bodies is less common. The sarcoid lesions also have to be distinguished from metastatic lytic neoplastic lesions which, by contrast, usually involve the pedicles and spinous processes multifocally.

Circulatory Disturbances

Blood Supply of Bones

For the present purpose we are mainly interested in the arterial supply of the long bones of the extremities. There is a separate arterial supply for the diaphyses, for the metaphyses, and for the epiphyses. The one or several larger nutrient arteries enter the bone through a grossly visible nutrient canal on the posterior aspect and branch into an ascending and descending ramus in the medullary cavity. The metaphyses have several smaller nutrient vessels entering through the cortex around the circumference. The epiphyses receive several smaller arteries, which branch from a vessel that also supplies the joint capsule and synovium. During the growing period, the growth plate completely separates the vascular territories of the metaphysis and the epiphysis. After closure of the growth plate, some connections between the two systems are reestablished, but the circulation, to a large extent, remains separate. The intraosseous arteries, being enclosed in a rigid compartment shielded from external pressure, are usually thin-walled. This is particularly true of the arterioles. The diaphysial cortex is, in part, supplied by ramifications of the nutrient vessels, in part by the periosteal vessels. The relative contribution of these two systems varies in different portions of the same bone. The epiphysial vessels, which form a system of arcades beneath the articular cartilage, contribute to the nutrition of the cartilage during the growing period, before formation of a more or less solid subchondral bony plate. The cortical bone has vascular supply through the Haversian and the Volkmann canals, which connect Haversian canals and also enter directly from the periosteum. Cancellous bone trabeculae are usually avascular, depending on the vasculature of the marrow spaces for nutrition. The sinusoidal veins of the marrow are numerous, thin-walled, and, in hemopoietic marrow, wide. They collect into a large, thin-walled vein, running lengthwise in the medullary canal. The venous return in part follows the nutrient artery, in part exits through multiple, circumferentially located, venous outlets in the metaphysial area. The epiphysial venous return drains into that of the adjacent joint capsule. On smaller and cancellous bones the blood supply is less complicated, except for those with growth plates, which always lead to separate epiphysial and apophysial vascular territories, at least during the growing period. The vertebral bodies show a radiant arrangement of larger veins, which converge medially and emit on the posterior surface of the vertebral body through two foramina, close to the midline. These segmental veins join the longitudinal vertebral plexus in the spinal canal, which is of some significance in the dissemination of infection or malignancy. The diploë of the cranial vault likewise shows large interconnecting venous channels, which drain through the parietal and mastoid emissary foramina but also have some connection with the large intracranial venous sinuses through small openings in the inner table.

Ischemia and Infarction

Pathology

Continued cortical remodeling during adult life leads to formation of more osteon fragments and interstitial lamellae, which are devoid of vascular channels. With increasing age more and more of these fragments become necrotic (Müller, 1926). In arteriosclerotic peripheral vascular disease, extensive portions, especially of the cortex, can become necrotic (Jaffe and Pomeranz, 1934; Sherman and Selakovich, 1957). Although microscopic study of the intact fresh specimen will reveal these changes, they are demonstrable on X-ray and on dry bone only if reaction in the

adjacent living bone, in the form of bone resorption or bone formation, has occurred. In many instances, such reactions to cortical necrosis do not occur and therefore will not be recognizable in an archeological specimen.

In infarction, larger areas of the fatty bone marrow and the intervening bone trabeculae undergo necrosis, presumably due to interruption of the circulation. The lesion is almost exclusively observed in the long bones of the extremities, mostly in adults. The areas involved are principally the diaphysial and metaphysial marrow of the femur, tibia, or humerus. The epiphyses, particularly the head of the femur or, less commonly, of the humerus are also the sites of avascular necrosis.

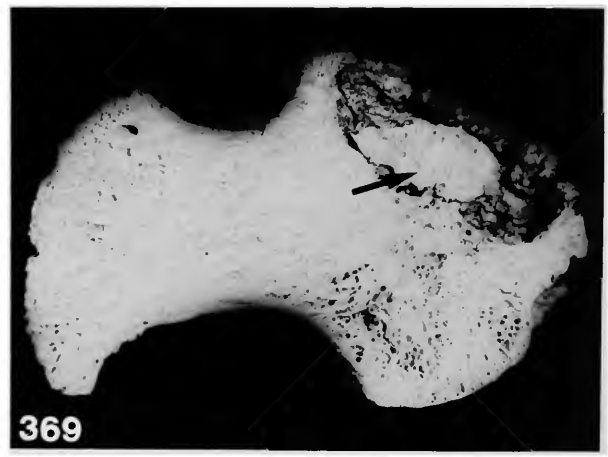
An infarction in the shaft of a long bone undergoes calcification, particularly at the interface between living and dead marrow, due to conversion of the liberated fat into calcium soaps. In fact, it is this change that makes the lesion visible on X-ray as a circumscribed lesion, often of up to 10 centimeters in length, showing a radiodense shell around the whole lesion and between its individual components. There is little or no change in the overlying cortex. It is questionable whether calcium deposits could be preserved in archeological material. Also, the sifting in of mineral particles into the marrow cavities of interred bones may mimic infarction on X-ray.

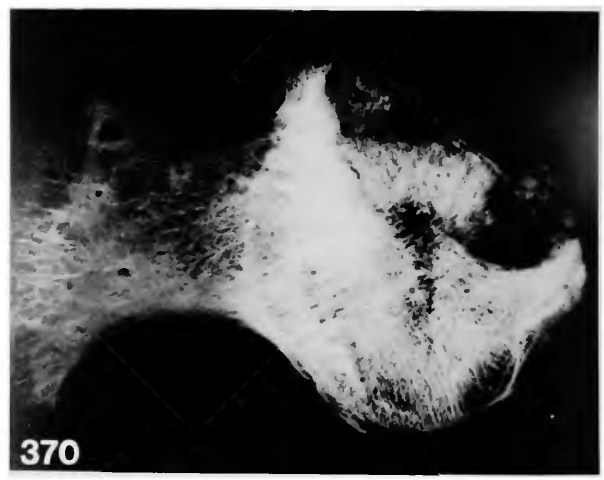
In the majority of present cases a definite cause for the infarction cannot be stated. Some known causes of circulatory impairment are blood sludging in hemoglobinopathies, nitrogen gas embolization in decompression disease, and arteritis in systemic lupus erythematosus. Circulating lipase, a fat splitting enzyme, in extensive pancreatic fat necrosis may also cause necrosis of medullary fat tissue (Scarpelli, 1956).

Necrosis of Femoral Head

Pathology

Subcapital or transcervical fracture of the neck of the femur frequently is complicated by aseptic necrosis of part or all of the femoral head (Figures 369, 370). This is due to the peculiarity of the blood supply to the femoral head, which consists of three groups of vessels: (1) the artery of the ligamentum teres, supplying a portion of the upper part around the fovea; (2) the inferior metaphysial vessels coursing beneath the synovium on the inferior aspect of the femoral neck





Figures 369, 370.—Aseptic necrosis of femoral head: 369, Proximal right femur, anterior view, showing collapse and cavitation of femoral head with sequestrum of necrotic bone (arrow). 370, X-ray, showing reactive osteosclerosis on the border of living and dead bone. (69-year-old male with bilateral aseptic necrosis of femoral head for several years; USPHS surgical specimen 2427 of 1975, courtesy of Dr. B. Ragsdale, Armed Forces Institute of Pathology, Washington, D. C., U.S.A.)

and supplying the inferior portion of the head; and (3) the lateral epiphysial vessels, which enter the head within one centimeter distal to the margin of the articular cartilage, supplying most of the femoral head and communicating especially with the vessels of the ligamentum teres (Trueta and Harrison, 1953). These are, apparently the most important vessels. If they are severed by the fracture, aseptic necrosis always develops, whereas the common severance of the inferior metaphysial vessels bears no consistant relationship to necrosis (Claffey, 1960). The area around the fovea is often spared if the ligamentum teres is intact.

If the fracture is survived for a long time, inactivity osteoporosis characteristically will be present in any surviving part of the femoral head and in the femoral neck, while the necrotic portion maintains the trabecular pattern as of the time of fracture, if no revascularization and new bone deposition has occurred. The necrotic portion appears dense on X-ray for several reasons: (1) the above-mentioned inactivity atrophy and osteoporosis in the adjacent surviving bone, (2) the deposition of calcium into the necrotic tissue of the fatty marrow, (3) the compression of dead bone trabeculae into a smaller area during collapse, and (4) the deposition of new bone on dead bone in the process of repair.

Traumatic dislocation of the hip also can be followed by aseptic necrosis. This is seen more frequently in posterior dislocations, especially if they are complicated by acetabular fractures. In traumatic dislocations the ligamentum teres is usually torn, and it is particularly an area on the top of the femoral head that may become necrotic and demarcated as a sequestrum surrounded by a sclerotic rim on its base, resembling a focus of osteochondritis dissecans.

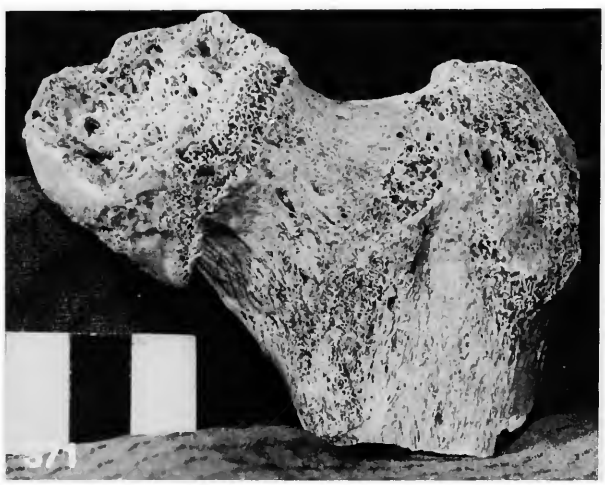
The so-called idiopathic aseptic necrosis of the femoral head is not common. About 50 percent of the cases are bilateral. Approximately 80 percent of the cases occur between 30 and 60 years of age, and approximately 80 percent are in males (Jaffe, 1972:634).

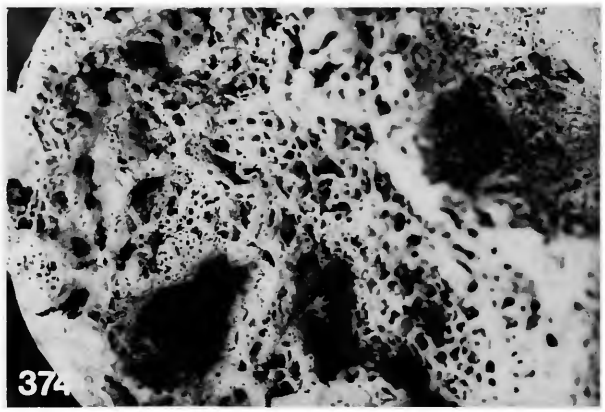
The necrosis usually begins in the weight-bearing area of the femoral head, beneath the articular cartilage. Its further development and gross and radiologic appearances are not different from those of known etiology, such as sickle cell anemia or Gaucher's disease. In systemic lupus erythematosus, a disease of the connective tissue of the body, unilateral or bilateral necrosis of the femoral head is not uncommon (Dubois and Cozen, 1960).

Paleopathology

In the paleopathological literature, circulatory disturbances have received little attention. Gray (1968) describes two possible cases of bone infarction in Egyptian archeological material. The first of these is a mummy (his No. 6) dated to the Greek period (ca. 330 B.C.). The mummy is currently located in the Horniman Museum in London. Gray found radiodensities in the distal femora and proximal and distal tibia, all of which he attributes to bone infarction. The figures of the X-ray films show radiodense lines circumscribing normal or radiolucent areas. The remainder of the skeleton is normal. The second case is a mummy (his No. 7) dated to the Roman period (first to second century A.D.). The distal portion of the femur has a large, oblong radiodensity aligned with the long axis of the bone. Gray attributes this condition to a solitary bone infarct or enchondroma. While these may very well be examples of bone infarction, all of the lesions illustrated in the published figures are suspicious. All the changes seen on the X-ray film could be due to dried overlying soft tissues or, depending on burial conditions, to infiltration of the tissue by soil constituents. Final judgment on these cases should be postponed until these possibilities are eliminated.

A possible case of necrosis of the femoral head is seen in a proximal fragment of a left femur (Figures 371–374) excavated in an American Indian site in Arkansas, USA (NMNH 255142). The archeological age is uncertain. Most of the superior portion of the femoral head was destroyed antemortem creating two large confluent





FIGURES 371–374.—Necrosis of the femoral head of left femur fragment: 371, Anterior view. 372, X-ray of anteroposterior view; note the radio-dense sclerosis inferior to the necrotic defect. 373, Anterosuperior view of reactive bone. 374, Detailed view of reactive bone showing the thickened trabeculae. (NMNH 255142.)

depressions. The smallest of these is superior, lateral, and continuous with the femoral neck. The largest depression involves about one-half of the area of the femoral head. There has been considerable bony reaction in the depressions, in which the exposed trabeculae have become greatly thickened indicating a long-standing condition. On the anteroinferior margin of the articular surface there is a bony projection about one centimeter square extending inferiorly. There is noticeable periosteal bone deposition on the femoral neck, suggestive of a low grade inflammatory condition. This raises the possibility of a septic condition contributing to the necrosis of the femoral head. However, periosteal reaction to an aseptic inflammation arising from trauma is also possible.

Perthes' Disease and Slipped Femoral Capital Epiphysis

PATHOLOGY

Perthes' disease is represented by extensive aseptic necrosis of the epiphysis of the femoral head. It is an uncommon disease occurring unilaterally in 90 percent of the cases. It usually begins between 5 and 9 years of age and involves

boys four times as often as girls (Jaffe, 1972:566). The condition has been found in different races but is rare in Negroes.

In the course of the disease the necrotic epiphysis increases in radiodensity, contrasted with focal radiolucency of the area of the femoral neck bordering the growth plate. Later the head flattens, due to a combination of compression fracture and lack of enchondral growth. The basal bulge of the flattened head leads to subperiosteal and enchondral bone formation, thickening the neck of the femur. The end result, after revascularization, is a mushroom shaped femoral head with overhanging margin but without significant dislocation of the center of the femoral head from the axis of the shortened femoral neck. Early severe degenerative arthritis modifies the appearance and can make the differentiation from the endstage of slipped femoral epiphysis difficult or impossible. In the acute stage, the differentiation from tuberculous coxitis and from aseptic necrosis in Gaucher's disease may be uncertain in dry bone.

Although slipped femoral capital epiphysis is not primarily an aseptic necrosis, it is best to discuss it in this connection. The primary pathology of this lesion is a stress fracture between the metaphysial side of the growth plate and the neck of the femur. This allows medial posterior and downward displacement of the head of the femur and, not uncommonly, leads to some degree of aseptic necrosis in the epiphysial bone. The disease is uncommon and occurs more frequently in boys than in girls. The age of manifestation ranges from 10 to 16 years in boys, 9 to 14 years in girls (Jaffe, 1972:577). Kelsey, Keggi, and Southwick (1970) found Negroes almost twice as often affected as Whites.

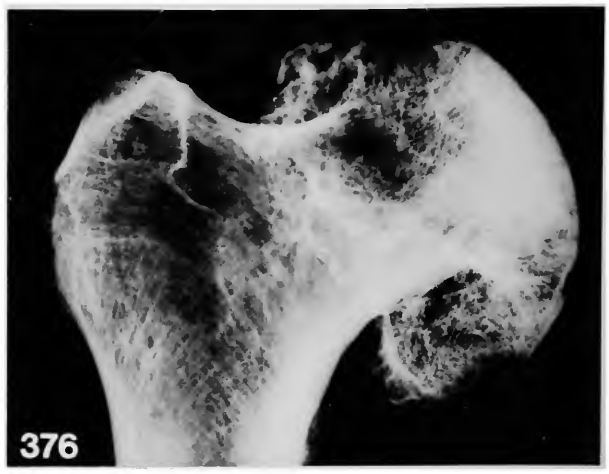
Since the growth plate remains with the epiphysis, the bone of the epiphysis is little altered, except in cases with extensive aseptic necrosis. The proximal end of the femoral neck, however, shows irregularities due to the fracture with subsequent abrasion and resorption. In dry bone, since the joint capsule and the cartilage are missing, the appearance of the proximal surface of the femoral neck would be the main clue. In the healed stage, the head, united with the neck in the slipped position, shows some dislocation of the center of the head towards the axis of the neck. The neck is always short and thick, reflecting both the attrition in the fracture area and the loss of enchondral growth for several years. As in Perthes' disease, early and severe degenerative arthritis complicates the picture. Occasionally, upward dislocation of the femur is observed, creating a new "acetabulum" on the lateral aspect of the ilium, while the slipped head is held in the anatomical acetabulum by the ligamentum teres (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952 (1):450–454).

Paleopathology

A right femur from the Valley of Chicama in Peru provides a possible example of Perthes' disease. This case is from a miscellaneous group of femora all accessioned as NMNH 265331 at the National Museum of Natural History, Washington, D. C. The archeological age is unknown. The epiphyses of the femur are fully fused indicating full adulthood. The femur is 383 millimeters in length and gracile, suggesting female sex. The diaphysis, distal metaphysis, and the joint surface are normal. The maximum diameter of the femoral head is 60 millimeters, much larger than expected for a female. Part of the femoral head was damaged postmortem, although enough remains to indicate the nature of the pathology.

There is a large circumscribed porous lesion covering more than half of the remaining joint surface of the femoral head (Figures 375–377). This porosity has completely obliterated the depression for the ligamentum teres. There is a depressed groove at the boundary with normal bone. The margins of the joint surface are characterized by exuberent bony overgrowth creating a mushroom-like appearance. This overgrowth extends well over the femoral neck. There are some bony outgrowths on the superior portion of the neck, but otherwise the neck appears normal.

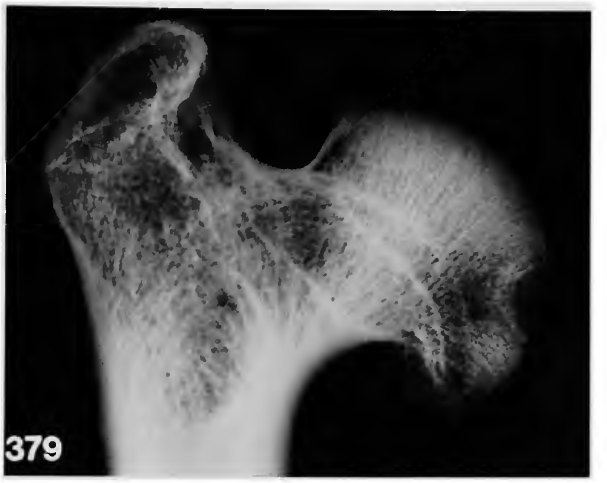
The X-ray film shows considerable thickening of the trabeculae under the porous lesion. This is indicated by a radiodense zone on the medial



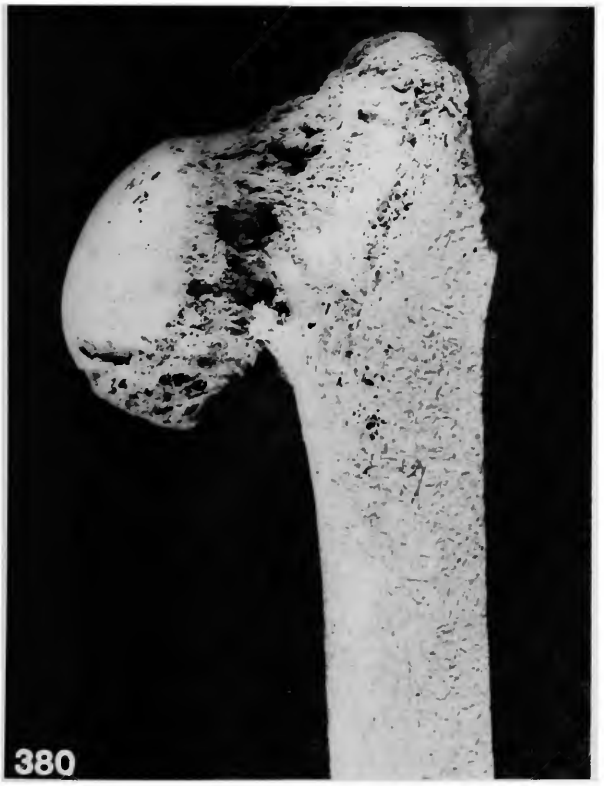
FIGURES 375–377.—Perthes' disease in a right femur: 375, Posterior view; notice extensive development of periarticular lipping. 376, X-ray of anteroposterior view; note the relatively normal length of the femoral neck. 377, Medial view of the femoral head; note the enlargement in diameter and extensive porous degeneration. (NMNH 256331.)

aspect of the femoral head. There are small radiolucent areas in the lateral head region and in the femoral neck. There has been considerable reduction in the mediolateral diameter of the head. This added to the marked overgrowth at the joint margins creates the false impression of the head having been forced into the femoral neck. However, the normal length of the neck and the overgrowth of the joint margins make it clear that the pathological process involves collapse of the superior medial portions of the femoral head followed by bony overgrowth on the articular margins.

The criteria for differentiating Perthes' disease and slipped femoral capital epiphysis can be seen in a right femur from the miscellaneous femora from Chicama, Peru (NMNH 265331). The maximum length of this femur is 370 millimeters, although this is misleading due to the inferior displacement of the femoral head. The bone is moderately robust, suggesting male sex. The diaphysis, distal metaphysis, and the joint surface are normal except for a moderate degree of middiaphysial, anteroposterior flattening. This is due in part to the abnormal gait induced by the



FIGURES 378, 379.—Slipped femoral capital epiphysis in a right femur: 378, Anterior view showing inferior displacement of capital epiphysis. 379, X-ray of anteroposterior view demonstrating shortened femoral neck. (NMNH 265331.)



FIGURES 380, 381.—Slipped femoral capital epiphysis in a left femur: 380, Anterior view; note that the superior articular surface is below the level of the greater trochanter. 381, Posterior view. (HMCS GR 1582.)

defective femoral head. The femoral head is displaced inferiorly (Figures 378, 379) so that the superior margin is about 15 millimeters lower than the greater trochanter. There is no evidence of porosity. Indeed the joint surface is smooth and intact. The depression for the ligamentum teres is well defined, unlike that of the head in Perthes' disease. Its position relative to the joint surface is much nearer the inferior, posterior margin of the joint surface than normal. The ligament attachment was maintained after the epiphysis slipped. Growth continued in the epiphysis but predominantly on the anterior and superior aspects. Ultimately, the head reunited with the neck. The femoral neck is abnormally short and thick resulting from the loss of the growth plate activity when the epiphysis slipped.

The X-ray film shows an even, well-organized, trabecular structure with no unusual radiologic abnormalities. It is possible to identify the growth plate of the femoral head in the film and see that the mediolateral diameter of the head is relatively normal. In marked contrast with Perthes' disease, the femoral neck is almost nonexistent on the superior aspect and greatly shortened inferiorly.

Another example of slipped epiphysis is seen in a left femur of a specimen from the Historical Museum in Chur, Switzerland (HMCS GR 1582). The specimen is from the archeological site at Bonaduz in Canton Grissons, Switzerland. The epiphysis has slipped inferiorly and the femoral neck is shortened (Figures 380, 381).

Other Vascular Disturbances of Epiphyses and Apophyses

There are a number of skeletal areas that are subjected to severe mechanical stress, which may lead to vascular disturbances of chronic trauma or a combination of both. This is particularly true of the skeletons of growing children and adolescents.

Köhler's Disease of the Tarsal Navicular Bone

PATHOLOGY.—This bone is in such a key position in the vault of the foot that its blood supply to the ossification center may be impaired in the growing child, leading to aseptic necrosis, usually with flattening of the bony center and increased density on X-ray. This flattening may be due in part to compression of the necrotic bone, in part merely an effect of arrested enchondral ossification in the necrotic area. Over the years more or less complete repair can be observed, secondary to revascularization. The disease is uncommon and occurs four to five times as often in males as in females. The onset of the disease is around 3 years of age, usually several months earlier in females in whom this ossification center develops correspondingly earlier (Jaffe, 1972:606).

Osteochondritis Dissecans

PATHOLOGY.—Osteochondritis dissecans is a type of aseptic necrosis characterized by the formation of a small, in profile triangular, sequestrum consisting of articular cartilage on one surface and necrotic subchondral cancellus bone. The disease occurs in the adolescent and young adult and is much more common in males than in females. Familial occurrence has been observed (Stougaard, 1964). The knee is the affected joint in 90 percent of the cases, and the most frequent location is the lateral portion of the articular surface of the medial femoral condyle. Other large joints are only rarely involved.

In the course of the disease, the necrotic fragment is extruded into the joint cavity and becomes a loose osteocartilaginous body, which often enlarges due to circumferential growth of the surviving cartilage. The cartilage may calcify, but the bone fragment remains dead and unaltered in size and shape. The subchondral osseous defect of the condyle closes over with a thin layer of bone but always remains a depression on the bony articular surface.

PALEOPATHOLOGY.—Wells (1961) describes an archeological skeleton from a Bronze Age cemetery (1600 B.C.) at Long Crickel, Dorset, England. The skeleton is that of a young female about 16 years of age. The abnormality consists of deficient bone on the anterosuperior margins of the third and fourth lumbar vertebral bodies. About one-third of the affected body is missing. Wells attributes this condition to adolescent osteochondritis.

FIGURE 382.—Osteochondritis dissecans of the medial right femoral condyle with severe arthritic degeneration of the joint. Specimen is a Late Saxon period burial from Thorpe St. Catherine, Great Britain. (Photograph reproduced from Wells 1974b, fig. 4, with permission from Mrs. Calvin Wells.)

However, a congenital malformation of the Morquio or Hurler type should be considered, as should trauma.

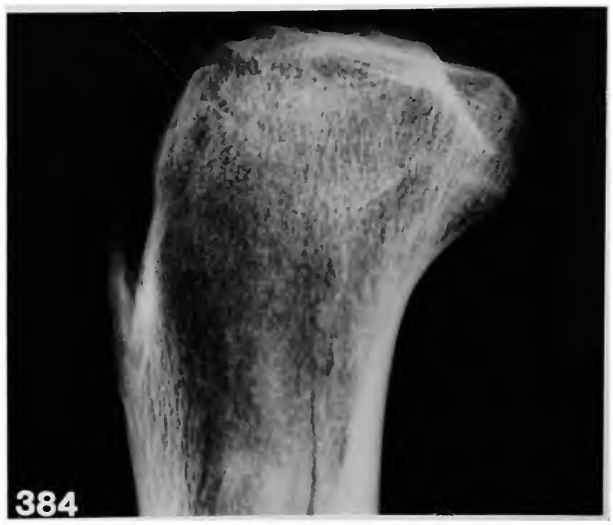
Wells (1962b) reports a case of osteochondritis dissecans in a Late Saxon burial excavated near Norwich, England. The specimen is male and the abnormality occurs on the medial condyle of the left femur. The defect is a large circumscribed depression in the joint surface about one centimeter in depth. Additional evidence of osteochondritis dissecans is reported in skeletons of Romano-British date from the Ieklingham cemetery in England (Wells, 1976). Several additional purported cases of osteochondritis dissecans in ancient British skeletal material are described by Wells (1974b). Figure 4 of his report shows a fairly certain example of osteochondritis dissecans of the distal femoral joint surface (Figure 382). In this report Wells expresses the opinion that osteochondritis dissecans is more common in Romano-British and Anglo-Saxon skeletons than skeletons of Bronze Age people in the British Isles.

Osgood-Schlatter's Disease of the Tibial Tubercle

PATHOLOGY.—The tibial tubercle is the site of the insertion of the patellar tendon. It develops from one or several apophysial ossification centers which, during the growing period, are separated from the proximal tibial metaphysis by a cartilage plate. There is considerable variation in this process but the powerful pull of the quadriceps may lead to partial avulsion of the tendon insertion with fragmentation of the apophysial center and ultimate marked nodular irregularity of the tibial tubercle after fusion with the metaphysis. This condition is essentially of traumatic origin and aseptic necrosis plays a minor, secondary part, if any. It is more common in boys than in girls. It does not occur before 8 years of age in girls and 10 years of age in boys and does not commence in either sex after 14 years of age (Jaffe, 1972: 599). More than 20 percent of the cases involve both knees.

PALEOPATHOLOGY.—Wells (1968) reports a possible case of Osgood-Schlatter's disease in a Late Saxon (ninth or tenth century A.D.) burial in Norwich, England (Norwich Castle Museum No. 15, 953 [9]). The affected bone is a left tibia from a male skeleton containing a defect of the tibial tuberosity. The defect consists of an oblong depression 30 millimeters long, 20 millimeters wide, and 10 millimeters deep.

Another possible case of Osgood-Schlatter's disease is found in the miscellaneous long bones from Chicama, Peru (NMNH 265330-661). This specimen is a right tibia (Figures 383, 384) of a fairly small adult, probably a female. The archeological age is unknown. The diaphysis, distal metaphysis, and distal joint surface are normal. In the region of the tibial tubercle there is a large irregular spur, which projects toward the knee joint. Both medial and lateral to the spur are zones of periosteal reactive bone. These are well healed indicating that the condition was not active at the time of death. The proximal joint surface of the tibia is abnormal. The bony surface for the attachments of the cruciate ligaments is poorly defined. The lateral and medial joint surfaces are poorly delimited. The medial joint surface extends posteriorly creating an irregular surface. There is a sharply defined narrow depression 11 millimeters long by 3 millimeters wide and 4 millimeters deep on the anterior lateral edge of



FIGURES 383, 384.—Possible Osgood-Schlatter's disease of the tibial tubercle in a right tibia: 383, Anterolateral view showing bony spur; note defect (arrow) inferior to joint surface, which may have been the result of a crushing injury. 384, X-ray of mediolateral view of proximal tibia. (NMNH 265330-661.)

the lateral joint surface. Grossly this defect appears to have resulted from a crushing injury to this portion of the joint. Reactive bony spurs in the region of the tibial tubercle and reactive bone deposition are compatible with Osgood-Schlatter's disease.

Freiberg's Disease of a Metatarsal Head

PATHOLOGY.—This lesion involves the head of the second, less commonly the third, metatarsal. The lesion consists of subchondral necrosis and infarction of the metatarsal head, which may separate from the shaft, similar to the fragment in osteochondritis dissecans. The second metatarsal is usually the longest and bears the burden of pressure against the ground, especially when the heel is unduly elevated.

The lesion usually occurs in adolescents but may be observed in adults. Bilateral involvement occurs. There is a concave defect in the articular bony surface of the metatarsal head with a sclerotic base. The metatarsal is somewhat shortened, the head broadened transversally, and the distal portion of the diaphysis thickened, with disappearance of the usual necklike constriction (Köhler, 1923). There is a corresponding change in the configuration of the articular surface of the adjacent basal phalanx.

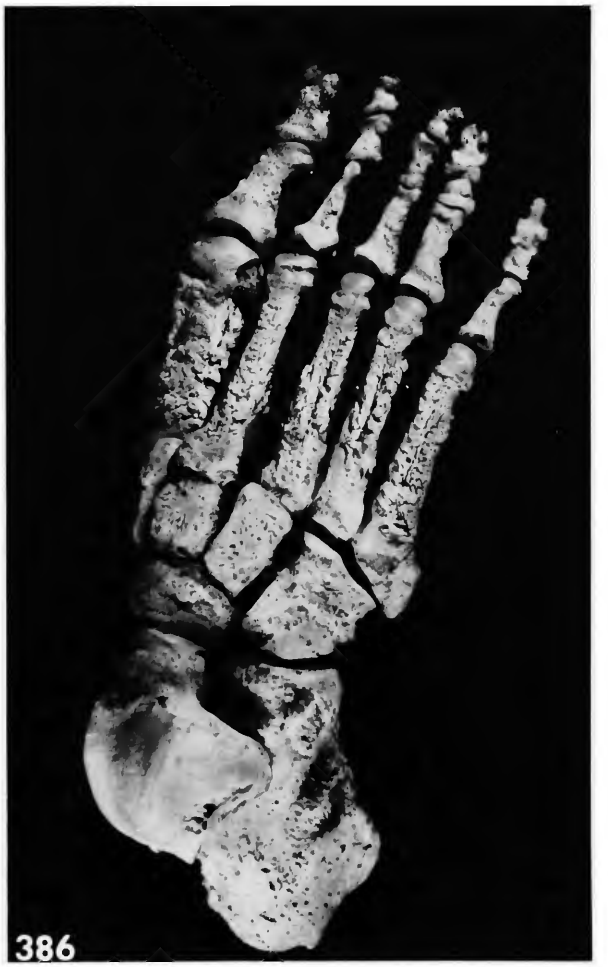
Kienböck's Disease of the Carpal Lunate Bone

PATHOLOGY.—The carpal lunate forming the center of the proximal carpal chain bears the brunt of the mechanical impact transmitted to the radius. The disease consists of necrosis and fragmentation of this bone, mainly on a basis of chronic trauma. The condition is not rare, more common in males, occurring between 20 and 40 years of age, with predilection of the right hand. Radiologically, areas of increased density and lucency alternate, and in the dry state bone areas may actually be fragmented after disappearance of interposed fibrous and cartilaginous tissue. More rarely the carpal navicular bone may be similarly affected (Preiser's disease).

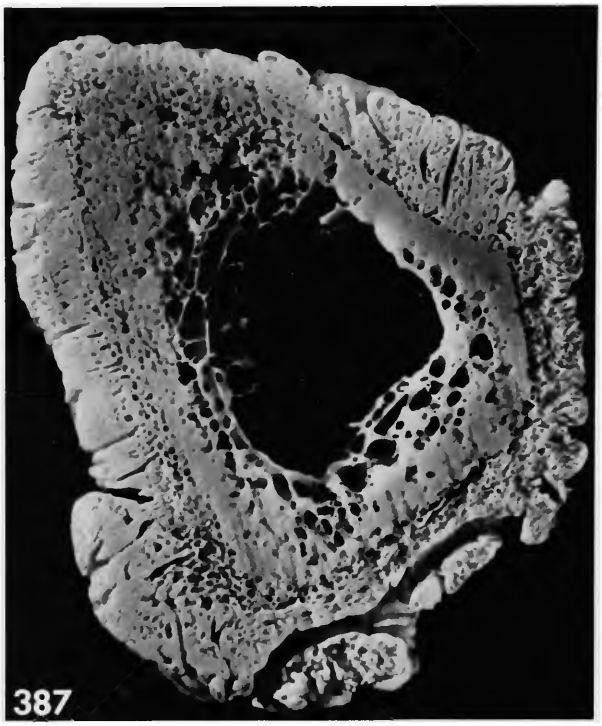
Pulmonary Osteoarthropathy

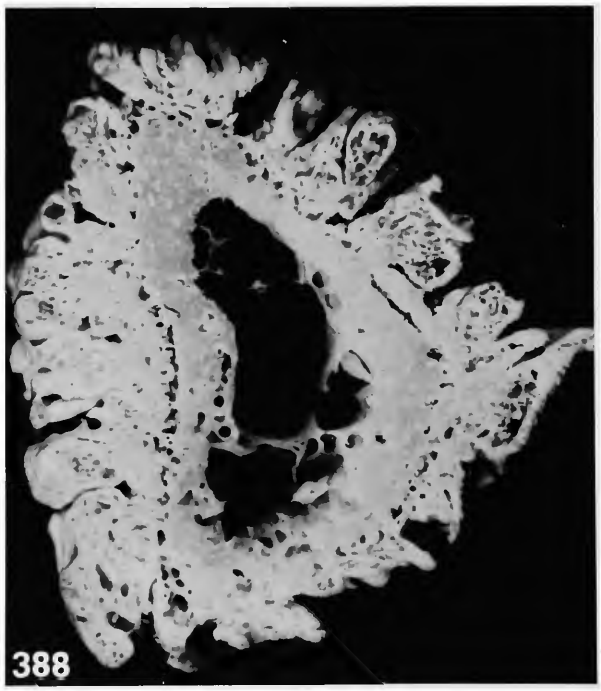
Pathology

Pulmonary osteoarthropathy is a somewhat obscure affection of the skeleton in persons with inflammatory or neoplastic, primary or metastatic, pulmonary lesions. Rarely pleural, cardiac, or mediastinal lesions may also lead to this condition. Although the skeletal involvement is initiated by a lymphocytic infiltration of the periosteum, it is presently thought that altered neurocirculatory reflexes are probably the underlying cause. In the living patient, the skeletal changes often disappear after removal of the pulmonary lesion. The concomitant arthritic changes and the clubbing of the fingers and toes only affect



Figures 385, 386.—Pulmonary osteoarthropathy: 385, Right radius and ulna showing marked, diffuse periosteal hyperostosis with pronounced vascular grooving. 386, Right foot, dorsal view; note periosteal hyperostosis of metatarsals and phalanges. (58-year-old female with large, solitary metastasis of breast cancer in lung; IPAZ 6649, autopsy 1259 of 1961.)





FIGURES 387, 388.—Pulmonary osteoarthropathy: 387, Cross-section of tibia. 388, Cross-section of fibula, enlarged to show periosteal bone deposition and cortical bone resorption. (58-year-old female with large, solitary metastasis of breast cancer in lung; IPAZ 6649, autopsy 1259 of 1961.)

the soft tissues and, therefore, need not be discussed here.

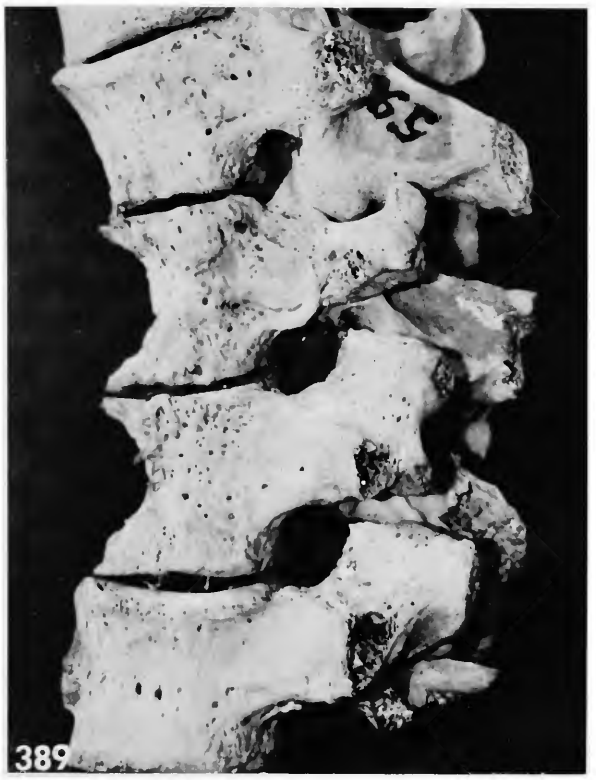
The skeletal lesion consists of a strictly symmetrical periosteal bone deposition on the shafts of the long bones of the extremities. The new bony shell is usually thickest at middiaphysis, tapering towards the metaphysis and sparing the bony ends and tendon insertions. The bones most frequently and most severely involved are the bones of the forearms (Figures 385–388) and lower legs (radius, ulna, tibia, and fibula). Femur and humerus show less involvement. Of the small tubular bones of hands and feet, the metacarpals and metatarsals are more affected than the phalanges (Figure 386), usually leaving the terminal phalanges, in spite of the clubbing, unchanged. The bones of the trunk rarely are affected and the skull is unchanged, except, infrequently, for bone deposition on the inner table (Jaffe, 1972: 290). The new bone is first fibrous, later remodeled into lamellar bone. It may be separated from the old cortex by a thin, fibrous layer. Its surface, on the dry bone, presents a rough aspect resembling a tree bark. It may be several millimeters thick at middiaphysis. In the late stages, the underlying cortex shows resorption with widening of the Haversian canals (Figures 387, 388) and, at times, endosteal resorption with widening of the medullary cavity. The carpal and tarsal bones merely show osteoporosis (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952 (1): 485).

Aneurysmal Erosion

Pathology

Bone is very sensitive to closely applied, pulsating pressure, as indicated by normal and abnormal vascular grooves on bony surfaces. An extreme case in point is the effect of the long-standing proximity of a large saccular dilatation of an artery (aneurysm) to bone.

The aortic arch is located immediately behind the manubrium of the sternum. Aneurysms in this area, mostly of syphilitic origin, can cause an erosion of varying depths on the posterior surface of the manubrium. Complete round perforation



FIGURES 389, 390.—Erosion of vertebral bodies by aortic aneurysm, probably arteriosclerotic; note the multiple deep scalloping defects partly showing a thin layer of reactive bone: 389, Lateral view. 390, Anterior view. Note the resistance to erosion of the areas adjacent to discs. (FPAM, Jubiläumspital 593.)

of the manubrium can occur. The descending aorta is closely attached to the vertebral column. Aneurysms of this portion, usually of arteriosclerotic etiology, may erode several vertebral bodies anteriorly. Since cartilage is not resorbed as readily as bone in response to pulsating pressure, deeply scalloped resorption defects are seen on several adjacent vertebral bodies while the endplates adjacent to the intervertebral discs are better preserved (Figures 389, 390).

The internal mammary arteries are closely attached to the posterior surface of the ribs, near the osteochondral junction. In congenital extreme narrowing of the aorta below the left subclavian artery (coarctation), the internal mammary arteries show marked compensatory dilatation. In this condition deep pressure grooves are seen on the ribs, near the osteochondral junctions.

Reticuloendothelial and Hemopoietic Disorders

In various disorders of the reticuloendothelial system, bone changes occur either due to accumulation of abnormal metabolites or abnormal proliferation of histiocytic cells.

Lipid Storage Diseases

Pathology

Gaucher's Disease

Gaucher's disease represents an inborn, genetically controlled, abnormality of lipid metabolism, in which abnormal cerebrosides are increasingly accumulated in histiocytes of the reticuloendothelial system, especially in the spleen, liver, lymph nodes, and bone marrow. The disease manifests itself in about one-third of the individuals in affected families. Both sexes are equally involved. The frequency of Gaucher's disease is about 30 times as high in Jews of central or eastern European descent as in Mediterranean Jews or in other populations (Jaffe, 1972:508). Although the disease may start in a family as a mutation, usually it seems to be transmitted as an autosomal recessive trait. Although the main involvement is extraskeletal, we are here only concerned with the bone changes.

The bone changes are due to continued accumulation over many years of cerebroside-laden histiocytes in the bone marrow. This accumulation may be diffuse or in the form of nodular aggregates. The deposition, although widespread throughout the skeleton, does not involve all bones equally. Diffuse infiltration in hemopoietically active bones (vertebrae, sternum, ribs, pelvis, and cranial vault) leads to widening of the marrow spaces and reduction of the number and diameter of bone trabeculae, giving a coarse, spongy appearance on X-ray and in dry bone. The cortices also appear thinned, but the periosteal surface remains smooth. This osteoporosis has no specific characteristics. The weakened porotic bone is subject to compression fractures in weight-bearing areas. This is especially observed in the lower thoracic and lumbar portion of the vertebral column. Anterior wedging with collapse and resulting kyphosis occurs (Pick, 1927:20–21).

In severe cases, changes in the femoral head are common. These manifest themselves as flattening of the curvature of the articular surface with lateral bulging and diminished height. This change may be brought about by interference with the precarious blood supply by pressure of accumulated Gaucher cells or by infractions of the porotic spongiosa or both. The head may be actually driven into the neck, which then appears shortened on external inspection. This deformity of the femoral head later leads to early development of degenerative arthritis in the hip joint because of incongruity of the two articular surfaces. These changes are very similar to other forms of aseptic necrosis of the femoral head, especially that due to Perthes' disease. Pathologic fractures also occur, mainly in the proximal femur, vertebrae, and ribs (Greenfield, 1970). The shafts of long bones, most often of the femur, may become the seat of nodular and diffuse infiltration with Gaucher cells, resulting in widening of the marrow space with increased diameter of the bone. This most characteristic change is often observed on the distal femoral metaphysis. Instead of the usual concave flare, it shows a straight contour or even slightly bulging outline (so-called Erlenmeyer flask deformity). This can be due, in part, to inhibited remodeling during the growth period or actual expansion of the thin metaphysial cortex in adult life.

The nodular infiltration, in addition to coarse porosis, may also lead to complete trabecular resorption in the affected area, giving a lytic appearance on X-ray and a cystic defect in dry bone. Characteristically, the diaphysial cortex

may show lamination and thinning, secondary to invasion of the Haversian canals by Gaucher cells (Pick, 1927:23). If this process is severe, a minimal amount of subperiosteal bone deposit can be observed, but usually the external cortical lamellae, although thin, remain smooth. Similar changes may be observed on the proximal tibial metaphysis or on the humerus, albeit less often. In long-standing cases, severe destruction of the proximal humerus with varus deformity is not uncommon. In these late cases, lytic skull and mandibular lesions and focal reactive sclerosis may occur (Rourke and Heslin, 1965). Bone marrow infarcts have been observed in various long bones (Arkin and Schein, 1948).

Niemann-Pick's Disease

Niemann-Pick's disease is a rare, congenital, familial disorder of the phospholipid metabolism, leading to progressive storage of phospholipids (mainly sphingomyelin) in the reticuloendothelial cells of the liver, spleen, lymph nodes, and bone marrow. There is no sex predilection and the inheritance seems to be recessive. About 50 percent of the affected individuals are Jews. The majority of the individuals die in infancy but survival to young adult age has been observed (Crocker and Farber, 1958:2–4).

Bone changes are mostly limited to reduction of number and size of trabeculae and cortical thinning of long bones. Occasionally, moderate widening of the distal femoral metaphysis is observed, not reaching the extent often seen in Gaucher's disease (Crocker and Farber, 1958:82). The generally poor health of affected infants may manifest itself in delayed appearance of secondary ossification centers. Superimposed deficiency in Vitamins D and C may add features of rickets and scurvy to the picture. In contrast to Gaucher's disease, collapse of the femoral head and focal lytic lesions do not occur (Gildenhorn and Amromin, 1961).

Other Lipidoses

Essential familial hypercholesteremia is a disorder of the cholesterol metabolism, inherited as a Mendelian dominant. The condition is characterized by elevated blood cholesterol levels and often leads to tumor-like deposits of cholesterol in subcutaneous and periarticular connective tissues or tendons (xanthoma tuberosum). There is no predilection of sex or race.

There seems to be no deposit of cholesterol in the bone marrow. Observed bone changes mainly consist of pressure erosions of phalanges of fingers and toes by adjacent xanthoma nodes (Merrill, 1920). This is apparently caused by the tightness of space in the subcutaneous compartments of digits, whereas much larger xanthomatous deposits in the vicinity of larger bones and joints do not usually indent the bones. The scooped-out cortical erosions may cross interphalangeal joints and simulate defects caused by gouty tophi (Gaál, 1933).

Lipid (cholesterol) granulomatosis (Erdheim-Chester's disease) is a rare disorder characterized by massive cholesterol deposition in the bone marrow and occasionally in other organs, but sparing the spleen. The condition remains asymptomatic into adult life. The reason for mentioning it here is that distinct bone changes have been demonstrated radiologically and anatomically (Chester, 1930–1931:577–592; Jaffe, 1972:537–541). The most marked lesions were found in the long bones of the forearms and lower legs, symmetrically. The changes consist of spotty and diffuse osteosclerosis, most pronounced in the metaphysial area but often also involving the epiphysis. The shaft cortex may be thickened but shows widened Haversian canals. The changes extend into the bones of hands and feet, most markedly so in the talus and calcaneus. Femur and humerus are less affected and the skull is normal. The sclerosis observed is due to both trabecular thickening and ossification of medullary cholesterol granulomas.

Histiocytosis X

Pathology

Histiocytosis X (Lichtenstein, 1953) is a term introduced to unite three different clinical mani-

festations of a non-neoplastic histiocytic lesion of unknown etiology: Letterer-Siwe's disease, eosinophilic granuloma of bone, and Schüller-Christian's disease. Although the clinical courses of these conditions are very different, transitions have been observed and the bone lesions are very similar, radiologically and histologically. The common denominator between the three conditions is proliferation of histiocytes in various tissues and organs. This may be diffuse or nodular, generalized or local, with varying admixture of eosinophilic leucocytes, lymphocytes, and plasma cells. Fibrosis and granulomatous aggregates of histiocytes, laden with neutral fat and some cholesterol, are present or dominant in chronic cases.

Letterer-Siwe's disease affects infants and children, mostly below 2 years of age. It involves most organs and tissues of the body and frequently takes an acute course with fatal outcome. Eosinophilic granuloma of bone represents the most common histiocytosis developing in children and young adults and is characterized by solitary or multiple bone lesions. The course is often self-limiting and benign. Schüller-Christian's disease is characterized by multiple bone lesions often occurring subsequently over periods of years in children, adolescents, and young adults. Lipid granuloma and fibrosis are an essential part of this condition. The ultimate prognosis is often unfavorable because of involvement of the brain, pituitary, lungs, or heart in some cases. Encroachment upon the pituitary often leads to severely retarded growth, approaching dwarfism (Chiari, 1931:430).

Although, as stated, the bone lesions are very similar in all three conditions, their distribution may be somewhat different. Any bone may be involved, but skull lesions are most common. Second in frequency are lesions in flat bones. Vertebrae, ribs, and long bones are not uncommonly involved. The small bones of hands and feet are usually spared. The lesion generally presents as a central, purely lytic, lesion without sclerotic margin or reactive bone formation. The small lesions are round or oval, later acquiring undulating or "geographic" borders.

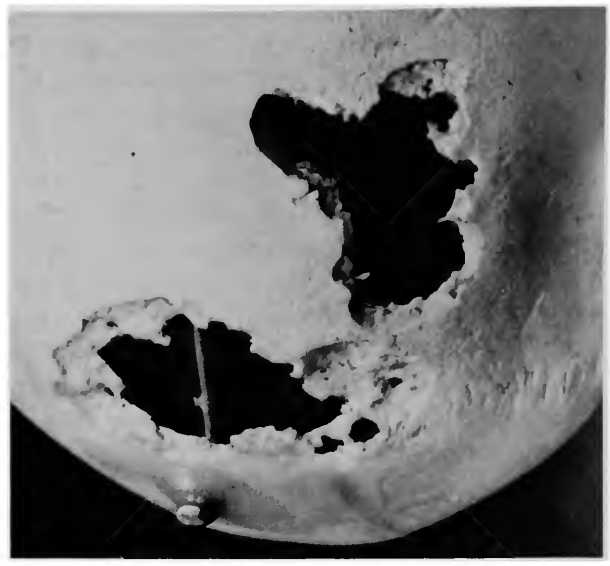


FIGURE 391.—Histiocytosis X, cranial vault with multiple penetrating defects showing geographic contours and little reactive bone. Multiple destructive lesions were found in many bones. (3½-year-old male, 1½ years known duration, IPAZ autopsy 1328 of 1955.)

In Letterer-Siwe's disease multiple skull lesions are usually present, involving the cranial vault and base, especially the sphenoid. In some cases facial bones are involved. In eosinophilic granuloma the most common lesion is a solitary, purely lytic, round or oval defect in an area of the cranial vault, showing a beveled edge and, occasionally, a central sequestrum. In Schüller-Christian's disease, large, multiple, confluent cranial defects are often seen (geographic skull) (Figure 391). The lesions of the cranial vault, even after destruction of both tables, are usually devoid of periosteal reactive bone. In eosinophilic granuloma, destructive focal involvement of the mandible is not uncommon, elevating the teeth above, creating the appearance of "floating" teeth on X-ray.

Lesions of the vertebral bodies, especially in small children, often lead to collapse, giving the picture of a vertebra plana.

The involvement of long bones is also primarily subcortical, mostly metaphysial, less common middiaphysial, and rarely epiphysial. If the overlying cortex is destroyed, subperiosteal reactive

bone formation does occur. Rib lesions may circumferentially erode the old cortex and expand the diameter, eliciting some sclerotic response in the new cortex. Pathological fractures can occur in long bones and ribs.

Anemias

Pathology

In various, mostly hemolytic, anemias bone changes occur secondary to the need for expansion of the spaces occupied by hemopoietic marrow (Caffey, 1937:293, 294).

Thalassemia

Thalassemia is a hemolytic anemia caused by a genetic defect in the hemoglobin structure. The severe manifestations of thalassemia major are usually restricted to homozygotes as regards the abnormal gene. Heterozygotes show thalassemia minor with little or no clinical, and certainly without skeletal, manifestations. Moderately severe cases occur (thalassemia intermedia), which may be due to a different abnormal gene. Recent advances in hemoglobin chemistry have shown that there are two major and several minor genetic abnormalities producing thalassemia. The classical thalassemia, affecting mainly central and eastern Mediterranean populations (Italians, Greeks, North Africans, but not French and Spaniards) is of the beta-type, characterized by the limited ability to synthesize the beta-chain of the globin molecule. The other large area of the world affected by thalassemia is India, Thailand, Malaysia, China, and the Philippines. In these areas, beta-thalassemia, but probably of a different genetic background, and alpha-thalassemia, in which the alpha chain synthesis is genetically inhibited, occur side by side. Less frequent instances of various types of thalassemia occur in a worldwide distribution in almost all ethnic and racial groups. The North and South American Indians, who apparently completely lacked a thalassemia gene, are an exception. All American Indian instances are due to admixture of White or Negro genes. A detailed discussion of the geographic, ethnic, and genetic problems of this disease complex has been given by Rucknagel (1966). Although extremely interesting, it is of little significance to the present problem since, regardless of genetic background, the clinical and skeletal changes are the same. A geographic distribution in different populations is given in Table 11 from Rucknagel (1966). The percentages

Table 11.—World distribution of alpha and beta thalassemia in indigenous populations (from Rucknagel, 1966) (dash = no data; ? = questionable data)

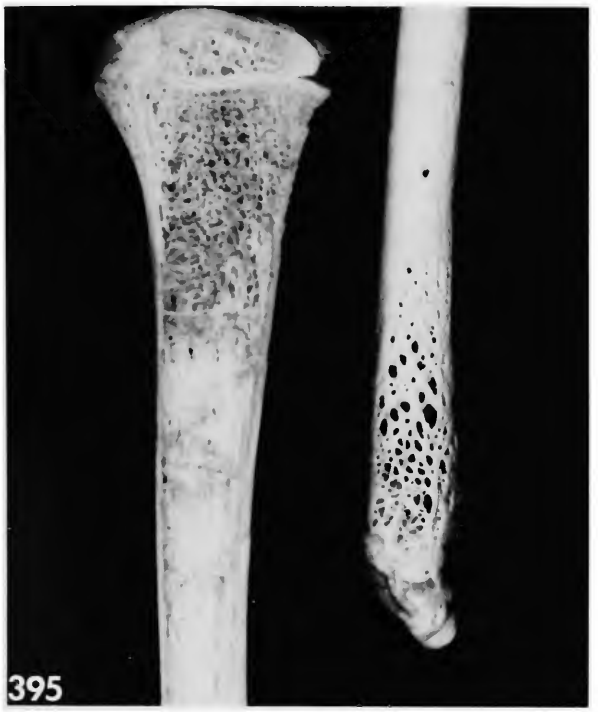
Geographic area	*Alpha* (%)	*Beta* (%)
Southern Europe		
Italy	0.2	0–20
Sicily	–	3–13
Sardinia	1	4–38
Greece	4–7	6–14
Portugal	–	0.6
Turkey	?	?
Africa		
Algeria		1–15
Morocco (Jews)		3
Egypt		0.3
West Africa	2–10	2
Middle East		
Israel (Samaritans)		14
Lebanon and Syria		5
Asia		
India (Madras, Castes)	–	15
Thailand (Valley)	3–5	5
Thailand (Hill tribes)	–	1–10
South Vietnam	–	2
China (southern)	–	3–5
Malaysia	1.5–2	
Indonesia	0.5	–
South Pacific		
New Guinea	–	0–8
New Britain	–	4–6
Philippines	?	1
Saipan		1
America		
North and South American Indians	0	0

FIGURES 392–394.—Thalassemia major; frontoparietal segment of cranial vault: 392, External view. 393, Endocranial view. 394, Cross-section showing widening of the diploic spaces and honey-combed buildup of subperiosteal bone replacing the outer and inner tables. (8-year-old Thai, studied by Putschar in 1962 at Department of Pathology, Medical School, Chiengmai, Thailand.)

quoted include homozygotes and heterozygotes, and it is understood that there is more than one genetic type of beta-thalassemia.

Thalassemia major results in a very high mortality in infancy and childhood and a limited adult life span in the few survivors. The heterozygote without anemia presumably has a selective advantage in malaria-infested areas, which would explain the geographic distribution. However, the evidence is more circumstantial and less clear-cut than in sickle cell anemia (Rucknagel, 1966). In thalassemia major, bone changes always develop and become radiologically obvious after the first year of life. Intermediate thalassemia may show skeletal lesions; thalassemia minor does not. The bone lesions in thalassemia are entirely due to increased spatial demands for the hyperplastic erythropoietic marrow. In keeping with the general distribution of hemopoietic marrow, the entire skeleton of the child is affected, more or less uniformly, while, in the adult, characteristic bone changes remain only in areas of permanent hemopoietic activity.

The most severe changes are in the skull. In children, the diploë of the cranial vault expands, leading to marked thickening of the cranial vault, usually beginning in the upper portion of the frontal bone (Figures 392–394). The trabeculae of the diploë are reduced in number with thickening and later radial rearrangement of the remaining trabeculae. The external table is progressively eroded and later completely destroyed. This is accompanied by honeycombed compartments of subperiosteal new bone, harboring hyperplastic marrow. The destruction of the inner table comes much later and is always much more limited than that of the outer table. On X-ray, in these severe cases, the honeycombed compart-



ments give the appearance described as "hair on end." The facial bone, especially the maxilla and zygoma, are expanded in their external dimensions, showing thin cortices and loose spongiosa, causing a mongoloid appearance with prominent cheek bones. Development of maxillary and sphenoid sinuses as well as of the mastoid cells is inhibited and delayed (Caffey, 1957a, 1972 (1): 89). Ethmoid sinuses are better developed, since there is little marrow in these bones (Baker, 1964). The involvement of the maxilla and, to a lesser extent, of the mandible leads to disorderly eruption of the teeth and malocclusion of the jaws. In general, the reduction of the total mass of trabecular and cortical bone leads to arrangement of residual trabeculae along stress lines to give maximal mechanical stability with minimal encroachment on marrow space. This is most pronounced in ribs, where the cortex can be completely missing and the trabeculae show diagonal arrangement with right angle crossings (Figures 395, 396). There is a linear reinforcement on the concave surface in response to respiratory bending stresses. The diameter of the ribs is expanded. Similar expansion and alteration of the trabecular pattern are seen in flat bones (pelvis, scapulae) showing a fan-like pattern on X-ray.

In addition to porosis, the vertebrae show decreased height, increased width, and cupping of the end-plates. Actual compression fractures occur, especially in the lower dorsal and lumbar areas (Caffey, 1972 (2):1393). In children, metacarpals, metatarsals, and phalanges show expansion, diagonally crossing trabeculae, and reticulated thin cortices.

Long bones of the extremities show marked widening of the medullary cavity accompanied

FIGURES 395–396.—Thalassemia major: 395, Longitudinal cut (*left*) of proximal tibia showing widening of epiphysial and metaphysial marrow spaces and of medullary cavity, marked thinning of cortex, and lines of arrested growth. External view (*right*) of distal radius with lace-like reduction of metaphysial cortex. 396, Rib, endothoracic surface, showing extreme lacey cancellization of cortex. (8-year-old Thai, studied by Putschar in 1962, Department of Pathology, Medical School, Chiengmai, Thailand.)

by thinning of the cortex, most pronounced in the femur. In the metaphysial area, the cortex may assume a reticulated appearance with markedly enlarged vascular foramina, harboring hyperplastic marrow. There is often inhibited remodeling of the distal femoral metaphysis, leading to a widened contour without the usual concave flare, closely resembling the so-called Erlenmeyer flask deformity seen in Gaucher's disease. Multiple "lines of arrested growth" are frequently present, in keeping with the usual inhibited growth and delayed epiphysial closure observed in this disease. In a smaller percentage, premature and irregular fusion of the growth plate occurs, especially of the proximal humerus, leading to significant medial tilting of the humeral head. In the series published by Currarino and Erlandson (1964), 12 of 79 thalassemia patients showed premature fusion of growth plates beginning after 10 years of age and producing shortening and deformity. Of these, nine concerned the proximal humerus (three bilateral), three the distal femur (one bilateral). The general reduction of growth may lead to an infantilistic, dwarfed skeleton (Caffey, 1972 (2):1284).

In adults, the tubular bones of hands and feet resume normal contours and may develop some osteosclerosis subsequent to the replacement of erythropoietic by fatty marrow (Caffey, 1972 (2): 1284). The widening and reticulation of the ribs remains mostly in the posterior portions and rarely may lead to a tumor-like expansion of erythropoietic marrow covered by a thin, expanded, cortical shell, projecting into the thoracic cavity. Changes in the vertebrae and, to a lesser extent, in the cranium remain. Pathological fractures, especially of the femur, occur in adults more frequently than in children.

Double heterozygous combinations with other hemoglobin abnormalities occur, but only in thalassemia-hemoglobin S combinations are bone infarctions observed (Middlemiss and Raper, 1966: 698). Pure thalassemia creates no circulatory obstructions and does not have an increased rate of osteomyelitis. In the differential diagnosis of dry skeletal parts, other hemoglobinopathies, congenital spherocytic anemia, and iron deficiency anemia have to be considered. The skull changes of thalassemia occasionally resemble those in sickle cell anemia but the extensive lesions of the rest of the skeleton do not. Cyanotic congenital heart disease (Ascenzi and Marinozzi, 1958) and, rarely, polycythemia (Dykstra and Halbertsma, 1940) can mimic the skull changes in thalassemia. Congenital hemolytic anemia and iron deficiency anemia never show bone changes of such severity.

Sickle Cell Anemia and Its Genetic Variants

A variety of normal and abnormal hemoglobins can be identified by electrophoresis. At birth, fetal hemoglobin F amounts to about 60–80 percent of the total and remains in significant amounts until about 6 months of age. It is rapidly replaced by adult hemoglobin A to a level below one percent, but there exists genetic abnormal persistence of hemoglobin F. Hemoglobin S is an abnormal hemoglobin, responsible for the sickling phenomenon. Another abnormal hemoglobin is C. A further abnormality of hemoglobin is caused by the thalassemia genes (Thal). All these genetic factors are realized in the various genotypic combinations of any two of them.

Sickle cell anemia patients have a genotype SS while individuals with sickle cell trait have genotype SA. The gene for hemoglobin S is almost exclusively limited to the Negro race with a slight scattering in adjacent Mediterranean populations (southern Italy, Greece, Armenia). Other genetic combinations (SC, and SThal) represent the genetic variants of sickle cell anemia. In persons with two abnormal hemoglobin genes, hemolytic anemia will develop in intermittent crises, and the erythrocytes will assume sickle shape under hypoxemic stress, due to crystallization of the abnormal hemoglobin within the cell. The result is constantly increased demand for red cell production to compensate for the cell loss in hemolysis. During crises, the misshapen red cells conglutinate and cause vascular obstructions, leading to areas of ischemic necrosis. Although the genetic abnormality is present at birth, the clinical man-

ifestation is prevented before 6 months of age due to the protective effect of the high concentration of fetal hemoglobin F before that age. The mortality of infants and children with sickle cell anemia is high.

In persons with sickle cell trait (SA), only extreme hypoxemic stress can produce hemolysis and infarctions. Persons with the genetic combination of sickle cell trait and persistent fetal hemoglobin (SF) are usually free of anemia. Of considerable interest is the fact that hemoglobin S is more resistant to attack by malaria parasites. This offers survival advantage to carriers of hemoglobin S, which in turn leads to increasing frequency of hemoglobin S in inhabitants of the malaria infested areas of tropical Africa. The frequency of hemoglobin S varies in different Negro populations. In North American Negroes, 7.5 percent show the sickling trait but only 1 in 40 (2.5 percent) of these suffers from sickle cell anemia (Lewis, 1942:234–235). Of course the American Negroes are a dislocated group of mixed genetic background. In Panama, Tomlinson (1945) found at autopsy that 9 percent of individuals with sickling trait actually showed clinical sickle cell anemia. The sex distribution rate of the sickling trait in 3000 determinations in Panama (representing autopsies, hospital patients and village populations), was 7.2 percent in males and 11.4 percent in females. In spite of a high frequency of hemoglobin S in African Negroes, clinical sickle cell anemia is uncommon. A survey of over 2000 Yorubas in Ibadan (Nigeria) showed the following distribution of hemoglobin genes: normal 70.9 percent, carriers of AS 23.9 percent, AC 4.3 percent, SC 0.7 percent, and CC 0.2 percent (from Cockshott, 1958).

The skeleton may be involved in sickle cell disease in three different ways: (1) changes secondary to increased demand of space for hemopoietic marrow; (2) sequelae of vascular obstruction of smaller and larger blood vessels; and (3) secondary infections superimposed on ischemic areas. Although a variety of bone changes have been described in sickle cell anemia, it should be emphasized that, as a rule, they are neither common nor obvious nor specific. The most careful and detailed study of bone changes in this disease has been published by Diggs, Pullian, and King (1937) in a series of their own cases. Their series comprised 39 Negro patients with active severe sickle cell anemia, ranging from 1 to 51 years of age. This includes eight autopsies at which special attention was paid to bone changes. The frequency and degree of bone involvement increased with age beginning in children past 5 years of age. The bones most likely to reveal alterations are the skull, vertebrae, tibia, and fibula, with the skull changes appearing the earliest. They found in their series that the majority of patients showed no alteration in size, shape, and density of their bones on X-ray. There is general agreement among all observers that bone changes are not related to the severity of the disease.

The first and most conspicuous changes are observed in the skull. The diploë may be thickened, and there may be diminished definition of the outer table on X-ray. The radiation density is diminished, sometimes with coarsened trabeculation arranged perpendicular to the inner table. Complete destruction of the outer table with so-called "hair on end" arrangement of bony septa is uncommon. It was present in only 1 of 39 cases in the series of Diggs, Pullian, and King (1937). They found the thickening of the diploë to be bilateral, symmetrical, and usually limited to the parietal bones, less often involving the frontal and occipital bones. The maximal thickness occurs in the vertex, tapering to normal towards the temporal area. Skull changes are most often observed in older children and in young adults. Occasionally the osteoporosis may lead to small focal lytic lesions resembling myeloma. The facial bones showed normal appearance on X-ray in their cases. Sarjeant (1974:166) states that the zygoma may be enlarged and the orbital roof thickened, but the frontal sinuses develop normally, although the other sinuses may be delayed and hypoplastic. In the mandible, reduction and coarsening of trabeculae is observed, accompanied by prominence of the lamina dura of the alveoli and thinning of the cortex. The vertebrae

show rarefaction of the spongiosa in the vertebral bodies. In adults, depression of the central portion of the end-plate with intact peripheral portion is observed, which is somewhat different from the "fish vertebra" appearance in postmenopausal osteoporosis. Widening and flattening of vertebral bodies, especially in the lumbar area, is seen in about 30 percent of adult patients (Jaffe, 1972: 700). The ratio of height to width of vertebral bodies in anterior-posterior views on X-ray ranged from 1:1.4 to 1:2.8 with a normal of 1:1.4 for Negroes of comparable age (Diggs, Pullian, and King, 1937).

The scapula, sternum, pelvic bones, and ribs (flat bones) may show increased lucency and coarsening of the trabeculation. However, in the series reported by Diggs, Pullian, and King (1937), only slight osteoporosis without widening of the bones was observed.

In long bones, the most marked changes occur in the tibia and fibula. Hemopoietic marrow may occupy Haversian resorption spaces in the cortex, especially near the medullary canal (Diggs, Pullian, and King, 1937). In adults with advanced disease, endosteal reactive lamellar bone and occasional bony plugging of segments of the marrow cavity can occur (Diggs, Pullian, and King, 1937). In these cases neither the medullary cavity nor the diameter of the shaft is widened. On the contrary Diggs, Pullian, and King (1937) found the cortex thickened and the medullary cavity actually narrowed. In their series they found 11 cases, mostly adults, with marked changes of long bones. The hyperplastic marrow also may produce multifocal spotty radiolucency in the metaphysial areas. The medullary cavities of metacarpals may be widened and vascular foramina enlarged (Cockshott, 1958). The changes produced by sickling and blocking of blood vessels are essentially ischemic infarctions, which may be large and located in the medullary cavity, small and spotty in the metaphysis, or involving portions of shaft cortex due to blockage of Haversian vessels. Generally, the large medullary infarcts are indistinguishable from those produced by other causes. As in those, they also tend to collect calcium salts around the necrotic focus, which is readily visible on X-ray. The metaphysial foci may appear as focal increased densities alternating with spotty lucencies of hyperplastic marrow. In fact, some of the bony plugging of the medullary cavity, as described by Diggs, Pullian, and King (1937), may well represent healed infarctions. The cortical infarcts may reveal a lytic intracortical separation on X-ray. Infarctions of the cortex of short tubular bones of hands and feet are not uncommon in infants and small children. These may elicit moderate periosteal reactive bone formation and spotty densities and lucencies in the course of repair (Jaffe, 1972:697–703). However, Diggs, Pullian, and King (1937) found no bone changes in hands and feet in their cases, but in some instances epiphysial necrosis may lead to shortening of individual fingers (Sarjeant, 1974:175–177).

Of special significance is the occurrence of aseptic necrosis in the head of the femur in sickle cell anemia and its genetic variants. This is the most common site of infarction. Apparently, it is even more common in the variants, particularly in SC patients, than in classical SS anemia (Cockshott, 1958). Golding, MacIver, and Went (1959) reported from Jamaica only 5 instances in 51 SS patients, but 13 in 19 SC and in 1 of 2 SThal patients. Chung and Ralston (1969:38) quote the incidence of aseptic necrosis of the femoral head from a literature survey as 0–12 percent in SS but 20–60 percent in SC disease. The lesion occurs mostly in adolescents near the time of fusion of the proximal growth plate. It resembles Perthes' disease, however with two differences: The lesion occurs several years later and it lacks metaphysial changes adjacent to the growth plate. Furthermore, Perthes' disease is uncommon in Negroes (Chung and Ralston, 1969:51). If the necrotic focus is small, hugging the base of the articular cartilage, it may simulate an unusually large osteochondritis dissecans or Perthes' disease (Middlemiss and Raper, 1966:695). In a series of 13 cases reported by Chung and Ralston (1969: 38), four showed bilateral hip lesions and six of these patients also showed aseptic necrosis of the

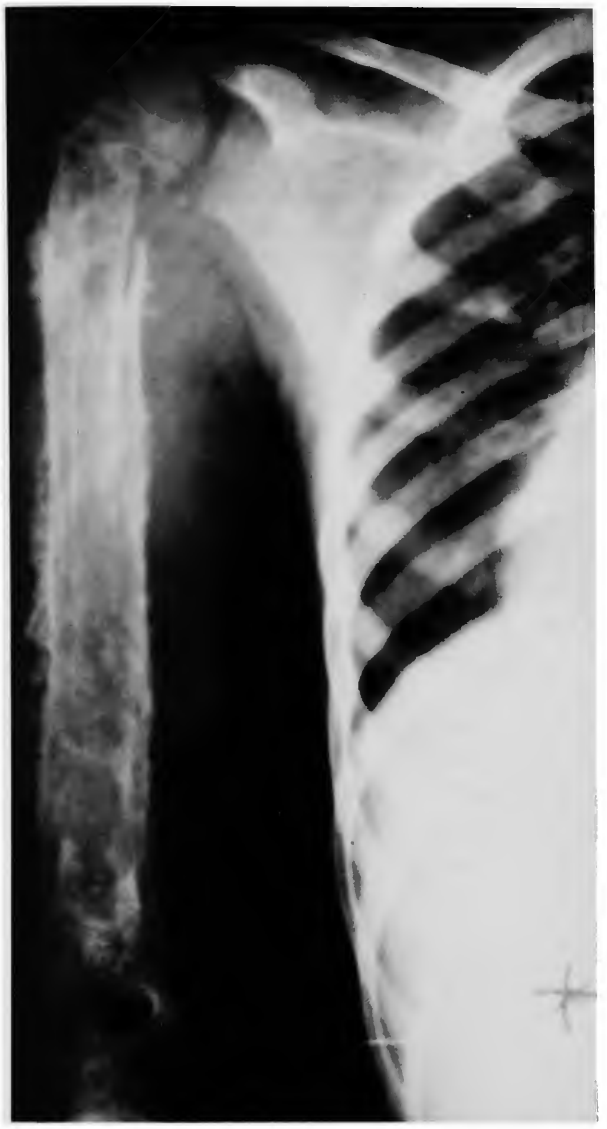


FIGURE 397.—X-ray of right humerus, of sickle-cell anemia with *Salmonella* osteomyelitis. (Nigerian negro, courtesy Dr. Stanley Bohrer, Ibadan, Nigeria.)

humeral head; others had infarcts in long bones. These additional necrotic foci mostly concerned older patients with classical (SS) sickle cell anemia rather than genetic variants. Occasional unusual locations of aseptic necrosis, such as elbow, knee, sacroiliac joint, and ankle, have been observed in SS disease (Chung and Ralston, 1969: 40). Secondary infections are not uncommonly superimposed on ischemic necrotic foci, which permit colonization of bacteria if circulating in the blood stream in only small numbers. In these infections, intestinal pathogens, especially strains of *Salmonella*, make up an unusually large percentage (Figure 397). As in ordinary types of osteomyelitis, the metaphyses of growing long bones are predilected. In contrast to purulent osteomyelitis, there is often destruction of the growth plate, which may lead to Coxa vara at the hip joint (Middlemiss and Raper, 1966:696). Pathological fracture through the osteoporotic bone also occurs.

In general, sickle cell anemia diminishes the rate of growth and frequently delays the closure of growth plates. For additional statistical and anatomical details consult Diggs (1967) and the recent survey of Sarjeant (1974:165–192).

Hereditary Spherocytosis (Congenital Hemolytic Anemia)

This is a genetically determined hemolytic disorder, characterized by the globular shape of the erythrocytes. It is uncommon in Negroes (Kline and Holman, 1957). The disease manifests itself at various ages. Bone changes are uncommon and slight, usually limited to the cranial diploë, rarely affecting long bones. The cases with onset in infancy or early childhood may show the most marked changes (Moseley, 1963:10–11).

Iron-deficiency Anemia

Bone changes in iron-deficiency anemia are uncommon. If such changes occur, they are usually limited to the skull vault, showing expansion of the diploë, vertical striation and occasional "hair on end" appearance on X-ray (Moseley, 1961, 1963, 1966; Lanzkowsky, 1968). The patients reported as showing the bone changes mentioned have been Indonesian (Eng, 1958), Negroes (Burko, Mellins, and Watson, 1961), or prematures and twins (Shahidi and Diamond, 1960). The facial bones and long bones are usually spared and pneumatization of the paranasal

sinuses is not delayed (Moseley, 1963:5). However, in some Turkish patients, enlargement of the facial bones, delayed pneumatization, and coarse porotic trabeculation around elbows, hands, and feet have been observed (Aksoy, Çamli, and Erdem, 1966).

In other conditions, such as cyanotic congenital heart disease with secondary polycythemia (Ascenzi and Marinozzi, 1958) or in polycythemia vera (Dykstra and Halbertsma, 1940), expansion of the cranial diploë due to hyperplasia of the hemopoietic marrow has been described.

Erythroblastosis Fetalis

In some infants with this disorder, usually based on RH incompatibility between the RH negative sensitized mother and the RH positive fetus, bone changes have been described. They consist of alternating band-like areas of increased and decreased radiodensity, especially in the metaphysis of the most rapidly growing bones (distal femur and proximal tibia). These changes were present in 20 of 110 cases (Brenner and Allen, 1963) but are not specific. They reflect disturbances of the late fetal enchondral ossification with delayed resorption of calcified cartilage cores in primary trabeculae (Follis, Jackson, and Carnes, 1942). The only reason to mention this finding is that similar changes can be seen in the newborn in congenital syphilis.

Paleopathology

Because of the problems in differentiating various types of anemia in dry bone specimens, I shall discuss here the paleopathology of all the anemias. This, of course, does not preclude an attempt to establish a probable specific diagnosis for a given case but does serve to emphasize the importance of other variables, such as ethnohistorical data on diet and endemic disease, in reaching a diagnosis.

An important basic distinction in the anemias is between genetic and acquired anemia. Some of the genetic anemias, such as sickle cell anemia and possibly thalassemia tend to be influenced by the distribution of malaria, since the abnormal hemoglobin in such anemias reduces the severity of the symptoms of malaria. Acquired anemia is a response to one or more of several variables including nutrition and debilitating diseases and thus occurs in any human population irrespective of the presence of malaria.

Most of the inferences made about the presence of anemia in paleopathological specimens are based on lesions of the skull thought to be associated with this disease. The lesions are found on the skull vault, primarily the outer surface of the parietal bones, and on the orbital roof (Figure 398). They consist of porous periosteal bone deposition in these areas of the skull. Henschen (1961: 724) attributes the earliest awareness of these lesions in clinical cases to Rokitansky (1844) and Virchow (1848). Several descriptive terms have been used for this condition including cribra crania, symmetrical osteoporosis, and spongy hyperostosis. I shall use the term "porotic hyperostosis" introduced by Angel (1966).

The relationship between the calvarial and orbital lesions has never been clear. Virchow (1874: 61–62) believed that both lesions are part of the

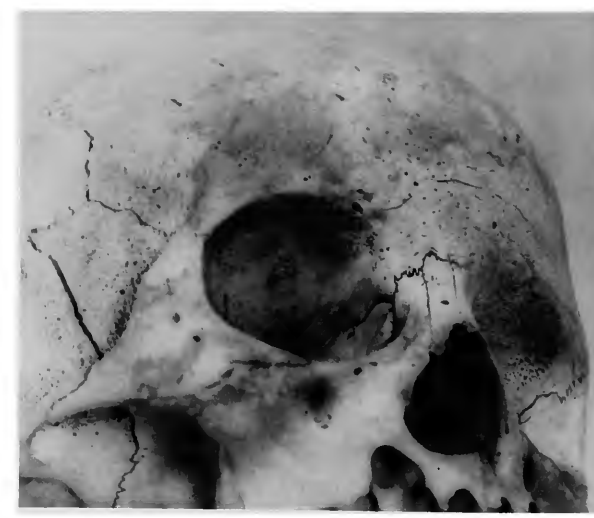


FIGURE 398.—Porous, reactive bone on the orbital roof. (Child about 12 years of age from Pachacamac, Peru, NMNH 266599.)

same disease process. Hrdlička (1914) indicated that the calvarial lesions begin in the orbit. Møller-Christensen and Sandison (1963:180) note the possibility of lacrimal gland involvement in orbital lesions and suggest mumps as a possible cause. Local inflammation of the orbital roof from such a cause probably would not affect the cranial vault. The implication of this opinion is that orbital lesions are not necessarily associated with calvarial lesions. My own experience is that one often finds both types of lesion in a single skull, but one also finds each type of lesion occurring by itself and occasionally in association with morbid conditions other than anemia.

Several morbid conditions other than anemia have been proposed as the cause of porotic hyperostosis. Williams (1929:854–855) suggested rickets as the most likely cause but also suggested that abnormal conditions created by artificial deformation of the skull could produce similar lesions. Earlier, Hrdlička (1914:58) expressed the opinion that the condition was not rachitic but more likely a "toxic" rather than a nutritive or degenerative disorder. Henschen (1961:724, 729) examined 2000 modern skulls from Sweden and found no evidence of porotic hyperostosis. However, he did find four such lesions in some mid-nineteenth century Swedish skulls. He suggests that the lesions may have been caused by nutritional conditions or chronic infection.

While infectious and metabolic disease may be implicated in porotic hyperostosis of the skull, anemia has received much more attention in the paleopathological literature as a possible cause of this condition. Vyhnanek, Kolar, and Stloukal (1963) describe two medieval skulls of children from central Europe with porotic hyperostosis. The authors conclude that they are examples of hemolytic anemia but are not more specific. Zaino (1964:403) proposes that the porotic hyperostosis, which is common in pre-Columbian skulls from Peru, is due to thalassemia.

Jarcho, Simon, and Jaffe (1965) report a case of porotic hyperostosis from a Pueblo Indian site in the American Southwest. The authors conclude that bone changes suggest the presence of some type of hemolytic anemia, probably thalassemia, in the pre-Columbian New World. The problem in inferring the presence of thalassemia in the ancient New World from skeletal lesions is that there is no evidence of this gene in post-Columbian unmixed American indigenous populations.

Moore (1929) summarizes the modern clinical findings in sickle cell anemia, which include the thickened hair-on-end appearance in roentgen films. He briefly describes a modern skull with similar lesions. He attributes this example of porotic hyperostosis to sickle cell anemia and offers the opinion that the morbid condition was widespread. As with thalassemia, however, there is no evidence of this genetic blood abnormality in modern unmixed indigenous populations in the New World.

In the Old World, Angel (1964, 1966) has proposed that porotic hyperostosis found in ancient Greek skulls is evidence for the presence of thalassemia in antiquity. Here, however, the evidence for the genetic variant in modern ethnic groups is unquestioned, making such an inference much more plausible.

Moseley (1965) added acquired anemia to the list of possible morbid conditions producing porotic hyperostosis. On the basis of his clinical experience as a radiologist he proposed to differentiate thalassemia major from other genetic and acquired anemias because of its involvement of the face and long bones. He expressed the opinion (1965:141) that porotic hyperostosis seen in skulls from Peru and Yucatan is due to iron-deficiency anemia. This diagnosis was also applied to a Bronze Age (1650–1550 B.C.) skull of a 6-year-old child from Wales with porotic hyperostosis, described by Cule and Evans (1968). Sir Arthur Keith, who also saw the skull, attributed the lesions to rickets (in Wheeler, 1923:21). The published photographs, as well as Keith's description, also suggest congential syphilis as a possibility. The presence of hypoplastic enamel defects in the teeth is also compatible with a diagnosis of congenital syphilis but could also occur in anemia, particularly if related to poor nutrition.

El-Najjar, Lozoff, and Ryan (1975) studied the expression of porotic hyperostosis in archeological skeletal samples from the southwestern United States. In their study they found an association between presumed dietary factors and the frequency of skeletons having porotic hyperostosis. The authors conclude that acquired anemia due to inadequate iron absorption is the most likely causative factor for porotic hyperostosis in skeletons from the southwestern United States. Mensforth, Lovejoy, Lallo, and Armelagos (1978:38) report an association between evidence of infectious disease and the occurrence of porotic hyperostosis in prehistoric skeletons from Ohio, USA. They suggest that illness and nutritional stress are important factors stimulating iron-deficiency anemia.

Two specimens from the National Museum of Natural History, USA, provide useful insight into the expression of porotic hyperostosis. The two cases are both from the American Southwest (NMNH 327074 and 327107) and are skeletons of young children from the Pueblo Bonito ruin, Chaco Canyon, New Mexico. This site is associated with the Pueblo III cultural period and dated between A.D. 919 and 1130 (Seltzer, 1944: 25), thus making it clearly precontact. During this period skulls show marked artificial deformation characterized by occipital flattening. The dental age of the first specimen (NMNH 327074) is about 1½ years (Figures 399–402). The lesions in this specimen are primarily porous but with some labyrinth-like lesions as well. The affected area involves the frontals including the orbital roofs but predominantly involves the outer table of the parietal bones. The inner table is not affected. The specimen is characterized by severe occipital flattening and the lesion encroaches only slightly on the deformed part of the skull. The facial bones and mandible are not markedly affected, although the region surrounding the zygomaticofacial foramen on the zygomatic bone suggests an inflammatory reaction. The lateral X-ray film shows the perpendicular striations found in many examples of porotic hyperostosis. A postmortem break through the lesion in the right parietal reveals an intact inner table, greatly enlarged diploë with the virtual elimination of the outer table.

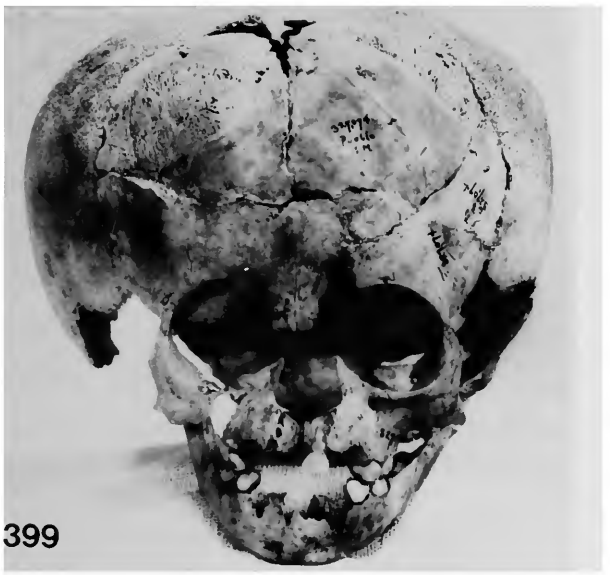
An X-ray film was prepared of the extant long bones including major portions of both femora, right tibia, both humeri, and ulnae. This film reveals a relatively enlarged marrow cavity and greatly diminished thickness of the cortex in all long bones.

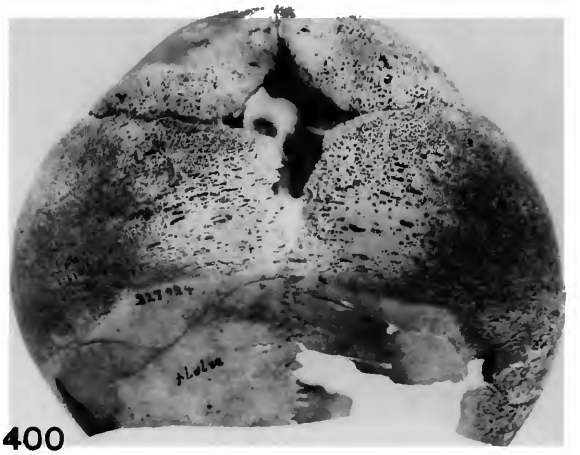
The second specimen (NMNH 327107) has a dental age of about 2 years. The state of preservation is not as good. Like the previous specimen the most severely affected region is the external table of the parietals (Figures 403, 404). The lesion does not cross the sagittal suture. The left frontal bone is also affected, with the disease process continuous across the coronal suture. Only portions of the right orbit are present. They reveal no evidence of porotic hyperostosis. The left temporal bone is abnormal with an irregular surface, which is thickened and slightly porous.

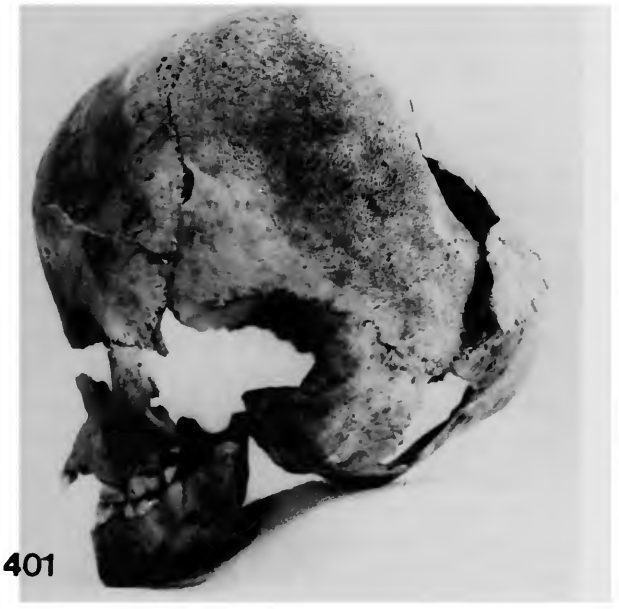
Two features distinguish this specimen from the preceding one. First, the lesion does occur in the region of deformation. This may be related to the fact that the occipital deformation is not as severe. Second, unlike the preceding specimen, there appears to be slight deposition of reactive bone on the inner table. The abnormal bone is concentrated in the region of the anterior fontanel with reactive tissue apparent at its sutural borders. There is a solitary lesion on the left parietal boss. A lateral X-ray film of the skull reveals the vertical striations in the area where porotic hyperostosis is most pronounced.

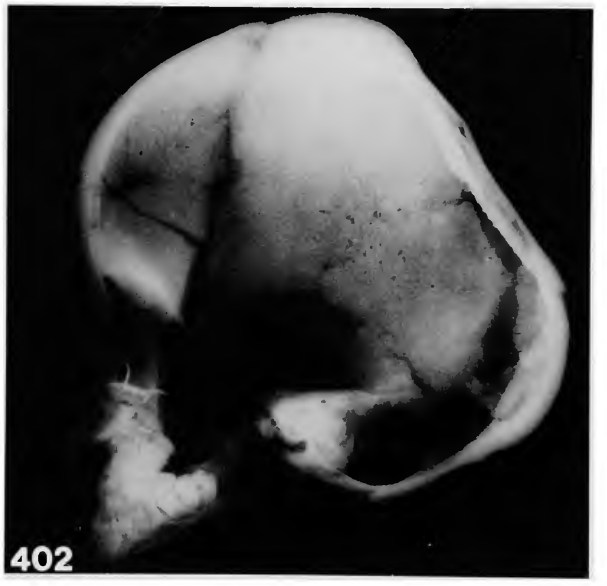
Also apparent in the film is the absence of the outer cortex in the porous area. The lesion itself is slightly different from the first specimen in being more variable in appearance. Beginning on either side of the sagittal suture, the parietal lesion has a narrow zone of finely porous bone. This quickly merges with bone that is labyrinthian in gross appearance. Continuing in a lateral direction the lesion is characterized by a circular porosity, which becomes finer and less pronounced in the area below the temporal muscle. Here the bone takes on the lumpy quality like that seen in the left temporal bone.

Like the first specimen, the long bones are





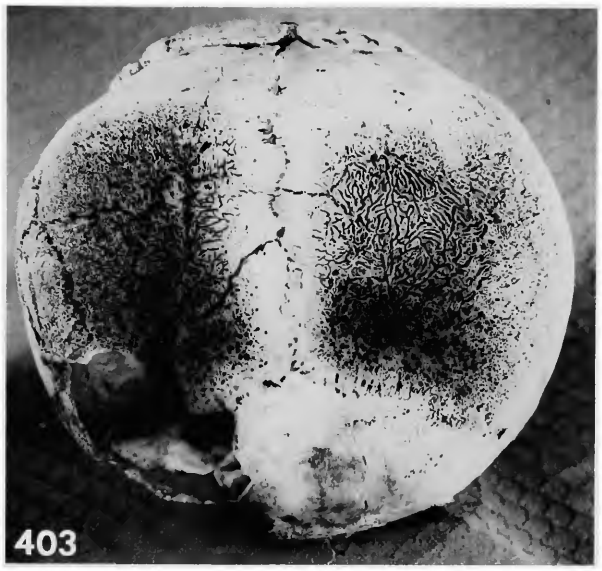




FIGURES 399–402.—Porotic hyperostosis of skull vault: 399, Facial view. 400, Porous lesions of skull vault. 401, Lateral view of skull. 402, X-ray of skull, lateral view. (1- to 2-year-old child from pre-Columbian site of Pueblo Bonito, New Mexico, USA, NMNH 327074).

affected by the disease process. An X-ray film of the complete right femur and the partially complete tibiae, left femur, and humerus reveals a generalized enlargement of the medullary cavity with the cortex much thinner than normal. Comparison of the abnormal femur with normal femora (Figure 405) of similar size from the same population reveals a cortical thickness of less than 1 millimeter for the abnormal specimen, while the normal femora are between 2 and 3 millimeters.

As El-Najjar, Lozoff, and Ryan (1975) argue, if some porotic hyperostosis is caused by nutritional anemia in which iron is deficient, one might find evidence of other metabolic problems resulting from iron deficiency. Von Endt and



FIGURES 403–405.—Bone marrow reactions in a skeleton with porotic hyperostosis: 403, Porous labyrinth-like lesions of the skull vault. 404, Broken section of parietal demonstrating expansion of diploë. 405, X-ray of femur of specimen (center) compared with the femora of two normal children. Note the expanded marrow space and thinned cortex. (2-year-old child from the pre-Columbian site of Pueblo Bonito, New Mexico, USA, NMNH 327107.)

Ortner (1979) hypothesized that dietary iron deficiency might also affect the production of two amino acids, hydroxylysine and hydroxyproline, which are found in bone protein. They reason that, since iron is a factor in the synthesis of hydroxylysine and hydroxyproline, these two amino acids might be deficient in the bone protein of children suffering from iron-deficiency anemia. Von Endt and Ortner (1981) evaluated this possibility using one of the specimens described above (NMNH 327107). They compared the amino acid residues of bone protein from an archeological skeleton of a child having porotic hyperostosis with a similar skeleton that did not have porotic hyperostosis. A bone protein sample from a modern child who died from accidental causes was used as an additional control. The hydroxylysine and hydroxyproline residues of the ancient and modern skeletal samples that did not have porotic hyperostosis were virtually identical. In the bone of the child having porotic hyperostosis there was 5 percent less hydroxyproline and 25 percent less hydroxylysine. The authors argue that these reduced concentrations support a diagnosis of iron-deficiency anemia in the skeleton with porotic hyperostosis.

Certainly a diagnosis of some type of anemia is a strong probability in the two cases described above. However, the specific type of anemia is more problematical. Moseley (1963:6, 1966:128) reports that the long bones are not affected in

iron-deficiency anemia. This conclusion is not supported by Lanzkowsky (1968:24), who found widened medullary spaces and thinned cortices in the postcranial bones, particularly the metacarpals and phalanges. This difference of opinion clearly indicates that differential diagnosis of anemia in dry skeletal specimens may not be possible without considering other variables, including biochemistry and paleoepidemiology.

There is the further problem of differentiation between porotic hyperostosis from anemia and similar lesions seen in infectious disease and metabolic disease. I have already discussed porous expansive lesions of the skull in infectious disease (p. 137). Similar lesions in scurvy and rickets will be reviewed in the section on metabolic disorders (p. 270). Briefly, porous lesions of the skull in infectious diseases tend to be periosteal. They tend not to involve an expansion of the marrow space or destruction of the outer cortex. The lesion typically is superficial to normal bone. In scurvy the bony reactions thought to occur could easily be confused with those of anemia or infectious disease. It is possible that in scurvy the outer cortex remains intact and no expansion of the diploë would be expected. The porous, expansive lesions in rickets are morphologically distinct from anemia. Although the lesion is porous and expansive, the pores in the skull are much finer and could not be confused with porotic hyperostosis associated with anemia. Indeed one should keep in mind that lesions attributed to metabolic diseases may, in fact, be due to undiagnosed anemia, perhaps associated with metabolic disease.

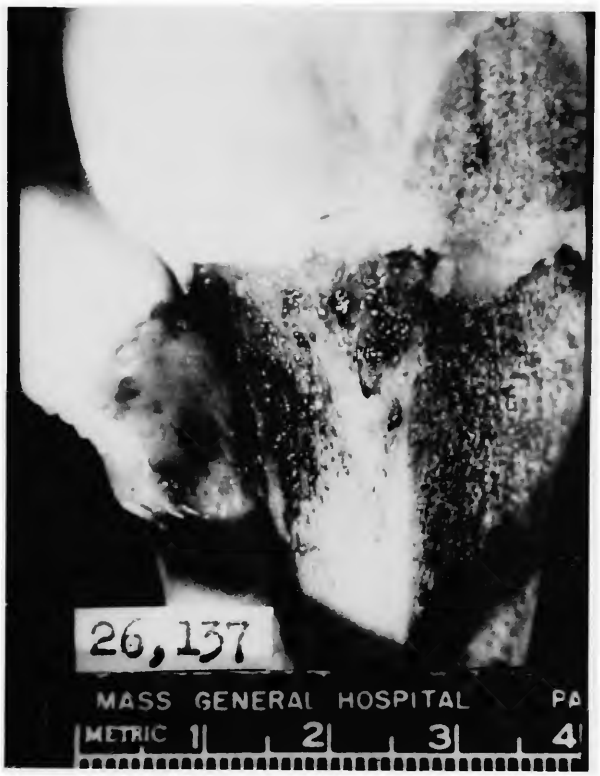


FIGURE 406.—Acute lymphatic leukemia; distal femur showing metaphysial cortical erosion due to leukemic deposits. (6½-year-old female, MGH autopsy 26137.)

Leukemia

PATHOLOGY

Leukemias are cancers of the myeloid and lymphoid hemopoietic cells of the bone marrow and occur in acute and chronic form in children and adults. The tumor cells extensively replace the normal bone marrow throughout the skeleton. As far as bone changes are concerned, only the acute leukemias of childhood need to be considered here.

FIGURE 407.—Sarcomatosis of cranium and many bones in lymphoma; frontal view of skull. Notice the destruction of the frontal and periorbital area. (About 20-year-old male, with generalized lymphadenopathy, ANM 2039 from 1883.)

In the growing bone of children, the replacement of the normal marrow cells by tumor cells and their subsequent proliferation lead, not rarely, to recognizable alterations of the bone structure. The most frequent lesion is a narrow radiolucent line on the metaphysial side of the growth plate. This change is not specific and resembles, to some extent, the lucent metaphysial zone in scurvy. Another alteration seen in acute childhood leukemias concerns the cortical surface of the metaphyses. Normally, in these areas, osteoclastic resportion of the modelling process creates a rough and somewhat grooved or porous cortical surface. In acute leukemia, these areas of the periosteum may be colonized by tumor cells emanating through the vascular foramina. This leads to widening of the vascular foramina and exaggerated grooving and porosity of the metaphysial cortical surface (Figure 406). This may be the most characteristic bone lesion of acute leukemia. Occasionally, widespread nonspecific subperiosteal bone deposits are observed over thinned cortices of long bones and ribs. Bone changes in adult leukemias are too uncommon and uncharacteristic to warrant discussion in this context.

In malignant lymphoma, bone lesions, if present, resemble lytic or sclerotic metastases of carcinoma. Occasionally diffuse subperiosteal involvement with cortical erosion occurs (Figure 407).

Myeloma

Pathology

Myeloma is a highly malignant tumor of plasma cells, usually arising in an area of hemopoietic bone marrow and, after varying time intervals, involving extensive portions of the skeleton.

The disease is relatively rare. There is a great prevalence of males over females affected. The tumor rarely occurs below 40 years of age with a maximal frequency between 50 and 70 years of age. There is no known racial or geographic prevalence.

The initial lesion usually arises in that part of the skeleton that contains hemopoietic marrow which, in the older adult individual, is the trunk skeleton and the proximal metaphyses, especially of the femur and the humerus. Single tumor cells disseminated through the blood colonize mostly in the areas of hemopoietic marrow. In the cranial vault and in portions of the long bones of the extremities, reactivated hemopoietic marrow appears, secondary to massive marrow involvement in the trunk. The facial bones and the small bones of the extremities are usually spared.

The initial lesion may remain localized and solitary for months or years, but ultimately dissemination to other parts of the skeleton almost always occurs. A typical location for this type of lesion is the proximal metaphysis of the femur and humerus (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952:1947). The solitary lesion is slow-growing, creating an osteolytic defect, ultimately eroding the cortex but permitting the new formation of an expanded bony shell with ridge-like internal reinforcements, giving the so-called soap bubble pattern on X-ray. Pathological fractures through the lesion are not uncommon. The lesion usually does not extend into the head of the femur. This distinguishes it from giant cell tumor and chondroblastoma. In dry bone, the solitary lesion could not be differentiated with certainty from unicameral bone cyst, chondromyxoid fibroma or nonossifying fibroma, except for the fact that these would usually occur in a younger age group. The differentiation of the lesion from solitary lytic metastasis of renal or thyroid carcinoma may be impossible. The most frequently observed and most characteristic manifestation is the disseminated form (multiple myeloma). The lesions start within bone marrow but ultimately scallop and destroy cortex. In most affected bones (vertebrae, ribs, sternum, clavicles, scapulae, pelvis, calvarium, and long bones) the individual lesions create punched out, purely lytic defects without reactive bony margins (Figure 408). Most are round, 3 to 10 millimeters in diameter. Individual large lesions are rare, with exception of the original ones; but small lesions can become confluent, often showing scalloped

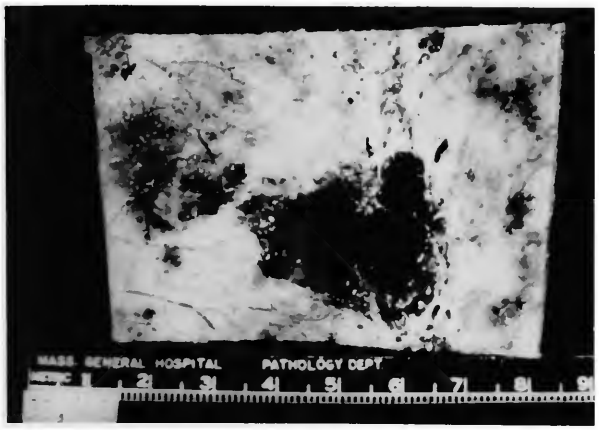


FIGURE 408.—Multiple myeloma. Endocranial view of cranial vault showing multiple lytic lesions destroying the internal table. (65-year-old male, MGH autopsy 33606.)

margins. In vertebral bodies this confluence of individual lesions is particularly common. The spinous processes are often involved. The destruction of the vertebral spongiosa often leads to collapse of multiple vertebrae, frequently with deep cupping of the end-plates due to expansion of the intervertebral discs (Figure 409). Wedge-shaped vertebrae, kyphosis, and scoliosis are common. The ribs also frequently show multiple transverse fractures and a coarsely reticulated irregular pattern of the few remaining trabeculae. Pathological fractures on long bones are also not uncommon. The differential diagnosis between disseminated multiple myeloma and osteolytic metastatic cancer, particularly of the breast, may be impossible. The male prevalence in myeloma and the female predominence in breast cancer are helpful in the differentiation. Furthermore, in metastatic carcinoma, even if predominantly lytic, some of the lesions usually show osteoblastic response, at least in the vicinity. Multiple myeloma frequently involves the glenoid fossa of the scapula and the lateral portion of the clavicle and disseminates into radius and ulna, a condition which is uncommon in metastatic carcinoma (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952:951). The presence of a larger, older lesion also helps in the differentiation from metastatic carcinoma. Rarely, multiple myeloma may leave the bone structure unchanged or produce only a

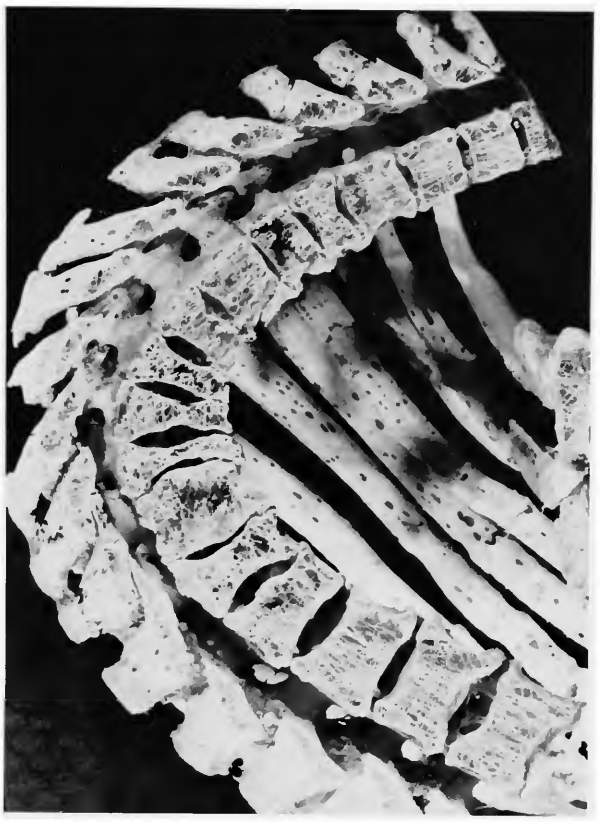


FIGURE 409.—Multiple myeloma. Bisected spine and ribs with severe osteoporosis, compression fractures, kyphosis. Notice disseminated small lytic lesions in ribs and spinous processes. (77-year-old female, IPAZ autopsy 1703 of 1954.)

pattern of diffuse osteoporosis without distinct lytic lesions. These cases cannot be recognized on the dry bone.

PALEOPATHOLOGY

The identification of myeloma in paleopathological specimens is complicated by the close resemblance between bone lesions of myeloma and those of some types of metastatic carcinoma. All the accounts of myeloma in the paleopathological literature are of the multiple type, and I shall limit the following discussion to that expression of the disease.

Ritchie and Warren (1932) report a case of multiple lytic lesions in a skeleton of a pre-Columbian Indian (ca. A.D. 800–900) from New York

State, USA. The skeleton is that of an old male. The roentgen films show multiple lytic lesions of the skull, which vary from a few to 25 millimeters in diameter. Other postcranial bones show similar lesions, with the long bones less affected then the axial skeleton. They suggest that prostate carcinoma is a possible cause but indicate that multiple myeloma is more likely, citing the opinions of H. U. Williams and R. M. Moodie in support of this diagnosis. The age and sex of the individual is compatible with a diagnosis of multiple myeloma. However, the large variation in the size of the lytic foci is more compatible with metastatic carcinoma.

Williams, Ritchie, and Titterington (1941) report a case of multiple lytic lesions in the skeleton of a 10-year-old pre-Columbian child excavated near Rochester, New York, USA. The lesions vary between 3 and 10 millimeters in diameter in almost every bone recovered. The authors suggest multiple myeloma as a probable cause, but this is unlikely because of the young age. A more likely cause would be one of the histiocytoses, such as Schüller-Christian's disease.

Fusté (1955) describes a neolithic human skull from the Pyrenees that had multiple, lytic lesions. The lesions were roughly circular with irregular borders associated with reactive bone. Either multiple myeloma or metastatic carcinoma is possible, although the presence of reactive bone is more likely in metastatic carcinoma.

Nemeskéri and Harsányi (1959) describe the skeletal remains, from the tenth to eleventh century site of Kérpuszta, of a male about 50 years of age (burial no. 38, catalog no. 5293) which contain multiple lytic lesions. Their figures 9–12 show longitudinal sections of humerus, femur, and innominate bones, all of which have lesions including a fairly large lytic lesion in the innominate. Although a scale is not provided, this lesion appears to be in excess of 2 centimeters in largest diameter. Roentgen films of the skull show multiple lytic lesions of fairly uniform size, none appearing to be in excess of one centimeter. There seems to be evidence of bony reaction surrounding some lytic foci, although sand infiltration could be the cause. The large size of some lesions and the possibility of bony reaction would, in my opinion, tilt the scale in favor of a diagnosis of metastatic carcinoma rather than myeloma. Indeed, Brothwell (1967a:337), in commenting on this and other similar cases, offers the opinion that skeletons with multiple lytic lesions should be considered cancer metastases, until more convincing evidence can be provided for a diagnosis of multiple myeloma.

Wells (1964b) reported on two medieval skulls with multiple lytic lesions. The skulls were from Kent and Suffolk, England, and both were adults of unknown age. Wells was reluctant to offer a firm diagnosis but suggested metastatic carcinoma and multiple myeloma as likely, with myeloma the more probable.

Brooks and Melbye (1967) describe a pre-Columbian skeleton from Missouri, USA, with multiple lytic lesions. The skeleton is that of a female over 40 years of age. Photographs of skull and femora reveal considerable variation in the size of the lesions. Some on the femora are at least half the diameter of the femur (about 15 mm). They attribute the probable cause of the lesions to multiple myeloma. The age and sex of this specimen is more compatible with a diagnosis of metastatic carcinoma, such as breast cancer, but certainly multiple myeloma is a strong possibility as well.

Morse (1969:25, 144) briefly discusses a possible case of multiple myeloma from a site on Santa Cruz Island, California, dated between A.D. 300 and 1450. The specimen is in the Lowie Museum (No. 12-4356), University of California, Berkeley, USA. There are multiple, coalescing, lytic lesions on the skull and postcranial bones, features that are compatible with multiple myeloma. The skeleton is reported to be a female with an estimated age of 45.

Morse, Dailey, and Bunn (1974) published a report on four paleopathological specimens from Florida and Mississippi, USA, with lesions that they attribute to multiple myeloma. All were dated to the pre-Columbian period. The first of these is a skull (their specimen number 1) from

the Sowell Mound thought to be a male about 45 years of age. A photograph shows a large circular lytic lesion 24 millimeters in diameter on the occipital bone. There are other lytic lesions in the skull ranging from 2 to 8 millimeters in diameter. The age and sex of this case are compatible with a diagnosis of multiple myeloma; however, metastatic carcinoma is equally possible and is more compatible with the large variation in the size of the lesions.

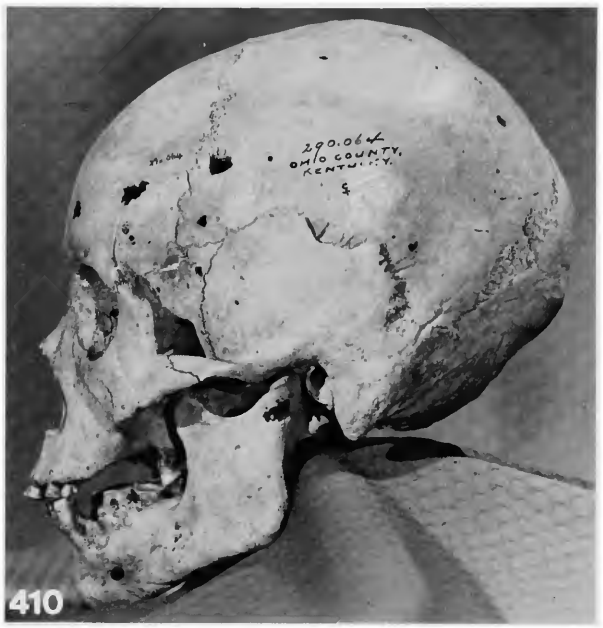
Burials 1 and 2 from the Calico Hills mounds in Jefferson County, Florida, USA, both show multiple lytic lesions of the skeleton. Burial 1 is reported to be a female about 25 years of age. The lytic lesions range in size from 1 to 13 millimeters. The published roentgen picture reveals some lesions on the skull, but major involvement is on the innominates and the distal femur. The latter in particular shows multiple coalescing lytic lesions with some evidence of bony circumscription, although sand infiltration could be creating the appearance of reactive bone around the lesion. The age and sex of the individual, as well as the possibility of bony reaction to the disease process, argue more for metastatic carcinoma rather than multiple myeloma in this case.

Burial 2 from the Calico Hills site consists of several postcranial bones and fragments of the skull. The authors report that the skull appears to be female and 25 years of age or less. A roentgen film of the skull shows multiple lytic lesions with no evidence of bony reaction. However, a photograph of the skull (Morse, Dailey, and Bunn 1974, fig. 4) shows considerable porous bony reaction of the external skull surface in areas surrounding lytic foci. In this case the lytic lesions are compatible with multiple myeloma, but the age and sex favor a diagnosis of metastatic carcinoma. Infectious disease should be considered as well.

The fourth case of purported multiple myeloma is from the Magnum Mound in Clarbourne County, Mississippi, USA. Burial 4 from this site is a female postcranial skeleton of about 35 years of age. The vertebrae, ribs, pelvis, and right scapula show multiple lytic foci. The skull (4) thought to be associated with the postcranial bones shows a normal outer cortex (Morse, Dailey, and Bunn, 1974, fig. 6). The roentgen film reveals lytic lesions in the diploic space sometimes penetrating through the inner table. There is a large eroded surface on the inner table, which the authors feel is antemortem. They suggest a large myelomatous area could have destroyed the inner table. However, they indicate that this large lesion is not typical of myeloma. The long bones are unusually light suggesting osteoporosis.

Three paleopathological specimens from the National Museum of Natural History, Washington, D.C., provide further insight into the problems of differentiating between multiple myeloma and osteolytic metastatic carcinoma. The first of these is a female skeleton (NMNH 290064) from the Indian Knoll site in Kentucky, USA. Most of the artifacts from this site are dated in the Late Archaic period (ca. 3000–1000 B.C.); however, some components date to the Late Woodland period (ca A.D. 800–1700). Thus the archeological age of this specimen remains obscure but with a strong probability of being Archaic. The age of the individual cannot be determined precisely. The skull sutures had just begun to fuse on the endocranial side, there is little evidence of arthritis, and tooth wear is marked. However, in this population tooth wear typically is marked in adults. These criteria suggest an age in the 30- to 40-year range. The disease process consists of multifocal, mostly lytic lesions distributed in the skull, mandible, axial skeleton, and the proximal left femur (the right femur is missing). The bones of the hands and feet are unaffected except for a slight, superficial osteoporosis of the superior surface of the calcanei. The gross lesions vary in size from being barely detectable to about 15 millimeters in diameter. Most of the cranial lesions (Figure 410) penetrate both tables, with no clear pattern regarding which table is most extensively affected. The lytic process is more extensive in the diploë, suggesting that the marrow was the focal point for the disease process. The roentgen films of the skull and long bones reveal additional lytic foci that are not visible from the outside. The

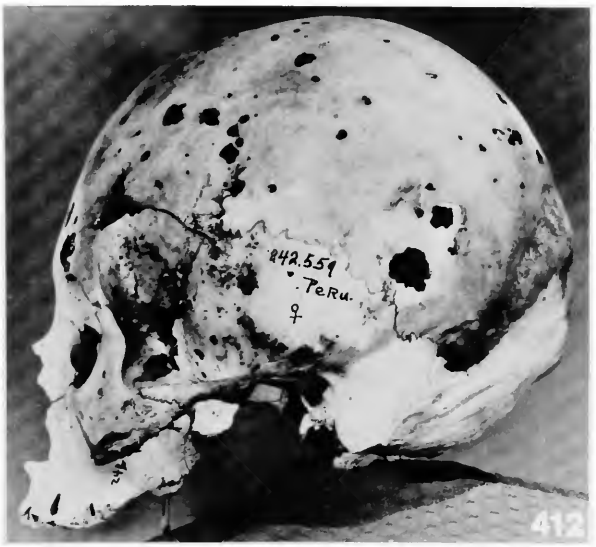
scapulae are present and both show multiple lytic foci (Figure 411), although the glenoid fossa is normal. However, unlike lesions elsewhere in the skeleton, there is a slight osteoblastic response adjacent to several of the lytic foci. With this exception, there is no osteoblastic reaction to any of the other lytic lesions in the skeleton. This suggests a rather acute disease process. Steinbock (1976:381–384) has reported on this case and concludes that "the size, location, and appearance of the destructive lesions in this Archiac Indian are highly indicative of multiple myeloma rather than metastatic carcinoma." While conceding that multiple myeloma is a strong possibility based on some aspects of the gross and roentgen film morphology of the lesions, I feel that an even stronger case can be made for metastatic carcinoma. The lesions are somewhat more variable in size than typical for multiple myeloma. Peripheral bone reaction on the scapulae is more characteristic of metastatic carcinoma as is the age and sex of the skeleton.

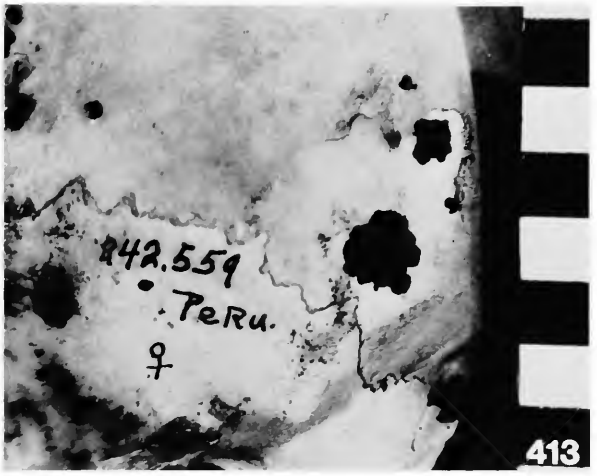


FIGURES 410, 411.—Multiple lytic lesions of the skull and postcranial skeleton: 410, Left lateral view of the skull and mandible. 411, Posterior view of left scapula with lytic lesions and porous bone hypertrophy (arrow) adjacent to lytic focus. (Adult female from Indian Knoll, Kentucky, USA, NMNH 290064.)

Another possible case of multiple myeloma is a female skull from Peru (NMNH 242559). This specimen is fully adult and probably in the 30- to 40-year age range. The archeological age is unknown. The external gross aspect of the entire skull, except the face, is characterized by several scalloped, lytic lesions (Figure 412), which vary in size from pin holes to 15 millimeters in diameter. There is no evidence of bony circumsciption (Figure 413) either by inspection or on roentgen films. The lesions are characteristic of both myeloma and metastatic carcinoma and, again to the extent that age and sex are useful variables in differential diagnosis, metastatic carcinoma would appear slightly more probable.

The third case from the National Museum of Natural History, Washington, D.C., USA, is also a female skull from Peru (NMNH 242578). Age at death is, like the two previous cases, in the range of 30 to 40 years. The archeological age is unknown. Unlike the lesions in the preceding two cases, the external appearance of the lesions is much less obvious. The lytic lesions, which do penetrate the surface, are small holes typically 1–2 millimeters in diameter. On the internal table the lesions are somewhat more pronounced. All bones of the skull are affected, but the facial bones and occipital bone show much less involvement. The greater wings of the sphenoid and the body of the sphenoid are markedly affected with much of the latter totally destroyed. In the X-ray





FIGURES 412, 413.—Multiple scalloped lesions of the skull: 412, Left lateral view of the skull with lytic lesions of varying size. 413, Detail of lytic lesion demonstrating the presence of spongy bone which indicates a lack of circumscription. (Adult female from Caudivilla, Peru, NMNH 242559; scale in cm.)

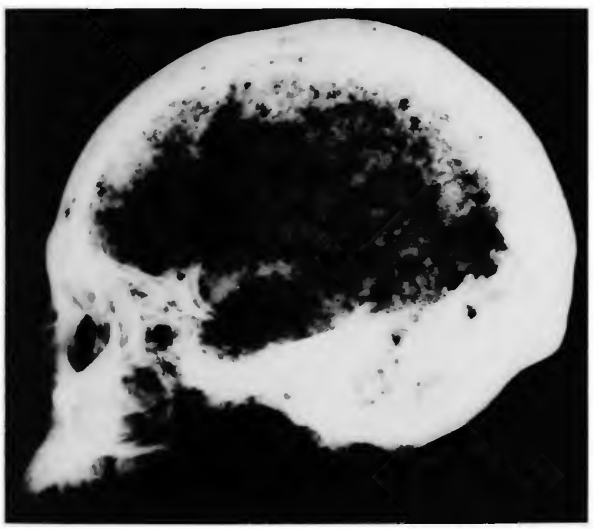


FIGURE 414.—X-ray of mediolateral view of multiple lytic lesions of the skull. Note the lack of sharply defined boundaries of the lytic foci. (Adult female from Peru, NMNH 242578.)

(Figure 414), it is apparent that the major focus of the lytic process is the diploë. There is no evidence of any osteoblastic reaction in any of the lesions. On the film a typical lesion consists of a lytic focus ranging in diameter up to 2 centimeters. Many of the lesions coalesce. The overall picture presented by this case is not typical of either multiple myeloma or metastatic carcinoma. However, Schinz, Baensch, Friedl, and Uehlinger (1951–1952:947, 949) briefly describe a case of atypical multiple myeloma in which "rather numerous individual foci are distinctly delimited, and a moth-eaten, finely mottled kind of osteolysis develops. . . . " This description and their published roentgen film views closely match the appearence of the Peruvian skull. However, Schinz, Baensch, Friedl, and Uehlinger caution that the case they describe is not easily distinguishable from metastatic carcinoma. Provisionally, however, it is useful to consider this skull as an example of atypical multiple myeloma.

Metabolic Disorders

Vitamin C Deficiency

Pathology

Scurvy is a disease caused by prolonged inadequate intake of vitamin C (ascorbic acid). It affects many organs and has a high mortality in severe cases. Ascorbic acid is essential in the formation of the collagen fibril polypeptide precursors. Since the organic matrix of bone consists mainly of collagen, scurvy manifests itself in diminished or absent bone matrix formation, most marked in the rapidly growing skeleton of the infant. Ascorbic acid also plays a role in the formation of the cement substance between vascular endothelial cells and of basement membrane. This explains the great tendency to hemorrhage in scurvy, occurring spontaneously and on minor traumatization. The skeletal manifestations, which alone concern us in this discussion, are most marked in infants and rather minor in degree in adults.

Adult scurvy has been recognized worldwide as a serious disease with high mortality. It occurred throughout history in populations deprived of fresh vegetables and meats due to war, famine, or prolonged trips at sea (e.g., voyages of discovery and polar explorations). Infantile scurvy, however, was not appreciated until well into the nineteenth century (Möller, 1862; Barlow, 1883).

Since vitamin C is destroyed by boiling temperature, its deficiency is prone to manifest itself in populations consuming mainly cooked foods, rather than in primitive peoples, who eat considerable amounts of raw food.

Infantile Scurvy (Möller-Barlow's Disease)

Since ascorbic acid freely passes from the mother through the placenta to the fetus, vitamin C deficiency does not exist at birth. Even if no, or almost no, vitamin C intake occurs after birth it takes several months before the deficiency manifests itself as a recognizable disease. Infantile scurvy is seldom observed before 4 months of age and reaches its maximum of occurrence between 8 and 10 months of age (Wimberger, 1925:288–307).

The skeletal changes are most marked in the areas of most rapid growth: costochondral junctions of ribs, distal metaphysis of femur, radius and ulna, and proximal metaphysis of humerus.

The skeletal changes in scurvy are twofold: those caused directly by the vitamin C deficiency, and secondary changes due to traumatic effects on the vulnerable scurvy bone. The primary lesion consists of the combined effect of severely diminished osteoblastic activity and continued osteo- and chondroclastic activity. This leads to a piling up on the metaphysial side of the growth plate of calcified cartilage septa, which are not or incompletely covered by bony matrix ("scorbutic lattice" of Park, 1954). The resulting mechanical instability of this metaphysial zone usually results in transverse fracture through this area with fragmentation of the lattice and of the sparse trabeculae, producing hemorrhage and dislocation of the epiphysis. Since not only enchondral but, equally, periosteal bone formation is inhibited, the cortex is thinner than normal, especially on the metaphysis of long bones.

The vascular vulnerability often leads to subperiosteal hemorrhages of varying degree and location. The most massive subperiosteal hematomas are usually observed in the weight-bearing long bones of the legs, especially the femur and tibia (Figure 415). Such hematomas usually begin at the level of a fractured metaphysis but may strip the entire shaft periosteum (Figure 416).

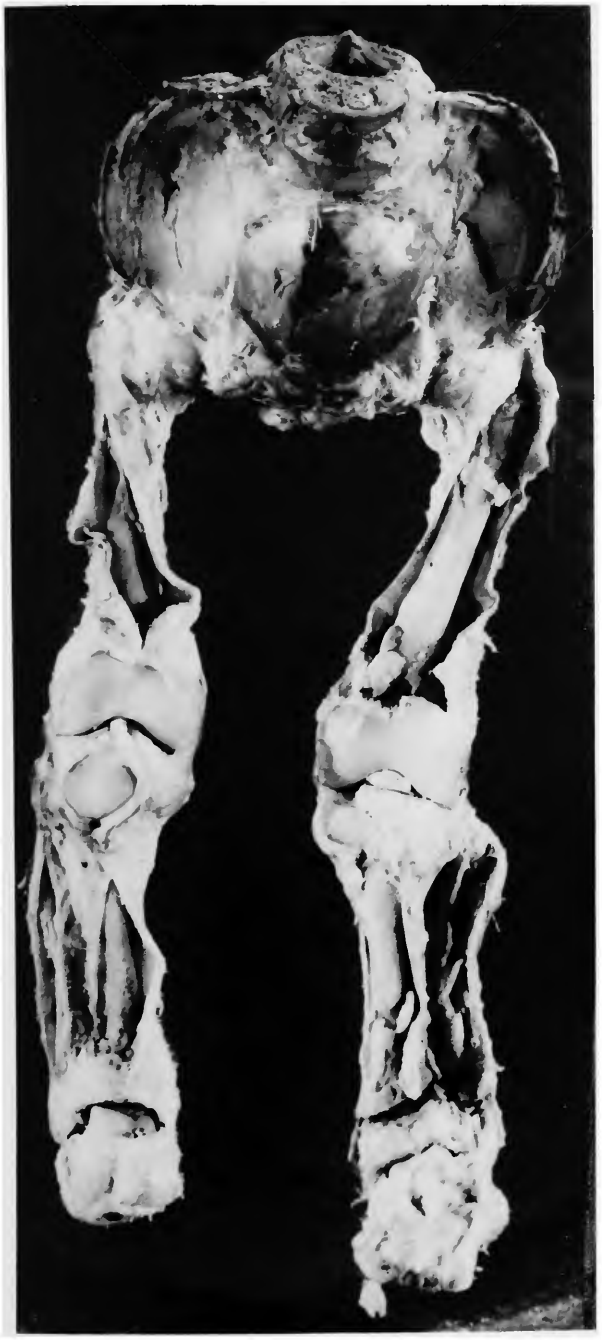


FIGURE 415.—Infantile scurvy with subperiosteal hematomas and transmetaphysial fractures of both distal femora and tibiae. Wet specimen with periosteum opened. (11-month-old infant, 6 months duration, PMWH W 3530.)

FIGURE 416.—Infantile scurvy, right bisected femur. Wet preparation showing massive subperiosteal hematoma and pathological fracture near the distal growth plate. (6- to 12-month-old infant, PMSG 9/856A.)

The elevated periosteum begins to produce reactive bone which, in the course of resorption of the hematoma, becomes attached to the cortical surface.

The changes observed in scurvy in different parts of the skeleton are as follows:

RIBS.—The bone adjacent to the osteochondral junction is often transversely fractured, causing an inward dislocation of the sternum and the rib cartilages, thus forming the palpable scorbutic "rosary."

LONG BONES.—In addition to the metaphysial fractures already described, the marked cortical thinning and deposition of reactive periosteal pumice bone, up to one centimeter in thickness (Schmorl, 1901:217), can occur. The proximal

metaphysis of the femur, if severely affected, caves in only beneath the head of the femur but not beneath the greater trochanter, because of the weight-bearing stresses transmitted through the hip joint (Erdheim, 1931b). This leads to a depressed angle of the neck of the femur. In healing, usually no major deformities remain, but collapsed metaphyses and dislocated epiphyses may become reunited in abnormal position. The secondary ossification centers of the epiphyses likewise show marked osteoporosis circumferentially surrounded by a layer of increased calcified cartilage at the growing periphery. This radiodense layer represents Wimberger's ring on the X-ray.

Flat Bones.—Subperiosteal hemorrhages also occur rarely on pelvic bones and scapulae (Fraenkel, 1929:228).

Skull.—Subperiosteal hemorrhages are common on the frontal bone and particularly on the portion forming the orbital roof. These hemorrhages are found more often on the orbital rather than on the endocranial surface and may lead to deposition of vertically arranged trabeculae of subperiosteal bone, covering the thin, partly resorbed cortex.

Fraenkel (1929:228–229) found involvement of the orbital roof in 4 of 34 autopsied cases of infantile scurvy. Hemorrhages also occur over the cranial bosses and over the maxilla and mandible (Fraenkel, 1929:229–230).

Healed Scurvy.—The fragmentation zone of the metaphysis undergoes slow remodeling, while normal new bone increasingly separates it from the growth plate. This shows on X-ray as a coarsely structured transverse band, which remains demonstrable for several years (Wimberger, 1925:288–307). Similarly, a double ring of radiodensity appears on secondary ossification centers in healing (Wimberger's ring).

Unfortunately, most of the changes of acute infantile scurvy have little chance of preservation in grave material due to the loss of the cartilage and soil disintegration of the infant bones. Possibly the changes of the orbital roof and remnants of incompletely remodeled fragmentation bands of healed scurvy may be of help in recognizing the disease.

Adult Scurvy

Already in older children, skeletal changes in scurvy are of a minor nature reflecting the slowed growth rate. In adults, skeletal changes are restricted to transverse fractures at the osteocartilaginous junctions of ribs (Aschoff and Koch, 1919:53).

Paleopathology

In view of the problems in diagnosing scurvy in dry skeletal specimens, it is not surprising that the paleopathological literature contains little evidence of this disease. Wells (1964a:118–119) states that edentulous jaws associated with alveolar osteitis often found in European medieval cemeteries are examples of scurvy. He further states that ossified subperiosteal hematomas of the long bones are often found in such cemeteries in association with exfoliation of teeth. Both these conditions, in Well's opinion, point to a high incidence of scurvy in European medieval populations.

Wilson (1967:193, 194) finds evidence of scurvy in some of the ancient texts of Mesopotamia and Palestine. The description of a disease known as "the stinking disease" combined with the loss of teeth, as well as the possible use of wine as a cure, certainly support a diagnosis of scurvy whose clinical features include foul breath and loss of teeth.

In the New World, early explorers suffering from scurvy encountered Indian tribes who treated the disease with a drink made by boiling bark and leaves in water (van Wersch, 1954:10–11). The extent to which this knowledge was widespread is not known. However, the existence of a treatment certainly suggests the presence of the disease.

Paleopathological evidence of scurvy in the New World has been reported by Saul (1972:56–66), who attributes the existence of expansive lesions of the diaphysis of long bones and the antemortem loss of teeth to scurvy. The skeletal material is from the pre-Columbian Mayan site called Altar de Sacrificios in Guatemala. Saul

supports his conclusion by citing sources that indicate that the amount of vitamin C in contemporary Maya Indian diet is deficient and in the past many Indians died from scurvy.

The lesions in Saul's figures are nonspecific and are more compatible with a diagnosis of infectious disease or trauma. The antemortem tooth loss seen in Saul's figures 40 and 41 is from a specimen whose age he estimates to be 60+ years. The pattern of alveolar resorption is more typical of tooth loss due to caries and abscess. The anterior teeth of the mandible are present as well, a condition that would be unlikely if the molar tooth loss was due to scurvy.

Hooton (1930:317) suggested that porotic hyperostosis found in the Pecos skeletal material from the American Southwest is due to scurvy or rickets. Cranial involvement in scurvy is not as common as that found in the long bones. Barlow (1883:169) however, noted porous lesions of the external skull vault in association with at least one case of scurvy and reported (1883:168) abnormal conditions in the crania of 8 cases in a sample of 31 scorbutic infants and children.

There remains a problem in ascribing porous lesions of the skull to scurvy. It is a well-documented fact that scurvy and rickets frequently occur in the same case (Follis, Jackson, and Park, 1940:745). The presence of one disease does seem to have an inhibiting effect on the expression of the other disease. The question of which disease is dominant is unresolved. Based on histological evidence, Follis, Jackson, and Park (1940:746) suggest that "vitamin D deficiency certainly inhibits vitamin C deficiency from expressing itself. . . . " In contrast Park, Guild, Jackson, and Bond (1935:286) offer the opinion that, since vitamin C deficiency inhibits osteoid formation, the expression of rickets would be inhibited by scurvy. It is also possible that scurvy could be found in association with anemia (Goldberg, 1963:51) and that porous lesions of the skull are caused by anemia rather than scurvy.

The experimental evidence suggests that hemorrhage from scurvy with the associated bone formation is seen only following trauma (Park, Guild, Jackson, and Bond, 1935:276; Follis, 1943: 580). In a subsequent publication Follis (1948: 140) states that "subperiosteal hemorrhages develop as a result of trauma and normal stress and strain." Whether the normal stresses and strains to which the head is subjected in the infant or young child is sufficient to trigger bleeding, which could give rise to a large porous lesion, must remain an open question. Although Fraenkel did not note skull vault lesions in his report of scorbutic autopsied cases, he did note the development of porotic, hypertrophic bone in the orbital roof (Fraenkel, 1929:228–229). The association of both these features with scurvy needs further confirmation. However, it would seem prudent to consider scurvy as a possibility when such a lesion is encountered in archeological specimens.

Vitamin D Deficiency

Pathology

Rickets

Rickets is a systemic disease of early childhood extensively affecting the skeleton, but without direct mortality. It is caused by inadequate availability of effective vitamin D, which is essential in the mineralization process of the organic matrix of bone (osteoid). Dietary vitamin D is transformed in the liver into the active compound.

Precursors of vitamin D taken with the food are transformed in the skin under the influence of the ultraviolet fraction of sunlight. This explains the prevalence of rickets in populations climatically (high latitudes) or mechanically (crowded cities, occlusive clothing) deprived of sunlight. However, it should be appreciated that the darkly pigmented races are not fully benefited by tropical sunshine, because most of the ultraviolet rays are absorbed by the skin melanin pigment. The main causative factor remains the inadequate intake of vitamin D and its precursors.

Rickets was not well documented by written descriptions until the sixteenth or seventeenth century. From then on, increasing attention has been paid to this common disease of early childhood, especially in northern Europe and North

America. There was a marked increase of frequency with industrialization and metropolitan crowding. Figures for autopsy material in Dresden, Germany, show that 89 percent of all children between 2 months and 4 years of age exhibited evidence of active or healed rickets (Schmorl, 1909:437). Clinically, 97 percent of young children had evidence of rickets in Vienna (Escherich, 1899, cited in Hess, 1929:48) and, as late as 1921, Hess found 75 percent of the children in New York City affected by rickets (Hess, 1921: 697). The disease occurs worldwide with varying frequency.

Rickets rarely begins before 4 months of age because vitamin D passes from the mother to the fetus through the placenta and is stored in the liver of the infant. The highest frequency is observed between 6 months of age and 2 years in the sunless winter months. Only a few new cases develop after 4 years of age. The manifestations are earliest seen and most marked on the rapidly growing areas of the skeleton: osteocartilaginous junctions of ribs, distal metaphysis of femur, radius and ulna, and proximal humerus (Wimberger, 1925:269–288). The changes at the growth plate consist of lack of preparatory calcification of the columnar cartilage and of the newly deposited bone matrix (osteoid). Since the proliferation of cartilage and the formation of bone matrix by osteoblasts continues at close to normal levels, both uncalcified cartilage, which cannot be resorbed, and unmineralized osteoid pile up next to the growth plate. In the ribs this results in a rounded nodular swelling of the osteocartilaginous junction, the "rachitic rosary," and, in long bones, in a broadening and cup-shaped depression of the metaphysial areas. Since the intramembranous bone formation is affected equally to the enchondral one, the rapidly growing and expanding cranial vault of the infant becomes increasingly replaced by nonmineralized osteoid giving rise to areas of thinning and softening (craniotabes). This actually precedes recognition of the changes in the long bones in acute rickets (Hess, 1929:208).

The changes observed in rickets are threefold: direct effects of the metabolic disturbance, deformities secondary to the vulnerability and pliability of the poorly mineralized skeleton, and retardation of growth.

Rachitic and Postrachitic Changes in Individual Bones

Skull.—In active infantile rickets the cranial vault may develop thin and soft areas, especially in the posterior lateral portions of the parietal bones and of the occipital squama (craniotabes). This is due to the rapid remodeling of the infant skull to accommodate the growing brain, replacing mineralized bone with osteoid. This is not to be confused with the incomplete ossification of the entire cranial vault in premature infants or with the congenital lacunar skull often complicating spina bifida, in which sharply marginated multiple defects in various areas of the cranial vault are present. Craniotabes may lead to permanent posterior flattening or lateral and asymmetrical deformities of the skull, due to pressure of the head against the supporting surface in the lying position. The closure of the fontanels is delayed in rickets. In cases where long-standing thickening of the cranial vault is seen, especially in older children, mainly due to external (occasionally also internal), subperiosteal bone deposition, the external deposition often spares the center of the parietal and frontal tubera (Schmidt, 1929:32)(Figures 417–420). In this process of remodeling, the outer and often the inner table as well disappears, so that the entire thickness of the cranial vault has the porous appearance of diploë (Figures 421–423). The weight of the head may cause the bone surrounding the foramen magnum of the softened skull base to be pushed upward, diminishing or effacing the angle of the clivus (Schmidt, 1929:60). Similar subperiosteal deposits can occur on the facial bones (Figures 424–426).

Long Bones.—The frequent combination of hypovitaminosis D and general malnutrition leads to a combination of rickets and osteoporosis, resulting in the so-called porotic form of rickets,

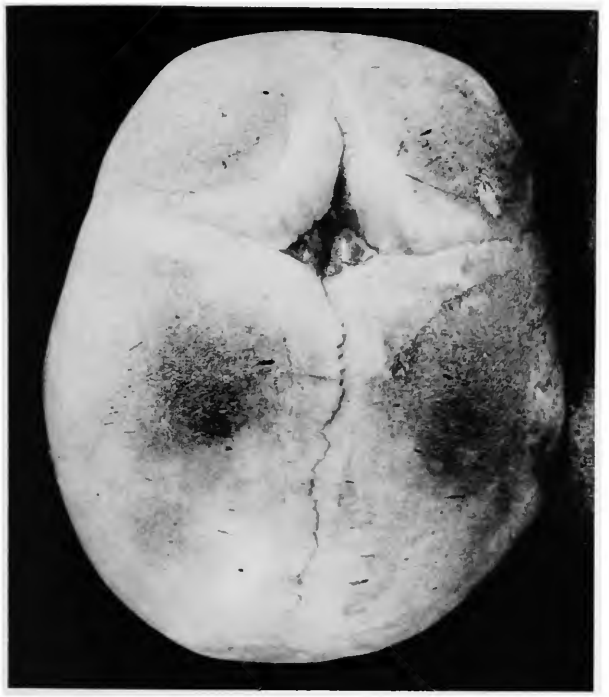
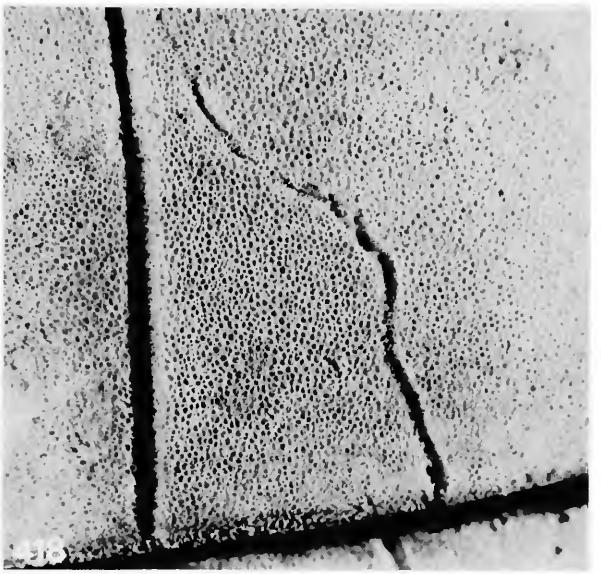


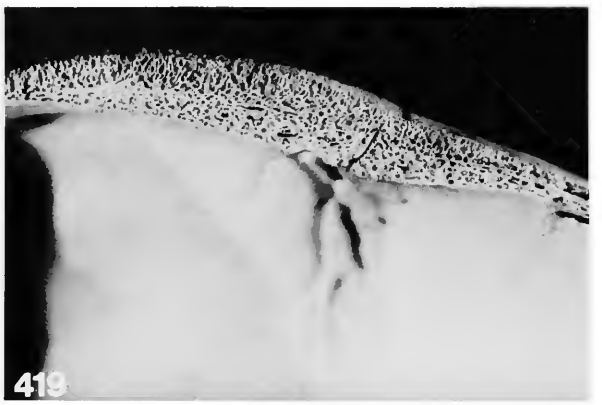
FIGURE 417.—Moderate rickets of cranium showing fine porotic bone deposition on frontal and parietal bosses. (7-month-old female, PMUG 2465, autopsy 6115 of 1874.)

characterized by brittle thin cortex and sparse cancellous trabeculae. In otherwise well-nourished infants, however, the deposition of massive osteoid, periosteally and endosteally, results in the plump bones with narrowed medullary spaces of the hyperplastic form of rickets. In the porotic form, stress fractures, especially in the diaphysis, are common, leading to axial deformities (Wimberger, 1925). In this type, bending deformities due to the flexibility of the undermineralized bones, with or without additional stress fractures, are common.

The growth is slowed down in active rickets, but the development of secondary epiphysial ossification centers is not delayed (Wimberger 1925). The most marked shortening is seen on the femur (Schmidt, 1929:73). The most characteristic change of a long bone is the flaring of the frayed metaphysial cortex and cupping of the end of the metaphysis.

Periosteal deposition of osteoid, which is mineralized in the healing process, is common. Gen-





FIGURES 418. 419.—Rachitic skull: 418, Outer table, parietal bone, showing porous periosteal bone. 419, Cut surface, showing subperiosteal fine cancellous bone externally and internally. (About 1-year-old female, FPAM 3874 from 1881.)

erally, these deposits are thickest at middiaphysis, giving the shaft the appearance of a column, without the usual tapering to the middle (Schmidt, 1929:33). These subperiosteal bone deposits show some characteristic distribution in different bones (Pommer, 1885). On the ribs, they are limited to the anterior surface and the margins, leaving the pleural surface free. On the femur, the deposits are heavier on the posterior than on the anterior surface. On the tibia, the

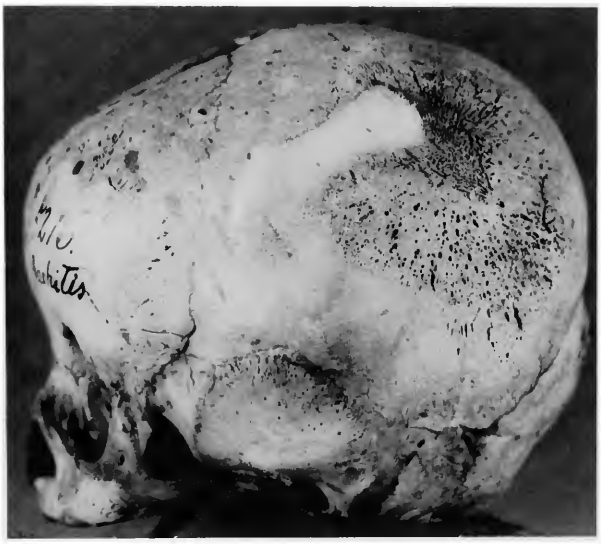


FIGURE 420.—Cranial rickets, left lateral view, showing porous subperiosteal bone deposition, mainly of parietal boss, focally transformed into a new outer table. (About 6- to 8-month-old female, DPUS 5216.)

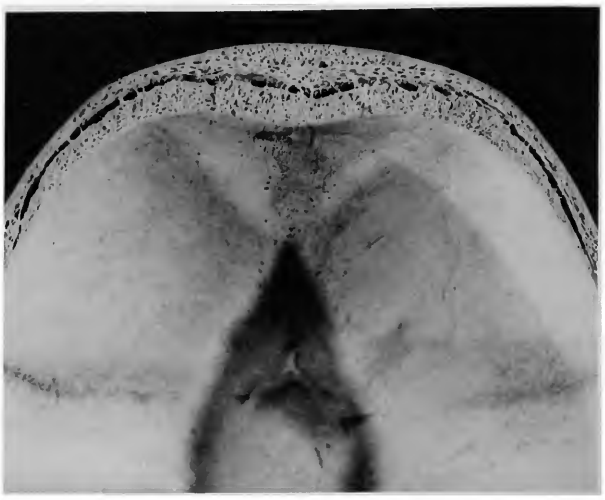


FIGURE 422.—Rachitic calvarium, cut surface, showing external and internal deposition of subperiosteal cancellous bone. (About 2 years of age, FPAM 5051 from 1889.)

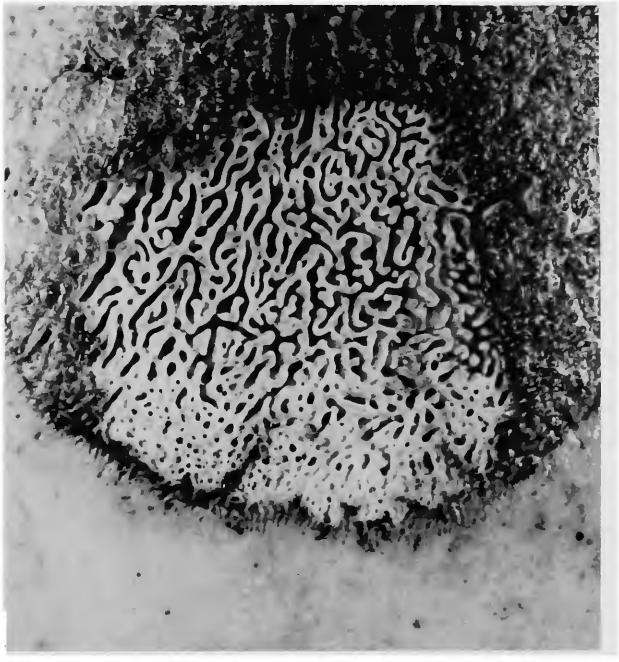


FIGURE 421.—Rachitic calvarium showing subperiosteal bone deposition with pronounced vascular pattern on right parietal boss. The dried periosteum appears dark. (7-month-old female with rickets and scarlet fever, DPUS 5212 from 1904.)

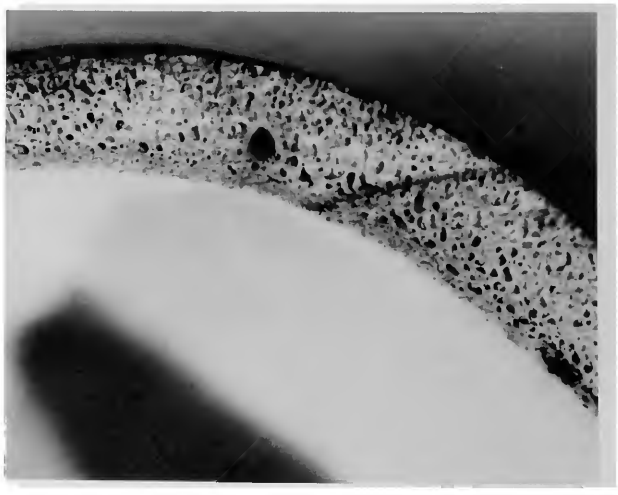
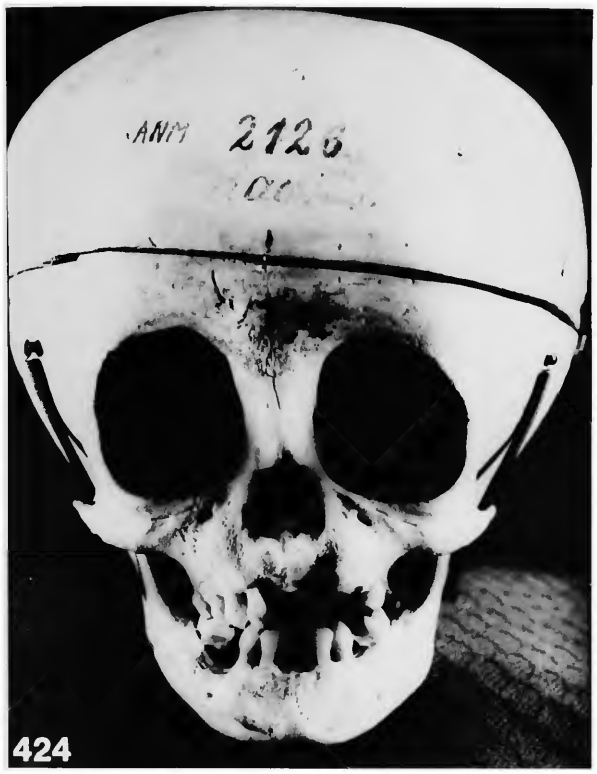
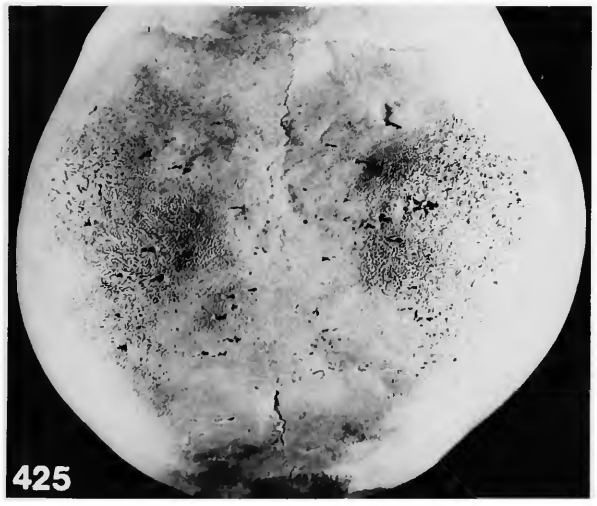


FIGURE 423.—Rachitic calvarium, cut surface, showing porous hyperostosis with obliteration of outer and partial obliteration of inner table. (2-year-old male, FPAM 2694 from 1858.)





FIGURES 424, 425.—Rachitic skull with slight hydrocephalus: 424, Frontal view, showing slight deposition of periosteal bone on glabella, maxilla, and mandible. 425, Top view of calvarium, showing periosteal bone focally consolidating into a new outer table on the parietal bosses. (3 to 6 years of age, ANM 2126 from 1866.)

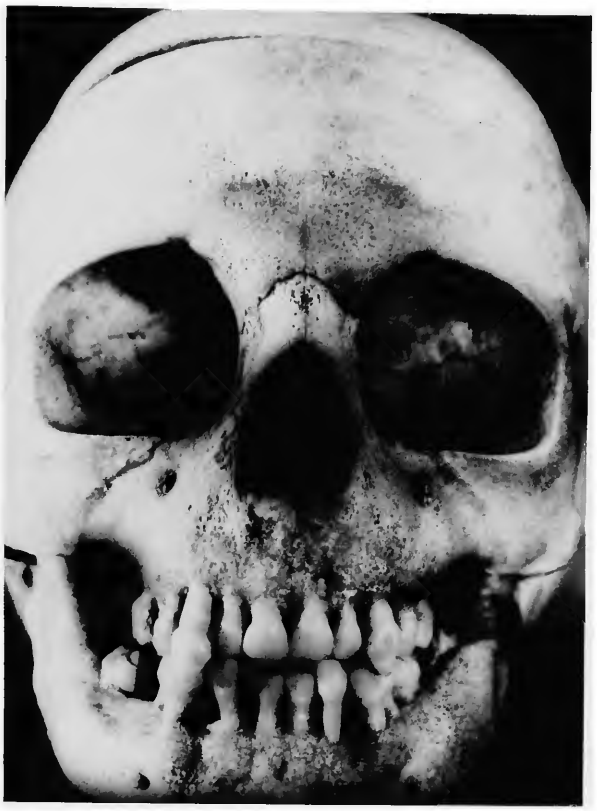


FIGURE 426.—Rachitic cranium, frontal view, showing fine subperiosteal bone deposition on glabella, facial bones, and orbital roofs. (3 to 6 years of age, ANM 2127 from 1866.)

deposits locate on the posterior and on the medial surface, leaving free the lateral surface that faces the fibula.

In severe and protracted or recurring cases, characteristic deformities develop, secondary to the static and dynamic stresses. After healing, these bending deformities largely remain for the rest of the life of the individual (Figure 427). When the deformities develop in the active phase, an alteration of the distribution of subperiosteal bone is observed. This alteration consists of increased deposition on the concavity of the deformity, in response to the altered stresses. These deposits may be locally accentuated by addition of callus over stress fractures, forming ridges or lumpy projections (Recklinghausen, 1910(1):213–215). Such fractures occur most often in the dia-

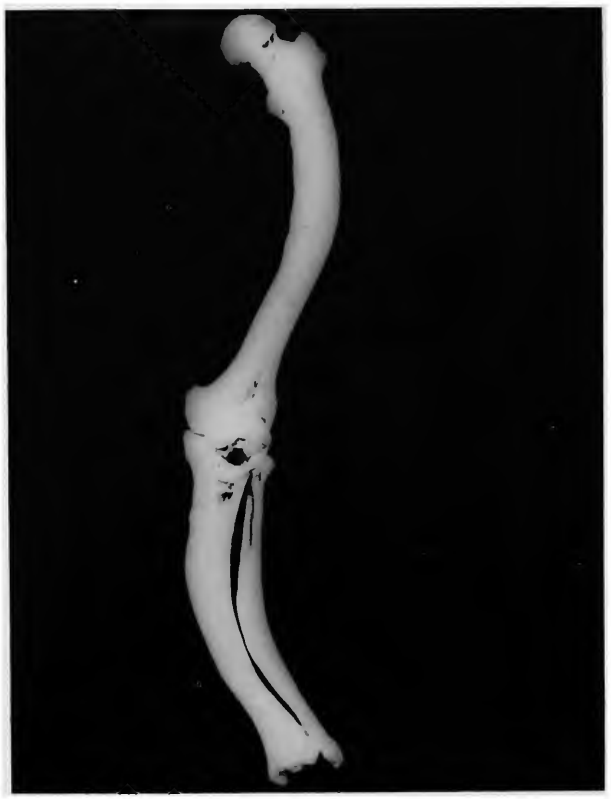


FIGURE 427.—Postrachitic deformity of left leg. (Adult, VM 106 from 1876.)

physis, seldom in the metaphysis (Wimberger, 1925) (Figure 428). They often transect only the cortex on the concave side and their margins may project into the medullary canal (Recklinghausen, 1910(1):213–215).

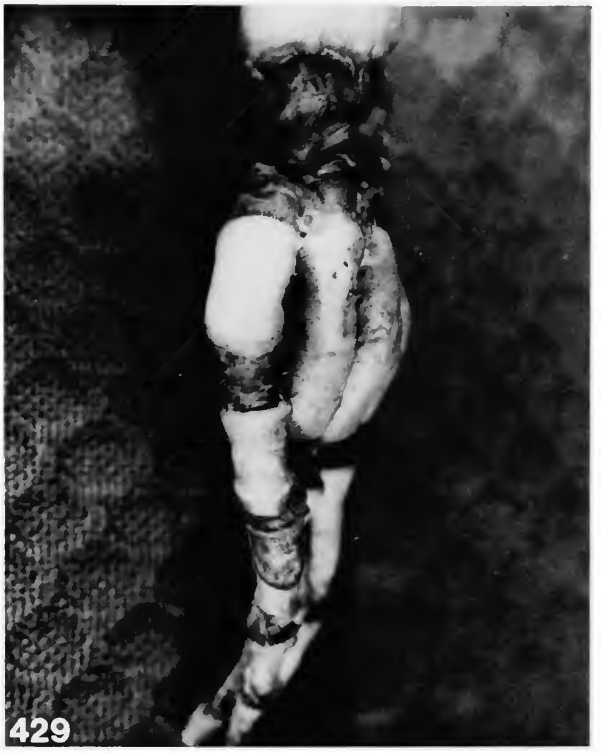
In the infant, before walking, bending deformities of the humerus and forearm occur, depressing the humeral head medially downward and bending the shaft laterally and forward (humerus varus)(Figures 428, 429). Later, after assumption of upright gait, deformities of the long bones of the lower extremity prevail. The femur usually shows downward bending of the neck (coxa vara), seldom the opposite increase of the neck angle (coxa valga). The femoral shaft usually bends at the lower metaphysis with an anterolateral convexity. The tibia also usually shows anterior bowing at the lower metaphysis, and the fibula has to follow with a similar deformity because of its firm fixation to the tibia. Rarely medial bending of the lower tibial metaphysis occurs (Schmidt, 1929).

Minor deformities can disappear due to corrective postrachitic growth. Major deformities become permanent and are followed by characteristic remodeling changes. The bent bones show a decreased transverse and increased anteroposterior diameter. The cortex of the convexity is markedly decreased and the cortex of the concavity is greatly increased. There is complete alteration of the trabecular pattern with new trabeculae crossing the medullary cavity in a radial arrangement, coverging to the concavity of the curve (Putschar, 1937:695–700). At this final stage it cannot be recognized whether the deformity was mainly caused by bending or by stress fractures or whether it occurred in the metaphysial or diaphysial portion.

RIBS.—The ribs may show flattening of their curves, secondary to bending of the rib cartilages at the osteocartilaginous junction. This leads to a forward bending of the sternum, giving the pigeon breast deformity (pectus carinatum). In severely malacic rickets, lateral depression of the rib contour may occur, secondary to the pressure of the arms (Putschar, 1937:600).

VERTEBRAE.—In severe rickets, the vertebrae may appear of decreased height due to compression, often combined with a deeper scalloping of the end-plate. Although kyphoscoliosis may develop after rickets, major abnormal curvatures are usually lacking in the active phase.

PELVIS.—The pelvis is more affected by altered growth than by mechanical deformation. The typical rachitic pelvis, during the active disease, is smaller and plumper than normal but does not show the disproportions of the adult postrachitic pelvis (Breus and Kolisko, 1900–1911(1):440–441). The typical postrachitic flat pelvis is characterized by an anteroposterior narrowing of the pelvic canal, mainly due to deficiency of growth of the iliac portion of the pelvic ring (Figure 430). The maximal growth period of this portion, especially from its posterior growth cartilage, falls into the infant period and is not made up later



FIGURES 428, 429.—Severe malacic rickets with very light, porous bones: 428, Both arms and shoulder girdle with bending deformities and healing stress fractures of all bones. 429, Close-up of left hand. (About 2 years of age, ANM 3334 and 3335.)

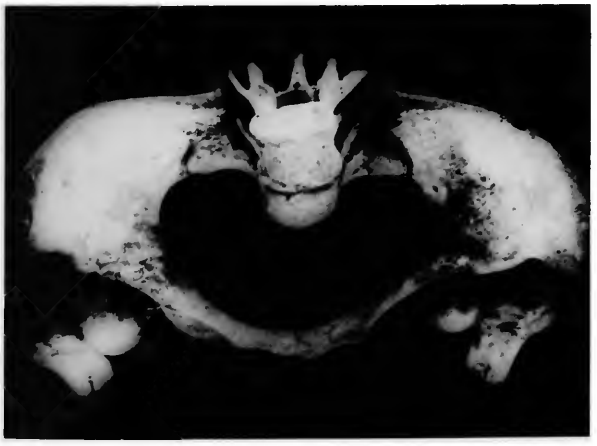


FIGURE 430.—Rachitic flat pelvis. (32-year-old female, VM 2 from 1860.)

while the maximal growth of the pubic bones falls into later childhood and adolescence. Therefore, the transverse diameter of the pelvic canal is not diminished and the pelvis appears flattened. Posteriorly also, the transverse diameter of the sacrum is not significantly affected (Breus and Kolisko, 1900–1911(1):469–476; Putschar, 1939: 483–492, 509–512). The sacrum tends to protrude more into the pelvic canal and the acetabula face more forward than normally.

In surveying all described findings, it seems obvious that many of the features of active rickets will not be recognizable in skeletal material after loss of the cartilage and disintegration of unmineralized osteoid. Craniotabes and the flaring distortion and cupping of metaphysial ends of

long bones and sternal ends of the ribs would have the best chance of being preserved. However, postrachitic deformities of the various bones of the skeleton would be readily recognizable.

Late Rickets

Rickets developing or recurring in later childhood and adolescence, at a time when bone growth has slowed down greatly, is more characterized by severe mechanical deformities than by changes at the growth plate resulting in deficient or altered growth. These deformities are essentially identical with those of adult osteomalacia.

Osteomalacia

The etiology of osteomalacia is identical with that of rickets—vitamin D deficiency. The difference in the manifestations is due to the fact that in the adult the skeleton is no longer growing; therefore, various changes on the cartilage and bones of the growth plate, which dominate the picture of rickets, are no longer present. As in rickets, the bone matrix formed during the disease remains uncalcified (osteoid). The first radiological changes are a diffuse diminished density of the otherwise normal-appearing skeleton, not distinguishable from osteoporosis. In dry bone the light weight and cardboard-like consistency is noted (Figure 431). Depending on the remodeling rate, it takes considerable time before the skeleton is sufficiently weakened to show radiolucent zones in mechanically stressed areas, due to replacement of the mineralized bone with osteoid (Looser, 1920; Milkman, 1930). In severe and protracted cases, mechanical deformation of the skeleton, due to fractures and pliability, dominate the picture (Figures 432, 433).

In the causation of osteomalacia, in addition to vitamin D deficiency, general malnutrition, especially deficiency of protein and fat, and low intake or loss of calcium and phosphorus, play an important role. This explains the great prevalence of the disease in the female, particularly between 20 and 40 years of age. Numerous closely spaced pregnancies and protracted lactation are a heavy drain on the mother of bone mineral and vitamin D. This loss to the fetal skeleton and, through the milk, to the infant is a significant factor in the pathogenesis of puerperal osteomalacia.

The disease used to be common in Europe in crowded cities and under poor economic conditions. Numerous cases have been reported from China and I have seen, even recently, many cases in India. There, skin pigmentation, shrouding clothing, and seclusion of women indoors (purdah) still contribute to the prevalence of the disease, in addition to the other factors enumerated above.

Although the deficiencies affect the entire skeleton, the most marked changes are observed in the bones that contain mostly cancellous bone and, therefore, have the highest remodeling rate. These are the ribs, sternum, vertebrae, and pelvis (Figures 434, 435). The skull and the solid cortices of the long bones of the extremities have a low remodeling rate. Therefore, deformities of osteomalacia rarely occur in these bones except in young, still growing individuals (late rickets and juvenile osteomalacia).

The specific change of osteomalacia is the deposition of unmineralized osteoid on the trabeculae of old bone and the replacement of resorbed old bone with osteoid. However, in contrast to rickets, subperiosteal deposits do not occur.

Looser-Milkman zones of great radiolucency, often followed by stress fractures only transecting the overburdened cortex, occur in characteristic locations and often symmetrically. The predilected areas are the ribs, the medial cortex of the neck of the femur and of the humerus, the pubic rami, and the lateral margin of the scapula. In many instances multiple such stress fractures occur in each area. In healing, internal and external callus is formed, which ultimately mineralizes.

The osteomalacic deformities are usually restricted to the trunk skeleton. The vertebrae are flattened with accentuated cupping of the endplates and dense trabeculae. There is angular kyphosis in severe cases, often at the level of the eighth or ninth dorsal vertebra, occasionally combined with a scoliotic curve (Jaffe, 1972:401). The

spinal deformity alters the shape of the thorax, which becomes deep and laterally narrowed. The ribs show a decreased curvature and the sternum is angulated and pushed forward. In very severe cases the ribs may, in addition, show inward bending, laterally, due to the pressure of the arms (Figure 436).

The most marked and most characteristic deformation is observed on the pelvis in puerperal osteomalacia (Figure 436). The fourth or fifth lumbar vertebral bodies descend and protrude into the pelvic canal. The iliac wings are rolled and folded inward. The pubic rami are pushed together with the symphysis pubis projecting, beak-like, forward. The ischial tubera are bent medially and the acetabular floors protrude inward. The sacrum is bent angularly forward in the lower portion (Figure 437). All these deformities narrow and partly obliterate the pelvic canal, reducing it to a cloverleaf-shaped space (Breus and Kolisko, 1900–1911(2):50–62; Putschar, 1939:535–540). Deformities of the long bones of the extremities are much less common. They consist most often of downward bending of the neck of the femur (coxa vara) or of the humerus. Deformities of the shafts of these bones are rare and due more to fractures than to pliability.

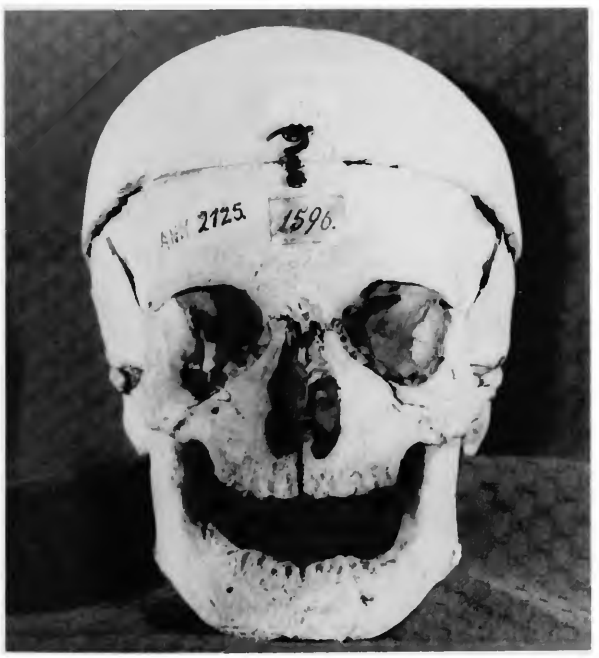
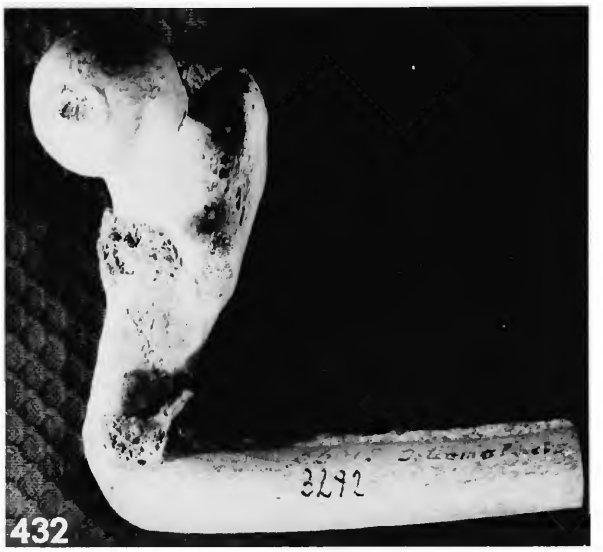
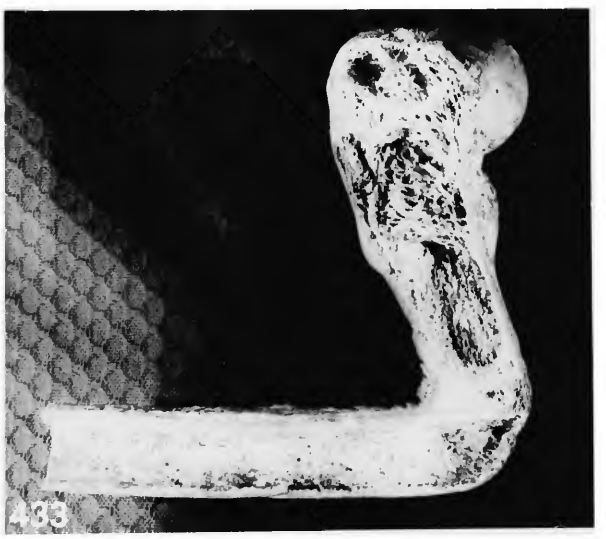
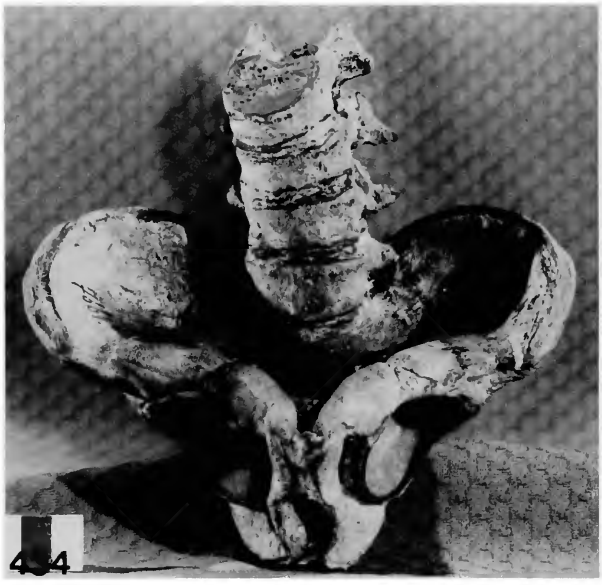


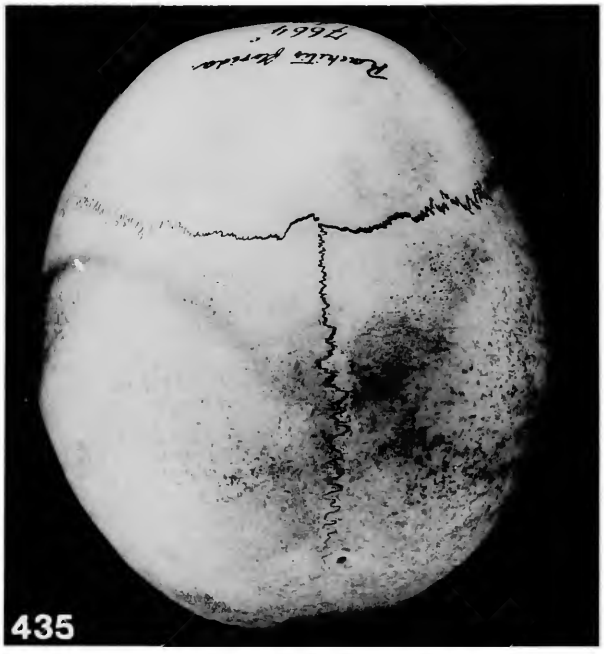
FIGURE 431.—Skull with severe active osteomalacia. Notice the diffuse porosity of the cortical surface; cardboard-like bones of very low weight. (20-year-old Italian male with multiple fractures, ANM 2125 from 1865.)





FIGURES 432, 433.—Angulated healed stress fracture in osteomalacia of right femur: 432, Surface view showing the unbroken cortex on the convexity and callus on the concavity. 433, Cut surface showing thin cortex and sparse cancellous bone. (ANM 3292.)





FIGURES 434, 435.—Active osteomalacia: 434, Pelvis and lumbar spine, showing maximal malacic deformity of the cardboard-like bones and delayed fusion of growth plates. 435, Calvarium, showing fine pitting and slight periosteal bone formation on the outer table. (18-year-old male with bending deformities of most bones, DPUS 7664c and 7664d from 1896.)

FIGURE 437.—Severe osteomalacia with angular kinking of sacrum from sitting. (43-year-old female, FPAM 5676.)

FIGURE 436.—Osteomalacic skeleton with severe pelvic and thoracic deformity. Notice the rib deformities caused by the weight of the arms, the angulated protruding sternum and the folded iliac wings. (Adult female, died after seventh pregnancy, PMES 1 (QAM(1).)

Although the osteoid of the active phase would not survive burial, the characteristic deformities that remain, even after healing and restored mineralization, will be readily recognizable.

Skeletal Changes Resembling Rickets and Osteomalacia

Excessive loss of dietary vitamin D due to inability of resorption from the intestines will lead to the changes of rickets or osteomalacia, depending on the age of the patient. Such conditions are celiac disease and nontropical sprue, chronic biliary stasis and biliary fistula. In the first, the fat-soluble vitamin D is eliminated with the fatty diarrheic stools; in the others, the bile salts necessary for the resorption of vitamin D are not present in the gut.

A group of syndromes characterized by various congenital defects in the renal tubules concerning excretion and reabsorption of different substances also, ultimately, affect the metabolic integrity of the skeleton (refractory or vitamin D-resistant rickets). The changes, when they occur, are essentially identical with rickets or osteomalacia, the only difference being that they do not develop until later childhood or adulthood. In those conditions in which various amino acids are lost through faulty renal excretion on a genetic basis, in addition to and preceding the manifestation of rickets or osteomalacia, one may observe severely inhibited growth (dwarfism) and severe osteoporosis leading to pathological fractures because of the inability to produce adequate amounts of organic bone matrix.

Paleopathology

Wells (1964a:116–117) expresses the opinion that medieval European towns and the "perpetual twilight of dark tenements," in the city slums, which were a prominent feature of cities during the Industrial Revolution, were the major factors in the marked increase of rickets associated with this phase of human history. Evidence of rickets in other geographical areas and other time periods is not common.

Hrdlička (1907:540) stated that in North America rickets did not occur in prehistoric indigenous groups. Snow (1948:508) describes a possible case of rickets in an 8-month-old infant (Burial 633, Indian Knoll site, Kentucky, USA). However, the description of features that lead to this conclusion is not convincing. Snow does not report any evidence of deformity in the long bones. He bases his conclusion on the presence of "swollen spongy extremities" in the smaller long bones and abnormal sternal ends of ribs. Since these are areas of active growth and consequent remodeling, they would normally appear somewhat spongy. In view of the lack of evidence of rickets in other North American skeletal series, this case should be reevaluated.

In the Old World, Wood-Jones (1910b:263) found no evidence of rickets in ancient Egyptian skeletal material. Virchow (cited in Pales, 1930: 40) thought that the distinctive features of European Neanderthal skulls were due to rickets. This possibility has not, however, received serious consideration by subsequent scholars who have studied the Neanderthal skeletons. In the Winchester Saxon collection of paleopathological specimens (BMNH CG 1966, no number) is a case of medieval healed rickets in a child about 6 years of age. The specimen consists of the right femur, tibia, and fibula. Anterior bowing is most pronounced on the femur and tibia (Figure 438). Another example of healed rickets is from the Royal College of Surgeons of England pathology collection at the British Museum (Natural History). The case is from the Ludgate Hill site in England (No. 1961, 5, 12, 5). The specimen is a right femur from an adult. There is marked anterolateral bowing with anteroposterior flattening and a buildup of bone along the linea aspera (Figure 439).

An example of rickets is found in the skeletal collections of the Historical Museum in Chur, Switzerland. The specimen is from the medieval archeological site of Bonaduz in canton Grissons, Switzerland. The skeleton in question is a male, probably over 50 years of age. The only evidence of deformity is seen in the tibiae. Both the right and left tibiae show bowing with an increased

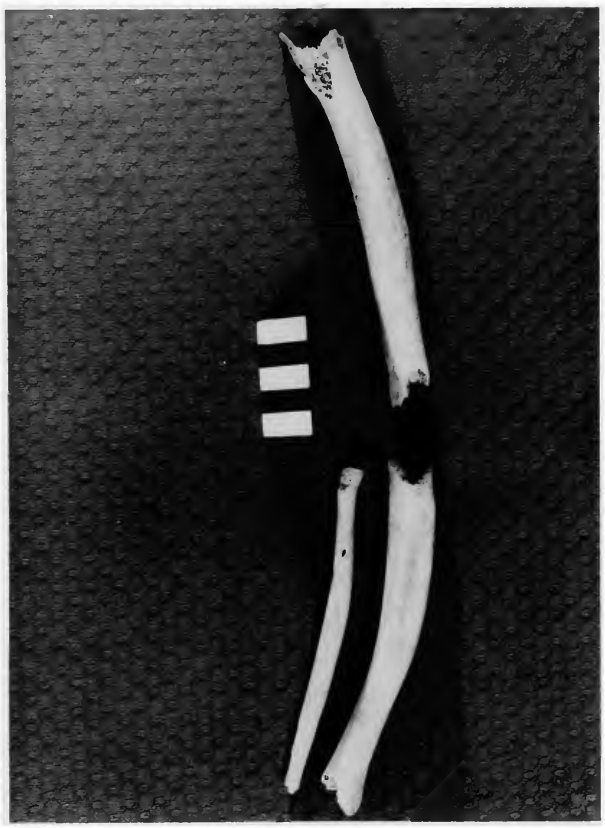


FIGURE 438.—Healed rickets in the right leg of a child about 6 years of age. Note that the interosseous line of the tibia is curved. (BMNH CG 1966, no number; scale in cm.)

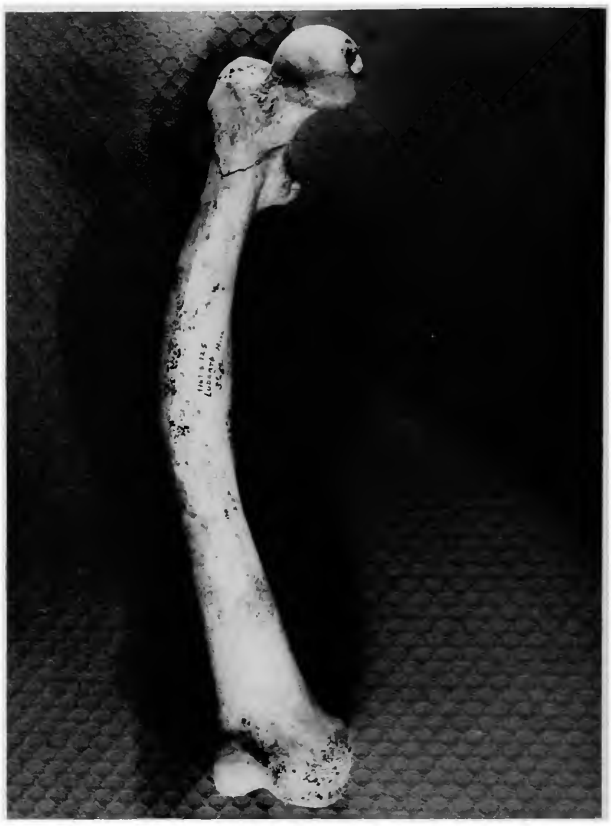


FIGURE 439.—Healed rickets in an adult femur. (BMNH, Ludgate Hill No. 1961, 5, 12, 5.)

anteroposterior diameter of the midshaft and a very prominent anterior edge (Figure 440).

Osteomalacia must certainly have occurred in ancient times although the evidence for it is scarce. A probable example of osteomalacia is seen in an isolated sacrum from the Twelfth Dynasty, rock tombs at Lisht, Upper Egypt (NMNH 256470 PI). The sacrum is small in size and unusually light in weight. It appears to be from an adult female skeleton. There is a 90 degree angle between the lower and upper portion of the sacrum (Figure 441) with the angulation primarily at the third sacral segment (cf. Figure 437).

Rowling (1967:275–278) describes a remarkable specimen from the Royal College of Surgeons of England pathology collection, which is stored at the British Museum (Natural History) (BMNH 178A). The specimen is from a predynastic Nubian and consists of the major bones of the arms and legs, the pelvis, and three lower lumbar vertebrae. The most striking feature of the specimen is the very extensive hypertrophic bone development on the femora. Rowling attributes this development to paraplegia with possible complications of rickets.

The authors have also studied this specimen during our survey of pathological specimens in 1974. In addition to the extensive hypertrophic bone associated with the femora, these bones show anterolateral bowing (Figures 442, 443). On the right femur, bowing is complicated by a

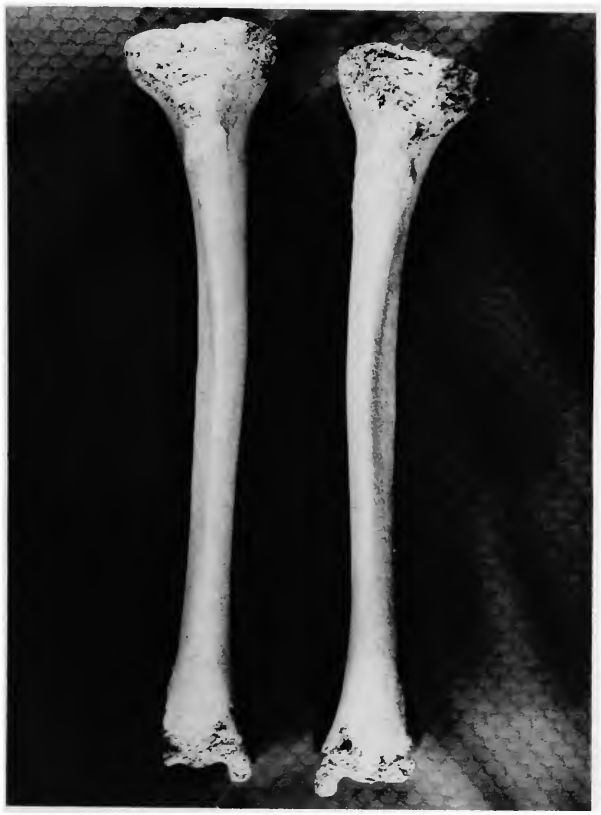


FIGURE 440.—Healed rickets in adult male tibiae. (HMCS GR 1352; no data.)

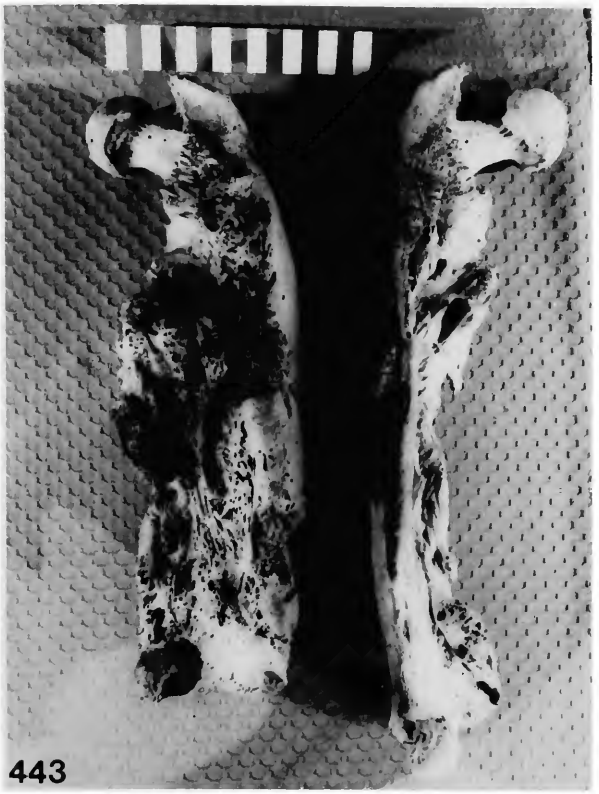
FIGURE 441.—Abnormally angulated sacrum probably due to osteomalacia. Isolated specimen from Twelfth Dynasty, rock tombs, Lisht, Upper Egypt. (NMNH 256470 PI.)

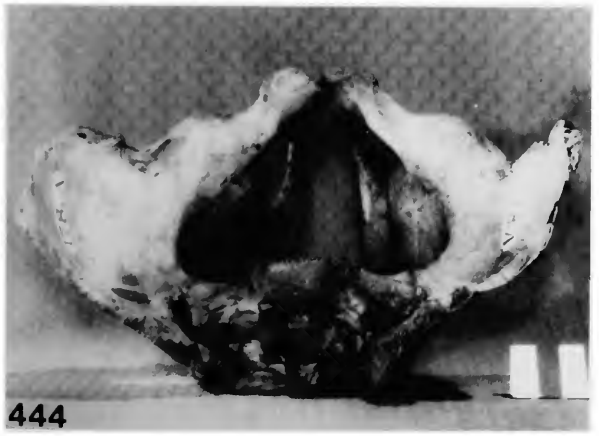
fracture with displacement and overlap of the broken ends. Both tibiae and fibulae show anterolateral bowing of a severe degree with extreme flattening, particularly of the fibulae, and partial synostosis between the fibula and tibia in the distal portion of the right side.

The humeri are straight but show a pronounced buildup of the deltoid insertion. Both radii and ulnae show severe lateral and anterior bowing with marked flattening and partial ossification of the interosseous membrane. Articular surfaces of all the long bones are normal. The pelvis is that of an adult male with a moderate deformity of the pelvic entrance, marked superior bulging of the acetabula, and greatly reduced subpubic angle (Figure 444). There is significant, periosteal, bony buildup, particularly on the lateral aspects of both iliac bones. The sacrum is angulated in its midportion anteriorly with a pronounced, smooth, bony ridge below the second intrasacral foramina.

The deformity of the long bones and pelvis illustrate typical changes in osteomalacia. The relationship of this disease with the exuberant development of bone, particularly on the femora, is not clear but is certainly not typical of osteomalacia. However, if Rowling's (1967:277) conclusion that the added bone is a result of paraplegia, osteomalacia could easily be a secondary effect of restricted outdoor activity due to paralysis. The unresolved problem with this hypothesis is that the bones of the leg show deformity indicating weight-bearing, which implies at least some ambulation involving the legs. While the sequence of events leading to this skeletal manifestation of disease may not be resolved, it is clear that osteomalacia did occur in geographical areas where it was theoretically unexpected.


442





FIGURES 442–444.—Deformed long bones and pelvis from osteomalacia with hypertrophic bone development of long bones perhaps developing as a complication of paralysis: 442, Deformity of ulnae, femora, and tibiae. The humeri are not bent. 443, Posterior view of femora. 444, Superior view of pelvis; note the deformity of the iliac bones and the greatly reduced subpubic angle. (BMNH 178A, Nubian Pathology Collection; scales in cm.)

Hypophosphatasia

PATHOLOGY

This is a rare condition, only fairly recently recognized (Rathbun, 1948), in which alkaline phosphatase is deficient in varying degrees on a genetic basis. Since alkaline phosphatase is an essential factor in the calcification mechanism, changes indistinguishable from ordinary rickets ensue in this condition. In severe cases, this deficiency is operative in the fetal period, resulting in the birth of a dwarfed infant with deformed long bones, a poorly mineralized cranial vault, and all the appearances of active rickets. These children usually live for only a few days or are stillborn. If the deficiency is of a lesser degree, rachitic changes may not develop until later childhood. These individuals survive with minor skeletal changes, of which osteoporosis is more obvious than rachitic stigmata. However, occasionally they do develop craniosynostosis (Jaffe, 1972: 440). (For a comprehensive discussion of this condition see Currarino, Neuhauser, Reyersbach, and Sobel, 1957).

Starvation

PATHOLOGY

Extreme prolonged malnutrition, especially protein deficiency, leads to a complete arrest of growth in the young individual that cannot adequately be made up later, resulting in stunted stature (Follis, 1948:9). In the adult, severe dietary restrictions can result in "hunger osteopathies," as observed after the First World War (Edelmann, 1919; Partsch, 1919; Beninde, 1920; Dalyell and Chick, 1921) and in inmates of European concentration camps in the Second (Maratka, 1946). The changes mainly affected the spine and occurred in both men and women, mostly in the second half of life. Such changes consisted of severe osteoporosis with some overlay of osteomalacia.

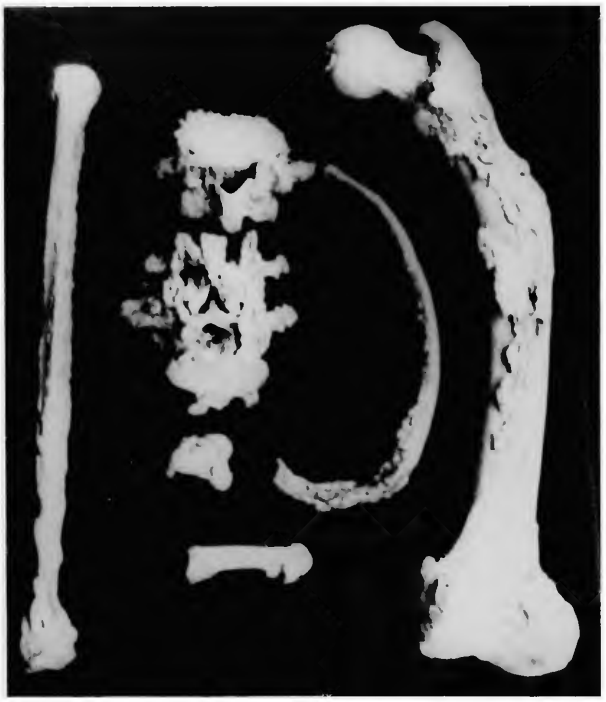


FIGURE 445.—Fluorosis, showing severe generalized periosteal hyperostosis of fibula, lumbar vertebrae, rib, talus, first metatarsal, and right femur, which shows a healed pathological fracture. (Adult Asian Indian male, studied by Putschar in 1965 at Department of Pathology, Guntur, India.)

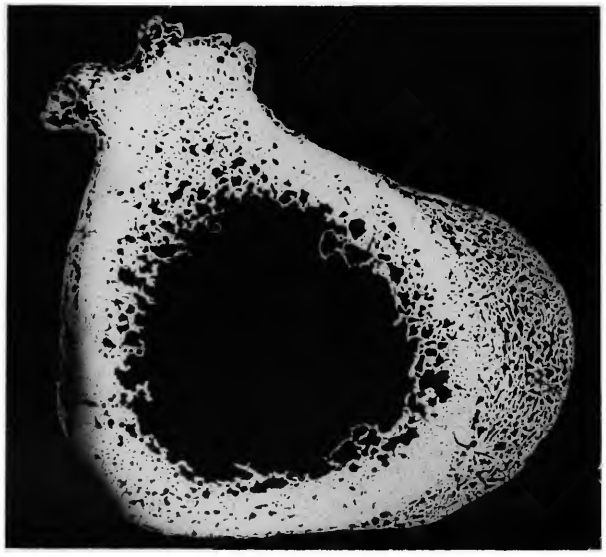


FIGURE 447.—Severe fluorosis. X-ray of cross-section of femur, showing massive subperiosteal bone deposition and some intracortical resorption. (Adult Asian Indian male, courtesy Dr. Niranjan Das Aggarwal, Rajendra Hospital and Medical College, Patiala, India.)

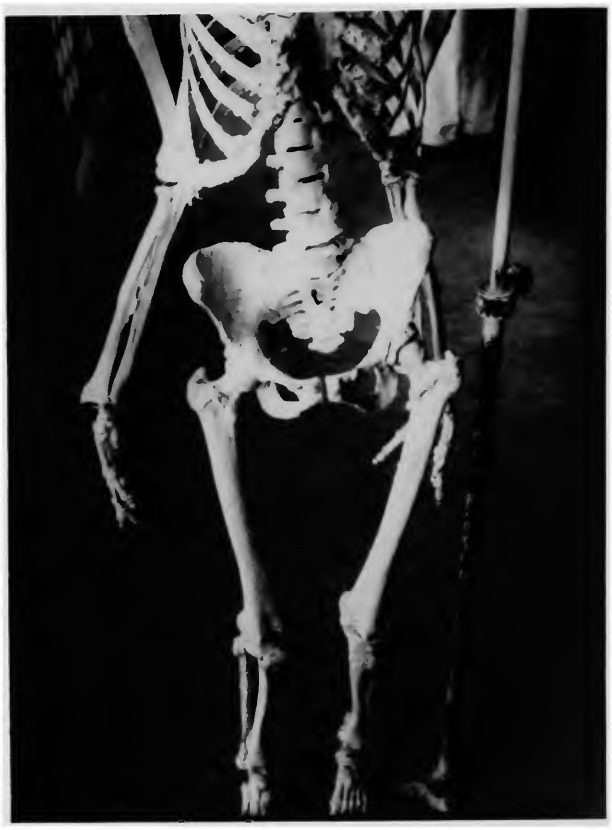


FIGURE 446.—Fluorosis with generalized periosteal hyperostosis. Notice the almost complete bony bridging of the interosseous membrane of the right forearm. (Adult Asian Indian male, studied by Putschar in 1965 at Department of Pathology, Medical School of Guntur, India.)

Fluorosis

PATHOLOGY

In certain areas of the world (Texas, USA, Iran, India, Taiwan) the surface water may have an unusually high fluorine content. The long-standing intake of high levels of fluorine can affect the structure and appearance of bone and teeth (Johnson, 1965). Fluorine can assume the position of the hydroxyl radical in the apatite crystal of mineralized tissues. In addition to the formation of abnormal apatite, fluorine has a toxic effect on the cells. The first noticeable effects of fluorosis are abnormalities of dental enamel, which, like bone, is mineralized by apatite crystals. It is mostly teeth of the second dentition, which are being formed during childhood, that

are affected. Such teeth show spotted enamel consisting of small, round, discolored and pitted areas in the otherwise shiny and white enamel surface. Bone changes are only observed in long-standing and severe cases (Flemming Møller and Gudjonsson, 1932; Sankaran and Gadekar, 1964). Fluorine stimulates periosteal and endosteal bone formation, but the bone produced is microscopically and mechanically abnormal. Externally, exaggeration of insertions of muscles and ligaments are noted by addition of bone-forming irregular excrescences (Figure 445). There is marked accentuation of the linea aspera of the forearm and lower leg (Figure 446). The ribs and vertebrae show ridges and icicle-like, bony excrescences into the intercostal muscles and paraspinal ligaments. There is considerable subperiosteal and endosteal bony accretion, often accompanied by increased resorption in the old cortex on long bones (Figure 447). The fluorosis bone is mechanically inferior and pathological fractures occur.

Osteoporosis

Pathology

Senile osteoporosis represents a condition of significantly diminished bone mass due to long-standing imbalance between bone resorption and bone formation. With increasing age, bone formation lags more and more behind bone resorption thus diminishing the entire bone mass. This leads imperceptibly into osteoporosis, which most observers arbitrarily define as a loss of 30 percent of bone mass or more. Osteoporosis usually does not manifest itself before the fifth decade and is more frequent and more severe in the female than in the male. This, at least in part, reflects the sharp drop in estrogens at menopause as compared with the slow steady decrease of testosterone in the aging male. However, comparative statistical studies between affluent industrialized populations and primitive populations requiring strenuous physical activity from both sexes, indicate that the mechanical forces play a great role in the development of senile osteoporosis. Severe osteoporosis, as measured by the incidence of fractures of the femoral neck, was highest overall and showed the greatest female prevalence in Sweden and the least in the Bantus (Chalmers and Ho, 1970).

Osteoporosis does not affect the entire skeleton evenly. Since the greatest physiological turnover of bone occurs in cancellous bone, the bones rich in spongiosa are most involved in osteoporosis. The bones of the trunk, such as the spine, ribs, sternum, and pelvis, are most severely affected by osteoporosis. Of the large bones, the heavily burdened femoral neck, which also mainly depends for its integrity on the trabecular architecture, shows the most characteristic changes. The skull, especially the calvarium, participates very little in senile osteoporosis. This may be, at least in part, due to the continued mechanical stresses of chewing.

The Cancellous Bones.—The vertebral bodies usually show osteoporosis first and most severely. The normal dense vertical and transverse trabecular system becomes reduced. In this process the transverse trabeculae are more affected than the vertical ones. The trabeculae are not only diminished in number but also in size. In late stages of osteoporosis the few remaining vertical trabeculae may become reinforced (sclerotic atrophy). Although the bone is normal in matrix structure and mineralization, the diminished quantity creates mechanical instability. If the intervertebral discs retain their normal turgor, they will bulge the thinned vertebral, bony endplates, creating the picture of so-called "fish vertebrae" (Figure 448). If the intervertebral discs are degenerated, this deformity of the vertebral body does not occur in spite of severe osteoporosis. Compression fractures resulting in flattened or wedged vertebrae are common, the latter resulting in secondary kyphosis. The other cancellous bones show similar changes characterized by rarefaction of the spongiosa and pronounced cortical thinning. The ribs also often exhibit multiple osteoporotic fractures which, however, heal with good callus response.

The Long Bones.—Since the physiological turnover in cortical bone is much slower and less than in cancellous bone, the long bones, with predominantly cortical bone, are later and less

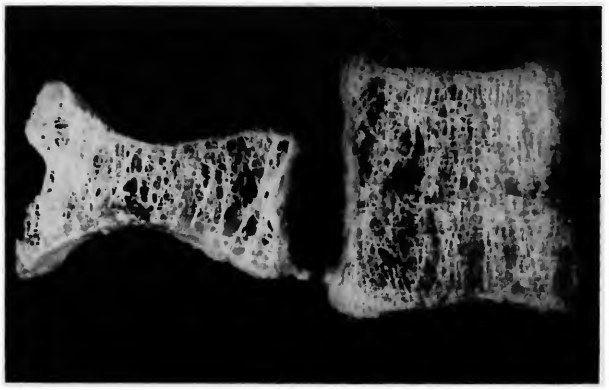


FIGURE 448.—Severe osteoporosis. Sagittal section of "codfish" vertebra (left) compared with normal vertebra (right). (Department of Pathology, MGH, no number.)

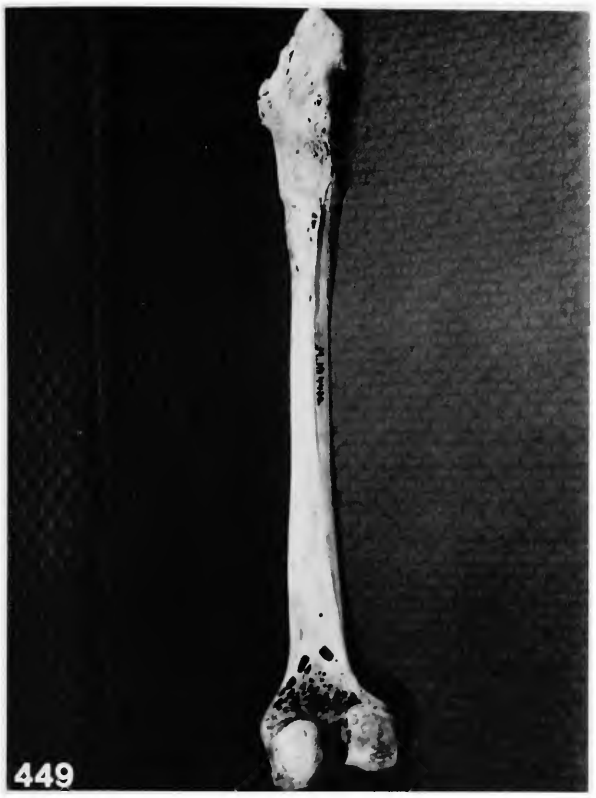
affected by senile osteoporosis. The resorption of the cortex proceeds in two patterns: the endosteal resorption, which leads to increased diameter of the medullary cavity, and the intracortical resorption, resulting in increasing numbers of unfilled or only partly filled Haversian resorption spaces. The latter process results in increasing porosity and lamination of the cortex which, in extreme cases, may assume a cancellous appearance, especially in advanced arteriosclerotic peripheral vascular disease.

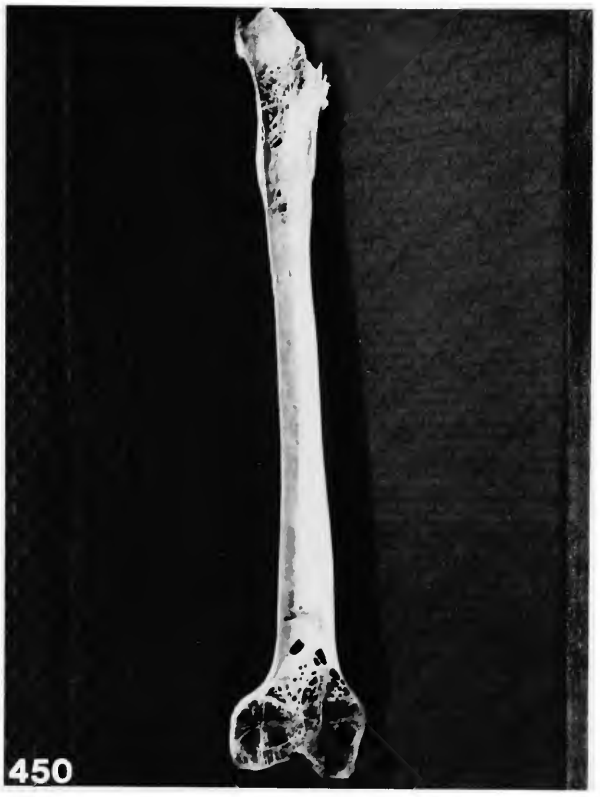
The frequency of pathological fractures of the neck of the femur (subcapital or transcervical) in advanced osteoporosis requires a discussion of the underlying structural changes in this area. The normal trabecular architecture of this eccentrically loaded, weight-bearing bone shows three trajectorial systems to distribute and absorb static and dynamic mechanical stresses: (1) The one system arising from the lateral cortex and (2) the system from the medial cortex (calcar), cross each other at right angles and terminate in the femoral head; (3) the third system, crossing the other two, arises from the area of the lesser trochanter, and terminates in the greater trochanter. It is particularly this third structure that first disappears in osteoporosis, accompanied by resorption of some of the other two trajectorial systems. This results in a roughly triangular area of the neck of the femur with sparse or absent bony trabeculae (Ward's triangle). This structural weakening precedes the pathological fracture, which often terminates in nonunion (pseudarthrosis) and aseptic necrosis of the femoral head. Impacted fractures may heal with shortening and widening of the neck of the femur.

Widespread osteoporosis may also be a secondary phenomenon in other conditions affecting the formation and resorption of bone. In osteogenesis imperfecta, inhibited osteoblastic activity but normal osteoclastic activity results in generalized osteoporosis. In Cushing's syndrome, the overproduction of adrenal corticosteroids likewise suppresses protein synthesis and, thus, bone matrix production. These conditions are discussed elsewhere (pp. 305, 337).

Localized osteoporosis is a frequent result of removal of mechanical stresses because of disuse, pain, trauma, infection or disturbed innervation (Figures 449, 450). In some cases posttraumatic osteoporosis can develop rapidly and out of proportion to the trauma (Sudeck's atrophy). This condition is, at least in part, caused by neurovascular changes affecting the circulation in bone, as demonstrated by the fact that the symmetrical uninjured area of an extremity may also show increased bone resorption.

Brief mention should be made of the occurrence of a peculiar symmetrical bilateral atrophy of the parietal bones. In this condition a large flat depressed area covered by a thin external table is observed. It is mainly the diploë that is diminished or absent. The outer and inner tables may be contiguous and the cranial vault transparent on transillumination. In a few cases the resorption is complete, creating a large defect surrounded by inward-sloping exposed diploë. This must not be confused with unusually large parietal foramina that show a smooth cortical margin (Goldsmith, 1922; Pepper and Pendergrass, 1936). The significance of the symmetrical parietal atrophy is not clear but is often thought to be related to senile osteoporosis (B. Epstein, 1953; Steinbach and Obata, 1957), while other observers attribute it to a developmental abnormality (Camp and Nash, 1944).





FIGURES 449, 450.—Extreme osteoporosis of right femur due to disuse and malnutrition. 449, External posterior view; notice reticulated perforations of metaphysial cortex. 450, Cut surface; notice paper-thin cortex and sparse trabeculae. (28-year-old female with tuberculosis of hip died in extreme cachexia, FPAM 4433, autopsy 81477 from 1884.)

PALEOPATHOLOGY

Osteoporosis, as manifested in the skeleton, involves two factors. The first of these is a reduction in the number and diameter of trabeculae accompanied by a thinning of cortical bone. The second factor is a change in osteon remodeling, in which the rate of refill of resorption spaces is diminished and the thickness of the osteon wall is reduced, leaving a larger central canal. Either one or both of these factors may be involved in what is seen as osteoporosis in dry bone specimens.

In an archeological case of osteoporosis the external appearance of the bone may be normal. Often the first indication that the disease is present in a skeleton is the greatly reduced weight of the bones. However, more subtle forms of bone loss have been studied by obtaining data on the relative amounts of bone marrow space and cortical thickness. Dewey, Armelagos, and Bartley (1969) studied bone loss in three Nubian skeletal samples. The authors found that osteoporosis in women began earlier in life among the ancient Nubians than in modern Western samples. They suggest that inadequate calcium intake and extended lactation, rather than hormonal conditions, were major factors in explaining the differences in the onset of osteoporosis.

Ericksen (1976) compared age-related changes in cortical thickness in three North American indigenous skeletal samples. In all three groups,

bone loss in females was much greater than in males. She also found a difference between populations in the rate of bone loss with age. She concludes that environmental factors, rather than genetics, is the major factor in the differences, and she speculates that nutrition and physical activity may be important.

Although the skull normally participates in osteoporosis to a lesser degree than other bones, alveolar resorption and thinning of the parietal bones do occur with increasing age. Lodge (1967) has described this condition in early Egyptian skeletal samples. He reports some support for an association between biparietal thinning and osteoporosis but concludes that other factors, such as a congenital predisposition, are probably involved. Stress from chewing may be another factor.

An interesting case of female osteoporosis was found in an Early Bronze Age shaft tomb (3150–3000 B.C.) at the site of Bab edh-Dhra in Jordan (NMNH, no catalog number, Tomb A 100E, Burial 2). The estimated lifespan of the individual was in excess of 50 years. The skeleton is from a secondary burial and many of the smaller bones are missing. The remaining bones are very delicate and fragments have been broken off.

The most noticeable changes are in the pelvis and vertebrae. The sacrum, for example, weighs 42 grams while another sacrum almost identical in size and from the same tomb chamber weighs 68 grams. This indicates a loss in bone mass in the osteoporotic sacrum of about 38 percent.

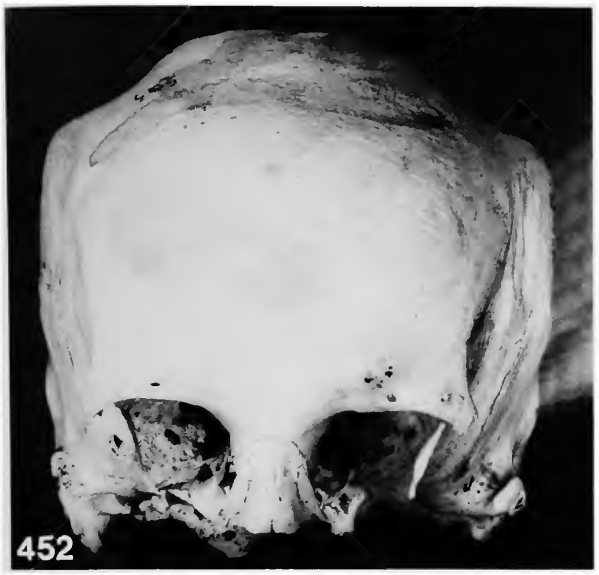
The gross size and shape of the vertebrae and pelvis are normal although there is a slight bulge in the pelvic canal from the femoral heads. The surface of the bone has patches of porous bone, particularly in areas adjacent to muscle or ligament attachments.

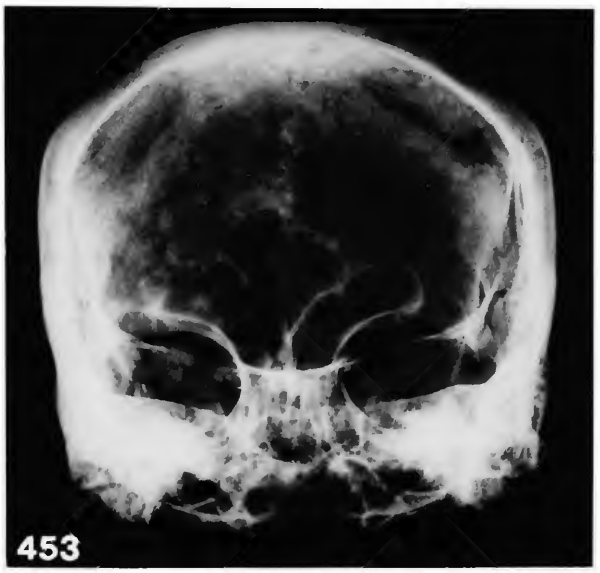
Although the long bones are less severely affected, roentgen films of the humeri show cortical thinning and loss of density characteristic of osteoporosis. An additional complication in the bones of the lower legs is the presence of osteomyelitis. This condition is severe on the left tibia and fibula, where there is also evidence of an overlying ulcer on the distal shaft of these two bones.

FIGURE 451.—Biparietal atrophy of the external table and diploë in an adult male skull from Peru, top view. (NMNH 294027.)

The bones of the skull and mandibles are normal given the age and sex of the skeleton. There has been little if any alveolar resorption and no biparietal thinning of the skull vault. Most teeth were lost or broken postmortem. Those that remain show unusually little wear but some evidence of caries.

Biparietal atrophy is not common in archeological skulls. Two examples from the collections of the National Museum of Natural History, Washington, D.C., USA, serve to document the presence of this condition in both the New and Old World. The first of these (NMNH 294027) is a male skull from near Santa Lucia in Peru (Figure 451). The archeological age is unknown. On the posterior portions of the parietals there is a large oblong depression in the outer table, measuring 55 by 40 millimeters on both sides. The inner surface of the inner table is unaffected. The edges of the depressed lesion are smooth although there is still evidence of the exposed spongy bone. This





FIGURES 452–454.—Biparietal atrophy in an old male skull from the Twelfth Dynasty near Lisht, Upper Egypt: 452, Facial view. 453, X-ray of anteroposterior view. 454, Posterior view. (NMNH 256186.)

suggests that the atrophic condition involves the outer table and diploë with, in extreme cases such as this one, some thinning of the inner table. There is no evidence of abnormal loss of bone thickness or density in other parts of the skull.

The second example is a skull from the Twelfth Dynasty site near Lisht in Upper Egypt (NMNH 256186). The skull appears to be from an old male. The oblong atrophic lesions are located in the posterior region of the parietals (Figures 452–454). The depressions are smooth but show remodeled spongy bone. The lesions are even larger than those in the skull from Peru. The morphology of the atrophic depressions suggests a process that primarily involves the outer cortex and the diploë. The remainder of the skull is normal.

Localized Hyperostosis

Pathology

Pregnancy Osteophyte

There are a variety of syndromes characterized by formation of mostly periosteal bone. The first of these is pregnancy osteophyte. During and shortly after pregnancy, one commonly observes a thin, chalky layer of surface parallel periosteal bone on the inner surface of the internal table of the cranial vault, especially in the frontal area, and to a lesser extent on the cranial base. This layer does not exceed 0.5 millimeter in thickness (Henschen, 1949:62–64). Concomitantly, there may be osteosclerosis of the diploë (Haslhofer, 1958). Burkhardt (1970:166–167) observed that the cranial vault is dense and heavy in pregnancy. These changes are apparently the result of altered pituitary hormone secretion.

Internal Frontal Hyperostosis

This is a common finding almost only occurring in aging females, particularly after menopause. Henschen (1949:85) estimates the female to male ratio of incidence close to 100:1. The lesion is usually restricted to the endocranial surface of the frontal bone but occasionally involves the temporal and parietal bone. The changes consist of marked thickening of the frontal bone, presenting a ridged buildup on the inner surface of considerable thickness. On cross-section much of the new bone is cancellous and has become part of the diploë. The inner surface shows a thin, but continuous, cortical layer. This alteration, which also is the result of changes in pituitary hormones after menopause, is frequent enough to serve as a means of sexing and aging in archeological material. (For a detailed discussion, see Henschen, 1949, and S. Moore, 1955.)

Leontiasis Ossea

This is a rare condition in which long-continued excessive bone formation on the cranial and facial bones leads to marked distortions of the normal features. The periosteal buildup and diploic sclerosis may be uniform (Figures 455, 456) or nodular and lead to narrowing of the orbits and of the paranasal sinuses (Figure 457). The changes would be obvious on dry bone and the only differential diagnosis would be Paget's disease. However, Paget's disease of such severity would not likely be limited to the skull. Microscopically, the characteristic mosaic pattern of Paget bone is not present in this condition. (For detailed discussion, see Knaggs, 1923–1924, and Jaffe, 1972:276–281.)

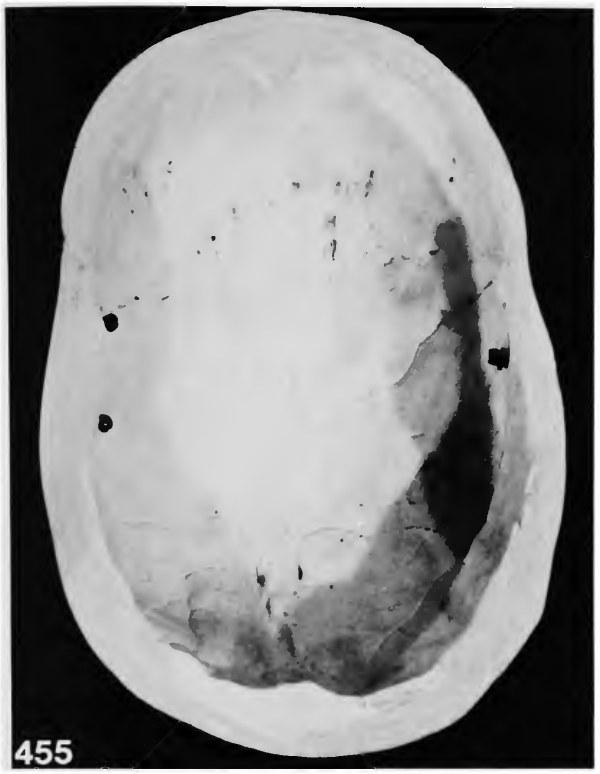
Infantile Cortical Hyperostosis (Caffey's Disease)

This is a disease affecting the skeleton of infants in the first year of life (Caffey and Silverman, 1945; Caffey, 1957b, 1972(2):1210–1222). The cause is unknown. It occurs sporadically or occasionally in several siblings. The lesion is characterized by massive deposition of layered periosteal woven bone on one or several bones. It is not a systemic disease. The bone deposits usually disappear spontaneously after weeks, months, or rarely years. The disease is rarely fatal. The most frequently involved bones are the mandible and the clavicle, followed in frequency by the long tubular bones of the extremities and ribs. The small tubular bones, most of the cancellous bones, and the skull are usually spared. The periosteal bone deposits on long bones are mainly over the diaphysis, less over the metaphyses, and spare the epiphyses. They are usually distinctly separated from the underlying cortex, which may become osteoporotic. The deposits can be so massive that they exceed the normal diameter of the involved bone. The surface of the deposits is rough and the contour uneven. If both bones of the forearm or lower leg are involved, the bony mantles of both may become fused.

Generalized Hyperostosis with Pachydermia

Pathology

This is a rare familial disorder characterized by excessive formation of dermal collagen and of periosteal bone. The disease usually begins



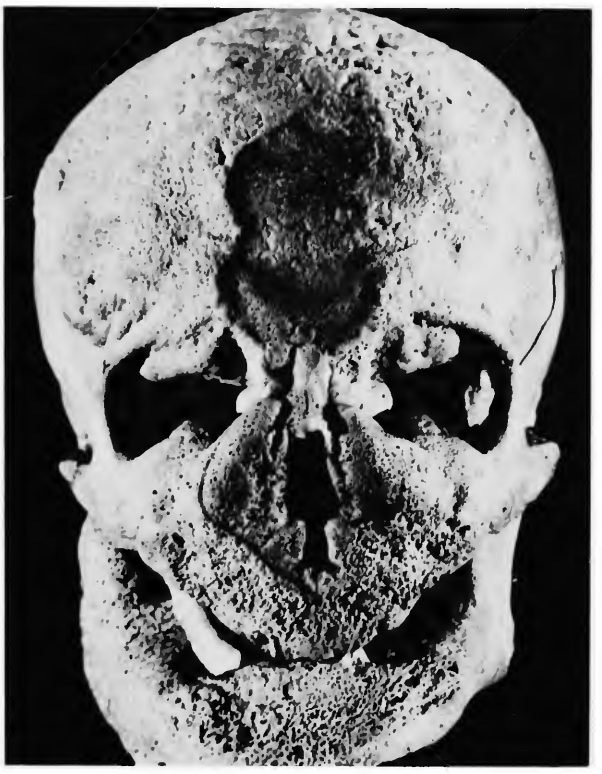
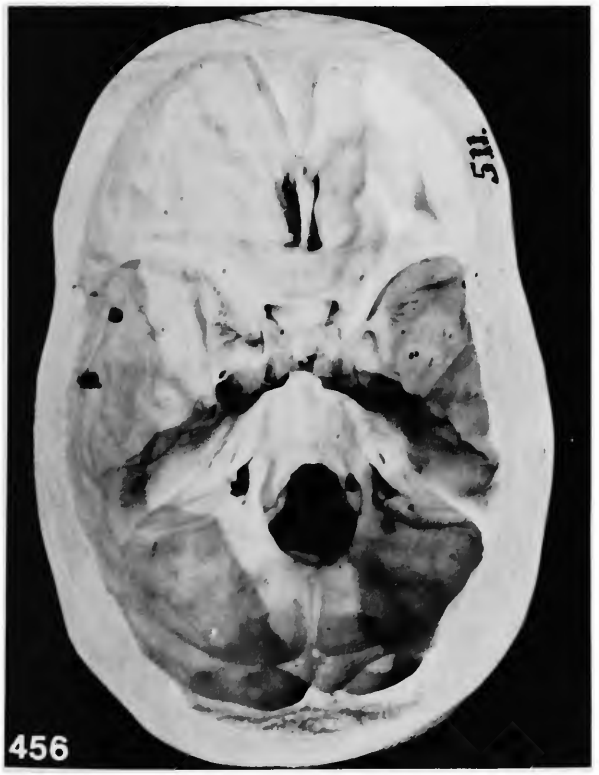
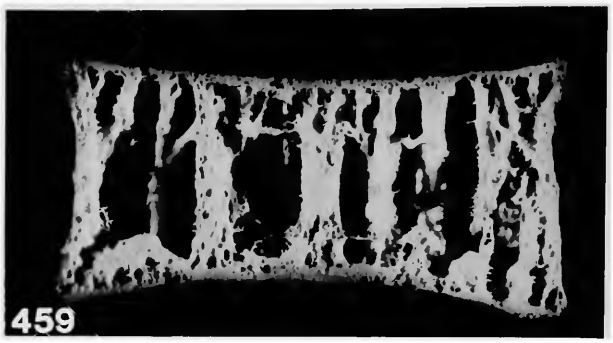


FIGURE 457.—Leontiasis ossea of cranium and facial bones, frontal view. Notice the massive deposition of periosteal bone. The paranasal sinuses were obliterated by bony overgrowth. (Adult Peruvian, WM S65.a from 1828.)



FIGURES 455, 456.—Leontiasis ossea. Notice thickening of cranial vault and ivory-like sclerosis of the diploë without encroachment on the cranial cavity; skull weight 2500 g. 455, Endocranial view of calvarium. 456, Endocranial view of base. (56-year-old female, FPAM, Jubiläumspital 511, from 1919.)



FIGURES 458, 459.—Generalized hyperostosis with pachydermia: 458, Anterior view of skeleton, showing excessive periosteal bone; notice sparing of joint ends and preservation of medullary cavities. 459, Vertical section of vertebral body, showing thickened, coarse trabeculae. (55-year-old male, IPAZ autopsy 1146 of 1940.)

around puberty and progresses through adult life. The outstanding feature in advanced cases is the markedly increased diameter of long bones, without alteration of the joint ends (Figure 458). The surface of the bones shows a rough irregular texture. On cross-section the new bone blends into the old cortex, which may show considerable porosis. The medullary canal is not obliterated. The skull usually shows only sclerosis of the skull base and of the diploë. The cancellous bones are less thickened and their trabecular architecture becomes coarse and exaggerated (Figure 459). The pelvis shows rough osteophytes and exostoses. In late stages spinal ligaments and interosseous membranes may be ossified. Large and small joints may become ankylotic. Carpal and tarsal bones may be transformed into solid bony blocks. (For a detailed discussion see Uehlinger, 1941.)

Paleopathology

Allison, Gerszten, Sotil, and Pezzia (1976) have described an archeological case of generalized hyperostosis with pachydermia in an adult female skeleton. The case is from a Huari-culture cemetery, which is dated about A.D. 1000 and is located in the Ingenio Valley, Ica, Peru. The bones of the skeleton are thickened and have greatly diminished marrow spaces. This has resulted in the bones being as much as 2½ times heavier than normal. Histologically the bone tissue, while thicker than normal, has good lamellar bone with normal osteon remodeling.

Endocrine Disturbances

The skeletal growth and maturation is mainly controlled by an intricate interrelationship between pituitary and thyroid. This interrelationship can be separated only in the animal experiment, revealing that the pituitary mainly controls growth and the thyroid maturation (Asling and Evans, 1956). Estrogens and androgens of testicular or adrenal origin stimulate longitudinal growth but hasten epiphysial closure. (For a general discussion of endocrine-skeletal relations see Putschar, 1960:405–414.)

Pituitary Disturbances

Pathology

The anterior lobe of the pituitary produces somatotrophic (growth) hormone, with direct action on skeletal growth, and thyrotropic hormone, which stimulates the production of thyroxin by the thyroid. Excessive production of growth hormone by hyperplasia or adenoma of the anterior pituitary produces characteristic skeletal changes besides effects on other tissues and organs not discussed here. Depending on the age of the individual affected by hyperpituitarism, either pituitary gigantism or acromegaly will result.

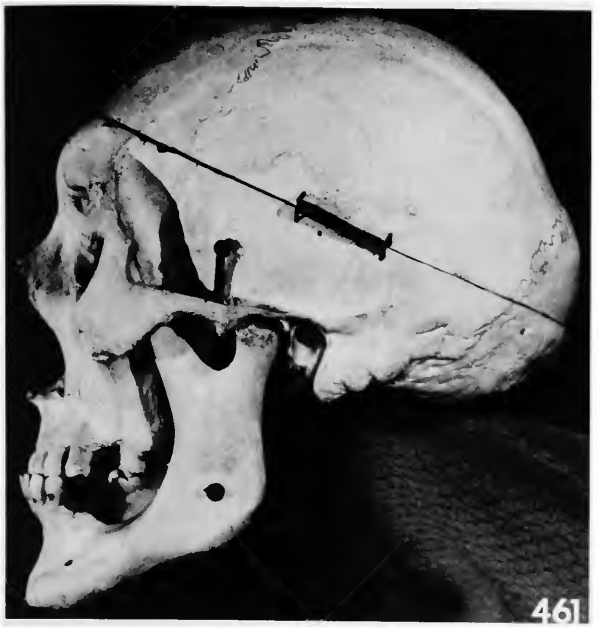
Pituitary Gigantism

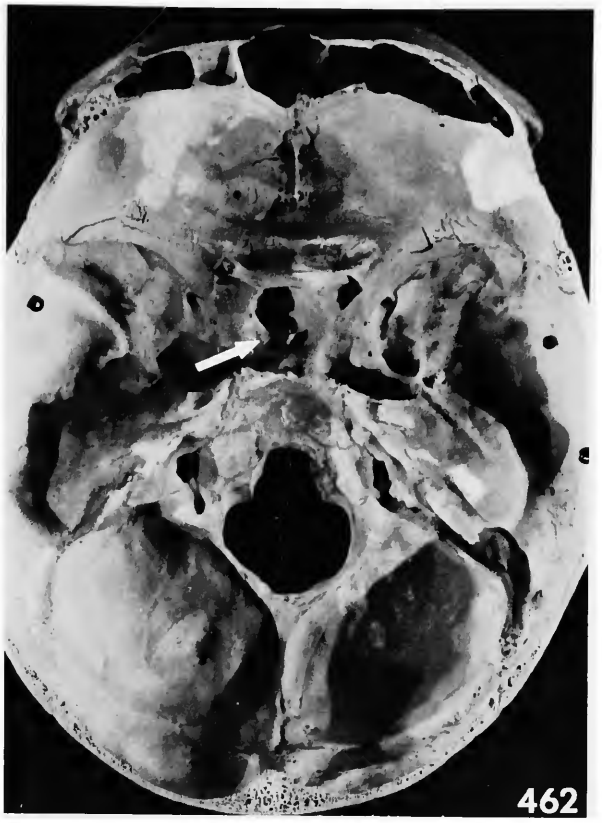
This is a very rare condition in which the continued excessive production of somatotrophic hormone during the growing period and beyond leads to gigantic proportions of the skeleton by overstimulation of the growth cartilages. In this condition, not only the total body height but also the length and diameter of all bones are enlarged. If the underlying cause was a tumor, the sella turcica is usually markedly enlarged with thinning or destruction of the anterior and posterior clinoid processes and, occasionally, with perforation of the bony floor of the sella. This condition results in an open communication between the sella and the cavum sphenoidale in the dry skull. In pituitary hyperplasia the contour of the sella may not be significantly altered. Continued hyperpituitarism beyond the growing period leads to a superposition of acromegalic features onto the giant skeleton (Figure 460).

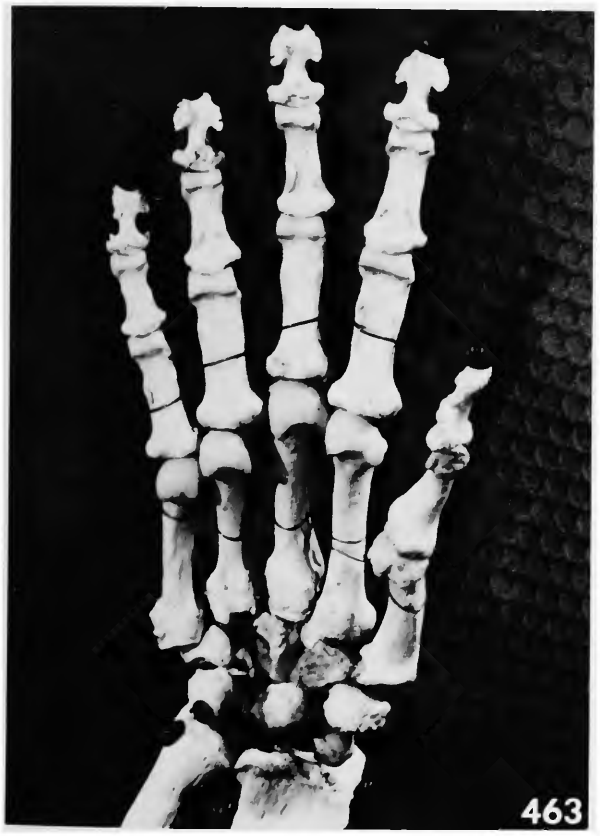
Acromegaly

This condition is much more common than pituitary gigantism and is usually the result of an acidophilic adenoma of the pituitary occurring in an adult (Figures 461–464). After closure of the

Figure 460.—Pituitary giant. (42-year-old Iranian male exhibiting giant stature and massive periosteal, especially cranial, bone apposition, Pahlavi University Medical School, Shiraz, Iran.)







FIGURES 461–464.—Acromegaly: 461, Skull, lateral view, showing prominent chin, malocclusion of teeth and prominence of inferior and superior nasal spine. 462, Skull base, showing enlarged frontal sinuses, enlarged sella (arrow) with resorption of anterior and posterior clinoid processes. 463, Right hand, showing extreme arrowhead tufting of terminal phalanges, and periosteal hyperostosis. 464, Lumbar vertebrae, showing marked subperiosteal apposition. (39-year-old male with pituitary adenoma, FPAM, Jubiläumspital 610.)

epiphysial plates, only the remaining cartilages and the periosteum can respond to stimulation by growth hormone. Therefore, there is no gigantism in acromegaly. The reawakened growth mainly affects the cartilages of the mandibular condyle, the rib cartilages, the synchondroses of sternum and pubis and, to some extent, the articular cartilages and intervertebral discs. The periosteal bone deposition is most marked at terminal points of bones and at normal prominences and insertion of tendons and ligaments. Usually, the skull shows enlargement of the sella, as described above, as a result of the pressure erosion of an intrasellar tumor (Figure 462). There is exaggeration of the supraorbital ridges and of the posterior occipital protuberance. The facial bones are enlarged and so are the paranasal sinuses. The cranial vault is thickened. The most marked changes affect the mandible. Since the cartilage of the mandibular condyle is a growth cartilage, marked elongation of the mandible with elongation of the ramus leading to pronounced prognathism and dental malocclusion result (Figure 461).

There is marked periosteal bony buildup on the chin, adding to the distortion of the mandible. Marginal buildup of bone on the alveolar process of the mandible and of the maxilla leads to separation between the teeth. The ribs show marked elongation, increasing the diameters of the thorax. The external relief on the bones is exaggerated on all prominent points, such as trochanters and linea aspera. The periosteal buildup is slow and consists of lamellar bone, intimately welded to the old cortex. The tufts of the terminal phalanges are characteristically enlarged with an arrowhead appearance (Figure 463). The changes of the articular cartilages are not directly visible on dry bone but the complicating degenerative arthritis is. The spine may show marked periosteal buildup with anteroposterior and lateral enlargement of the vertebral bodies, complicated by degenerative arthritic lipping (Erdheim, 1931a:203–210) (Figure 464). There is often kyphosis, mainly secondary to muscular weakness.

Hypopituitarism

Hypopituitarism only affects the skeleton significantly if the functioning pituitary is destroyed during the growing period. This is most often due to a tumor of developmental remains of Rathke's pouch (craniopharyngioma) developing within or above the sella in a child. Malformation or traumatic destruction of the pituitary is rarely the cause. Hypopituitarism in the adult produces no demonstrable skeletal changes beyond nondescript osteoporosis.

Pituitary Dwarfism

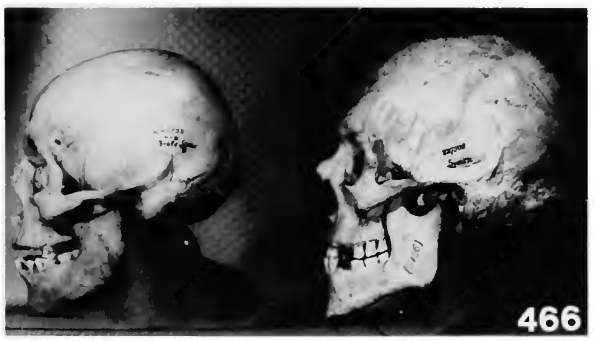
Deficiency of growth hormone in early life leads to pronounced inhibition of longitudinal growth, resulting in dwarfism of infantile proportions (Priesel, 1920). The synergism between pituitary and thyroid is demonstrated by the concomitant hypothyroidism secondary to the deficiency of thyrotropic hormone. Therefore, growth is not only stunted but the development of secondary ossification centers is markedly delayed, and the epiphysial and apophysial growth plates remain open long into adult life (Erdheim, 1916: 338–344, 356), although they do ultimately close in old age (Priesel, 1920:252). The cranial sutures also remain widely open into adult age. The sella may show widening, if the tumor was intrasellar, or an aperture at the floor as a remnant of a persistent craniopharyngeal canal. The skeleton is gracile in dimensions, the cortices are thin, and the spongiosa porotic and sparse. Although growth plates may remain, the metaphysial surface is usually closed by a thin layer of bone, indicating arrested growth. (For detailed bone measurements in hypopituitary dwarfs, see Priesel, 1920.)

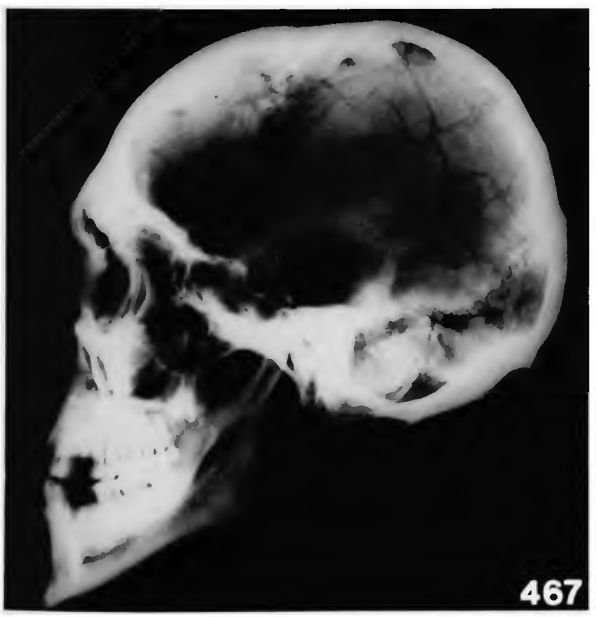
Paleopathology

Acromegaly

Brothwell (1965a:158) describes an ancient Egyptian skull in which the face is unusually long. The photograph of the skull (Brothwell, 1965a, pl. 4c) shows an enlarged mandible and

an apparent overgrowth leading to prognathism and malocclusion. Brothwell's diagnosis of acromegaly is almost certainly correct.





Morse (1969:26, pl. 22b) provides a brief summary of features associated with acromegaly accompanied by a lateral photograph and roentgen film of the skull and mandible of a Late Woodland (ca. A.D. 900) burial from Dick's mound in Illinois, USA. He describes the long bones as elongated and thickened and the lower jaw as lengthened. The photograph of the roentgen film and description of the long bones support a diagnosis of acromegaly.

The almost complete skeleton of an indigenous North American from the Midwest, USA (NMNH 227508) provides another possible example of acromegaly. The skeleton is from the historic period and is remarkable because of the massive size of both skull and postcranial bones. The maximum length of the skull is 201 millimeters, maximum breadth is 144 millimeters. The cranial capacity is estimated to be 1865 cubic centimeters, which is well within the normal human range but about 400 cubic centimeters greater than average. The gross morphology of the skull is rugged (Figures 465, 466) with pronounced and unusually high markings (near the sagittal suture) for the origin of the temporalis muscle. The mandible has slightly overgrown the development of the maxilla and has prominent bony projections on the anteroinferior border of the chin. The roentgen film demonstrates clearly the additional growth of the mandibular condyles and the increased angle between the ramus and mandibular body. The roentgen film (Figure 467) reveals a large but not abnormal pituitary fossa; thus the presence of a pituitary tumor is problematical.

The long bones are long and heavy. The right femur, for example, is somewhat damaged postmortem, but the estimated length is 545 millimeters. Stature estimates based on this length are

FIGURES 465–467.—Possible acromegaly: 465, Comparative facial views of normal (left) and possible acromegalic (right) skulls from the same skeletal sample. 466, Left, lateral view; note the projection of the chin in the skull on the right. 467, X-ray of lateral view of possible acromegalic skull. (Normal skull, NMNH 243703; possible acromegalic skull, NMNH 227508.)

189.75 centimeters (ca. 75 inches). This is certainly at the upper end of the normal range among American Indians but not abnormal. This suggests somewhat greater pituitary function during growth but with major morphological changes, as in the skull, occurring after the normal growth period had ended.

The skeleton appears to be fully adult and male with estimated age, based on the morphology of the fragmentary pubic symphysis, about 35 to 45 years of age. The ends of the ribs show evidence of secondary growth as would be expected in acromegaly. The bones of the hands and feet are large but of normal proportions, although the phalanges, particularly the terminal phalanges, show evidence of secondary growth and remodeling on the distal ends (tufting). The abnormal features of this skeleton are compatible with a diagnosis of acromegaly. However, the disease process does not appear to be as severe as cases encountered in the European pathology collections.

Pituitary Dwarfism

I know of no references on pituitary dwarfs in the literature on paleopathology; however, there is a probable case of this condition in the collections of the National Museum of Natural History, Washington, D.C., USA (NMNH 314306). This remarkable case is from the Hawikuh site in New Mexico, USA, which includes late precontact and early historical components. The skeleton is fragile and damaged but includes most of the bones.

The features of dental and skeletal maturation used in estimating age are affected by a deficiency of pituitary hormone. Therefore, the age of the skeleton cannot be determined with certainty. However, the second permanent molars have erupted. There is no evidence of a third molar in the left mandibular fragment; however, this absence might be due to agenesis rather than young age. All the teeth that are present are normal in size, which has created severe crowding in the small jaws.

In the postcranial skeleton most epiphyses are unfused. The primary elements of the innominate have fused, indicating an age in excess of 12 years. The ischiopubic ramus is unfused. The distal epiphysis of the humerus has fused. These features added to the dental eruption clearly indicate an age in excess of 12 years in a normal individual. Because of the delayed development and fusion of epiphyses in pituitary dwarfism, a minimal age in the twenties would seem more likely for this case.

The skull is somewhat deformed postmortem but is obviously much smaller than normal (Figures 468, 469). Maximum length is 145 millimeters, maximum width is 108 millimeters. Typical skull measurements for a 12-year-old child from the same site would be maximum length 160 millimeters and maximum width 127 millimeters. The relative proportions of the skull are normal although more typical of a child than a 20 year old. Unfortunately, the base of the skull has been damaged and lost, making observations on the pituitary fossa impossible.

The postcranial long bones are very slender and shorter than normal (Figures 470, 471). Stewart (1968:133) indicates that the approximate femoral length (without epiphyses) of a 12 year old is 310 millimeters. The femoral length of the dwarf is about 280 millimeters or 90 percent of the expected length of a 12 year old. If, as I think likely, the dwarf was fully grown the expected femur length would be about 370 millimeters and thus would have been only three-fourths as tall as normal. The torsion angle of the left femur is unusually large and probably would be associated with an abnormal gait during life.

The vertebrae are small but of normal proportions, although the bodies of thoracic vertebrae T5–T11 show a slight eccentric development to the right, which might be associated with a slight scoliosis. The sacrum and innominates are small but of fairly normal proportions, although the anteroposterior dimension of the pelvis is longer than would be expected. The pubic symphysis is poorly formed lacking the normal features of the symphysial face and the ridge formation. The bones of the hands and feet are of normal shape although very small.

The overall appearance of the skeleton conveys

FIGURES 468–471.—Pituitary dwarf: 468, Comparative facial views of normal (left) and dwarf (right) skulls: normal skeleton selected on basis of comparable dental and epiphyseal development and closure. 469, Right lateral view of skulls; note the prognathism of the dwarf skull. 470, Compartive anterior views of a right normal and abnormal femur and tibia. 471, X-ray of same long bones. (Normal bones, NMNH 308611; dwarf, NMNH 314306.)

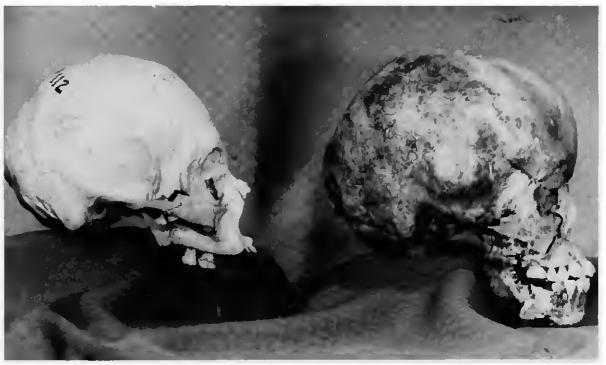


FIGURE 472.—Possible congenital idiot skull (left) compared with pituitary dwarf skull (right), right lateral view. Note the different contour of the frontal bones. (Congenital idiot, NMNH 379510; pituitary dwarf, NMNH 314306.)

the impression of diminished growth and development. The features of the face, particularly the large but depressed nasal aperature, are suggestive of features seen in achrondroplasia. The general morphology of the skull and long bones, however, would rule this possibility out. In general there has been diminished growth and delayed fusion of epiphyses, both of which are conpatible with a diagnosis of pituitary dwarfism.

Another specimen in the collection of the National Museum of Natural History, USA, is of interest in the context of pituitary dwarfism. The specimen consists of only the skull from Chilca, Peru (NMNH 379510). The archeological age is unknown. The skull was initially obtained by J. R. Wells who published his observations (1942). He reports that the skull was a microcephalic with a cranial capacity of 485 cubic centimeters. He concludes that the skull does not resemble that of a congenital idiot as described in the clinical literature.

Wells contributed the specimen to the National Museum of Natural History, USA, and Hrdlička (1943) added additional data and observations in a subsequent report. Hrdlička (1943:77) concluded that except for its small size, "the skull is unquestionably a 'normal' specimen, i.e. it shows nothing of any pathological nature."

The profile of the Peru skull reveals diminished development of the frontal region of the skull (Figure 472). A measure of this feature is the length of the arc from nasion to bregma. On the Peru skull this arc is 76 millimeters. This arc on the New Mexico pituitary dwarf is 100 millimeters. To the extent that diminished frontal bone development is indicative of brain abnormality, the Peru skull would appear to be a congenital idiot rather than a pituitary dwarf or microcephalic.

Other Endocrine Disturbances

PATHOLOGY

Hypothyroidism

Thyroxin, the thyroid hormone, is secreted by the follicular epithelium under stimulation by the thyrotropic hormone of the anterior pituitary. The effects of thyroxin are, in addition to the general stimulation of the metabolism and local effects on the skeletal maturation, a stimulating feedback on the pituitary to secrete somatotrophic hormone. The main skeletal alterations are seen in severe thyroxin deficiency.

The severe deficiencies occur on the basis of congenital absence of the thyroid (sporadic cretinism) or of suppressed thyroid function in infants of goiterous mothers who are hypothyroid themselves (endemic cretinism). This is mainly observed in mountainous areas of the world, where endemic goiters occur due to iodine deficiency.

Since in either form of cretinism the deficiency already operates through intrauterine life, the skeletal effects are similar to but more severe than those in primary hypopituitarism. Sporadic cretins show the most severe degree of drawfing because of complete absence of thyroxin. This is accompanied by great delay in formation of secondary ossification centers and permanently open epiphysial plates, resulting in a great discrepancy between radiological and chronological bone age. In endemic cretins there exists a gradient of deficiency leading to a subnormal stature of childlike proportions (de Quervain and Wegelin, 1936: 35–41). In these individuals the epiphysial plates

FIGURE 473.—Dysplastic femoral head with multiple ossification centers and short, plump neck, markedly delayed epiphysial closure in a 37-year-old female hypothyroid dwarf with nodular goiter; body length 114 cm. (FPAM, Jubiläumspital 869.)

ultimately close, albeit beyond middle age. The last to close is the spheno-occipital growth plate, resulting in a brachycephalic configuration. The cranial sutures also remain open longer than normal. Continued growth at the mandibular condyle may ultimately result in some degree of prognathism. The marginal ossification centers of the scapula and the ilium may form only incompletely and remain open (Looser, 1929).

Most epiphyses show multifocal irregular ossification centers, which later fuse (for details see Wilkins, 1941) (Figure 473). These abnormalities of the ossification centers are specific. The development of the ossification center of the head of the femur is not only delayed but often abnormal. The lack of bony support of the femoral head leads to deformation of the cartilage model, secondary to static and dynamic stresses. The end result is a misshapen flattened head with a mush-

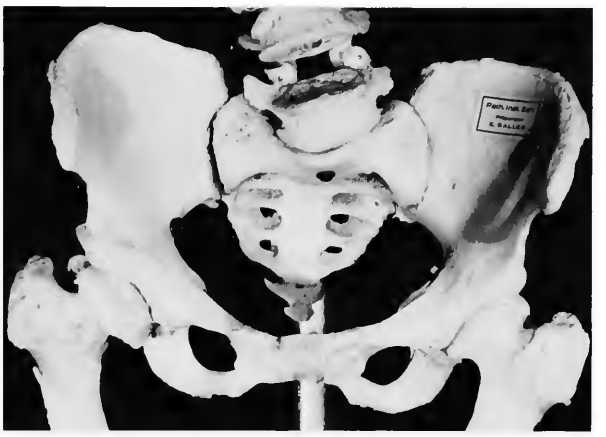


FIGURE 474.—Skeleton of a cretin showing typical hip deformation with mushroom-shaped femoral heads, coxa vara, and secondary degenerative arthritis, bilateral. (67-year-old female, IPAZ 65 from 1940.)

room-like deformity (not unlike the endstage of Perthes' disease), which fits badly into the acetabulum. However, in contrast to Perthes' disease, there is no aseptic necrosis. This misshapen "cretin hip" leads to early and severe degenerative arthritis (de Quervain and Wegelin, 1936) (Figure 474).

Hyperthyroidism

Excessive amounts of thyroxin, as observed in Graves' disease or toxic goiter, mostly occur in adults. Skeletal changes in this condition may be absent or limited to increased bone resorption, producing osteoporosis. Occasionally, these changes may mimic those in mild hyperparathyroidism and result in stress fractures (Askanazy and Rutishauser, 1933; Uehlinger, 1957). These findings would not be diagnostic of hyperthyroidism in dry bones.

Cushing's Syndrome

Of the different hormones produced by the adrenal cortex under stimulation of pituitary adrenocorticotropic hormone (ACTH), only the glucocorticoid cortisone needs to be discussed briefly in regard to its skeletal effects.

Cushing's syndrome may be caused by any of

the following: hyperplasia, adenoma, carcinoma of the adrenal cortex, or, less commonly, adenoma of the basophilic pituitary cells. The excessive secretion of cortisone leads to suppression of protein synthesis, including production of collagen. The ensuing osteoblastic suppression and scarcity of bone matrix production results in severe osteoporosis. The osteoclastic resorption is not increased, but it is not balanced by a corresponding amount of bone formation. The condition may be observed at any age. The severe osteoporosis is most marked in bones rich in trabeculae where the normal turnover is rapid. The vertebral column and ribs are most severely affected. The vertebral bodies show marked cortical thinning and severe reduction in the number and size of the trabeculae (Figure 475). The end-plates bulge inward due to the pressure exerted by the resilient intervertebral discs, creating the picture of biconcave, so-called "codfish" vertebrae. The vertebral bodies lose height and compression fractures are common. Anterior wedging with kyphotic deformity on the vertebral column occurs. The ribs become severely osteoporotic and may exhibit multiple pathological fractures (Sissons, 1956). The long tubular bones also show trabecular osteoporosis and endosteal cortical resorption, but the changes are less obvious and the outer cortical surface appears normal. Although this osteoporosis is not specific, severe changes of this kind and severity, particularly in a child or young individual, are very suggestive of Cushing's syndrome.

FIGURE 475.—Cushing's syndrome. Vertebra with extreme osteoporosis after 10 years of cortisone therapy. (51-year-old female, MGH autopsy 20884.)

Hypogonadism

Estrogen and testosterone both stimulate enchondral growth and skeletal maturation. In both sexes subnormal production of gonadal hormones will delay the appearance of secondary ossification centers and postpone the closure of epiphysial discs and sutures. Periosteal bone formation is also inhibited, resulting in a gracile skeleton with thin cortices. Since the period of enchondral growth is markedly lengthened, the normal skeletal proportions are altered due to disproportional length of the upper and lower extremities (eunuchoid stature) and elogation of the mandible, producing some degree of prognatism. Since castration of the male was practiced in various cultures, it is worth mentioning that in the early castrate (eunuch) the above-mentioned changes are present in a most marked degree, resulting in a tall, long-legged, gracile skeleton. Cessation of production of gonadal hormones in the adult, either by involution (menopause) or pathologic destruction of the gonads will often ultimately, over a period of about one decade, result in trunk osteoporosis in either sex (Nowakowski, 1955).

Hypergonadism

Excessive amounts of gonadal hormones produced by various neoplasms or other abnormalities will result in precocious puberty in affected children. Although the enchondral growth is stimulated, the early closure of the epiphysial plates results in a short but stocky skeleton (Wilkins, 1950:230; Prader and Maassen, 1953:148).

Hypoparathyroidism

The parathyroids control the level of ionized calcium in the blood plasma. This is accomplished by increased secretion of parathormone

in response to lowered blood calcium. Parathyroid hormone acts on the skeleton by stimulation of osteoclastic resorption, liberating calcium from the bone matrix.

Inadequate production of parathormone, due to congenital absence or acquired impairment of the parathyroids, results in hypoparathyroidism. In this rare condition, suppression of osteoclastic remodeling can lead to a degree of nondiagnostic osteosclerosis as a result of the continued osteoblastic endosteal and periosteal bone formation (Figure 476).

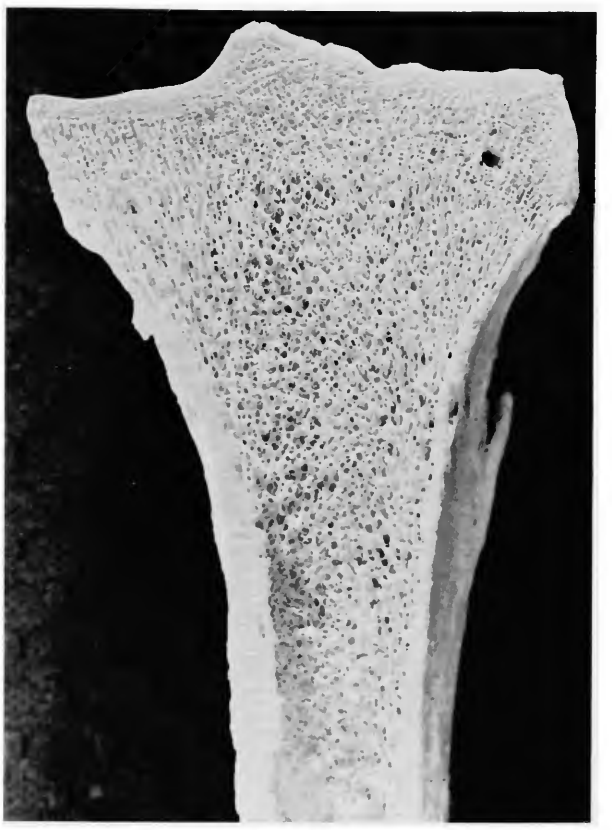


FIGURE 476.—Hypoparathyroidism. Frontally bisected proximal right tibia showing marked cortical thickening, some periosteal hyperostosis, and increased strong spongiosa. The femur showed the same findings. (62-year-old female hypothyroid with postsurgical hypoparathyroidism of 10 years duration, IPAZ 6168, autopsy 1265 from 1955.)

Hyperparathyroidism

Hypersecretion of parathormone leads frequently to marked skeletal changes. It occurs as a primary condition or secondary to renal impairment.

PRIMARY HYPERPARATHYROIDISM (osteitis fibrosa cystica generalisata).—This condition, first described by Recklinghausen (1891), is the result of hypersecretion by either a parathyroid adenoma of one gland or by diffuse hyperplasia of all parathyroids. The continued overstimulation of osteoclastic resorption results in osteoporosis and the continued loss of the liberated calcium through the kidneys in a negative calcium balance. The early changes are demonstrable only microscopically. They consist of osteoclastic intratrabecular resorption (Figure 477) accompanied by fibrous conversion of the bone marrow ("dissecting bone atrophy"). Only advanced cases will show significant radiologic and gross skeletal changes. As a direct effect of continued bone resorption exceeding bone formation, general osteoporosis develops. This resorption may be radiologically and grossly demonstrable, particularly on the cortical surface of phalanges, forming scalloped depressions. Similarly, the normally present thin layer of lamellar bone (lamina dura) lining the dental alveoli undergoes resorption, visible on X-ray and by direct inspection of the dry jaws. The osteoporosis of the spine may lead to biconcave deformity of the vertebral bodies. Cranial osteoporosis tends to diminish the distinction of an outer and inner table. There is

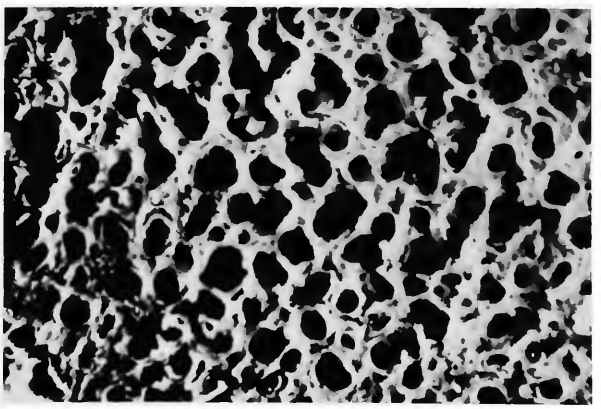


FIGURE 477.—Hyperparathyroidism. Vertebral spongiosa showing centrotrabecular resorption (dissecting atrophy). (70-year-old female, IPAZ autopsy 1204 from 1970; courtesy Prof. E. Uehlinger.)

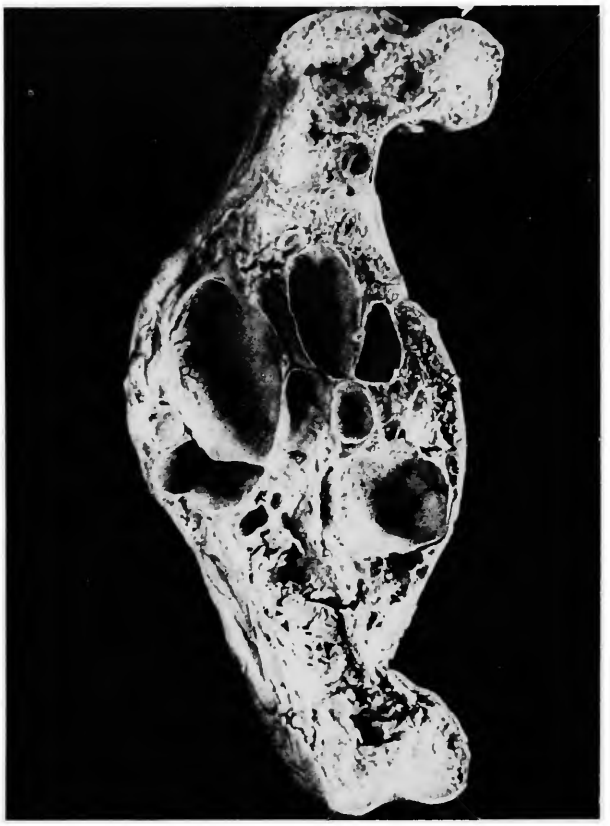


FIGURE 478.—Hyperparathyroidism (osteitis fibrosa cystica) in bisected femur.

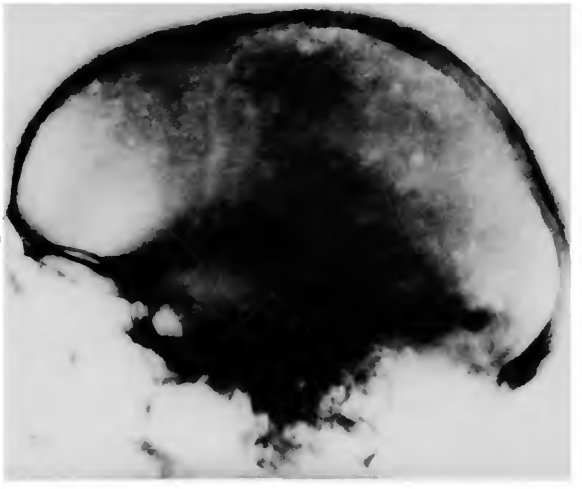


FIGURE 479.—Secondary hyperparathyroidism. X-ray of skull, showing "cotton-wool" appearance of cranial vault. (62-year-old female, IPAZ surgical specimen 12807154.)

transformation of the diploë into fine trabecular, poorly mineralized, cancellous bone. The major gross abnormalities observed are the result of the continued and repeated minor traumas acting upon the vulnerable, demineralized skeleton. These changes consist of cyst formations and proliferations of fibrous marrow ("brown tumors") secondary to hemorrhage and microfractures. These lesions appear on dry bone as cystic lytic defects with one or several cavities separated by bony septa and ridges (Figure 478). They may expand the affected area of the bone remarkably and lead to pathological fractures and deformities. These lesions can appear in any bone but predilect the mandible, the long and short tubular bones of the extremities, and the pelvis. In extreme cases the demineralized skeleton may become more pliable and deformed than in osteomalacia or fibrous dysplasia. The preservation of these changes in archeological material is unlikely.

SECONDARY HYPERPARATHYROIDISM.—In a variety of chronic renal diseases, parathyroid hyperplasia occurs as a response to disturbance of the metabolism of calcium and phosphorus. The majority of these cases concern children and will show, in addition to rickets-like changes (p. 273) and stunted growth, features of hyperparathyroidism. In adults, of course, alterations of growth no longer can occur and, therefore, the picture of renal osteopathy is a combination of hyperparathyroidism and osteomalacia. In rare instances of longlasting renal impairment in adults, all features of primary hyperparathyroidism, including skull changes (Figure 479), cysts, and brown tumors, can be observed (Jaffe, 1972:326).

Other Bone Diseases

Paget's Disease

Pathology

Osteitis deformans (Paget, 1877) is a chronic bone disease, which may affect a single, several, or many bones but never involves the entire skeleton. The etiology is completely unknown. It occurs commonly in populations of European descent but is very rare in Asians. The disease seldom appears before 40 and mostly not before 50 years of age. The frequency and extent of involvement increases with age. Males are more often involved than females. The disease is not rare in a limited form but widespread multiosseous involvement is uncommon. Schmorl (1932: 698–699) found an incidence of 3 percent of Paget's disease in a series of over 4600 autopsies of individuals above 40 years of age. The lesions of the 138 cases found in this study show the following distribution in the skeleton (in rounded off percentage figures listed in decreasing frequency): sacrum 57 percent, spine 50 percent (lumbar segment 26 percent, thoracic 17 percent, cervical 7 percent), femora 46 percent, skull 39 percent, sternum 32 percent, pelvis 30 percent, clavicle 18 percent, tibia 11 percent, ribs 10 percent, humerus 6 percent. These figures indicate the great predilection for the axial skeleton, which often is the only area involved.

The disease process can be best characterized as a pathological speedup and distortion of the normal remodeling mechanism. It always begins as a local process but may spread through an entire bone, including the epiphyses, clear up to and involving the subchondral bone plate of the joint. In the skull the process extends readily across suture lines. The initial change is excessive osteoclastic resorption accompanied by fibrous conversion of the bone marrow and hypervascularity. This is followed by osteoblastic overstimulation, producing irregular and excessive amounts of woven and lamellar bone. In the long protracted course of the disease osteoclastic and osteoblastic activities occur simultaneously, leading to excessive remodeling in affected areas, which results in the characteristic mosaic pattern in the microscopic picture. This picture consists of increased numbers of fragments of lamellar bone separated by cement lines. This pattern is present in trabecular and cortical bone. Since it is a feature of the microarchitecture of mineralized bone, it is recognizable in ground sections of dry bone (Figure 480). This is one instance where microscopic examination of archeological bone will be of great diagnostic significance.

The early lesions are predominantly lytic and osteoporotic; later, thickening of the cortex by endosteal and periosteal bone deposition and enlargement of the bones is observed. The trabecular architecture becomes accentuated and its usually smooth outline assumes irregular surface contours on X-ray. This alternation of resorptive and sclerotic areas gives a mottled picture on X-ray. The changes of the bone at different stages of the disease and at different locations in the skeleton present a variety of different pictures, necessitating the following, more detailed discussion of various areas.

The Skull.—The early Paget lesion of the skull is represented by the so-called osteoporosis circumscripta (Erdheim, 1935). This single, or sometimes multiple, lesion of the cranial vault is characterized by prevailing bone resorption, resulting in reduction of the trabeculae in the diploë and marked thinning of the inner and outer table, and creating an area of markedly increased radiolucency on X-ray. The thinned tables may exhibit a porous surface. The outline of the lesion is somewhat wavy, may cross suture lines and has a sharp interphase against adjacent uninvolved

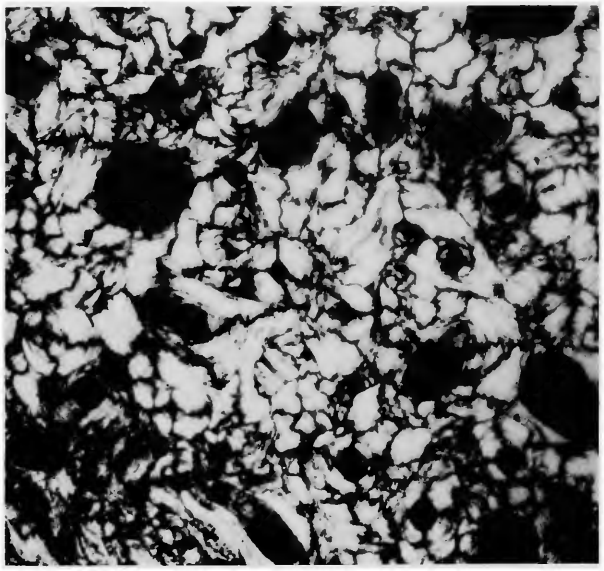


FIGURE 480.—Mosaic pattern with numerous cement lines in Paget's disease, femoral cortex. Ground polished section of undecalcified bone. Polarized light, approximately ×100. (Specimen courtesy Prof. E. Uehlinger.)

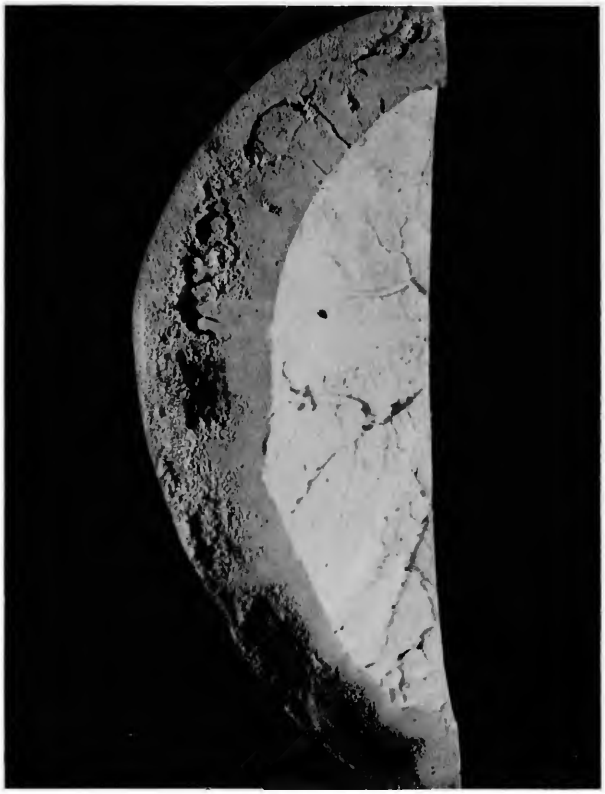
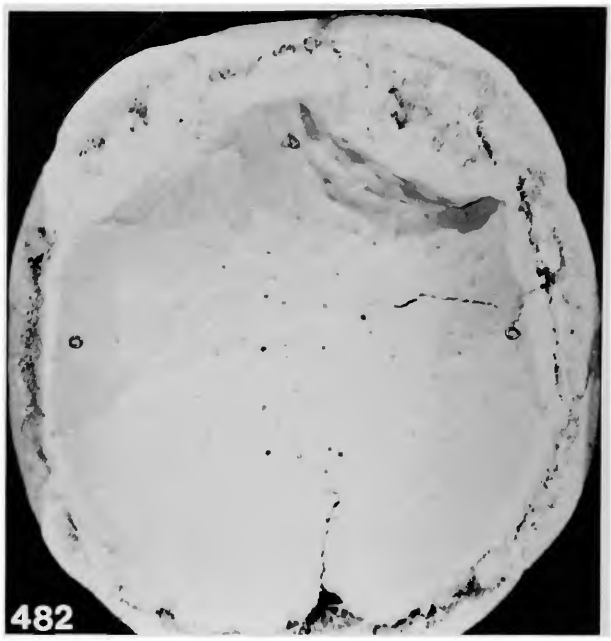
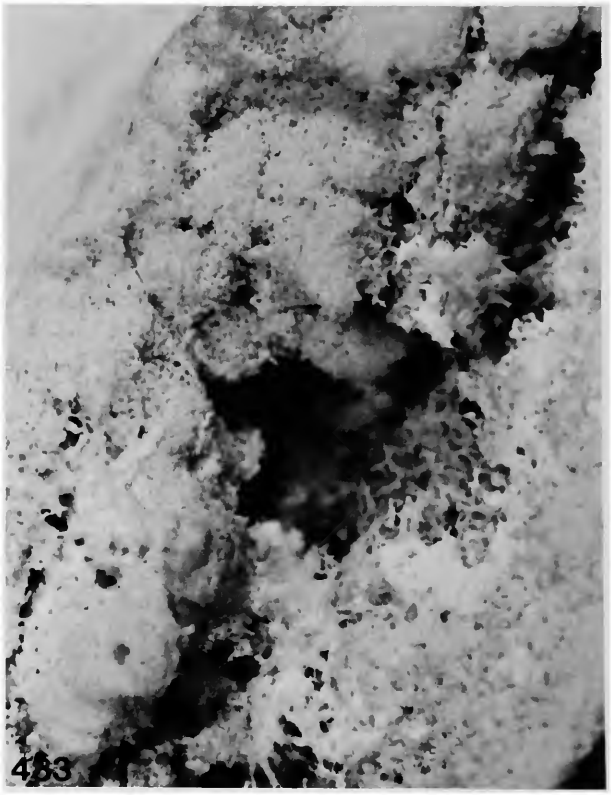


FIGURE 481.—Paget's disease of calvarium (original case), showing extreme thickening and sclerosis of cranial vault with nodular bony masses in the diploë. (68-year-old male, duration 22 years, WM H.S.62.1 from 1876.)





FIGURES 482, 483.—Paget's disease: 482, Cranial vault, endocranial view, showing thickening, especially of frontal bone, with nodular bony masses in the diploë. The bone is soft and pumice-like. 483, Close-up of frontal area with nodular diploë. (65-year-old female, polyostotic Paget, IPAZ 4566, autopsy 46 from 1940.)

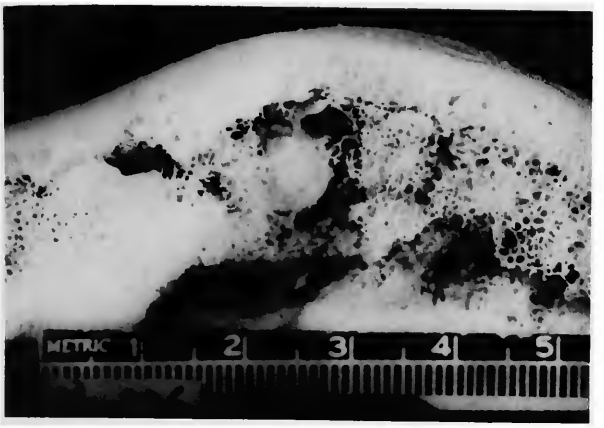
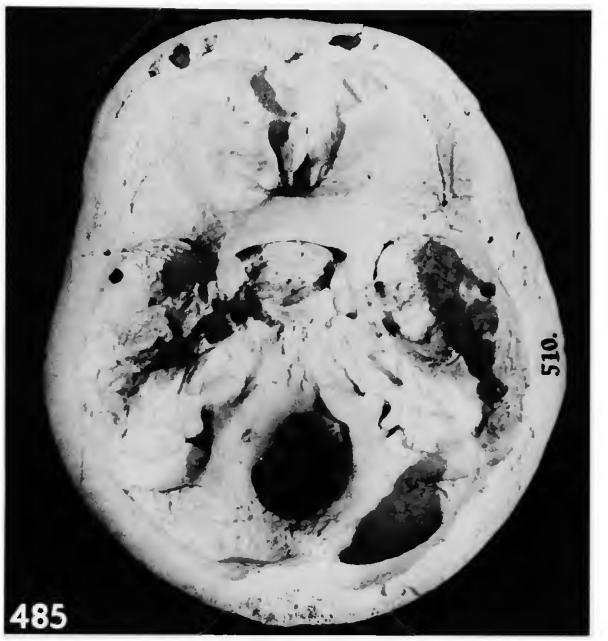


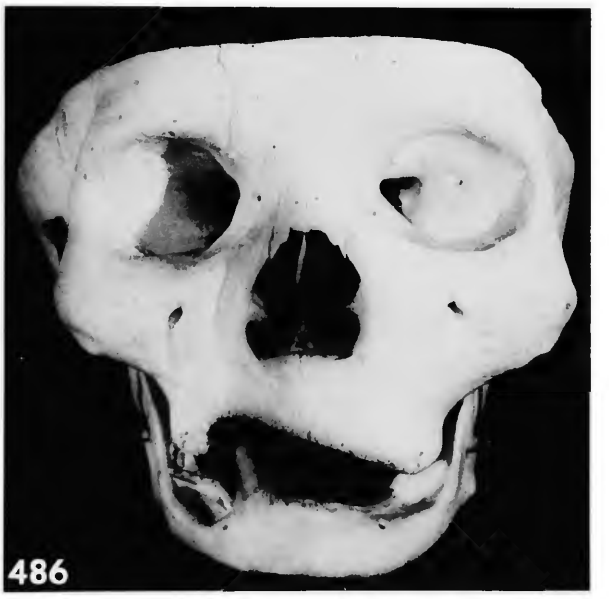
FIGURE 484.—Paget's disease of frontal bone with circumscribed nodular densities in diploë. (66-year-old female, MGH autopsy 34336.)

bone. Although this lesion may remain for a number of years, ultimately, characteristic blastic lesions of Paget will develop either in the skull or in other areas of the skeleton (Kasabach and Gutman, 1937:598).

The location and appearance of this lesion is distinct enough to make one suspect early cranial Paget's disease on a dry bone specimen. Changes of fully developed Paget's disease of the cranial vault may exhibit distinct thickening of the calvarium by endocranial and external bone formation. Such calvaria may reach several centimeters in thickness and there may be considerable encroachment on the intracranial space (Figure 481). The cross-section of the calvarium shows areas of laminated thickening of the tables and alternation of porotic and sclerotic areas in the diploë, presenting the characteristic appearance of finely porous pumice bone (Figures 482, 483). In the diploë, ball-shaped masses of sclerotic Paget bone occur, exhibiting greatly increased radiological density (Figure 484). These masses are more common and much more dense than those seen occasionally in fibrous dysplasia.

The skull base may be involved (Figure 485). Thickening takes place mostly on the endocranial surface with encroachment upon the cranial cavity. The cranial air sinuses may also be impaired by the transformation and thickening of their





FIGURES 485, 486.—Skull in advanced Paget's disease: 485, Cranial base, showing massive endocranial hyperostosis and sclerosis of thickened cranial vault. 486, Frontal view, showing diffuse hyperostosis of facial bones with porous subperiosteal bone deposition. (75-year-old female, FPAM, Jubiläumspital 510, from 1917.)

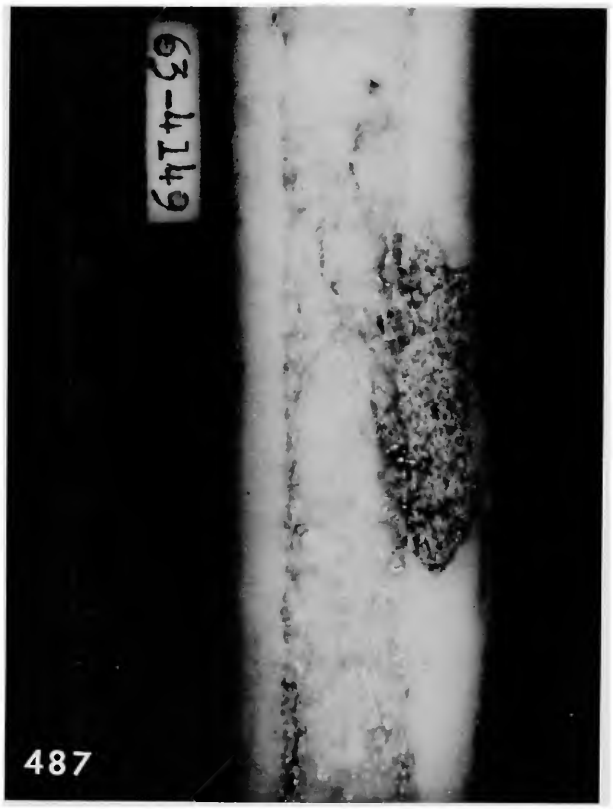
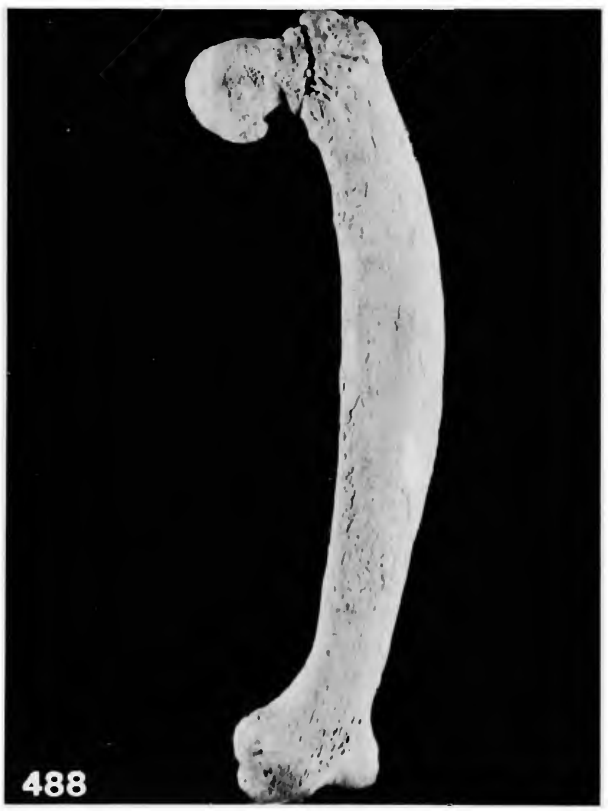
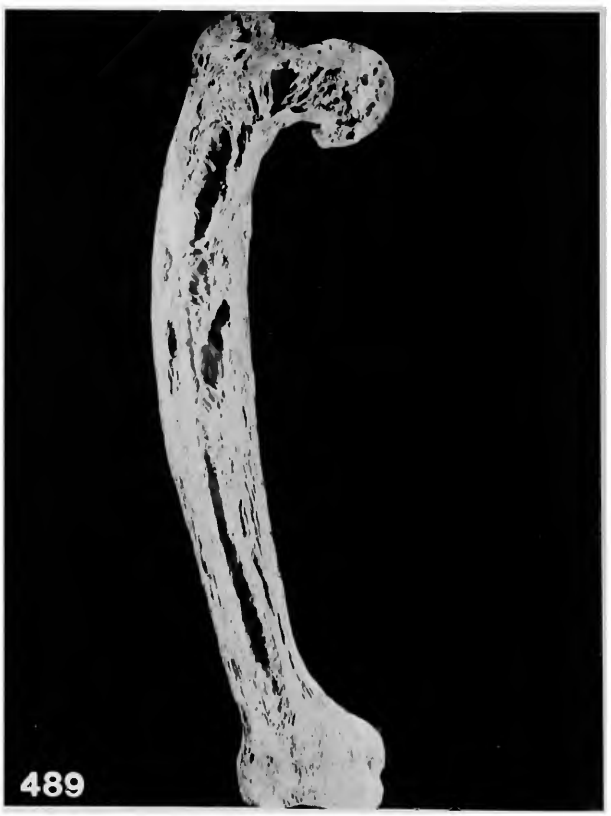


FIGURE 487.—Early Paget's disease. Bisected tibia showing lytic transcortical lesion (dark) with wedge-shaped progressive margin. (77-year-old female, MGH surgical specimen 4149 of 1963.)

walls. The cortex of the petrous bone may participate in Paget's disease but the otic capsule is usually spared. This is in keeping with its lack of remodeling. The facial bones are usually spared. If the facial bones are involved, subperiosteal deposition of pumice-like bone may disfigure the face and resemble leontiasis ossea (Figure 486). The jaws also ordinarily do not participate in the disease, although abnormal deposits of radiodense cementum around some dental alveoli is not uncommon in Paget's disease.

THE LONG BONES.—The femur and the tibia are the most common sites of involvement. The





FIGURES 488, 489.—Left femur, Paget's disease: 488, external view, showing bowing and thickening. 489, Cut surface, showing cortical thickening, narrowing of the medullary canal and irregularly coarsened cancellous bone. (64-year-old male, 10 years duration, WM HS62.3)

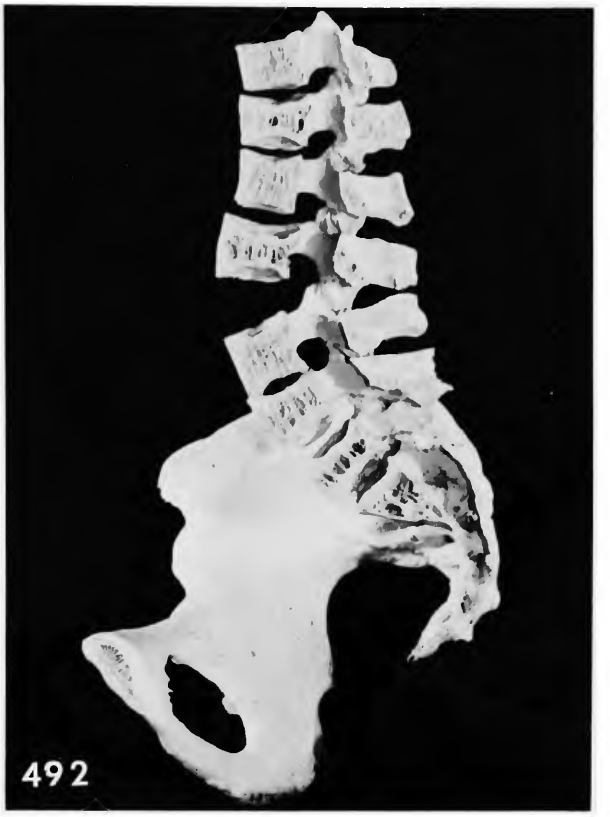


FIGURE 492.—Paget's disease of lumbosacral spine, complicated by secondary hyperparathyroidism. Midline cut showing the characteristic coarse trabeculation of Paget's disease and the fine pumice bone deposition along the endplates, particularly of the fifth lumbar and first sacral vertebra, characteristic of hyperparathyroidism. (71-year-old female, IPAZ autopsy 358 of 1960.)

early lesion in these bones also is represented by a purely lytic osteoclastic resorption involving the entire thickness of the cortex and showing a sharp wedge-like border against uninvolved bone (Figure 487). This lesion alone is not diagnostic in dry bone and can, even on X-ray, be mistaken for a tumor metastasis. The later blastic phases show marked thickening and lamination of the cortex. The bones become plump by periosteal bone formation. Bowing, especially of the weight-bear-

FIGURES 490, 491.—Paget's disease of tibia showing pumice bone subperiosteally and transcortically: 490, Surface. 491, Cut surface. The spongiosa is normal. (Studied by Putschar in 1940 at Charleston General Hospital, Charleston, West Virginia, USA.)

ing bones, occurs mainly due to complete or incomplete transverse pathological fractures, demonstrating the mechanical inferiority of Paget bone (Figures 488, 489). The spongiosa of the epiphyses and metaphyses shows characteristic coarsening and focal deposition of pumice bone (Figures 490, 491). The medullary cavity is preserved. Secondary degenerative arthritis over severely affected epiphyses is common. Although, occasionally, any bone may be affected by Paget's disease, the fibula and the small tubular bones of hands and feet are usually spared. The same is true of the tarsal and carpal bones.

THE SPINE.—The axial skeleton is most often the site of Paget lesions. The frequency of involvement decreases steadily from the sacrum to the cervical spine. Within a vertebra, the body is most markedly affected. The lesion may extend into the neural arches but often spares their posterior portions and the spinous process. The vertebral bodies show laminated thickening of the end-plates and the cortex, with porotic reduction of the trabeculae in the center (Figure 492). These trabeculae are reduced in number but thickened with uneven surfaces (Figure 493). Compression fractures of severely affected vertebrae are not uncommon. Frequently, although several vertebrae show the disease, they are separated by completely normal vertebrae.

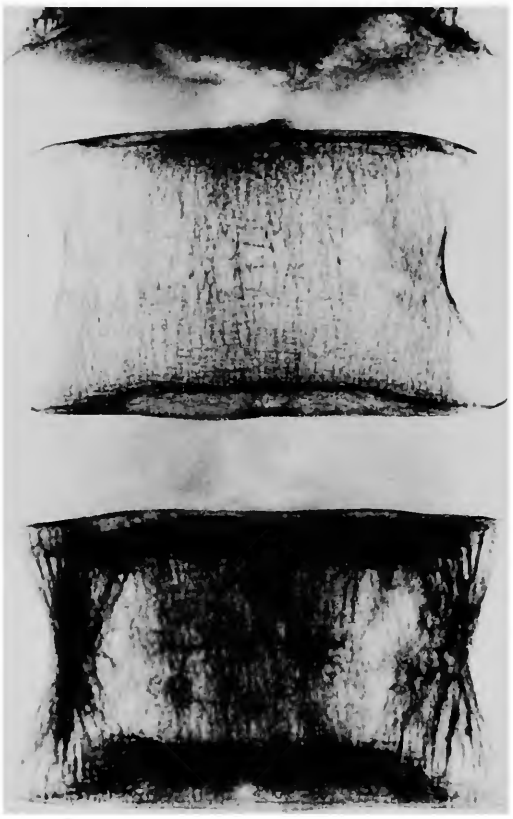


FIGURE 493.—X-ray of Paget's disease of vertebra with adjacent normal vertebra (above). Note increased density and coarsening of the spongiosa of the involved vertebra. (European X-rays show densities as black and lucencies, white; studied by Putschar.)

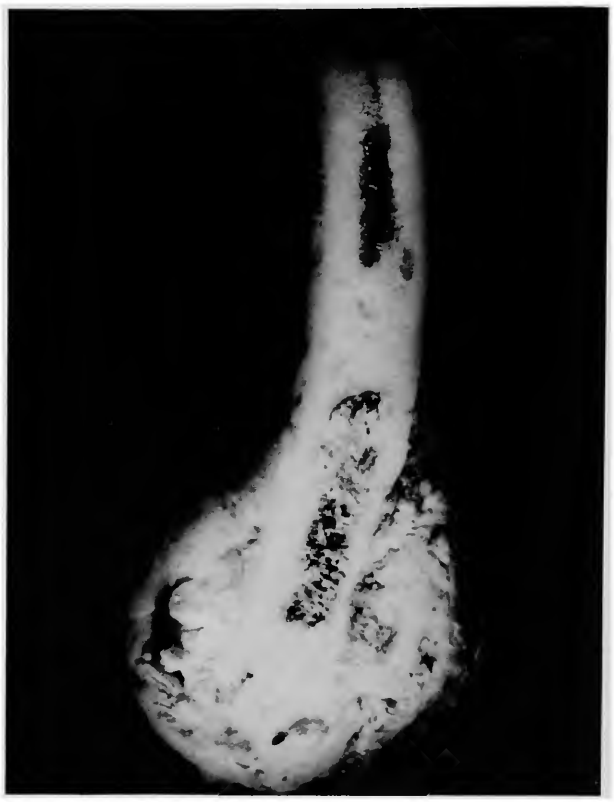


FIGURE 494.—Osteosarcoma in Paget's disease of right distal femur. (80-year-old female, MGH surgical specimen 13584 from 1963.)

THE FLAT BONES.—The ilium is often involved if there are lesions in the sacrum. The sternum rates high in the Schmorl (1932) series. The disease usually involves the manubrium or the body

but does not cross the synchondrosis. Ribs are not uncommonly affected and may become markedly thickened.

PAGET SARCOMA.—In longstanding active Paget, especially of the long bones and calvarium, a sarcoma may arise in one or several areas (Figure 494).

PALEOPATHOLOGY

The earliest reference to Paget's disease in archeological human remains is a report by Pales (1929:263–267), which includes a brief discussion of a femur from the Neolithic site at Lozere in France. The specimen is part of the collections of the Museum of Natural History of Paris. There has been some postmortem damage to the joint surfaces, but otherwise they appear normal. There is considerable anterior curvature of the diaphysis with a marked expansion of the cortex. The X-ray film indicates that the cortical bone in the diaphysis is reduced in density. The line drawings, published in Pales (1929) as figures 1 and 2, portray a gross morphology compatible with a diagnosis of Paget's disease. No histology was reported, making this diagnosis probable but not certain. The antiquity of this specimen would certainly make restudy with appropriate histology desirable.

Five purported cases of Paget's disease in prehistoric skeletons from the Illinois River Valley in the United States were published by Denninger (1933). In all cases the long bones have a thickened cortex. In some of the cases the skulls were thickened. In no case was there any report of the histology. In view of the many disease conditions that can produce the lesions described by Denninger, including periostitis and osteomyelitis, these specimens would need to be restudied before the diagnosis of Paget's disease can be accepted.

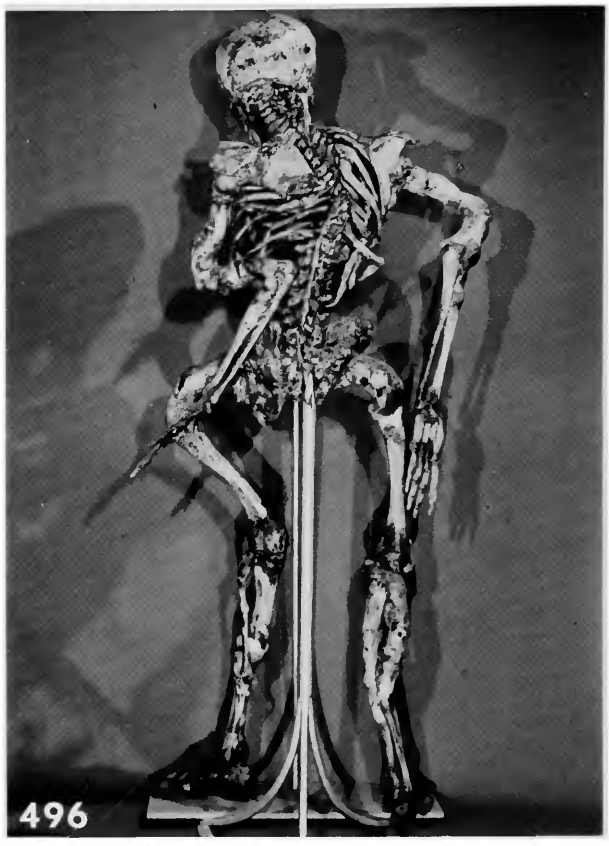
Fisher (1935) reports a purported case of Paget's disease in a skeleton from a prehistoric mound in Crawford County, Wisconsin, USA. The specimen consists of portions of both tibiae and a part of the lower jaw. Morse (1969:57–58) also discusses this case noting that the jaw fragment is normal. He also notes that one of the tibiae was sent for study to Dr. Lent C. Johnson at the Armed Forces Institute of Pathology in Washington, D.C. Johnson found no histological evidence of Paget's disease in the bone.

Wells and Woodhouse (1975) report a case of Paget's disease in a skeleton from an Anglo-Saxon burial ground (A.D. 950) in Durham, England. The skeleton is male with an estimated age of 65 years. The authors indicate that the skull is unusually thick. The surface of most bones is rough and pitted. The spine exhibited kyphoscoliosis with evidence of osteoarthritis. The pelvis has the appearance of a slight osteomalacic deformity. The major bones of all the extremities are bowed. The left femur was fractured antemortem with poor alignment and considerable deformity with healing. The poor quality of the bone prevented the preparation of microscopic sections; thus the presence of the mosaic histological pattern could not be determined. However, the generalized distribution of abnormal bone, the long bone deformity, and the roentgen film appearance of the femora and vertebrae are compatible with a diagnosis of Paget's disease. The vertebrae show coarse vertical striations and discontinuity of affected vertebrae.

Fibrous Dysplasia

PATHOLOGY

Fibrous dysplasia is a bone disease of unknown etiology characterized by faulty differentiation of portions of the osteogenic mesenchyme into fibro-osseous tissue. It manifests itself in a single or in multiple bones, often limited to one limb or one side of the body (Lichtenstein and Jaffe, 1942: 814). The entire skeleton is never involved even in widespread manifestations, and there is no basic metabolic disorder (Figures 495, 496). The disease, especially the polyostotic form with widespread manifestations, usually begins in childhood and often stops progressing at the termination of the growth period (Uehlinger, 1960:283). Females are more often affected than males and may exhibit endocrine disturbances leading to precocious puberty and premature termination of



FIGURES 495, 496.—Polyostotic fibrous dysplasia showing "shepherd crook" deformity of both proximal femora and deforming involvement of many bones: 495, Anterior view. 496, Posterior view. (40-year-old female, DPUS 4016, French catalog 323a from before 1891.)

growth (Albright's disease) (Albright, Butler, Hampton, and Smith, 1937) (Figures 497–499).

The bone lesion consists of dysplastic proliferation of endosteal fibrous tissue, in which fine trabeculae of woven bone arise by direct fibro-osseous metaplasia, without differentiation of osteoblasts. Depending on the amount of bone and the degree of mineralization, the lesion may appear purely lytic, ground-glass, or dense on X-ray. It may ultimately involve most parts of a bone, sparing some portions and practically always sparing the epiphyses. In small bones, tumor-like expansion is observed after pressure resorption of the old cortex. However, at least a thin shell of newly formed periosteal bone surrounds the lesion at all times. This results in a cyst-like lesion confined by a ridged, thin, bony shell, particularly in small and flat bones, such as ribs and pelvis. The weakened bony structure permits mechanical deformities with or without major pathological fractures (Figure 497). The recognition of the lesion in dry bone would depend on at least partial preservation of the fine or coarse woven bone trabeculae (Figure 500). Admittedly such preservation may be unlikely and differentiation from various other lesions, especially giant cell tumor, unicameral cyst, and nonossifying fibroma may be impossible in a single lesion. Hyperparathyroidism should be possible to rule out on the basis of absence of metabolic bone disease in uninvolved parts of the skeleton.

Monostotic Fibrous Dysplasia

This condition occurs most frequently in a rib, the proximal femoral metaphysis, the jaws, or the tibia. Other bones are less frequently involved (Schlumberger, 1946a, 1946b). In dry bone, differentiation from eosinophilic granuloma and enchondroma may be impossible, especially in the rib. In the mandible, or less often in the maxilla, giant cell reparative granuloma or eosinophilic granuloma may give a similar picture. Probably the most characteristic solitary lesion is that of the proximal femoral metaphysis. The dysplastic focus often weakens the subtrochanteric area resulting in the typical "shepherd crook" deformity, while the head and neck of the femur are more nearly normal.

Polyostotic Fibrous Dysplasia

In involved limbs the disease affects the proximal bones earlier and more severely than the distal ones. The proximal femoral metaphysis is the most frequent location (Figures 501–503). In 36 cases of polyostotic fibrous dysplasia, the involvement of different bones was as follows: femur 36, tibia 24, fibula 10, metatarsals 10, toes 8, humerus 12, radius 9, metacarpals 8, fingers 6, pelvis 15, skull base 6 (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952(2):1199).

The lesions in the cranial vault and face are asymmetrical and often unilateral (Figure 498). Rounded masses of woven bone can be found in the diploë, similar to those seen more frequently in advanced Paget's disease. Lesions of the skull base tend to be sclerotic and inhibit pneumatization of the paranasal sinuses (Figure 499).

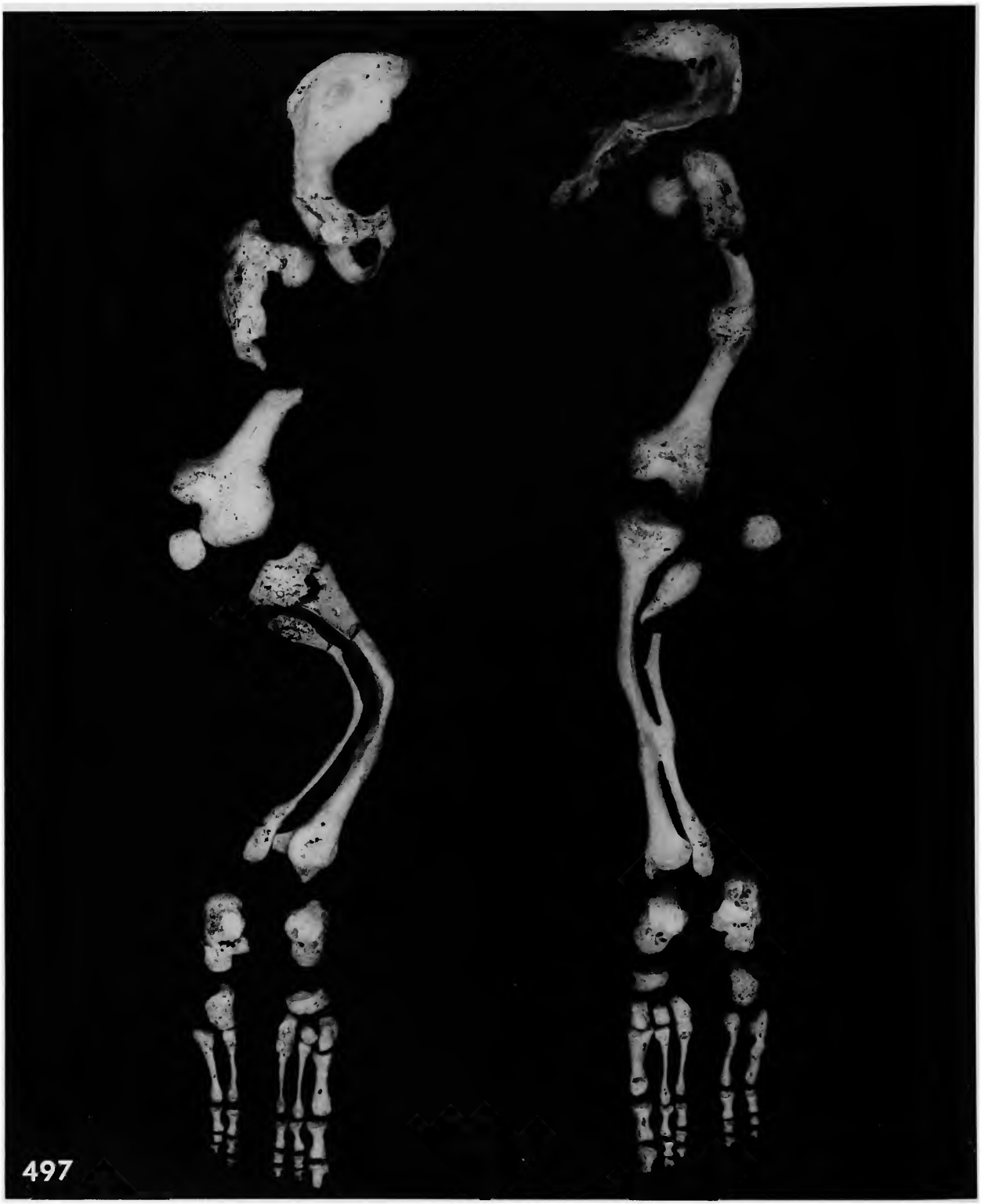
The skeletal distribution of the lesions, especially in the limbs, is similar to that of Ollier's enchondromatosis; but, in the latter the distal tubular bones of the extremities (metacarpals, metatarsals, and phalanges) are most frequently and more severely involved. The presence of the cranial changes discussed above would also rule out enchondromatosis.

PALEOPATHOLOGY

Denninger (1931) provides a description of a probable case of polyostotic fibrous dysplasia in an adult male skeleton from a pre-Columbian site in Illinois, USA. The disease affected the bones of the left side of the body. The left side of the skull was deformed and there was the characteristic shepherd's crook deformity of the left femur. The cortex of the bowed upper end of the left femur is thin. This region has multiple cyst-like lesions revealed in the roentgen film. Postmortem breakage reveals the poorly organized bone partially filling the marrow space. The right femur is not deformed but exhibits some pathological changes on the proximal end. In the region of the greater trochanter, there are perforations of the cortex with poorly organized, spongy bone apparent through the perforations. Similar lesions are found on both tibiae and fibulae. The bones of the left foot show increased porosity of the cortical surface with cortical perforations and spongy bone apparent in the first and second metatarsals. Perforations and spongy bone development are also seen in the bones of the pelvis primarily on the left side. The ribs of the left side are similarly affected as is the seventh cervical vertebra and many of the thoracic vertebrae. Denninger's description of this case leaves little doubt that this pre-Columbian Indian skeleton is an example of polyostotic fibrous dysplasia.

Wells' (1963b) report of an adult skeleton from an Anglo-Saxon site in Britain, dated in the seventh century A.D., provides another example of probable fibrous dysplasia. Of the bones recovered, only the left humerus shows evidence of the disease. The normal axis of the bone has been deformed producing anterior bowing. The cortex of the diaphysis is enlarged with the focus being near midshaft. The longitudinal cut section reveals extensive development of fine spongy bone in this region with cystic cavities between the spongy bone and the cortex.

A possible case of fibrous dysplasia in a young adult female skull from Egypt (NMNH 256360) was brought to my attention by Dr. Eugen Strou-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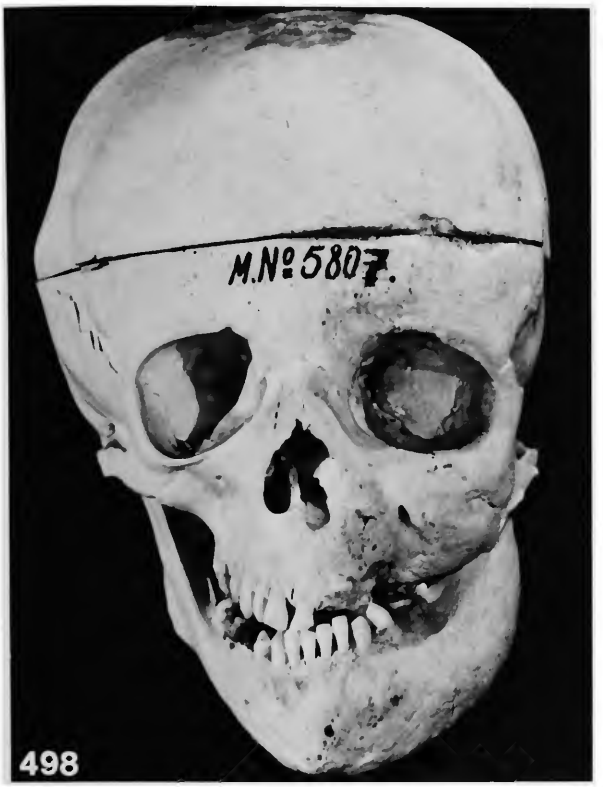
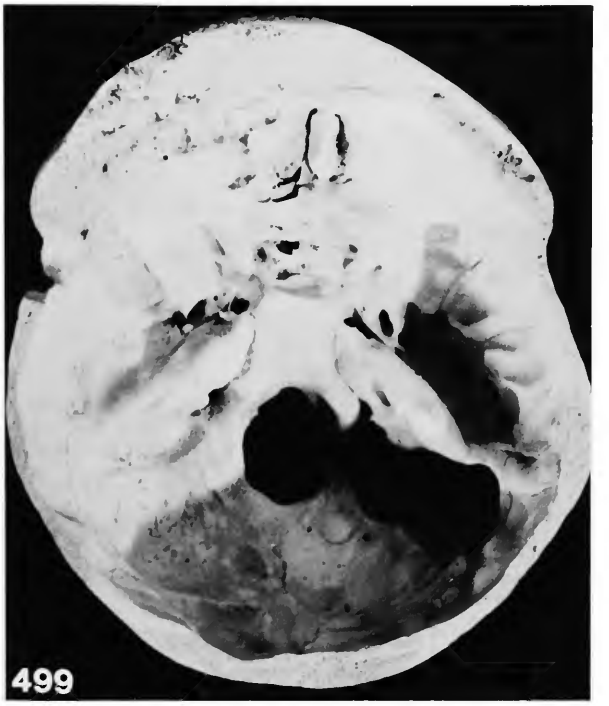




FIGURES 497–499.—Polyostotic fibrous dysplasia with precocious puberty and skin pigmentations (Albright's disease): 497, Both lower extremities with multiple deformities and fractures. 498, Skull showing left-sided involvement and deformity. 499, Skull base shows massive frontal involvement, somewhat resembling Paget's disease. (16-year-old female, 161 bones were affected, FPAM 5807, autopsy 331 from 1949, St. Pölten Hospital, Austria.)

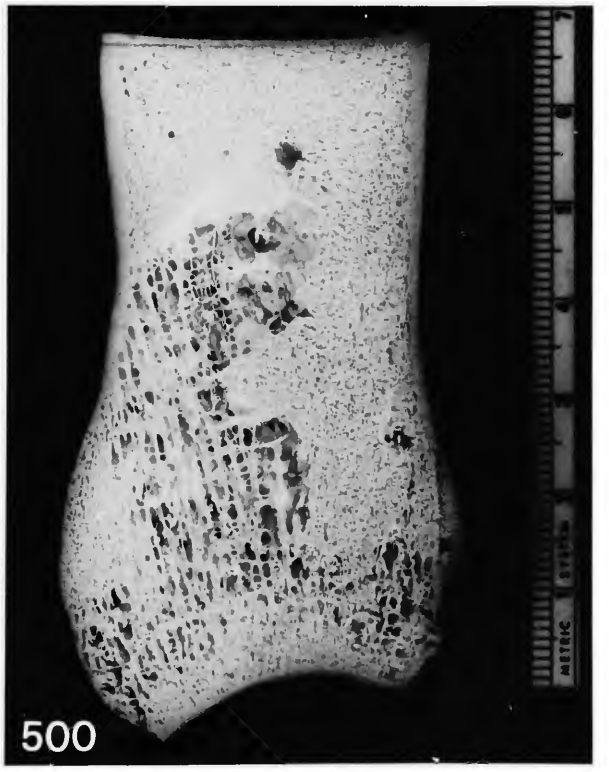
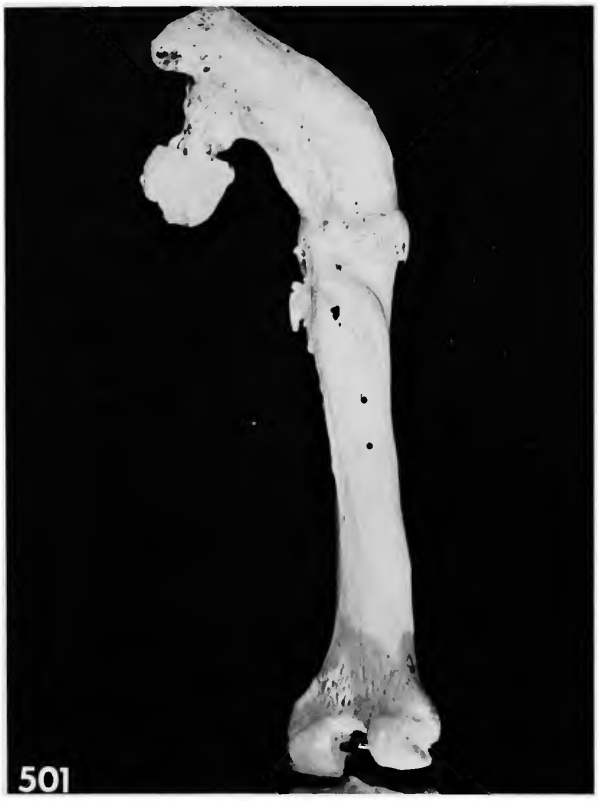
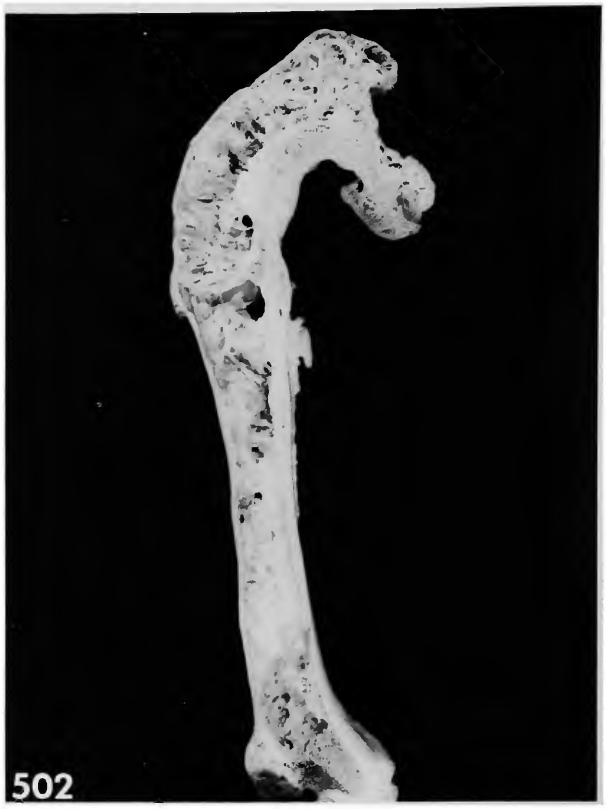
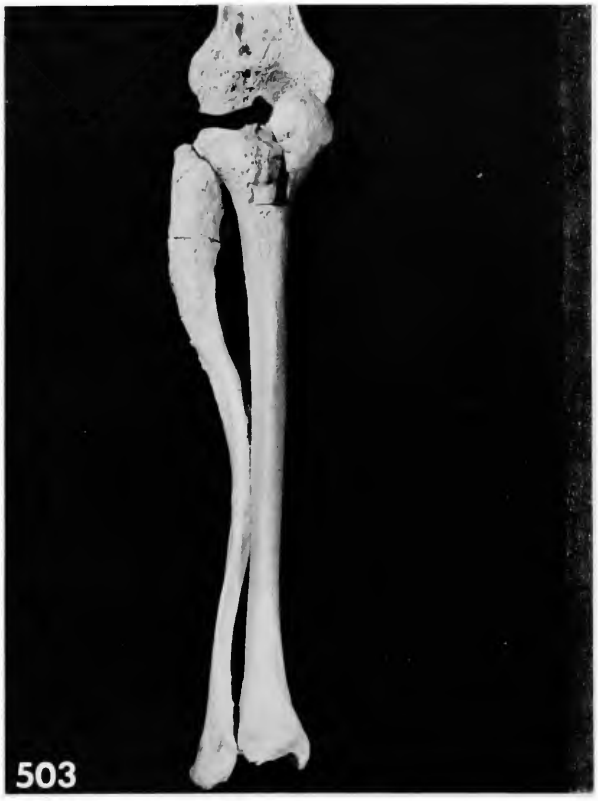


FIGURE 500.—Polyostotic fibrous dysplasia of bisected distal tibia, showing finely porous abnormal bone replacing cortex and spongiosa. (40-year-old Chinese male; studied by Putschar in 1960 at Department of Pathology, National University, Taipei, Taiwan.)







FIGURES 501–503.—Polyostotic fibrous dysplasia: 501, Right femur, posterior view, showing typical "shepherd crook" deformity. 502, Cut section, showing cystic changes and pumice bone filling the medullary cavity. 503, Right lower leg, showing marked involvement of fibula. (63-year-old male, with involvement of many bones, IPAZ autopsy 953 from 1938, skeleton 80.)

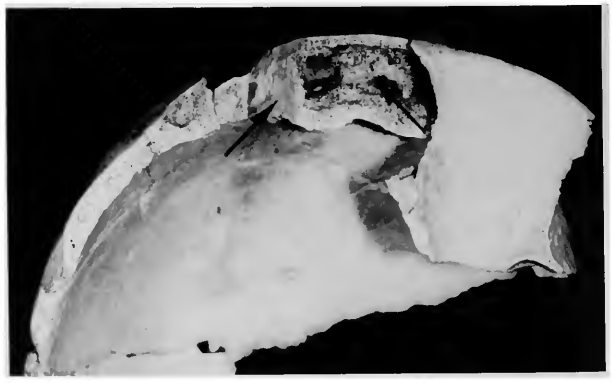
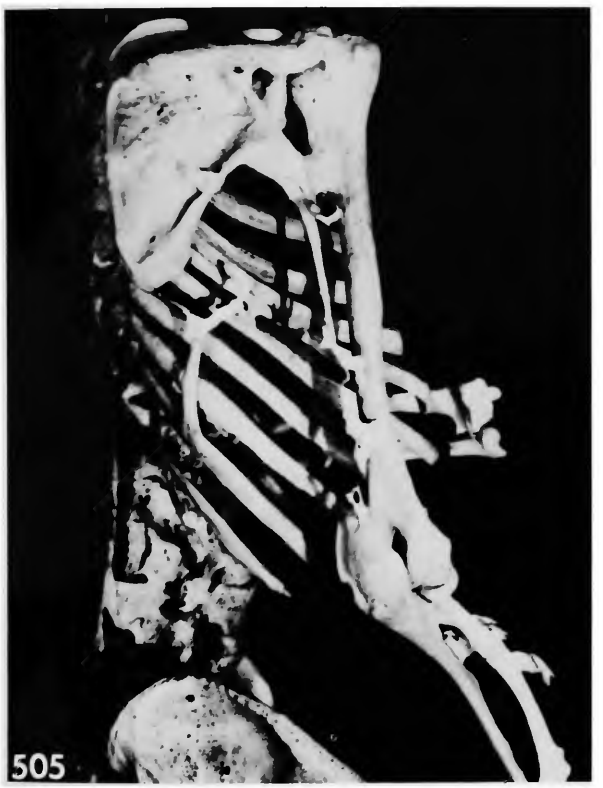


FIGURE 504.—Fibrous dysplasia in the region of the frontal sinus. Note roughened texture and nodular fiber bone proliferation (arrows). (NMNH 256360.)

hal during his research on the Egyptian skeletal collection in the National Museum of Natural History, USA. The skull is from the rock-tombs at Lisht, a Twelfth Dynasty site near Matanieh in Upper Egypt. The skull is fragmentary but includes much of the vault and face. The abnormality is limited to the frontal bone in the region of the frontal sinus. Indeed it was a postmortem break in the skull in this region that revealed the presence of dense round nodules of bone near the frontal sinus and coarse bony spicules covering the internal bony surface of the sinus cavity (Figure 504).

The gross morphology of the lesions is compatible with a diagnosis of either Paget's disease or fibrous dysplasia. The young adult age estimate would, however, make Paget's disease unlikely. A ground section through one of the circular nodules of bone reveals a mixture of well-organized lamellar bone and poorly organized woven bone. There is no evidence of the characteristic mosaic pattern associated with Paget's disease. The combination of fiber bone proliferation, the lack of mosaic pattern in the histology, and the young age of the specimen support a diagnosis of fibrous dysplasia, although a very mild expression of this



FIGURES 505, 506.—Myositis ossificans progressiva: 505, Thorax, lateral view, showing bony bridging along fascial planes. 506, Pelvis and right thigh, anterior view, showing extensive fascial ossification. (48-year-old female, IPAZ autopsy 2406 from 1968.)

disease. Greenfield (1975, fig. 3:106) has published a documented case of fibrous dysplasia showing similar nodular proliferation of bone in the frontal sinus revealed by the roentgen films.

Myositis Ossificans Progressiva

Pathology

This is a rare hereditary and familial disease (Uehlinger, 1936). The name is really misleading because it is not an inflammatory disease and it does not primarily affect the muscles but the fascias, tendons, and ligaments. Although the disease may not manifest itself until a child is several years of age, its congenital nature is attested by frequent concomitant congenital anomalies. These are shortness, usually bilateral, of the first metatarsal and/or the first metacarpal. At times the second phalanx of the fifth finger is missing. The disease usually begins to manifest itself in the neck-shoulder area and progresses distally. It begins as fibroblastic proliferation followed by metaplastic ossification of the fibrous tissue. The end result is formation of bony prongs and spicules following the outline of muscles from origin to insertion and immobilizing intervening joints. The ossification of spinal ligaments may be followed by ankylosis of spinal and costovertebral joints. The disease progresses into adulthood, resulting in almost complete skeletal immobility (Figures 505, 506). The limitation of respiratory motion and the bridging of the temporomandibular joint may contribute to death.

Neuromechanical Deformities

Pathology

Kyphosis

Kyphosis is the pathological increase of the normal slight anterior concave curvature of the thoracic spine. The deformity is caused either by changes in the intervertebral discs (primary kyphosis) or of the vertebrae themselves (secondary kyphosis) (Putschar, 1937:675–681). The primary kyphosis separates into two groups: the juvenile and the senile form.

Juvenile Kyphosis (Scheuermann's disease).—This deformity develops in adolescents with great predilection of the male sex (Scheuermann, 1921). The apex of the curvature in this deformity usually falls in the area of the eighth to tenth thoracic vertebra. The underlying cause is probably extrusion of nucleus pulposus material, mostly into the adjacent vertebral bodies (Schmorl's nodes) followed by anterior narrowing of the disc space and subsequent growth disturbance in this area of the end-plate, resulting in some degree of wedging (Schmorl, 1930; Schmorl and Junghanns, 1971: 345–354). The location of the wedge vertebrae usually coincides with the apex of the curvature. Since, of course, the curvature cannot be appreciated on the skeleton, the presence of one or several adjacent wedge vertebrae and of round or oblong defects near the center of the vertebral end-plate, corresponding to the location of the disc herniation, would be the main findings. If this type of deformity is long survived, secondary sclerotic changes and marginal anterior lipping may be added to these findings.

Senile Kyphosis.—This is a very common spinal deformity, developing in the fifth decade and increasing in frequency and degree with age. The underlying cause is degeneration and attrition of the intervertebral discs, particularly of their anterior portions, in the physiologically kyphotic thoracic segments. The apex of the curvature in senile kyphosis is in the upper thoracic spine, and wedging of vertebral bodies is either absent or slight. The main features recognizable on skeletal material are osteosclerosis of the anterior portions, especially the end-plates, of affected vertebrae with marginal lipping and, sometimes, anterior fusion of several segments (Figure 507, left). Disc herniations are not a characteristic part of this condition. The anterior marginal lipping, in contrast to spondylosis deformans, arises directly from the vertebral end-plate and is usually of rather moderate dimension. In advanced cases the anterior portions of the interver-

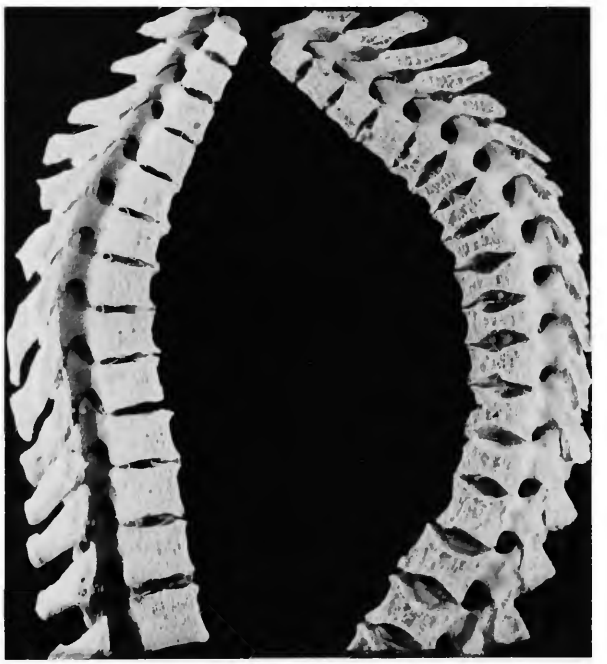


Figure 507.—Kyphosis. Two bisected spines. Left: Senile kyphosis with anterior compression of intervertebral discs and bony bridging. Right: Severe postmenopausal osteoporosis with kyphosis, showing biconcave "codfish" vertebrae. (IPAZ 6017: left, 71-year-old female, autopsy 385 from 1955; right, 65-year-old female, autopsy 403 from 1955.)

tebral disc may be replaced by continuous bone trabeculae and the lipping may be completely smoothed down by modeling resorption.

SECONDARY KYPHOSIS.—This deformity is secondary to lowered mechanical resistance of the vertebrae. A variety of conditions, especially osteoporosis (Figure 507, right), osteomalacia, hyperparathyroidism, and Paget's disease, may present this complication. In this condition wedge vertebrae, due to compression fractures, are common. The intrinsic changes of the underlying bone disease would help in recognition of the deformity as secondary. This is especially true of kyphosis due to a congenital anomaly of the spine, such as dorsal hemivertebra.

Scoliosis

The vertical position of the spinal column in the human depends on a precarious balance of neuromuscular control and fibrous supportive structures. Scoliosis is the term used for lateral deviations of the spinal column from the midsagittal plane. The factors leading to this deformity are various and usually not obvious. An exception to this is scoliosis due to congenital maldevelopment of one or several spinal segments, especially lateral hemivertebra or segmental disarrangement. However, the pathogenesis need not concern us here since the observable bone changes, which develop secondary to the deformity, are all essentially the same. Scoliosis, with or without a significant kyphotic component, often starts in childhood and progresses throughout the growing age and early adult life. This factor is of importance because the bone changes are, to a large extent, the result of altered growth and modeling under abnormally directed static and dynamic stresses (Putschar, 1937:649–675).

The deformity usually shows a double curvature, which permits the position of the head close to the midsagittal plane despite the lateral deviation of the spinal column. These deviations are not purely lateral but include a considerable element of rotation toward the convexity (Figure 508). This, as well as abnormal pull of tendons and ligaments, accounts for the bone changes by means of modified growth and modeling. Since, in the dry skeleton, the continuity of the vertebral column is not maintained, alterations of individual vertebrae are important for the recognition of the deformity. The transverse processes of the thoracic vertebrae are deflected backwards on the convex and forwards on the concave side. The transverse processes of the lumbar spine are short and plump on the convex side and long, slender, and pointed on the concave side. The spinous processes usually are deflected towards the concavity in the lumbar spine. The vertebral bodies show lateral wedging at the apex of the curvature in addition to rotation and torsion. The latter is reflected in a diagonal pattern of the vertebral cortex, secondary to the altered course of the fibers of the anterior spinal ligament. The spongiosa architecture of the laterally wedged vertebrae usually shows sclerotic reinforcement on the overburdened concave side and reduction in number and size of trabeculae on the unburdened convex side. The areas of the neural arch and of the diarthrodial intervertebral joints show the least degree of deformation. In long-standing severe scoliosis, additional bone changes may occur secondary to degenerative processes in the eccentrically stressed intervertebral discs. These changes are most marked laterally on the concave side of the wedged vertebrae and follow the same pattern as described in senile kyphosis. Bony fusion across the narrowed intervertebral space occurs in the apex of the curvature. The overexpansion of the disc on the convex side may lead to bony replacement of the disc itself. Ossification of various spinal ligaments and, occasionally, ankylosis across the diarthrodial joints can occur, especially on the concave side of the apex of the curvature.

The ribs, which are firmly attached to the spine by two joints and to the sternum through the rib cartilage, must adapt their shape and curvature to the spinal deformity. Generally, they are spread on the convexity and pressed together on the concavity of the spinal deformity. These changes include alterations of the size and loca-

FIGURE 508.—Severe thoracic and lumbar kyphoscoliosis. Notice the structural torsion of the vertebrae, the fusion on the concavity and the new joint formations between compressed ribs. (VM, no number.)

tion of the costovertebral articular facets and of the angulation of the neck of the ribs. Length, width, and curvature are altered by modified growth and modeling. If the curvature is extreme, ankylosis of costovertebral joints and formation of new, joint-like, irregular areas between compressed ribs can occur (Figure 508).

A special type of kyphoscoliosis can occur in neurofibromatosis, a condition characterized by formation of multiple nerve sheath tumors on spinal and peripheral nerves. In this disease, the maximal scoliotic deformity more frequently involves the cervical spine. The tumors of spinal nerves in this condition lead to smooth widening of interspinal foramina in the affected area. This feature would permit recognition of such a scoliosis as due to neurofibromatosis even on the dry skeleton.

Postparalytic Deformities

Neuromuscular paralysis occurring in childhood and adolescence can affect the skeleton in various ways. Paralysis of different muscle groups can unbalance the spinal column and cause postparalytic scoliosis. The examination of such a spine will not reveal the pathogenesis of the deformity. Since the growth and surface modeling of a bone are influenced by muscle pull and weight-bearing, the lack of either or both will alter its size and shape. Paralysis of an extremity results in smooth and slender shafts because of reduction of periosteal bone formation (Figure 509). The lack of muscle pull and weight-bearing is most pronounced when it affects the hip joint.

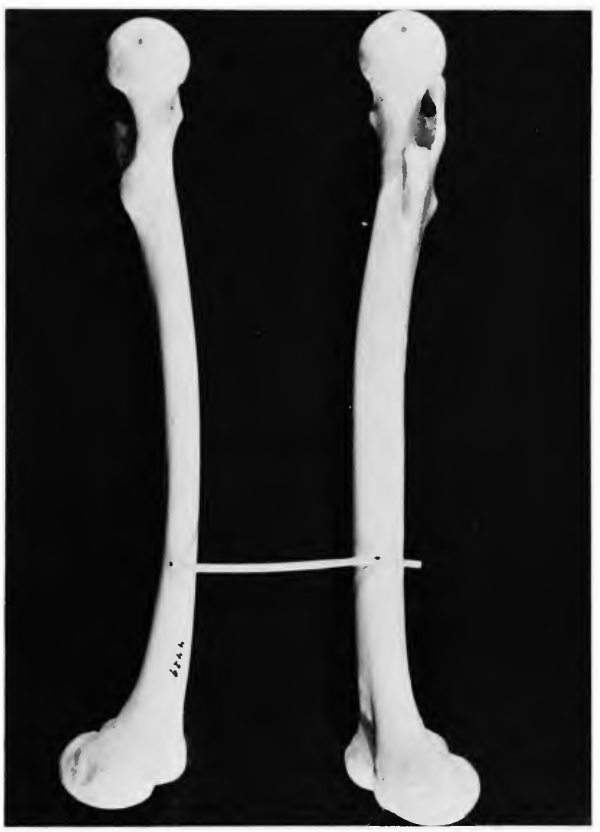


FIGURE 509.—Femora from individual with poliomyelitic paralysis of left leg. Notice small diameter and minimal surface relief of the affected left femur. (19-year-old male, died 15 years after onset of paralysis, IPAZ 4429, autopsy 468 from 1939.)

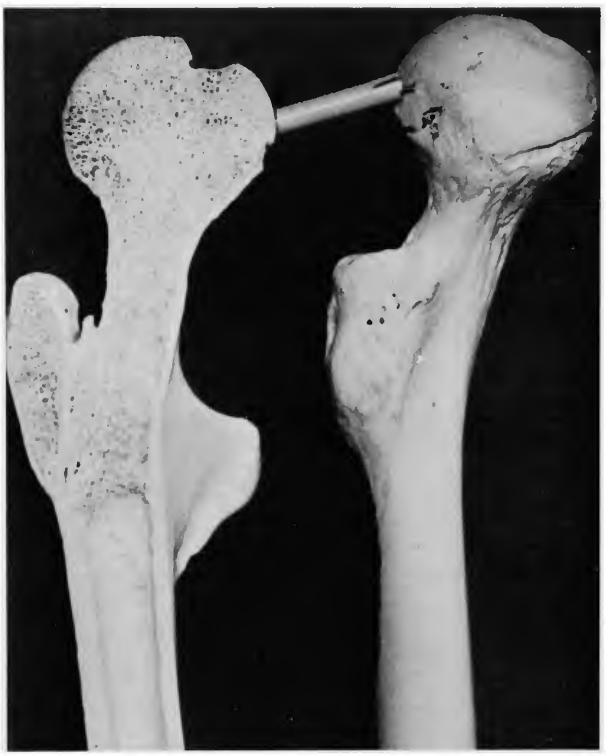
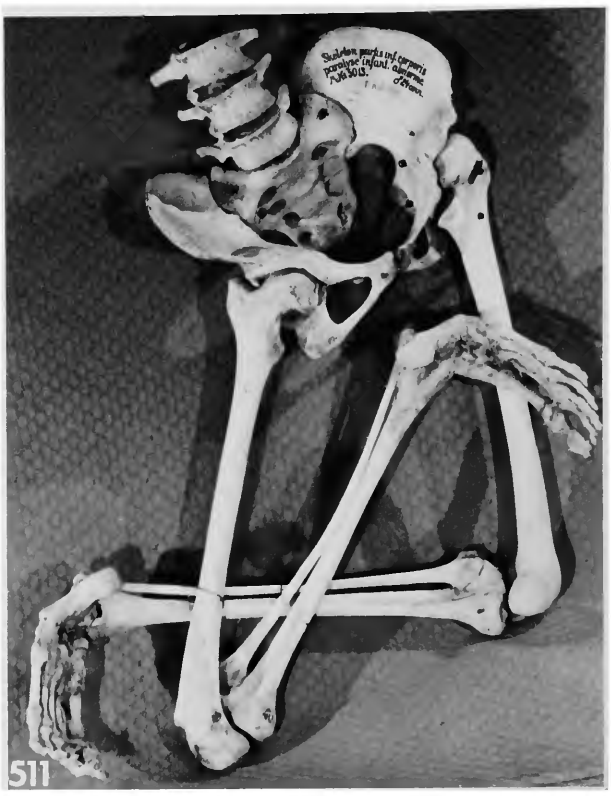


FIGURE 510.—Postparalytic deformity of proximal right femur showing extreme coxa valga, flattened, elongated lesser trochanter, slender femoral shaft and small femoral head. (14½-year-old female, 9 years after acute poliomyelitis, IPAZ 4337, autopsy 1372 from 1938.)

The paralyzed hip develops a much steeper angle of the femoral neck, which is slender and elongated (paralytic coxa valga) (Figure 510) (Putschar, 1937:701–705). The most common postparalytic foot deformity is a clubfoot (pes equinovarus), which may be combined with an exaggerated plantar arch and uptilted calcaneus (pes cavus) (Figures 511, 512).



FIGURES 511, 512.—Postparalytic deformities of distal skeleton, showing lumbosacral scoliosis, flaring ischia from sitting, flexion contracture of both knees, bilateral clubfoot with paralytic pes cavus deformity: 511, Overall view. 512, Dorsal view of left foot. (24-year-old male, many years after poliomyelitis, FPAM 5013, autopsy 88103 from 1880.)

PALEOPATHOLOGY

Scoliosis

References to spinal deformities in the literature on paleopathology are rare. This is due, in part, to the fact that vertebrae are not as well preserved as many other parts of the skeleton. Another problem is that kyphosis and scoliosis are most noticeable in an articulated spine and thus would tend to be overlooked in archeological specimens unless the defect was noticed during

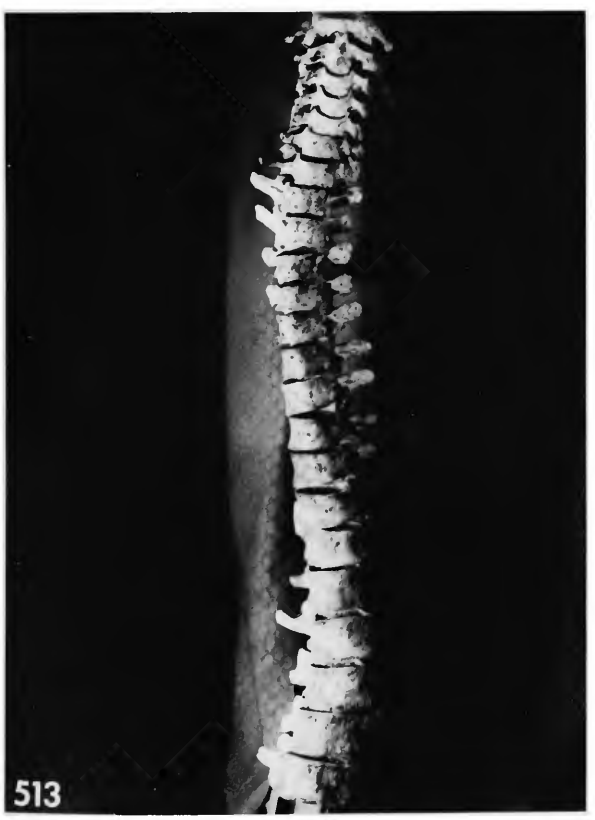
excavation. (For some examples of kyphosis induced by infectious destruction of vertebral bodies, see pages 125, 145, 166).

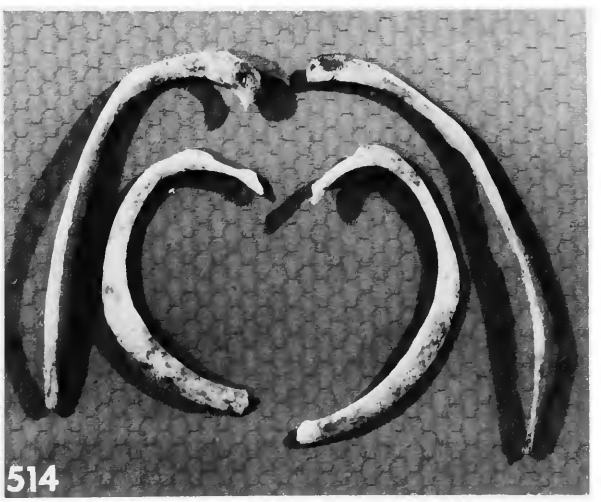
Brothwell (1961:327–328) reviewed five published and unpublished cases of scoliosis in British skeletons. One of these is from the Beaker period (ca. 2000 B.C.). Brothwell attributes the scoliosis in this specimen to congenital problems in which additional centers of ossification developed as semivertebrae. The anterior view shown on Brothwell's plate 8d clearly shows the lateral deviation that resulted from this abnormal development.

An interesting case of scoliosis comes from the archeological site of Hawikuh located near the modern village of Zuni in New Mexico, USA. The site includes pre-Columbian and historic components. During the early historic period the site was used as a base of operations for the Spanish explorer Francisco Vasquez de Coronado and a Franciscan mission was established there in 1630 (Smith, Woodbury, and Woodbury, 1966: 1). The site, including the cemetery, was excavated between 1917 and 1923. Skeletal material from both the pre- and post-contact period was recovered from the cemetery.

The skeleton in question (NMNH 314297) is from that portion of the cemetery associated with the historic period. However, the skull is morphologically different from the Indian Christian burials. J. Lawrence Angel (personal communication) suggests that the ethnic affinity of the skull is Spanish and that this skeleton may be the remains of one of the priests massacred during the Pueblo Revolt of 1680.

The estimated age of the skeleton is between 25 and 35 years. The sex is clearly male. There is no evidence of deformity on the skull although there is considerable antemortem tooth loss, including all incisors. The upper right canine was





FIGURES 513, 514.—Scoliosis: 513, Spinal column exhibiting an S-shaped lateral curve. 514, Second and tenth ribs showing asymmetrical development in response to scoliosis. (Adult male, NMNH 314297.)

lost shortly before death as evidenced by the reactive bone still in evidence in this part of the maxilla. Caries is well advanced in the upper left first molar although the corresponding molar on the right is worn but not decayed. Only the lower right third molar is present in the mandible, with clear evidence that most of the molars were lost antemortem.

The upper extremities have a slight lateral deviation of the distal humeri and considerable bowing of both radii and ulnae. The major abnormality is seen in the vertebrae, which exhibit an S-shaped scoliosis (Figure 513). The ribs (Figure 514) and manubrium are also abnormal reflecting the conditions on the vertebrae. The cervical vertebrae and first two thoracic vertebrae are normal. The first lateral deviation to the right begins at T-3 and continues to T-8 where the lateral deviation begins to be oriented toward the left, continuing until L-2 when the lateral orientation shifts back toward the right. The sacrum shows minimal evidence of abnormality and the pelvis appears normal. The major long bones of the lower limbs have a slight lateral bowing of the distal tibiae and fibulae. The combination of scoliosis and slight deformity of the limbs suggests the possibility of rickets in early childhood as the cause of scoliosis.

Postparalytic Deformities

Wells (1964a:92) reports the presence of a left humerus and radius, which is shorter and lighter than the right humerus and radius in a Neolithic skeleton from Sussex, England. He notes a similar deformity in a Bronze Age skeleton from Norfolk, England. Both of these cases are probable examples of postparalytic deformity.

Mitchell (1900) has described a predynastic Egyptian skeleton in which the left femur is shorter than the right. He attributes this condition to poliomyelitis. Elliot-Smith (1912) noted the existence of a deformity of the left foot in the Nineteenth Dynasty Egyptian mummy of Pharaoh Siptah. Initially he attributed the deformity to poliomyelitis but in a subsequent publication (Elliot-Smith and Dawson, 1924:100, 157) he refers to this specimen as an example of clubfoot.

Skeletal Dysplasias

The skeletal dysplasias can be grouped into two categories: (1) those affecting the proliferation and biology of cartilage and (2) those affecting the formation and/or resorption of bone.

As far as the group of cartilage dysplasias is concerned, a great number of syndromes have been described. Most of them are very rare and terminate fatally during infancy or early childhood. Others are survived but fail to show diagnostic features in the macerated skeleton that would permit classification beyond the term "dysplasia." For these reasons only classical achondroplasia, with typical disproportional dwarfism and some mucopolysaccharidoses will be discussed here in some detail. The dysplasias of bone formation and resorption, however, are better characterized and, in most instances, due to the tissue involved, exhibit identifiable skeletal changes. For this reason, even rare entities have been included in this discussion.

Achondroplasia

Pathology

Achondroplasia is a familial or sporadic congenital disorder of cartilage growth. This condition is the cause of the most common form of dwarfism. In many cases, affected persons can reach adult age. The basic defect is a severe inhibition of cartilage proliferation, thereby limiting the enchondral growth in all areas of the body. The enchondral ossification of the limited amount of proliferated cartilage proceeds normally and so does the intramembranous and periosteal bone formation. Adult achondroplastic dwarfs usually reach a height of 130 centimeters or less (Mørch, 1941:118). The dwarfism is disproportionate (Figure 515). This is due to the fact that the bones with the fastest growth and the least number of growth plates are most severely shortened. Thus, the trunk, although shortened, is least affected because of the great number of growth plates in the spine. The extremities are much more severely shortened and, in the extremities, the fast growing long bones, which have only two growth plates, are most markedly affected. The femur is more shortened than any other bone, followed by the humerus, and the bones of the lower legs and forearms. The fingers and toes with their numerous growth plates are less affected. The body disproportions in the achondroplastic newborn are about the same as in the adult. The discrepancy between longitudinal growth, controlled by cartilage proliferation, and circumferential growth, controlled by periosteal bone formation, results in changes in the shape of the bones (Figure 516). The diameter of the diaphyses and the cortical thickness are close to normal. The epiphyses and metaphyses are usually disproportionately wide as compared to the length of the bones (Figure 517). The degree of penetration varies from case to case and milder forms are observed. For details the reader is referred to the monograph of Mørch (1941), who studied more than 100 achondroplastic individuals. The skull typically is brachycephalic but large and bulbous. This is due to the discrepancy of the short skull base, which depends on enchondral growth at the intersphenoidal and spheno-occipital synchondroses, and the normal sized cranial vault, which is intramembranous in its development. The more normally large cranial vault over the short skull base exhibits an increased curvature, which is responsible for the bulging forehead. On the other hand, the shortness of the skull base is responsible for the markedly depressed bridge of the nose. Mørch (1941: 118) found this typical skull deformity in 80 percent of his cases. However, the characteristic shortness of all four limbs was present in all cases.

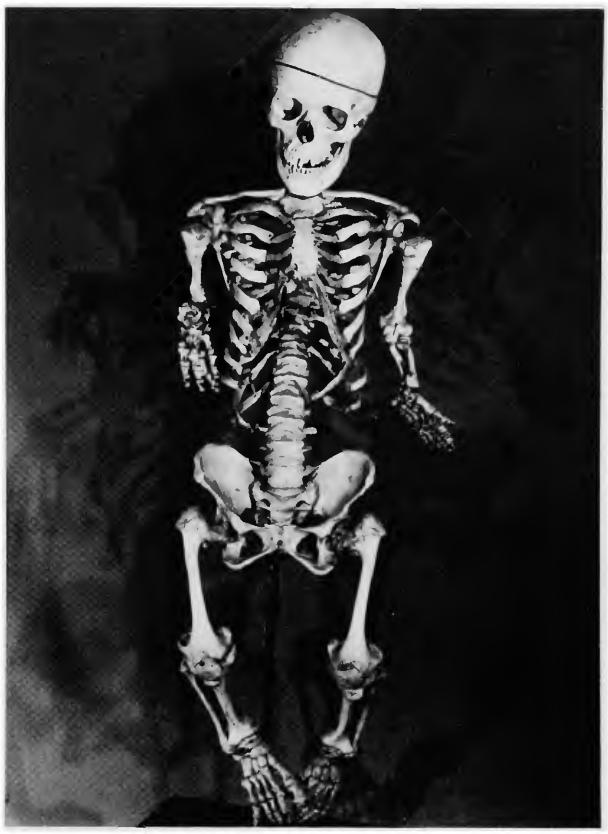


FIGURE 515.—Achondroplastic dwarf. Notice the short and thick bones; the rudimentary development of humeral and femoral heads with varus deformity is unusual. (Skeleton of adult female, who died in childbirth; PMUG 2668, autopsy 15050.)

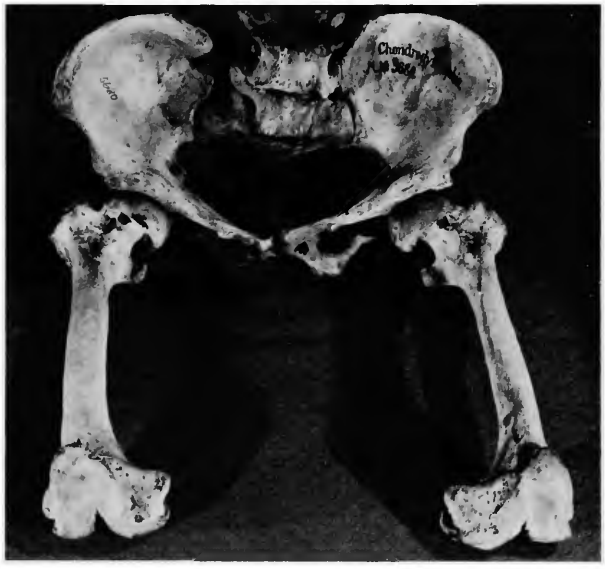


FIGURE 517.—Hyperplastic achondroplasia of right femur. Notice the short and thick diaphysis and the enlarged flaring metaphyses. (Newborn, WM S 59.3 from 1880.)

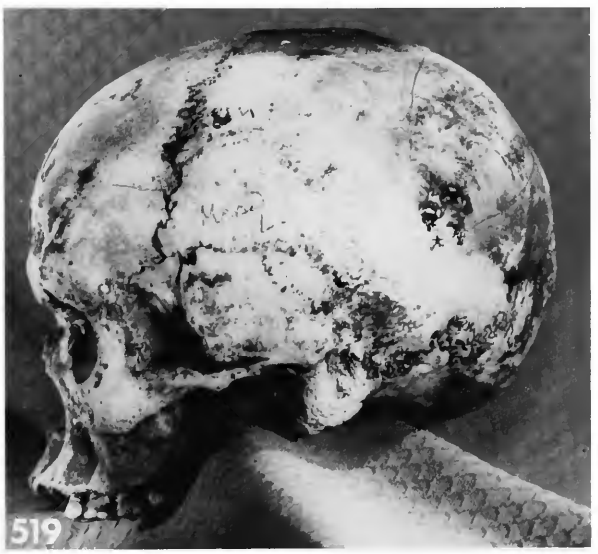
All patients also showed limitation of the extension of the elbow joint to 150 degrees and increased lumbar lordosis. About one-third showed lower thoracic kyphosis and approximately one-half revealed the typical "trident" hand, in which the fingers tend to be of more equal length than normal, with tapering terminal phalanges. The pelvis can be severely narrowed in all dimensions, particularly in the diameter between symphysis pubis and sacral promontory (Figure 516). This may create an insurmountable obstetric difficulty in the female achondroplastic dwarf in primitive populations, resulting in the death of the mother and the fetus (Figure 515).

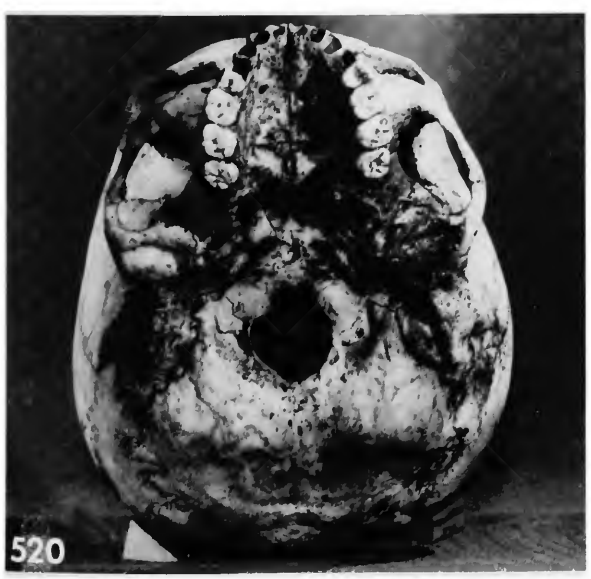
FIGURE 516.—Achondroplasia, showing narrow pelvis with flaring iliac wings and thick, short femora with accentuated muscular insertions. (Adult, probably female, FPAM 5680.)

Paleopathology

Seligmann (1912) published photographs and a brief description of a skull found by Sir Flinders Petrie in the temple of Thotmes IV at Thebes in Egypt (Eighteenth Dynasty). He states that the abnormality seen in this skull could be the result of cretinism or achondroplasia. In Seligmann's opinion the skull had poorly developed nasal bones, which led him to conclude that cretinism was the cause of the skull abnormality. Keith (1913:195–200), in his general discussion of achondroplasia, includes a description of this skull concluding that it was achondroplastic. The skull was from a female individual about 25 years of age. Keith's profile drawing shows shortening of the skull base with otherwise relatively normal development of the cranium, a condition that is characteristic of achondroplasia.

Bleyer (1940) concludes that a predynastic (ca. 5000 B.C.) skeleton from the Budari district in Egypt was achondroplastic. He also attributes the





FIGURES 518–520.—Achondroplastic skull from the Fourth Dynasty Egyptian tomb of King Mersekha: 518, Facial view. 519, Left, lateral view; note the depression in the region of the nose. 520, Skull base; note the abnormal shape of the foramen magnum and the short basioccipital. (BMNH AF.11.4/427.)

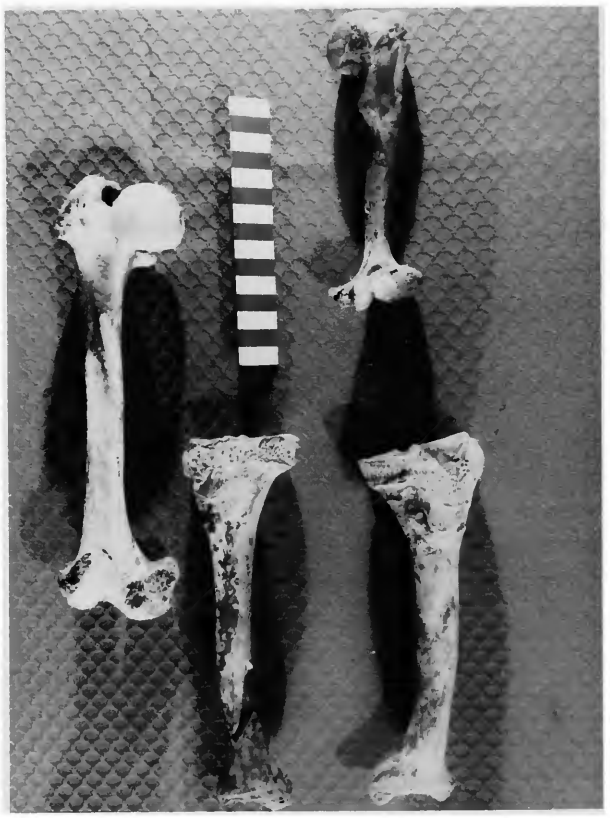


FIGURE 521.—Achondroplastic long bones (left humerus, right femur, and both tibiae) from the Fourth Dynasty Egyptian tomb of King Mersekha. Notice the short length but relatively normal diameter of the shafts and epiphyses. (BMNH AF.11.4/427; scale in cm.)

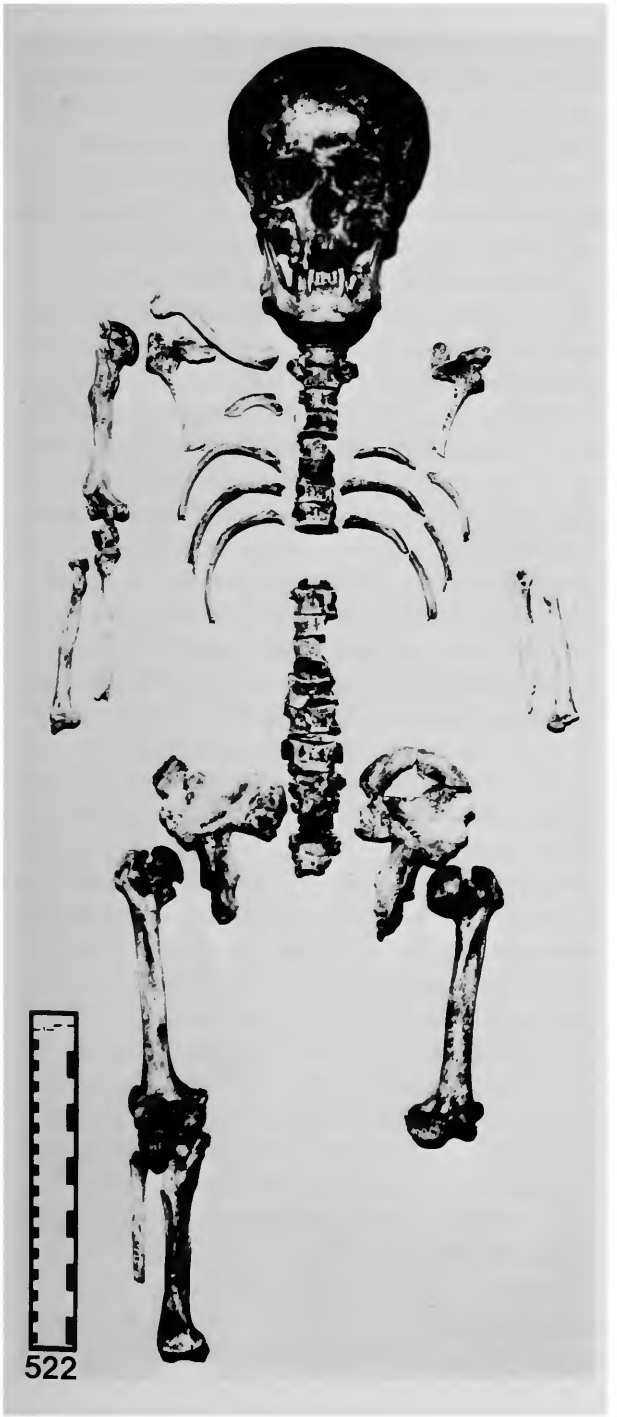
abnormality of a humerus found in the First Dynasty Egyptian tomb of King Zer to achondroplasia.

Brothwell (1967b:433) briefly describes two achondroplastic skeletal specimens from the Fourth Dynasty Egyptian tomb of King Mersekha. The first case consists of long bones only. The second specimen (BMNH AF.11.4/427) includes the skull, the left humerus, right femur, and both tibiae. I have studied and photographed the latter specimen, which exhibits the classic features of achondroplasia. The skull vault is of normal size (Figures 518–520). However, the skull base is shortened as evidenced by the very short basioccipital (Figure 520). The shortened skull base contributes to the appearance of a depression in the middle third of the face. The nasal bones and the frontal processes of the maxilla are broad, and the short face is accentuated by the prognathism of the alveolar portion of the maxilla. All adult teeth have erupted but have little wear, suggesting that the individual was a young adult. The long bones are very short (Figure 521) with the femur about 235 millimeters in maximum length. The epiphyses and apophyses are all fused, indicating adulthood. The tibiae have slight medial bowing of the distal half. The humerus is robust with the abnormal joint morphology associated with achondroplasia. The diaphyses of all the long bones have near normal diameters, indicating virtually normal periosteal bone formation.

In the New World, Fowke (1902:372) describes a skeleton, less than 152 centimeters (5 feet) in stature (undated but probably pre-Columbian) from mound number 4 near Waverly, Ohio, USA. The long bones were thick with extraordinary development of the areas of muscle attachment. The skull was thick and had a full and high forehead. Snow (1943:10) concluded that the features in Fowke's description were compatible with achondroplasia, and I would agree that such a conclusion is reasonable.

Snow (1943) provides unequivocal evidence of achondroplasia in indigenous New World skeletal material. Two dwarf skeletons were recovered during excavations of thousands of skeletons from the cemetery of an Indian village site at Moundville, Alabama, USA. No European or Colonial American trade goods were recovered during excavations indicating that the site was pre-contact.

The first of the two dwarf skeletons to be excavated was an adult female with a stature estimated to be about 125 centimeters. The posterior portion of the skull was not recovered. The forehead is prominent. The region around the nose is depressed, and the maxilla and mandible are prognathous. All the adult teeth had been lost antemortem. The postcranial bones exhibit the classic features of achondroplasia. The vertebral column, because of the multiple growth centers,



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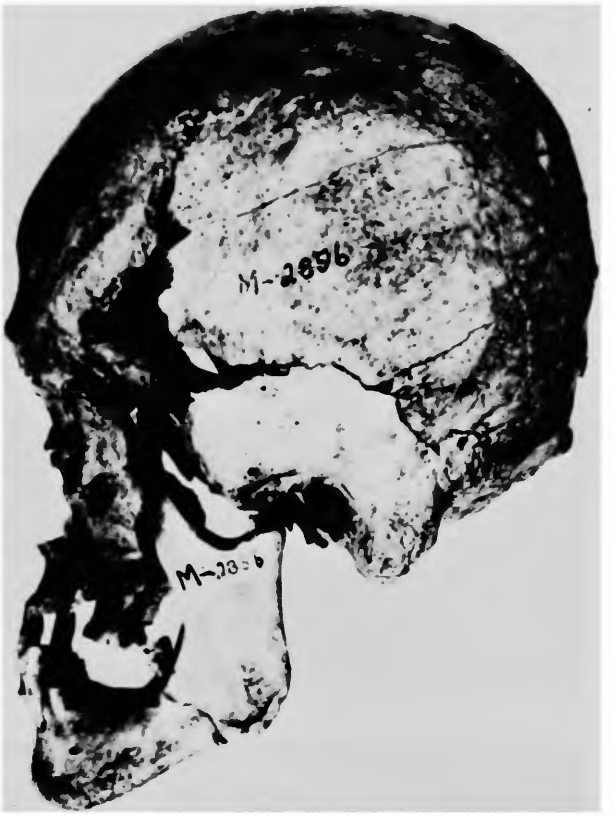

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FIGURES 522, 523.—Achondroplastic male skeleton from a pre-Columbian archeological site at Moundville, Alabama, USA: 522, Front view of skeleton (scale in cm and inches). 523, Left lateral view of skull. (Photograph from Snow, 1943, figs. 4, 6; courtesy Dr. Joseph Vogel and the Natural History Museum, University of Alabama.)

is less affected than the long bones. The bones of all limbs are short but robust.

The second skeleton is a male dwarf excavated five years later but from the same area of the mound (Figure 522). The reported stature of the male dwarf is about 150 centimeters, but this estimate is probably too large. The skull (Figure 523) is virtually complete and exhibits abnormal

features similar to the female dwarf. The cranium is large (Snow estimates cranial capacity to be 2011 cc), but the skull base is short, creating a midfacial depression. The individual was an adult but had lost many teeth to decay antemortem. The long bones are short but robust. The humeral and femoral heads are abnormally displaced distally. In the femora, the greater trochanter is at the same level as the femoral head. The morphological features of both dwarf skeletons are compatible with a diagnosis of achondroplasia.

FIGURES 524, 525.—Achondroplastic skull from ossuary IV, Accokeek, Maryland, USA: 524, Left lateral view; note the concave profile of the face. 525, Base of skull; note the abnormal foramen magnum and short basioccipital. (NMNH 379527.)

Hoffman (1976) provides a description of an archeological achondroplastic skeleton from Sacramento County, California, USA. The specimen is dated between A.D. 1500 and 1800. The skeleton is nearly complete and is currently part of the Lowie Museum of Anthropology skeletal collection (University of California, Berkeley, California, USA). The cranium is large and bulbous. The midfacial region is depressed and the cranial base shortened. The vertebrae are small and kyphotic at T11 and T12. Vertebral bodies of T10–L1 have projecting osteophytes creating a beak-like lateral appearance. The long bones are very short but robust exhibiting an external morphology common in achondroplasia. The kyphosis associated with the lower thoracic and upper lumbar vertebrae is also suggestive of Hunter's disease.

An isolated skull from ossuary IV at the Ferguson Farm, Accokeek, Maryland, USA, is probably another New World example of achondroplasia. The site is Late Woodland with no evidence of European trade goods associated with the ossuary. This suggests a pre-contact date. The skull vault is of normal size. The face is fragmentary, although there is little doubt that the region of the nasal bones was depressed (Figure 524). The base of the skull exhibits an abnormal foramen magnum and an abnormally short basioccipital (Figure 525).

Mucopolysaccharidoses

PATHOLOGY

The mucopolysaccharidoses are a group of diseases characterized by biochemical abnormalities in the mesenchymal cells, mainly fibroblasts and chondrocytes elaborating mucopolysaccharides. In these abnormal conditions, various mucopoly-

saccharides are, in part, accumulated in these cells, interfering with their normal function. Since mucopolysaccharides form a large part of cartilage matrix, cartilage proliferation and, therefore, skeletal growth and development are variously affected in these conditions. The base of these conditions is an inborn, genetically controlled, biochemical abnormality, which often manifests itself in several members of one family. Presently, six different mucopolysaccharidoses have been recognized (McKusick, 1972:521–686), of which only three are sufficiently well characterized in their skeletal manifestations to warrant a brief discussion here: Hurler's syndrome (gargoylism), Hunter's syndrome, and Morquio-Brailsford's syndrome.

HURLER'S SYNDROME (mucopolysaccharidosis type I).—This syndrome is transmitted as an autosomal recessive trait affecting children of either sex. The time and degree of its manifestations vary. Often it becomes apparent in infancy or early childhood and few individuals live beyond 10 years. Skeletal changes have even been observed in the newborn (Caffey, 1951, 1952). The main substance accumulated in fibroblasts and cartilage cells is chondroitin sulphate B. The abnormality involves many tissues and organs of the body, but we are here only concerned with the bone changes. In typical cases the affected growth cartilages lead to markedly stunted growth or dwarfism. Also, the development of secondary ossification centers is delayed and morphologically abnormal (Jaffe, 1972:545). Among the many deviations from the normal size and shape of bones observed, the changes of the spine and of the hands are the most typical. The spine shows a characteristic ossification defect at the dorsolumbar junction. This may affect the twelfth thoracic but usually involves the first and often, to a lesser degree, the second lumbar vertebral body. The cartilage model of any involved vertebra is ossified only in the posterior and inferior portions, giving the impression of a beak-like inferior projection on the lateral view of the X-ray or of the dry bone. This lack of anterior ossification at the dorsolumbar junction leads to a sharply angulated kyphosis at this level. The hand shows marked widening of the metaphyses of metacarpals and phalanges adjacent to the growth plate, with tapered narrowing on the other end. Thus, the enlarged metacarpals taper proximally, the phalanges distally. As usual, the first metacarpal behaves like a phalanx. The distal phalanges are hypoplastic. The changes are less marked in the corresponding bones of the feet.

Early in the disease, the long bones show periosteal bone deposition followed by increased endosteal cortical resorption, leading to widening of the marrow space and cortical thinning (Caffey, 1951, 1952). The humeri often show varus deformities of the head. Other deformities are coxa valga with plump femoral necks, shallow acetabula and genu valgum due to lateral tilting of the maldeveloped distal femoral epiphysis (Jaffe, 1972:542–552).

The skull frequently shows premature closure of the sagittal suture, giving a laterally narrowed, anteroposteriorly elongated, configuration. The skull base in the area of the sella is elongated; the mandibular condyles are flattened or even concave (Aegerter and Kirkpatrick, 1968:138–152). The skull bones are thick and dense. The scapulae are often broad and thick, as are the ribs, with exception of their misshapen necks and their narrow osteocartilaginous junctions.

HUNTER'S SYNDROME (mucopolysaccharidosis type II).—In this syndrome, which was only recently differentiated from Hurler's syndrome, the skeletal changes are practically identical with those of Hurler's syndrome. The inheritance is sex-linked so that only males are affected. Survival into adult age occurs.

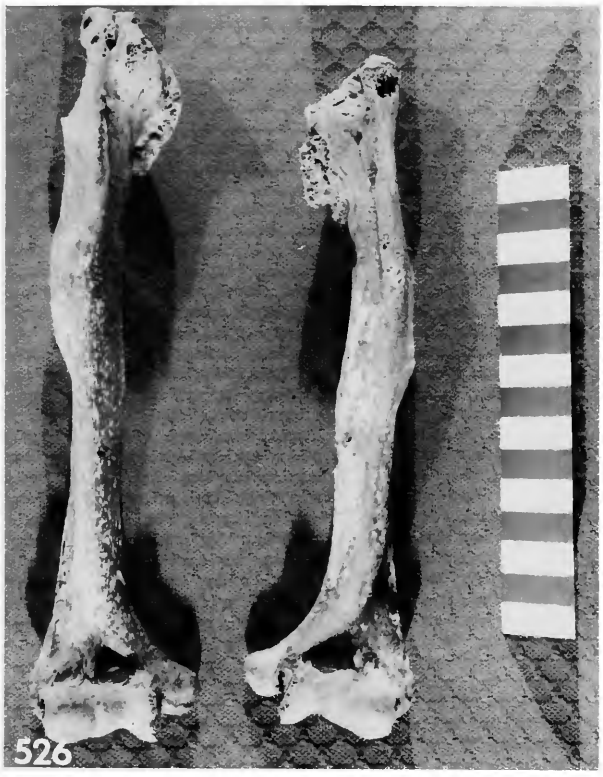
MORQUIO-BRAILSFORD SYNDROME (mucopolysaccharidosis type IV).—This syndrome is transmitted as an autosomal recessive trait. Both sexes are involved equally and there are usually multiple siblings affected in one family. The syndrome is not apparent at birth but begins to manifest itself in early childhood. The progressive accumulation of a mucopolysaccharide, probably mostly keratan sulfate, leads to skeletal changes and growth disturbances. The outstanding bone change is the flattening of all vertebrae with

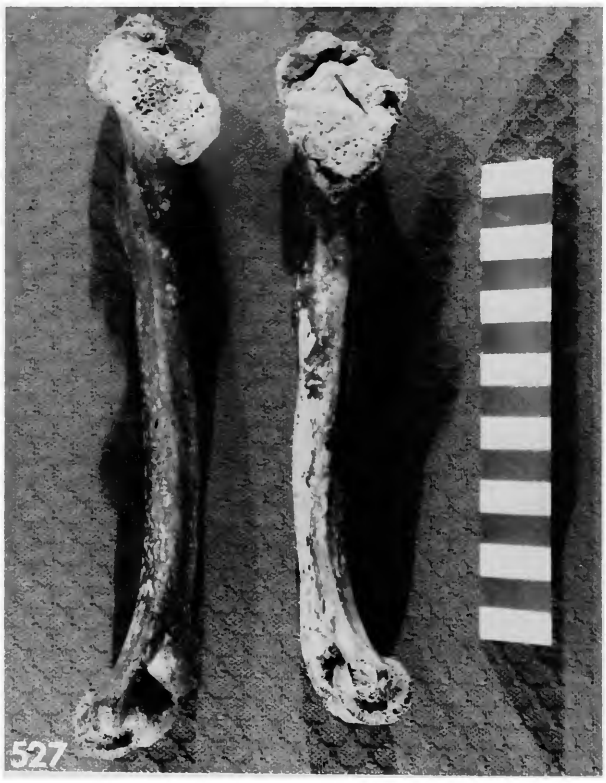
widening of the transverse and anteroposterior diameter. There is anterior wedging, especially at the dorsolumbar junction, leading to severe kyphosis. Subsequent to this deformity, the ribs show an altered configuration and the thorax is enlarged in its anteroposterior diameter. The sternum is angulated at the junction of the manubrium and corpus and protrudes forward. Growth is generally stunted, resulting in dwarfed stature. There is disproportionate shortening of the neck and trunk due to general flattening of the vertebral bodies (platyspondylia). The skull is normal and, therefore, relatively too large. The primary dentition is usually normal, but the permanent teeth are abnormal in shape and position (Aegerter and Kirkpatrick, 1968:141).

Secondary ossification centers are delayed and show abnormal configuration. The long bones may be significantly shortened and often show flaring of the metaphyses with lipping at the junction with the epiphysis. Similar spike-like projections occur on tarsal bones (Jaffe, 1972: 220). Hip deformities are common, consisting of large deep acetabula with irregular contour and delayed ossification of the femoral head (Jaffe, 1972:219).

Paleopathology

In the MacIvor collection stored in the British Museum (Natural History), there is a pair of deformed humeri (BMNH AF.11.3/75). There are no other bones associated with this specimen which is from early dynastic Egypt. Both humeri are abnormally short (Figure 526). The diaphysis is normal in diameter with a well-developed del-





Figures 526, 527.—Possible mucopolysaccharidoses in the isolated right and left humeri of an early dynastic Egyptian specimen: 526, Anterior view, showing the shortened length and abnormal development of the humeral heads. 527, Medial view, showing porosity and abnormal development of humeral heads. (BMNH AF.11.3/75; scales in cm.)

toid tuberosity. This combination of features is suggestive of achondroplasia. However, the humeral heads exhibit severe malformation of the articular surface with pitting of the subchondral plate particularly on the right (Figure 527). The left humerus is about 2 centimeters shorter than the right. Brothwell (1965a:164) includes an illustration of the right humerus in his discussion of congenital defects but does not attempt a specific diagnosis. The general appearance of the humeri is undoubtedly indicative of a chondrodysplasia, although the extreme abnormality of the humeral heads is not typical of achondroplasia. The external morphology of the humeral heads suggests an almost complete failure in the development of the epiphysis. This aspect is more compatible with conditions seen in the mucopolysaccharidoses. While the age cannot be determined, the fusion of the distal humeral epiphyses and apophyses indicate a minimum age of about 14 with a good probability of at least being a young adult. The age of the individual makes Hurler's syndrome a less likely possibility than either Hunter's syndrome or Morquio-Brailsford's syndrome.

Osteogenesis Imperfecta

Pathology

Osteogenesis imperfecta is based on an inborn deficiency of the mesenchymal cells in the production of mature collagen fibrils. It is often a familial disorder but also occurs as a spontaneous mutation. In severe manifestations of this disorder the collagenous fibrous tissue throughout the body is poorly developed. However, since the organic bone matrix almost entirely consists of collagen, skeletal changes dominate the picture.

The disease expresses itself in two forms: osteogenesis imperfecta fetalis (Vrolik type) and osteogenesis imperfecta tarda (Lobstein type).

Osteogenesis Imperfecta Fetalis.—In this condition the disease fully manifests itself in intrauterine life. Osteoblastic inhibition results in great reduction of enchondral and intramembraneous bone formation. The condition is often fatal at birth or in early childhood. Already in the uterus the fetus suffers numerous fractures, which show various stages of callus formation at birth. The newborn reveals markedly shortened extremities because of the accordion pleating of numerous transverse fractures of the long bones. The trunk is also shortened because of compression fractures of the vertebral bodies. Numerous transverse rib fractures exhibit multiple knobby swellings of fracture callus. The skull is soft and may show only very rudimentary ossification centers in the cranial vault. If the condition is survived for a longer time, multiple ossification centers may form, giving rise to Wormian bones. The bones show an extremely thin cortex and very sparse and thin cancellous trabeculae. Recognition of this type in skeletal remains would depend on ideal conditions of preservation. However, since the condition affects the entire skeleton, it could be surmised even from single bones.

Osteogenesis Imperfecta Tarda.—This condition is not manifest at birth but reveals itself during childhood and adolescence. The continued deficiency of bone formation results in frequent fractures, particularly of the long bones. Although normal bone formation is inhibited, trauma can overcome the osteoblastic inhibition and fractures heal. In some instances callus formation can be so excessive that it may be mistaken for an osteosarcoma. In these individuals, enchondral growth is normal, but periosteal bone formation is very limited. This results in bones of normal length but subnormal diameter of the shafts. The cortex is very thin and the cancellous bone sparse. The inhibited osteoblastic activity is also reflected in the delay of cortical remodeling. This can be demonstrated microscopically in undecalcified ground sections, revealing persistence of surface parallel lamellar bone and scarce osteonization. Multiple fractures may lead to severe deformities, especially of the weight-bearing bones of the lower extremities and the pelvis. However, some of the severe deformities are also due to bending by muscle pull and static forces, which the defective skeleton cannot resist (Figure 528). Any bone except the skull may be deformed. The tendency to develop new fractures usually

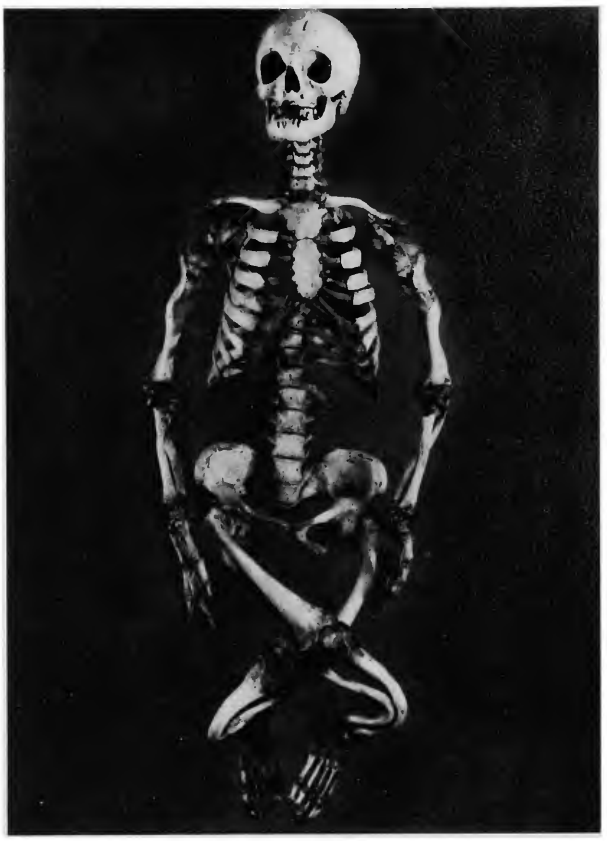


FIGURE 528.—Osteogenesis imperfecta tarda. Notice the severe bending deformities of both legs and the narrow pelvis. From a woman who died in childbirth. (PMES 1QAL(2).)

diminishes in adult age. Since odontoblasts produce the collagenous matrix of dentin, they are subject to the same suppression as osteoblasts. Teeth of the first and second dentition show deficient and discolored dentin, while the ectodermal enamel is normally formed. However, inadequate support of the enamel leads to chipping and early dental loss.

PALEOPATHOLOGY

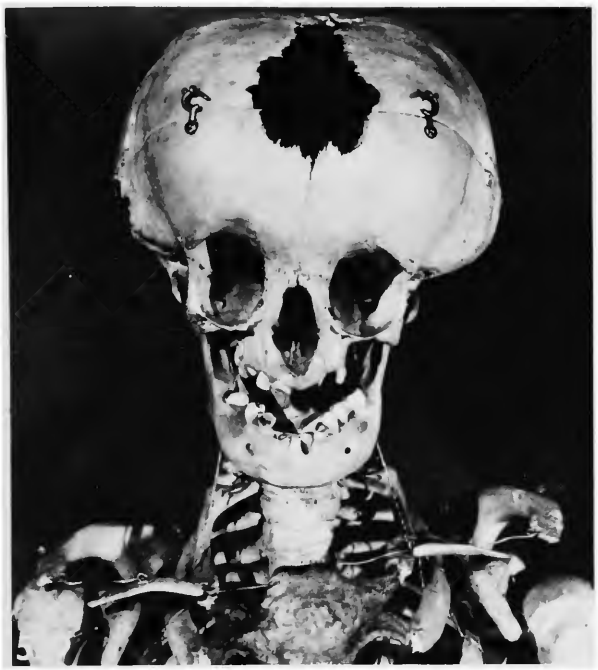
Wells (1965b) describes a burial from an Anglo-Saxon cemetery at Burgh Castle in Suffolk, England, dated about the seventh century A.D. The abnormality occurs in the left femur and consists of a 90-degree angulation of the proximal diaphysis. Wells estimates the age to be at least 18 years. The femur appears to have been fractured twice at the point of the angulation. Wells states that possible causes are rickets, fibrous dysplasia, and osteogenesis imperfecta. He rejects rickets, since that disease is unknown in other Anglo-Saxon burials, and fibrous dysplasia for unstated reasons, concluding that osteogenesis imperfecta is most likely. The age of the individual would indicate the tarda manifestation if osteogenesis imperfecta is the cause.

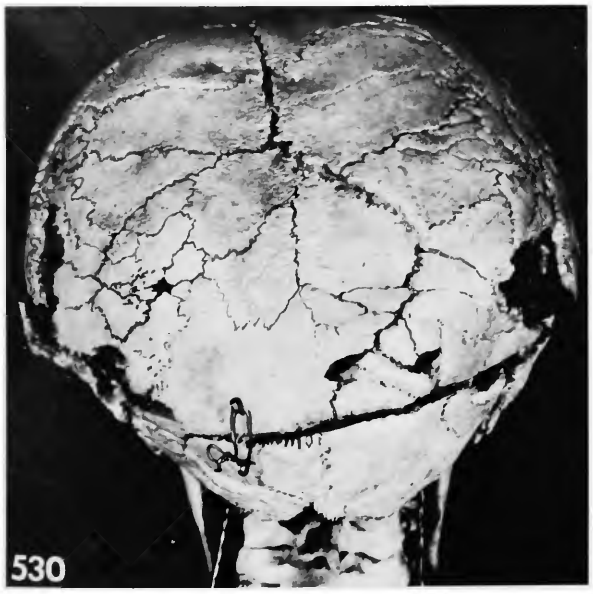
A more certain example of osteogenesis imperfecta is described by Gray (1970). The skeletal specimen is that of an infant from the cemetery of Beni Hassan on the east bank of the Nile at a site known as Speos Artemidos in Egypt. This site is dated to the Twenty-first Dynasty (ca. 1000 B.C.). The skeleton was found in a coffin and is currently stored in the British Museum, Department of Egyptian Antiquities (Registry No. 41603). The bones are friable and extremely light. The skull has an enlarged vault with vertically elongated eye orbits and multiple ossification centers. The teeth are also abnormal with poorly developed roots. The long bones are deformed. The bones of the lower extremities show well-marked anterolateral bowing. The cortex is thin and the spongiosa poorly developed. All of these features are compatible with a diagnosis of osteogenesis imperfecta.

Dysostosis Cleidocranialis

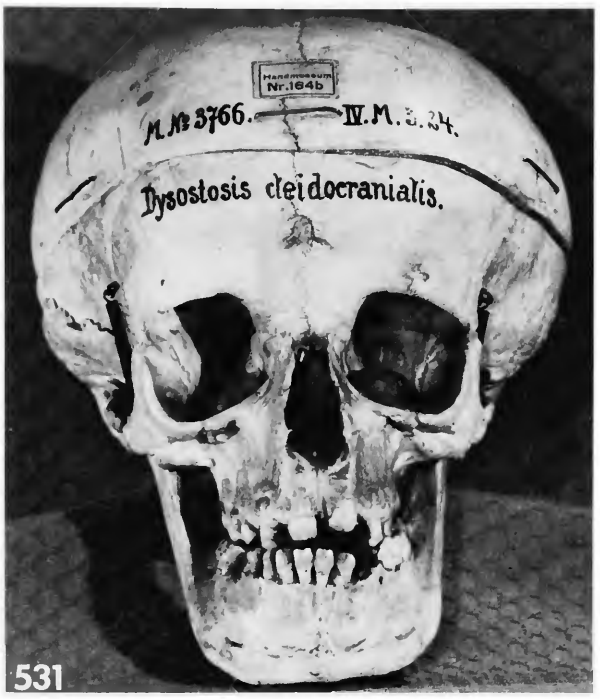
PATHOLOGY

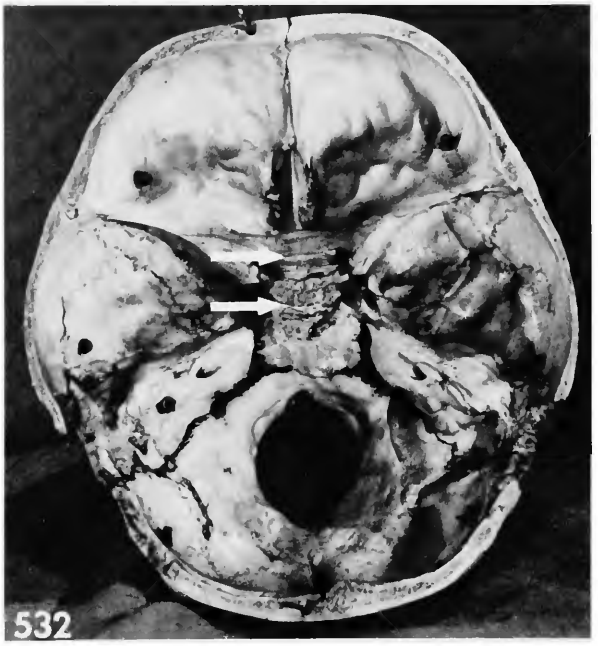
This is a congenital disorder of skeletal development mainly affecting intramembranous bone formation. The condition is usually familial but sporadic cases occur. The most marked changes concern the cranial vault and the clavicles (Figures 529, 530), both areas of intramembranous ossification. The skull is brachycephalic with increased transverse diameter and accentuated frontal and parietal bosses (Figure 531). The fontanels are large and persist long beyond the normal time for closure. The sutures are widely separated and remain open, including the metopic suture. The space between the sutures may





FIGURES 529, 530.—Dysostosis cleidocranialis: 529, Frontal view, showing enlarged frontal skull with wide-open anterior fontanel and rudimentary clavicles; notice delayed dental eruption. 530, Posterior view, showing Wormian bones. (25-year-old male, FPAM 5701 from 1909.)





FIGURES 531, 532.—Dysostosis cleidocranialis: 531, Frontal view, showing enlarged skull vault and delayed eruption of teeth. 532, Skull base, showing open intersphenoidal and spheno-occipital growth plates (arrows). (25-year-old male, FPAM 3766 from 1879.)

ultimately be occupied by numerous Wormian bones (Figure 530). The skull base may be slightly shortened and the basal growth plates fail to fuse (Figure 532). The facial bones may be small and the mandible may not be fused at the chin. Clavicles, which develop from three ossification centers, of which at least the middle one is intramembranous, are usually defective, unilaterally or bilaterally. They may be completely missing or, more often, only the lateral or only the medial third is developed. Failure to unite different portions of the clavicle may be observed (congenital pseudoarthrosis). In the rest of the skeleton, late and rudimentary ossification of the femoral head and neck may result in severe coxa vara deformity. The pelvic bones may show delayed or incomplete fusion of ischium and pubis. Dental development is affected, leading to delayed shedding of the primary dentition, delayed or incomplete eruption of the secondary dentition (Figure 531) and formation of dentigerous cysts. Since this disturbance does not interfere with survival, the adult skeleton may demonstrate changes that are fairly diagnostic of this anomaly.

FIGURE 533.—Osteopetrosis of both femora with pathological fracture of one. Notice the complete bony filling of the medullary canal and the lack of metaphysial remodeling. (15-month-old male, MGH autopsy 34143.)

Osteopetrosis

PATHOLOGY

Osteopetrosis is a congenital disorder of the skeleton characterized by severe inhibition of osteoclastic activity. The disease often is familial, occurring in several generations and in multiple siblings. There is no sex predilection. Parental consanguinity is common. The disease occurs in an infantile severe form leading to death in infancy or early childhood, and a mild form, starting in later childhood or adolescence and compatible with adult survival.

INFANTILE OSTEOPETROSIS.—This condition begins in fetal life. The basic defect is severe inhibition of osteoclastic activity without suppression of osteoblastic activity. This leads to preservation of primary trabeculae of enchondral growth containing cores of calcified cartilage. Continued deposition of bone on such trabeculae results in a jumbled, solid mass of bone and calcified cartilage, filling the bone marrow spaces and ultimately causing fatal anemia. All remodeling has to be initiated by osteoclastic resorption, but it is generally inhibited in this disease. This condition first becomes noticeable on the metaphysial side of the fast growing plates, such as the distal femur and proximal tibia; however, ultimately all bones show abnormal density. Since there is some fluctuation in the degree of osteoclastic suppression, parallel banding of denser and more lucent areas occur in the metaphyses. Corresponding to this are concentric layers of alternating degrees of density in ossification centers (Figures 533, 534). Ultimately, the whole medullary space is filled with the abnormal bone containing calcified cartilage (Figure 535). On the long bones the physi-

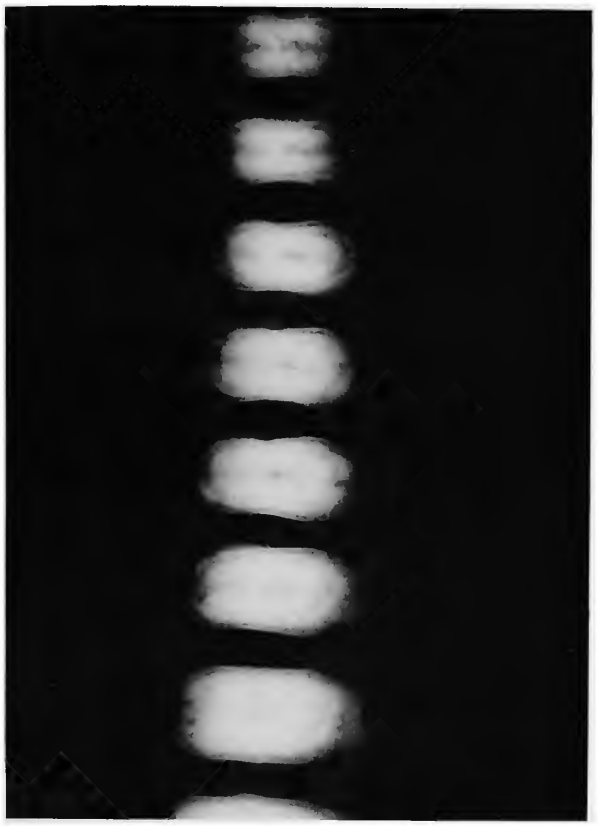


FIGURE 534.—Osteopetrosis. X-ray of vertebral bodies. Notice the great density and concentrical arrangement of bone. (15-month-old male, MGH autopsy 34143.)

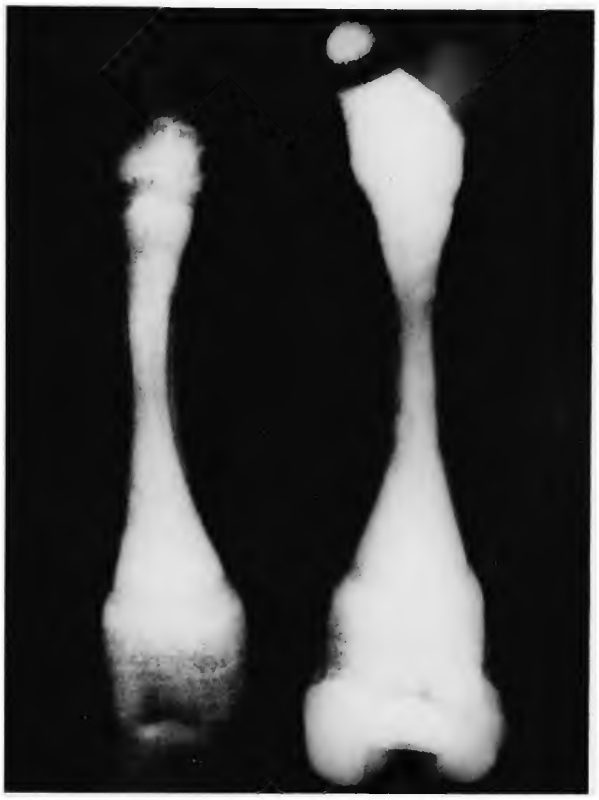


FIGURE 535.—Osteopetrosis. X-ray of femur. Notice the great density of the secondary ossification centers and the alternating bands of density and lucency in the unremodeled metaphyses. (15-month-old male, MGH autopsy 34143.)

ological remodeling of the metaphysis is inhibited, resulting in marked widening of this area instead of the usual flare. In spite of the excessive amount of bone present, its quality is inferior and a stress-oriented trajectorial trabecular system is not developed. Therefore, pathological, usually transverse, fractures commonly occur (Figure 533).

The skull shows thickening of the base and of the vault. In cases with severe anemia there may be expansion of the medullary space beneath the outer periosteum, giving a striated pattern on X-ray (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952, (1):699). The suppressed osteoclastic activity prevents the normal endocranial resorption and the necessary widening of the foramina for the passage of the various cranial nerves. The facial bones also become dense. The normally cancellous bones (vertebrae, ribs, sternum, pelvis) show marble-like density on X-ray without distinct trabecular pattern. The bones are heavy.

OSTEOPETROSIS TARDA.—In this milder form, the fetal encroachment on the bone marrow space does not occur and the remodeling defects on the long bones and the skull are slight or absent. However, the density and heaviness of many bones is apparent (Figures 536, 537), the inferior architecture of the bones remains, and repeated pathological fractures in adults are common. The delayed remodeling of the diaphysial cortex microscopically shows small osteons separated by residual periosteal and enchondral bone (Laubmann, 1935).

FIGURES 536, 537.—Generalized osteosclerosis, probably osteopetrosis: 536, Massive ivory-like osteosclerosis with occlusion of medullary cavity of distal right humerus. 537, Lower thoracic vertebrae with sclerotic obliteration of cancellous pattern. (45-year-old male with fractures and progressive anemia, WM S66.2 and S66.3 from 1932.)

Pyknodysostosis

PATHOLOGY

This hereditary congenital condition has some similarity to osteopetrosis, because the bones are unusually dense and easily fracture on minor trauma. However, the medullary cavities are not obliterated. The skull shows increased density, but the sutures remain widely separated and the fontanels open. The intervening portions of the cranial vault ultimately fill in with Wormian bones. An additional feature of this condition is tapering of the terminal phalanges with absence of the usual tufts. The individuals are frequently dwarfed, in part as the result of multiple fractures.

Metaphysial Dysplasia (Pyle's Disease)

PATHOLOGY

This is a familial hereditary disorder characterized by delayed remodeling of metaphysial cortical bone. The straight or even convex contour of the metaphysis is most marked on the distal femur and proximal tibia. Similar remodeling defects may be observed on the cranial base and facial skeleton, revealing thickened bones of increased density resembling so-called leontiasis ossea.

PALEOPATHOLOGY

Urteaga and Moseley (1967) provide an excellent report of a case of metaphysial dysplasia in a partial skeleton from a Mochica period (A.D. 200–800) burial from Peru. Unfortunately, except for the right humerus, the upper portion of the skeleton was destroyed by grave diggers. This humerus has an enlarged metaphysis, which extends toward the midshaft. The shaft itself is bowed laterally. The major abnormality is seen adjacent to both knees. In this region there is a pronounced failure of remodeling during the growth phase, which has created the classic Erlenmeyer-flask deformity of the metaphyses associated with metaphysial dysplasia. Both tibiae

FIGURES 538, 539.—Melorheostosis: 538, Left leg, lateral view, showing nodular and plaque-like periosteal hyperostosis. 539, Distal left femur, longitudinally cut section, showing medullary endosteal sclerosis and spotty epiphysial spongiosclerosis. (24-year-old male, IPAZ MB 6672 from 1964.)

are deformed with lateral S-shaped bowing. A longitudinal section through one femur and tibia reveals extensive cancellous bone development in the abnormal areas adjacent to the knee. The failure of cortical and cancellous bone remodeling in the metaphysial areas leaves no doubt about the diagnosis of metaphysial dysplasia.

Diaphysial Sclerosis (Camurati-Engelmann's Disease)

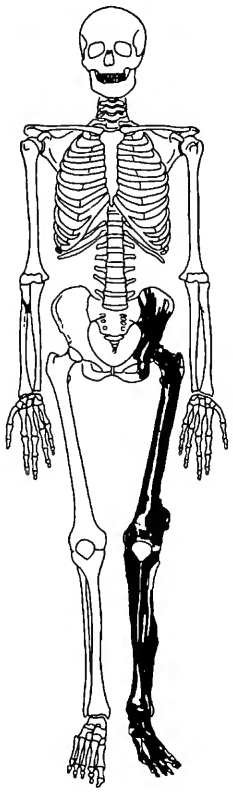
Pathology

This familial hereditary disorder is characterized by continued periosteal bone deposition over the diaphyses of tubular bones, sparing the metaphyses and epiphyses. This results in columnar or even fusiform shape of the long bones. The periosteal cortical thickening is most marked at middiaphysis and tapers towards both metaphyses. The cancellous bones and the skull are not involved. The disease usually manifests itself in childhood, but mild cases may not be discovered until adult age.

Melorheostosis (Leri's Disease)

Pathology

This is probably a developmental disorder usually only involving several bones of one extremity. The disease manifests itself usually in childhood and adolescence. The changes consist of streamlike deposition of dense bone on the periosteal and endosteal surfaces, resembling the distribution of wax running down on a candle (Figures 538–541). Usually a whole row of bones are involved in continuity, representing a ray of the extremity; for instance, humerus, radius, radial carpal, thumb, and index finger. Trunk and cranium are spared.



FIGURES 540, 541.—Melorheostosis: 540, Middiaphysial cross-section of left femur, showing periosteal, endosteal, and medullary bone deposition (24-year-old male, IPAZ MB 6672 from 1964). 541, Schematic drawing of the involvement of the left pelvis and leg in this case (courtesy Prof. E. Uehlinger).

Paleopathology

Lester (1967) provides a brief report of a skeleton found in an Ipiutak cemetery at Point Hope, Alaska. The specimen was dated by carbon-14 analysis to the fifth century A.D. The skeleton is male and reported to be in excess of 50 years. It is well preserved with most bones represented. The only bone affected by the disease process is the right fibula. The abnormality is characterized by a lumpy deposition of dense bone on the periosteal surface of the diaphysis. The metaphysis and joint surfaces appear to be spared. The medial aspect of the diaphysial cortex is normal. The external appearance of the abnormal bone and the dense roentgen-film appearance are compatible with a diagnosis of melorheostosis, although periostitis or a local osteomyelitis should be considered as well.

Osteopoikilosis

Pathology

This is a familial hereditary anomaly of the skeleton, which may manifest itself in several siblings. It has been found in the newborn. Since the anomaly is not visible on external inspection and is asymptomatic, only a chance X-ray will reveal its presence. The anomaly consists of multiple spotty condensations of dense lamellar bone in the spongiosa (Schmorl, 1931). The individual foci are one or several millimeters in diameter, round or oblong, and, in some areas, confluent. They appear on X-ray as foci of cortical density. They are most commonly found in the epiphyses and metaphyses of the long bones, the carpal and tarsal bones, and small tubular bones of hands and feet. There is, not uncommonly, symmetrical involvement. With the exception of the areas adjacent to the glenoid fossa and to the acetabulum, the bones of the trunk and, as a rule, of the skull are usually uninvolved.

This lesion is so characteristic that it should be readily identifiable on X-ray of dry bone provided that postburial sand infiltration, which could mimic the picture, can be excluded.

Osteopathia Striata

Pathology

This anomaly is similar to osteopoikilosis and likewise is not visible by external inspection of the bones. The lesion consists of parallel, streak-like condensations of cancellous bone, involving epiphyses and metaphyses of long bones. The skeletal distribution is similar to that in osteopoikilosis and mixtures of both anomalies have been observed.

Skeletal Malformations

Malformations of the entire skeleton are mostly related to incomplete twinning and usually are not compatible with prolonged postfetal life (Figure 542). The various forms of these malformations will not be discussed here. Less serious malformations of the skeleton are often due to failure of closure of fetal clefts or suppression of a portion of the skeletogenic mesenchyme. Many of these are compatible with life and maturity. Of these the most frequent and characteristic will be discussed here in their regional occurrence.

Skull

Pathology

The most frequent fatal malformation of the skull is anencephaly, a condition in which the cranial vault never developed and the rudimentary brain is directly exposed. This condition is not infrequently combined with partial or complete failure of closure of the neural canal (craniorachischisis). The skull base shows marked deformities of its constituent bones. The skull vault is absent and the orbits may be rudimentary.

A common malformation compatible with life is the uni- or bilateral lack of fusion of the globular and maxillary processes resulting in various degrees of hare-lip and cleft palate. The defect is located between the lateral incisor and the canine tooth.

Congenital herniations of meninges and brain will present on the skull bones as round, midline defects above the bridge of the nose or in the occipital area (Figures 543, 544).

Premature synostoses of one or several cranial sutures result in various characteristic skull deformities distinctly different from cultural, artificial deformations (Figure 545). The increasing pressure of the growing brain leads to separation of open sutures (Figure 546) and deep cerebral impressions on the inner table (Figure 547).

Paleopathology

Cleft Lip and Cleft Palate.—These abnormalities are uncommon in archeological skeletons. However, this does not mean that cleft lip and palate were rare in antiquity. Tretsven (1965: 229) found the incidence of cleft abnormalities among living American Indians in Montana to be higher than non-Indian people. He further notes (p. 236) that many American Indian groups have descriptive terms for cleft abnormalities in their native language. Mortality of infants with cleft lip and/or palate would have been high in antiquity. The defect inhibits effective nursing which would have contributed to early death. Also many infants with grossly observable deformities would have been killed.

Berndorfer (1962) has described a female skull about 25–30 years of age excavated in southern Hungary and dated to the fifteenth century A.D. The only abnormality of the skull is a poorly developed premaxilla and a small defect of the inferior aspect of the right pyriform aperture. The anterior alveolar region of the maxilla has a slight indentation suggestive of a cleft lip. The incisors are missing antemortem and the sockets for their roots are not present. Berndorfer rules out gingival atrophy following antemortem loss of the incisors, because the remaining teeth are normal. He does not consider cultural artificial extraction of the teeth as a possible cause of the slight defect in the alveolar bone. However, the abnormality of the pyriform aperture and subnormal development of the premaxilla would support a diagnosis of slight cleft lip.

Brooks and Hohenthal (1963) report the presence of cleft abnormalities in three indigenous skulls from two archeological cemeteries in California, USA. Two of the skulls are from the Newark site on the southeastern shore of San Francisco Bay. The skulls come from the Late Middle Horizon level dated by carbon-14 to 2340

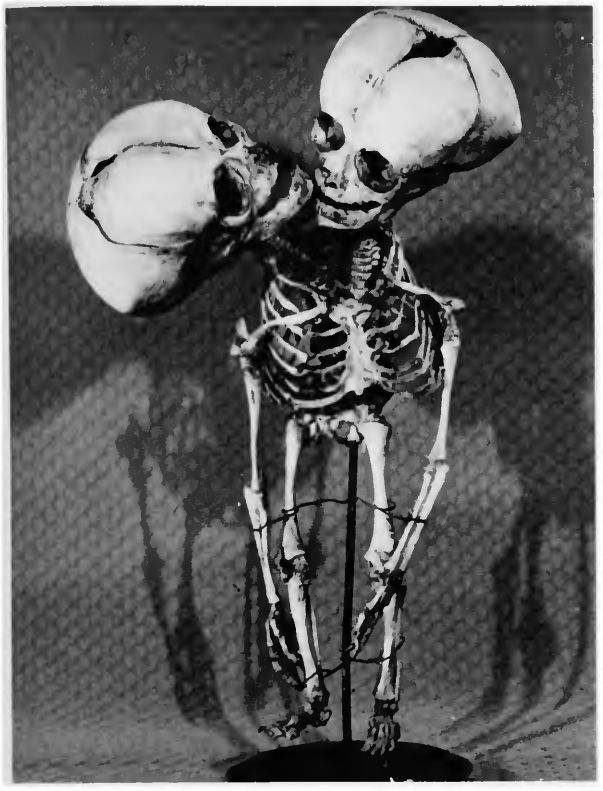
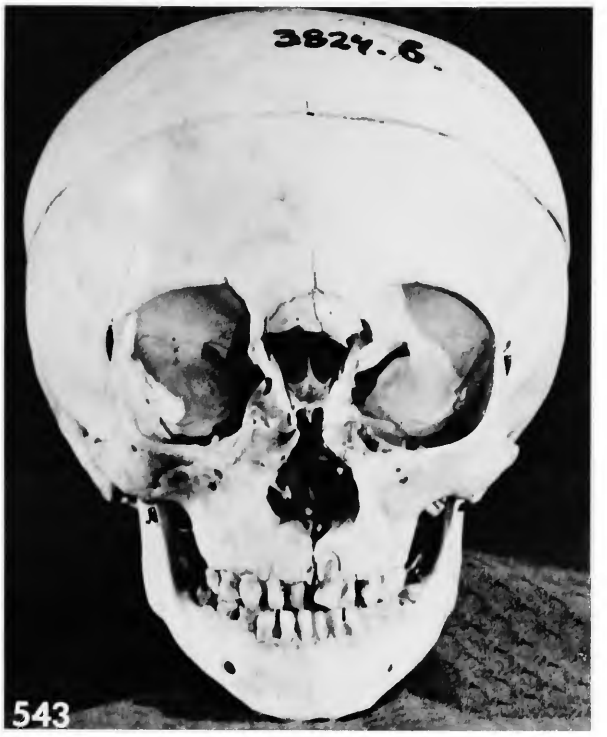
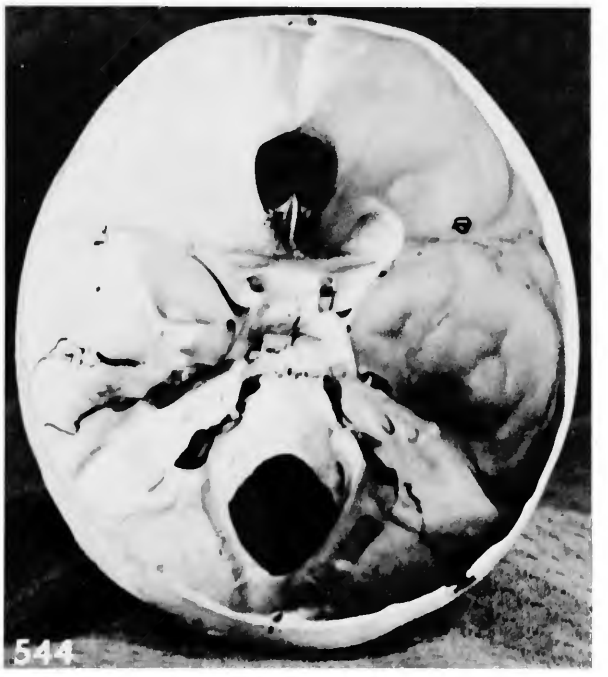


FIGURE 542.—Dicephalic infant with duplication of the spine. (Newborn, FPAM 1576.)

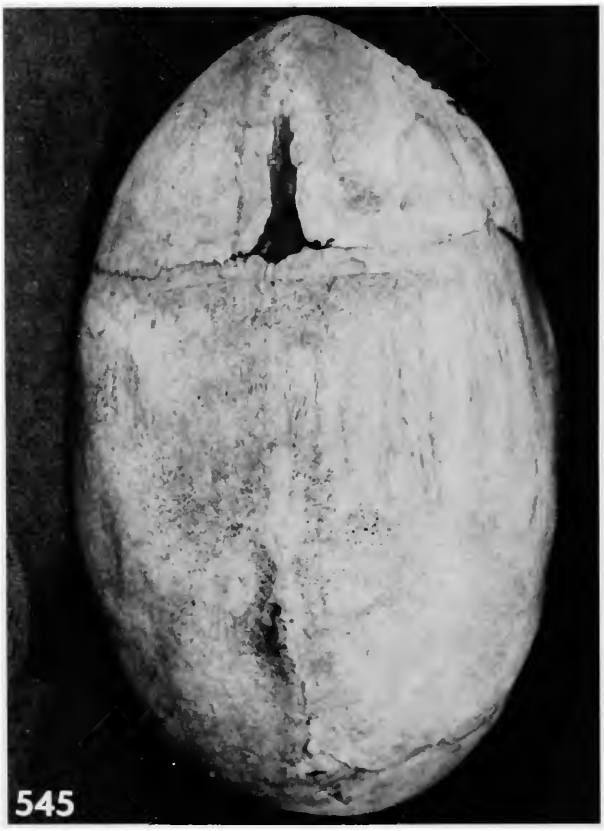
years B.P. The skulls are stored in the Museum of Anthropology at the University of California, Berkeley, USA (Nos. 8474 and 9859). The third skull (No. 22,117) is from a site in Sacramento County in California and is dated between 2000 and 4000 years B.P.

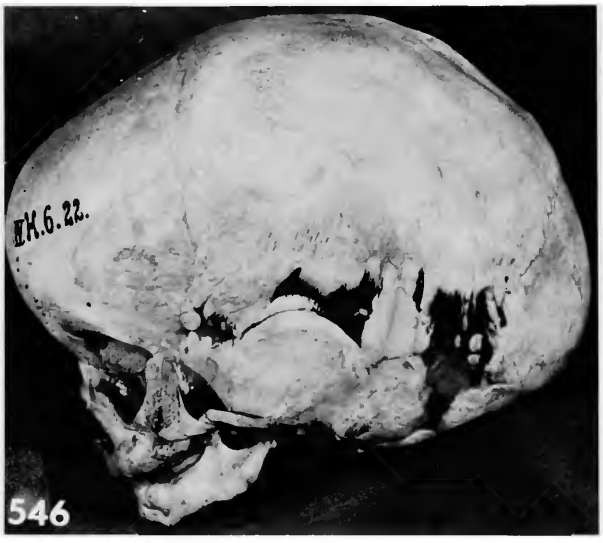
Skull No. 8474 is from a male about 30 to 40 years of age. Both the maxilla and palate have cleft defects. The upper right incisors were missing antemortem. The mandible is reported to be normal. Skull No. 9859 is also male with an estimated age between 22 and 25 years. The nasal bones are abnormal but the picture is obscured by postmortem damage. Both the maxilla and palate have cleft-type defects. The extensive nature of the facial abnormality added to the general porosity of the bones of the forehead and face are atypical of cleft palate and lip. The authors suggest the possibility of injury, although they conclude that cleft abnormality is the best interpretation of the defect. Cranium No. 22,117 is a





FIGURES 543, 544.—Anterior midline encephalocele: 543, Anterior view; notice flattened protruding nasal bones above defect. 544, Endocranial view; notice smooth-edged defect of ethmoid area. (5 years old, PMUG 3824, autopsy 17395 from 1890.)





FIGURES 545, 546.—Premature synostosis of sagittal suture with midline bony ridge and lateral defects along suture lines, secondary to intracranial pressure: 545, Top view. 546, Lateral view of skull. (9-week-old infant, FPAM 3576 from 1877.)

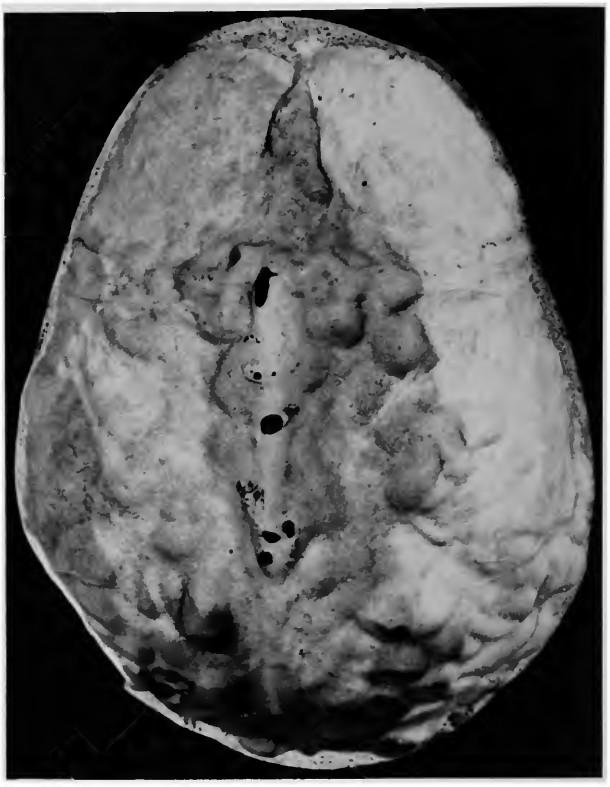


FIGURE 547.—Premature synostosis of the sagittal and mid-portion of the coronal suture, showing pressure atrophy with deep cerebral impressions and midline perforations of the cranial vault; endocranial view. (2-year-old male, FPAM 2118 from 1844.)

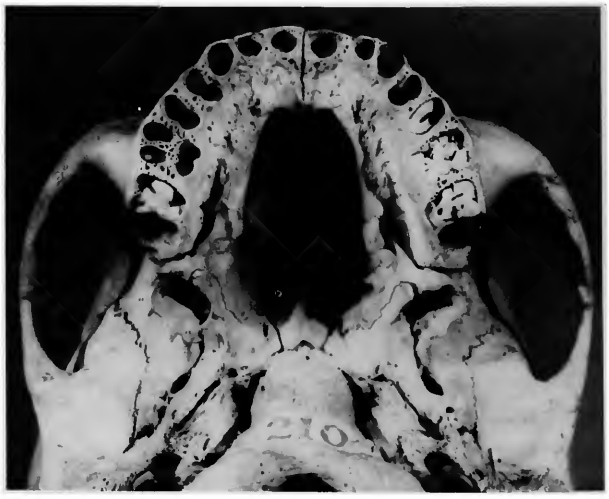
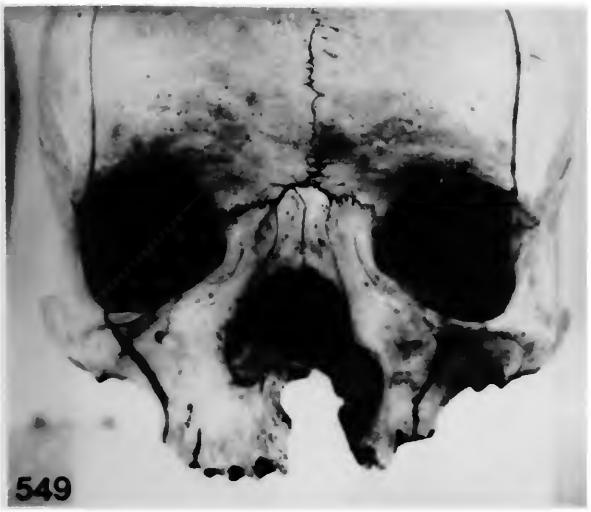
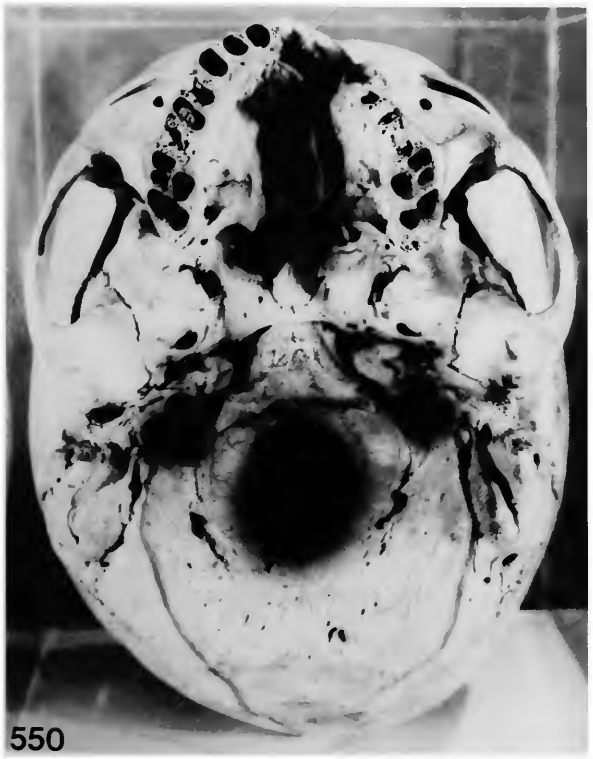


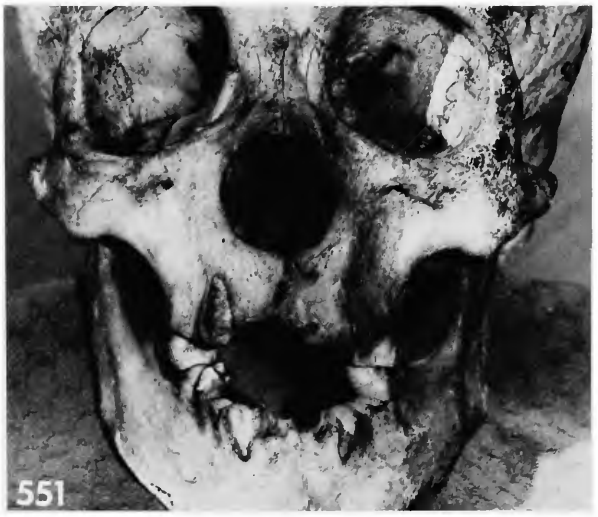
FIGURE 548.—Cleft palate of the central and posterior area. (Nubian skull, BMNH 210 72/291.)

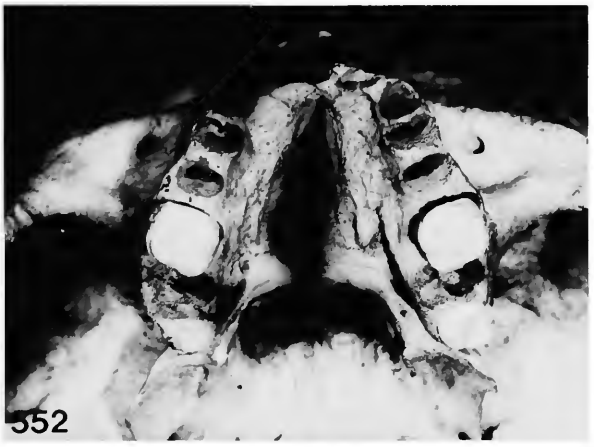




FIGURES 549, 550.—Cleft lip and palate in a skull from an indigenous South Pacific islander: 549, Facial view. 550, View of palate. (WM A1.3.)

female skull between 25 and 28 years of age. The skull has a marked unilateral loss of bone of the alveolar portion of the right maxilla. There are also extensive inferior bony projections on the

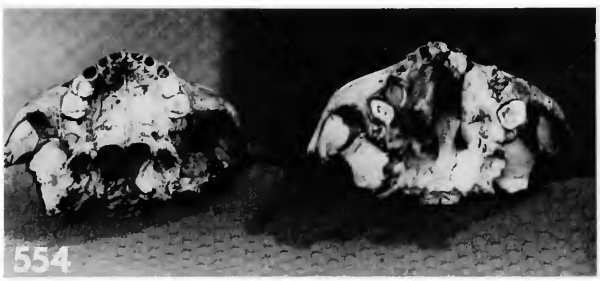




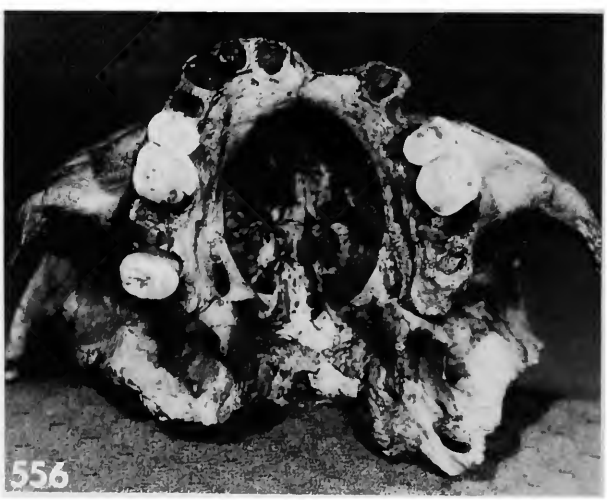
FIGURES 551, 552.—Cleft palate and possible cleft lip in a young adult, female skull from an archeological site in Colorado, USA: 551, Facial view. 552, View of palate. (NMNH 316482.)

maxilla near the suture with the zygomatic bone. The authors conclude that this is a case of unilateral hare-lip. On this specimen there is also evidence of porosity, which in my opinion is suggestive of a reactive response to injury and/or infection. Such a possibility should be kept in mind in studying paleopathological specimens with abnormalities of the maxilla and palate.

Derry (1938a) reports two examples of agenesis of the premaxillary region of the maxilla. The first example is a skull from a Twenty-fifth Dynasty Egyptian site on the east bank of the Nile.



FIGURES 553, 554.—Cleft lip and palate in the skull of an 8- to 10-year-old child from the Nasca region in Peru (right) compared with a normal skull (left) of about the same age from an archeological site in Kentucky, USA: 553, Facial view. 554, View of palate. (NMNH 293252, right; NMNH 243198, left.)



FIGURES 555, 556.—Cleft lip and palate in the incomplete adult skull from an archeological site in Kentucky, USA: 555, Facial view. 556, View of palate. (NMNH 243208.)

The skull is female, past middle age and normal except for the absence of the premaxilla. The hard palate is reduced in size and the maxillary incisors are absent antemortem. The provenience of the second skull is unknown. The skull appears to be from an adult female. As in the first skull the premaxilla is absent. The mandibles associated with the skulls project beyond the anterior border of the maxilla (mandibular prognathism) supporting the observation of subnormal development of the maxilla.

In the Nubian pathology collection of the British Museum (Natural History) there is a complete cranium of an adult female which exhibits cleft palate (BMNH 210 72/291). Most of the teeth have been lost postmortem, but judging from the intact dental alveoli the teeth were normal. The cleft defect is bilateral and involves only the central and posterior portions of the palate (Figure 548).

An example of both cleft lip and palate is seen in a skull from the Wellcome Museum of the Royal College of Surgeons of England in London (Figures 549, 550). The specimen (WM A1.3) is from an indigenous South Pacific islander. The right side of the alveolar arch is relatively normal, although there is a complete cleft through the left portion of the anterior maxilla, which is contin-

uous with the cleft of the left hard palate. The maxillary defect is continuous with the pyriform aperture.

Three specimens from the National Museum of Natural History collections at the Smithsonian Institution, Washington, D.C., USA, provide additional evidence for the presence of cleft abnormalities in New World indigenous populations. The first of these two specimens is a complete cranium of a young adult female about 18 to 20 years of age from southwestern Colorado, USA (NMNH 316482). The archeological age of the skull is unknown. The skull exhibits some occipital flattening, which is unrelated to the abnormality of the maxilla. Both maxillary central incisors and the right lateral incisor are absent antemortem. The sockets are absent and the alveolar bone is thin. Dental alveolei are present for the remaining teeth although the socket for the upper right canine is poorly formed with the anterior portion missing. There is a small cleft in the central portion of the intermaxillary suture (Figure 551). The nasal bones are depressed about 15 millimeters below nasion, as if the result of a healed blow in childhood. The palate is cleft primarily to the right of the midline with slight abnormality to the left (Figure 552). Both the palatine process of the maxilla and the palatine bone are involved although the defect is less severe in the posterior portion of the palate. The mandible is normal.

The second specimen is from an 8- to 10-year-old child from the Nasca region in Peru (NMNH 293252). The archeological age is not known. Only the anterior portion of the skull is present including the frontal, sphenoid, zygomatic, nasal, palatine bones, and the maxilla. The cleft defect is primarily on the left side (Figure 553, 554) and involves the alveolar and palatine process of the maxilla, as well as the palatine bone and the internal bones of the nasal cavity. The cleft of the alveolus and palate are continuous with each other and with the nasal cavity. There has been a lateral deviation of the intermaxillary suture toward the right side of the face.

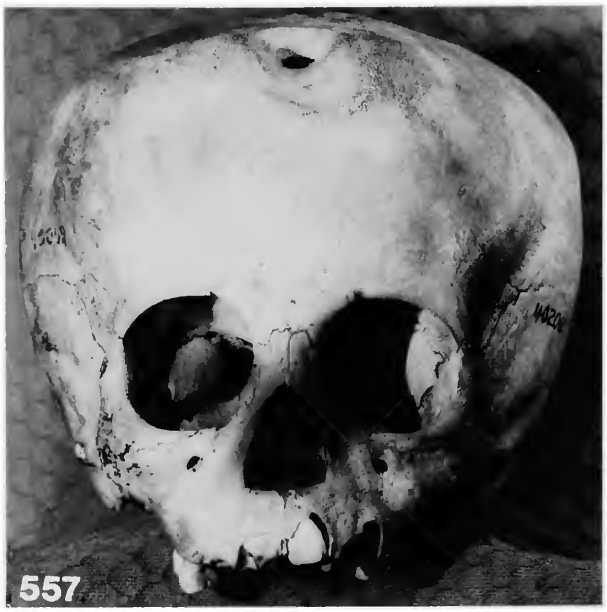
The final example of cleft palate is from an archeological site in Kentucky, USA (NMNH 243208). The archeological age is unknown. The alveolar portion of the left maxilla may have been affected as is suggested by the antemortem loss of the central and lateral incisors (Figures 555, 556). However the major defect is a large oval opening in the palate. The vomer and conchae are missing as well.

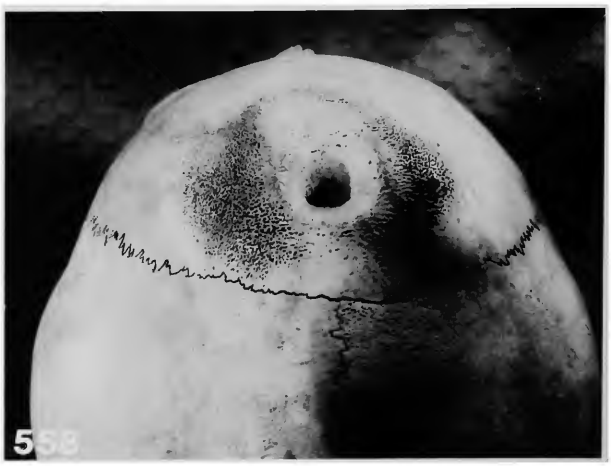
The above-published and original examples serve to document the presence of the cleft lip and palate in both New and Old World indigenous ancient populations. They also serve to illustrate the point that other conditions such as trauma and infection need to be considered in differential diagnosis.

Congenital Herniation.—Stewart (1975) has called attention to the misinterpretation of congenital herniations (dysraphism) of the skull. The defects in the bone produced by such herniations have been attributed to the practice of trephination (Powell, 1965; Sublett and Wray, 1970). Both examples, published by Stewart (1975), exhibit herniation very close to bregma. They have a sharply defined anterior border but a more gradual slope to the depression posteriorly.

Another example of this abnormality was brought to my attention by T. D. Stewart. The skull is from the skeletal collections of the Field Museum of Natural History (FM 40208) and is from a child 6 to 8 years of age from Ancon, Peru. The archeological age is thought to be between A.D. 1000 and 1200. The large abnormality is located on the frontal bone anterior to bregma (Figures 557, 558). The defect extends completely through both tables of the frontal bone. The margin is most sharply defined on the posterior edge with a gradual elongated depression extending anteriorly about midway toward nasion. Lateral to the defect are two areas of porous reactive bone. The defect itself is well circumscribed, exhibiting smooth compact bone throughout.

The nature of the soft tissue lesion, of course, is not known. Stewart (1975:437) notes that the prognosis in a living patient depends on the type of herniated tissue and the amount of herniation. The prognosis for survival to adulthood would be poor in a case where the herniation was as extensive as evidenced by the size of the skull defect.





FIGURES 557, 558.—Congenital herniation of the skull vault in a 6- to 8-year-old child from an archeological site in Ancon, Peru: 557, Facial view. 558, Top view; note the porous, reactive bone surrounding the herniation. (FM 40208.)

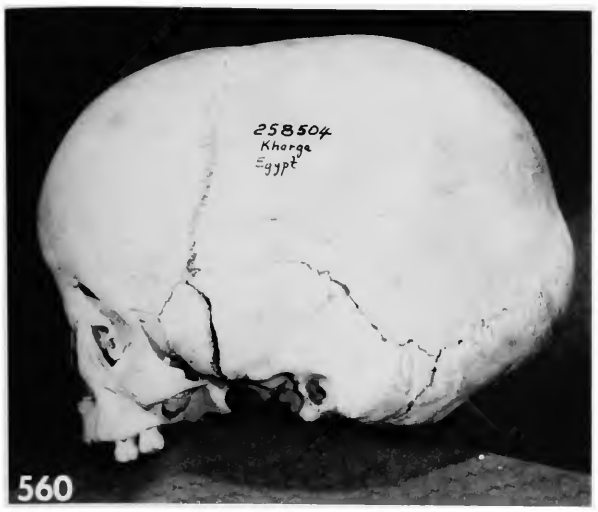
Thus, it appears that this abnormality may have led to the premature death of the child.

BIPARIETAL PERFORATIONS.—Broca (1875) has described another congenital defect of the skull vault in which there are unusually large perforations of the parietal bones often occurring as an enlargement of the parietal foramina. An example of this condition is seen in a skull from California, USA (NMNH 276981). The skull is that of an adult male with bilateral perforations of the parietals exceeding one centimeter in diameter (Figure 559). The perforations are located on the posterior portion of both parietals near the sagittal suture.

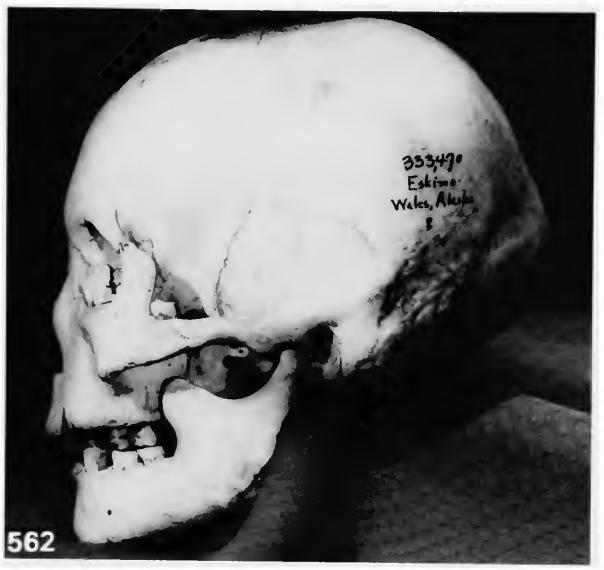
PREMATURE FUSION OF SUTURES OF THE SKULL (Craniostenosis).—This abnormality can change the normal shape of the skull. The changes in shape will depend on which sutures are involved and the age of onset of the fusion. For example, fusion of the sagittal suture in early childhood produces an elongated skull with a prominent forehead. Fusion of the coronal suture in early childhood results in a shorter skull but prominent development of the frontal region with a very high forehead. Several reports on craniostenosis in archeological skeletons have been published (e.g., Davis, 1865; Eiseley and Asling, 1944; Hohenthal and Brooks, 1960; Comas, 1965; Bennett, 1967; Stewart, 1972).

Of the many examples of craniostenosis in the collections of the National Museum of Natural History, Washington, D.C., USA, I have chosen

FIGURE 559.—Biparietal perforations in an adult skull from an archeological site in California, USA. (NMNH 276981.)

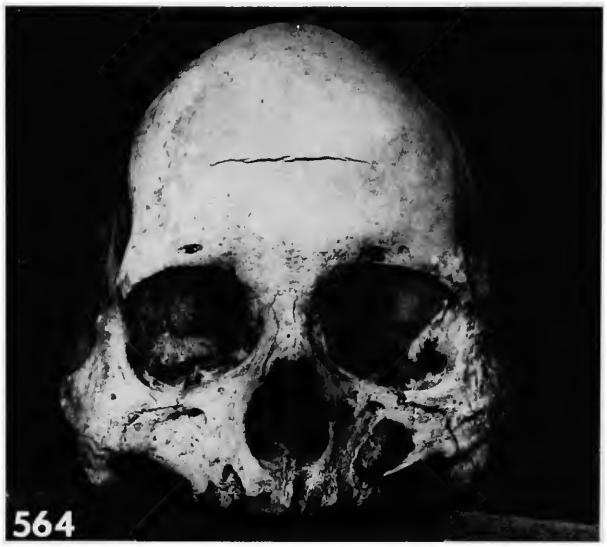


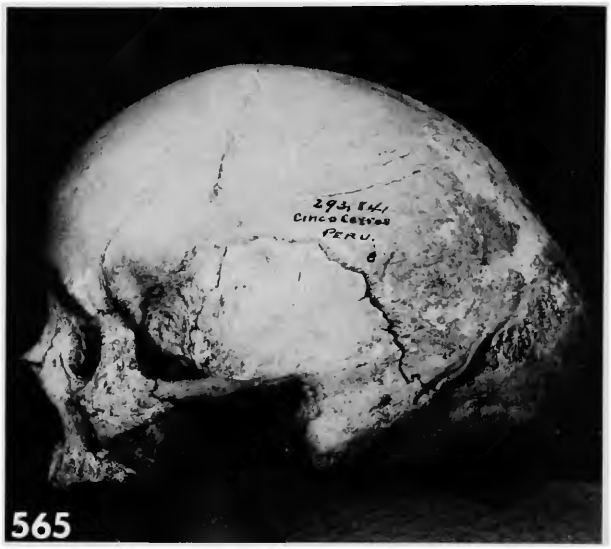
FIGURES 560, 561.—Premature fusion of the sagittal suture in the skull of a 3- to 4-year-old child from the Twentieth to Twenty-fifth Dynasty site of Karga in Egypt: 560, Left lateral view; note the somewhat abnormal length of the skull. 561, Top view, showing the fused sagittal suture. (NMNH 258504.)



FIGURES 562, 563.—Premature fusion of the coronal and sphenoparietal suture in a young adult skull from Wales, Alaska, USA: 562, Left lateral view, showing the complete fusion of the sphenoparietal suture and the slight saddle-depression near bregma. 563, Top view. (NMNH 333470.)

three cases to illustrate the relevant features associated with this abnormality. A child's skull (NMNH 258504) from the Twentieth to Twenty-fifth Dynasty site of Karga in Egypt illustrates premature closure of the sagittal suture (Figures 560, 561). This child was 3 to 4 years of age at the time of death. Except for the anterior 2 centimeters, the entire sagittal suture was fused. All other sutures, which are normally open at 3 to 4 years of age, are open. Even at this early age, the skull is slightly elongated with a slight axial





FIGURES 564–566.—Early premature fusion of the sagittal suture in an adult male skull from an archeological site in the region of Cinco Cerros, Peru: 564, Facial view, showing the keel-like vault of the skull. 565, Left lateral view. 566, Top view. (NMNH 293841.)

deviation of the skull suggestive of excessive growth on the right side of the skull vault.

Premature fusion of the coronal suture is seen in a female Eskimo skull from Wales, Alaska, USA (NMNH 333470). The skull is from an individual about 25 to 30 years of age at the time of death. Endocranially the coronal, sagittal, and lambdoid sutures are fused. Ectocranially both the sagittal and lambdoid have begun to fuse. Except for 2 centimeters on either side of the midline, the coronal suture is completely fused ectocranially with obliteration of the sutural lines (Figures 562, 563). The sphenoparietal suture is also fused and obliterated. In profile the skull vault exhibits a saddle-like depression just posterior to bregma. Otherwise, the shape of the skull

is normal indicating that the premature fusion of the coronal and sphenoparietal suture occurred after significant growth of the brain and skull had ceased.

Another example of craniostenosis in an adult skull illustrates the morphological features that occur when premature fusion occurs early in childhood or infancy. The skull is from Cinco Cerros, Peru, and is from an adult male (NMNH 293841). The archeological age is not known with certainty but is thought to be pre-Columbian. The skull is long and very narrow (markedly dolichocephalic) with the elongation primarily in the posterior portion of the skull (Figures 564–566). The sagittal suture is completely fused both endo- and ectocranially. The coronal and lambdoid sutures are completely fused endocranially and partially fused ectocranially.

HYDROCEPHALUS.—Although this condition is rarely a congenital abnormality, it does produce malformation of the skull that could be observed in archeological specimens; for this reason brief mention of this condition is included below. Derry (1913) provides a detailed report on an abnormally large skull and postcranial bones from the Roman period in Egypt. The bones are those of a fully adult male. Derry reports that the cranial capacity is at least 2900 cubic centimeters, which exceeds the normal range of cranial capacity by several hundred cubic centimeters. The lateral drawing of the skull exhibits relatively normal proportions for the face so that the skull abnormality is limited to the brain case. The postcranial bones exhibit abnormalities that Derry attributes to partial paralysis of the left side. Grim and Plathner (1952) describe a hydrocephalic skull from central Europe dated to the Stone Age. Armelagos (1969) reports the presence of a hydrocephalic skull of a child in Nubian skeletal material dated between A.D. 350 and 550.

Spine

PATHOLOGY

Complete lack of closure of the neural canal shows the two halves of the spinous processes and neural arches pointing laterally (complete spina bifida). Incomplete bony fusion of one or several spinous processes (spina bifida occulta) is common, especially in the sacral area (Figure 567). Lateral or dorsal hemivertebrae occur, causing abnormal curvature of the spine (congenital scoliosis and kyphosis) (Figures 568, 569). Segmental disarrangement of vertebral ossification centers may result in fusion of vertebral segments, particularly in the cervical area (Klippel-Feil syndrome). Symmetrical or asymmetrical sacralization of the fifth lumbar vertebra is common. Lumbarization of the first sacral vertebra is more rare. The sacrum and coccyx may completely fail to develop, resulting in close approximation of the posterior iliac spines.

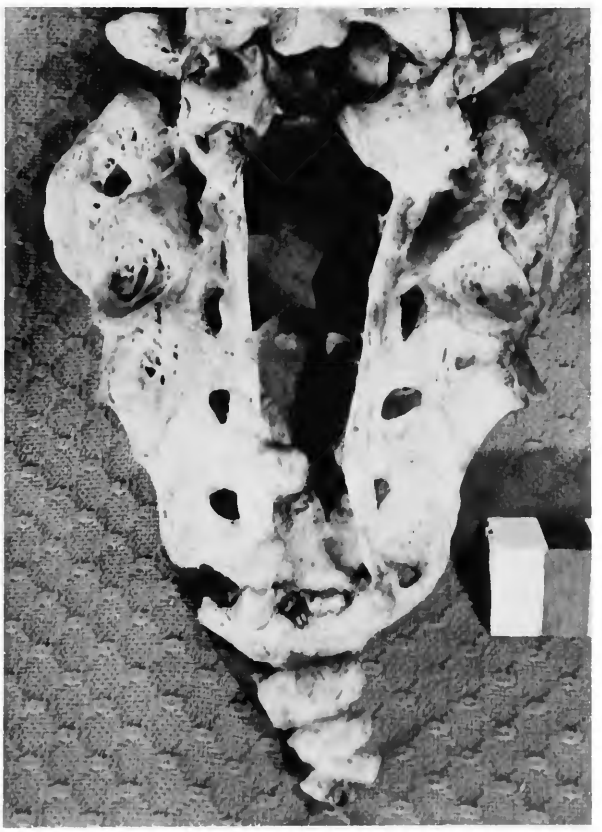
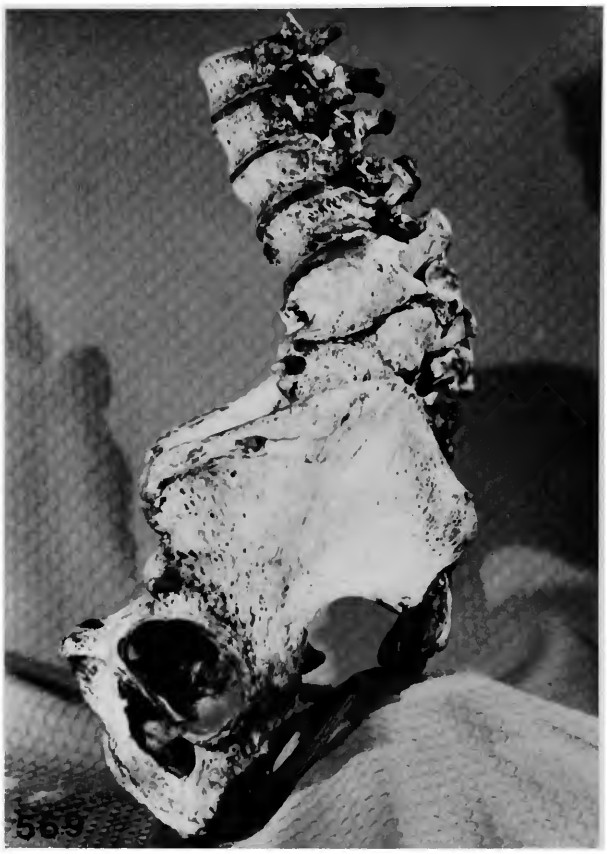


FIGURE 567.—Spina bifida of sacrum. (Young adult female, FPAM 2381.)



FIGURES 568, 569.—Supernumerary wedge vertebra between first and second lumbar with secondary scoliosis and right sacralization of fifth lumbar vertebra: 568, Anterior view. 569, Lateral view. (60-year-old male, FPAM autopsy 61729.)

PALEOPATHOLOGY

Spina bifida is a commonly mentioned abnormality in reports of archeological specimens. Indeed, references to this condition are so common that a review of all would be beyond the objectives of this book. Post (1966) has reviewed the existence of population differences in the frequency of spina bifida occulta in modern human groups. These differences suggest a genetic substrate in the expression of spina bifida. Ferembach (1963) reports an unusually high frequency of this abnormality in the sacra of a skeletal sample from a cave at Taforalt in northeastern Morocco. The site is dated to 10,500 to 12,070 years B.P. Ferembach calls attention to the problem of deciding what constitutes an abnormal condition in sacra since many sacra have some evidence of incomplete development of a neural arch. Until this problem is resolved, comparisons of the frequency of spina bifida as reported by different authors will be impossible.

I excavated an unambiguous example of sacral spina bifida from the Early Bronze Age site of Bab edh-Dhra in Jordan. The specimen is dated between 3150 and 3000 B.C. and is from the skeleton of a young male (Tomb A100E). In life the individual had probably suffered from tuberculosis, which resulted in destruction of the fourth lumbar vertebral body. This infectious condition is unrelated to the spina bifida occulta of the sacrum. The neural arch of the first sacral segment is divided at the midline of the spine (Figure 570). The arches of segment two through five never formed, leaving the canal exposed.

Other types of vertebral malformations are less

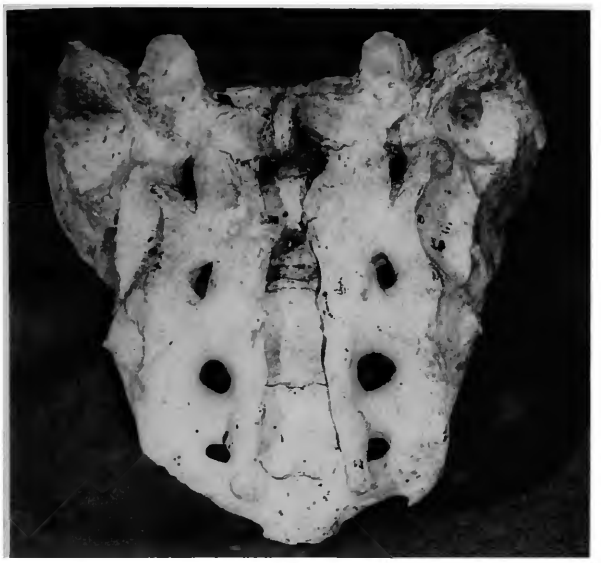


FIGURE 570.—Sacral spina bifida in a young adult skeleton from the Early Bronze Age site of Bab edh-Dhra, Jordan. (NMNH uncataloged, from tomb A100E.)

commonly reported than spina bifida. Barclay-Smith (1911) has provided a detailed report of multiple anomalies in the vertebral column of a young female skeleton from excavations at Sakkara in Egypt. The site probably dates to a period between 600 and 500 B.C. In this specimen there are eight cervical vertebrae. The first or atlas vertebra is fused to the base of the skull, the second or axis vertebra and the third cervical are fused together. The neural arch of C7 is divided through the spinous process and an extra cervical vertebra (C8) has an associated cervical rib on the right side. Damage to the left side precludes evaluation of the presence of a cervical rib on that side. The thoracic vertebrae are normal except for the diarthrodial joints between T11 and T12. The left joint has the morphology of a lumbar vertebra, while the right is a typical thoracic joint. The lumbar vertebrae exhibit a slight lateral curvature. The neural arches of L3 through L5 are divided at the lateral portion of the right lamina. L5 has an additional division of the spine creating a separate arch segment. The sacrum also has a separate neural arch segment including a portion of the left lamina of the first sacral vertebra (S1). Barclay-Smith (1911:170) suggests that the multiple anomalies were the result of early training and activity as a contortionist. A developmental malformation would appear to be more likely.

Jarcho (1965) has reviewed a vertebral abnormality, first described by MacCurdy (1923), which Jarcho thinks is an example of Klippel-Feil syndrome. The specimen is from Poricarcancha, Peru, and apparently is from an adult male. The sixth and seventh cervical and first thoracic vertebrae are fused. There are two hemivertebrae on the right side, the first between T3 and T4, the second between T4 and T5. These wedge vertebrae have resulted in a sharply angled scoliosis with T4 at the apex of the angle.

On the right side of the thoracic vertebrae there are 13 rib facets with the extra rib on the hemivertebra between the third and fourth vertebrae. On the left side there are the normal 12 facets. The first four ribs are fused near their proximal ends. The abnormalities of this case certainly suggest the congenital Klippel-Feil syndrome.

Another case of vertebral malformation is seen in a young adult female skeleton from the pre-Columbian site of Puye in New Mexico, USA (NMNH 262939). The skeleton is well preserved and normal except for the abnormal segmentation and fusion of the third through the fifth thoracic vertebrae. The spines, the diarthrodial joints, and the vertebral bodies of these vertebrae are fused (Figure 571). The left segment of T4 is fused to T3. Likewise, the left segment of T5 is fused to the right segment of T4. The right segment of T5 is also fused to T4, but the line of the early division between T5 and T4 is still apparent. Because there are no extra ossification centers and an abnormal segment is on each side, there is no scoliosis.

Another defect of the spine involves the separation of a major portion of the neural arch (spondylolysis) from one or more vertebrae. The defect separates the main part of the vertebra from the inferior facets and may permit the vertebral body to slide forward (spondylolisthesis).

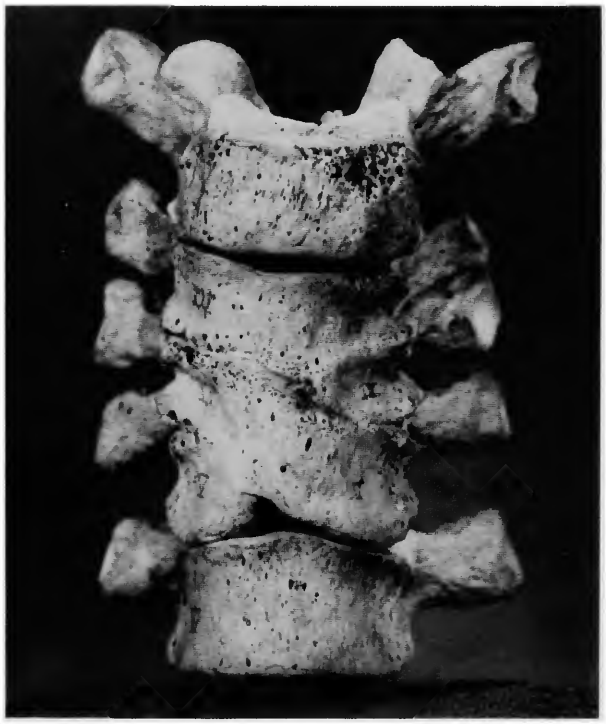


FIGURE 571.—Malformation of the third, fourth and fifth thoracic vertebrae from the prehistoric site of Puye, New Mexico, USA. (NMNH 262939.)

Stewart (1931) has reported on the frequency of this condition among Eskimo skeletal remains and found an unusually high frequency (27.4 percent) in this human group. Within the Eskimos he found that skeletons from the northern part of Alaska had a greater frequency than those from the southern part. Since the condition was thought to be congenital, Stewart attributed the differences to inbreeding of an isolated group for the northern skeletal sample or perhaps differences in origins for the two groups. Stewart (1956) later found an age-related association in which the incidence of spondylolysis increased with age and concluded that stress rather than genetics was the significant factor in the expression of separate neural arches. He did not rule out a genetic substrate but expressed the opinion that any genetic predisposition was related to erect posture rather than any specific genetic defect of the bone.

Lester and Shapiro (1968) in a study of another Eskimo skeletal sample also found a high incidence (40 percent) of spondylolysis and that the frequency of the defect increased with age. Curiously, in spite of the latter finding, the authors suggest that a hereditary weakness is an important factor in the expression of the abnormality. Spondylolysis is not limited to the New World although comparative data are not available for Old World groups. I have found two examples out of 92 burials in the Early Bronze I (3150 to 3000 B.C.) shaft tombs excavated at Bab edh-Dhra, Jordan, in 1977. Both examples are from adult males. One of these specimens is from the east chamber of tomb A100 (Figure 572). The age of the skeleton is in excess of 50 years. The arch defect is on the fifth lumbar vertebra. The arch is completely free of the vertebra, with the break occurring at the pars interarticularis. The broken edges of the bone exhibit considerable remodeling indicating that the break was long standing. The anterior surface of the vertebral body has considerable periosteal reactive bone, perhaps the result of periosteal activation due to anterior slippage of the vertebra. The inferior edge of the vertebral body has slight arthritic lipping as do the corresponding areas of the first sacral vertebra. The second and third thoracic

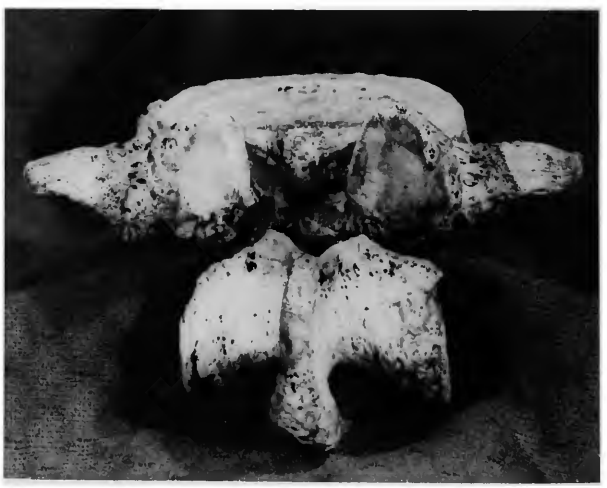


FIGURE 572.—Separate neural arch of the fifth lumbar vertebra in an adult male skeleton from the Early Bronze Age site of Bab edh-Dhra, Jordan. (NMNH uncataloged, from tomb A100E.)

vertebrae of this skeleton are fused. Fusion occurred at the spines, diarthrodial joints, and the lateral portions of the vertebral bodies. The disc space is preserved and the cause of the fusion is not apparent.

Ribs

Pathology

Rudimentary unilateral or bilateral supernumerary cervical ribs attached to the seventh cervical vertebra are not uncommon. A supernumerary thoracic rib is often attached to a lateral supernumerary hemivertebra. Segmental disarrangement may result in a forked rib with one cartilage. This abnormal rib is usually broader than normal.

Pelvis

Pathology

Failure of proper development of the anterior abdominal wall may leave the urinary bladder open (extrophy of bladder). In this situation, which is compatible with adult life, the symphysis pubis is absent and, although the pubic and ischial rami are properly fused, there is a gap of several centimeters between the two halves of the pelvis anteriorly (cleft pelvis) (Figure 573). Failure of development of the massa lateralis of the first sacral vertebra results in marked pelvic asymmetry (Figure 574).

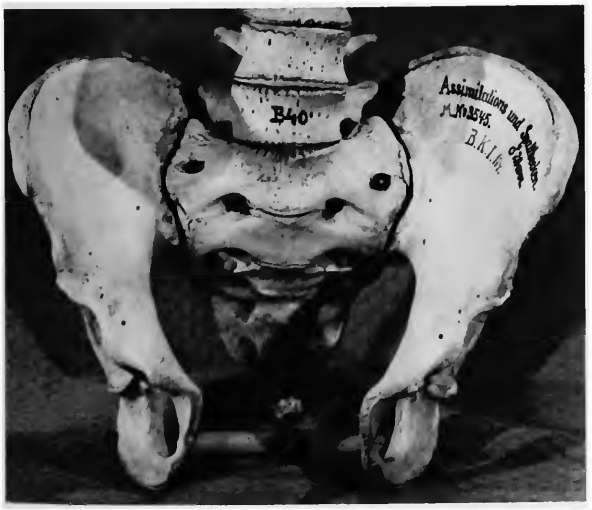


FIGURE 573.—Cleft pelvis of a patient with extrophy of the bladder and sacrum with 6 segments. (20-year-old male, FPAM 3545 from 1877.)

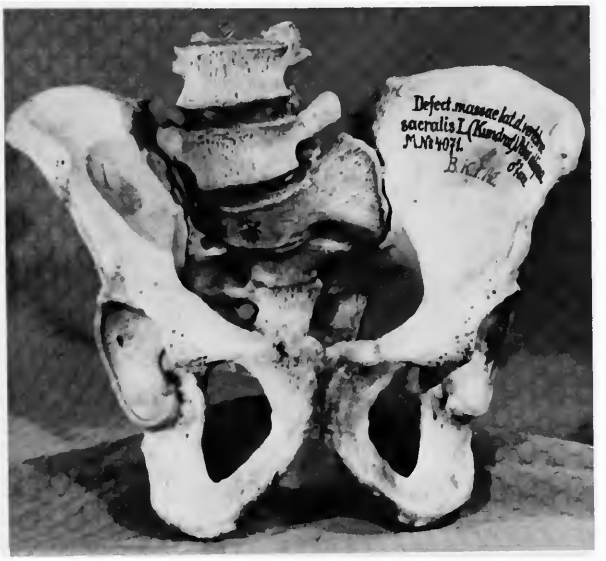


FIGURE 574.—Asymmetrical pelvis due to absence of right massa lateralis of first sacral vertebra. (About 30- to 40-year-old male, FPAM 4071.)

A hypoplastic shallow acetabulum on one or both sides leads to congenital dislocation of the femur upwards without disruption of the capsule or the round ligament. This is compatible with adult life and will result in formation of a bony reaction on the lateral cortex of the ilium, resembling a new acetabulum (Figure 575). Subsequent degenerative arthritis in this abnormal "joint" even will reveal eburnation of the bony surface. The femoral head is flattened and shows a vertical groove for the flattened round ligament. There is often a large marginal exostosis present, which points downward (Figure 576). The acetabulum is small, flat, and triangular, indicating that it never articulated with a mature femoral head.

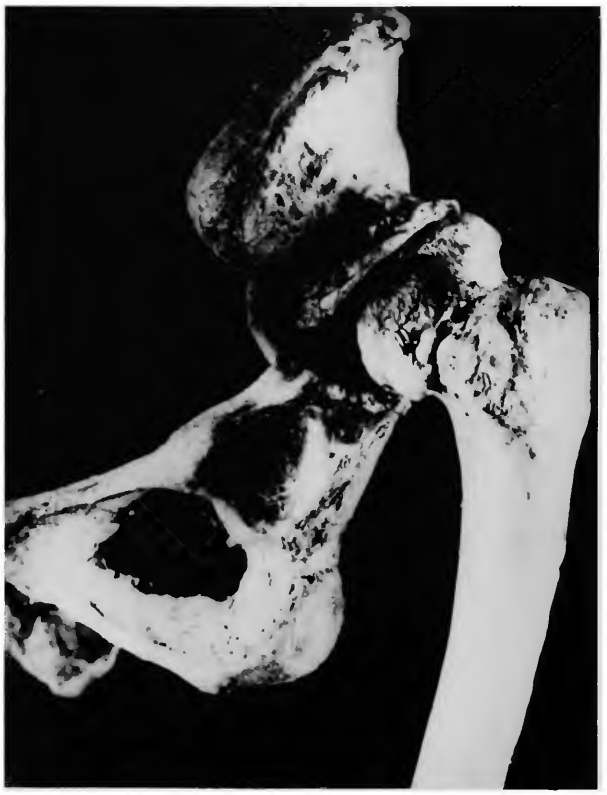


FIGURE 575.—Congenital dislocation of left hip showing rudimentary acetabulum and iliac neo-acetabulum with shelf-like roof. (Adult, VM 1694.)

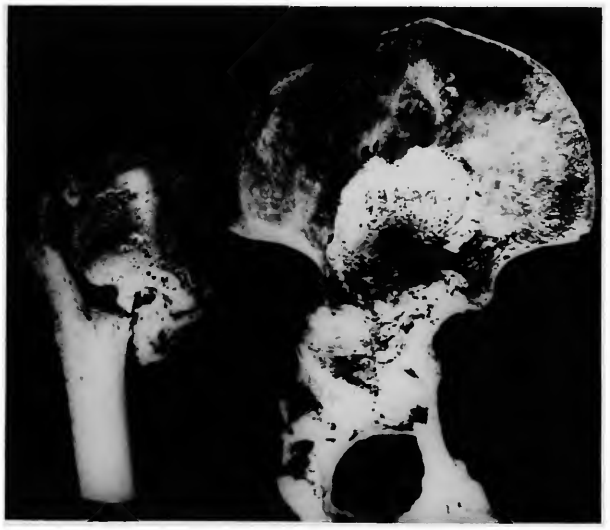


FIGURE 576.—Congenital dislocation of right hip with severe degenerative arthritis. Notice the rudimentary acetabulum, the neo-acetabulum on the iliac wing and the characteristic downward exostosis on the femoral head. (VM autopsy 224 from 1920, Roessle Collection.)

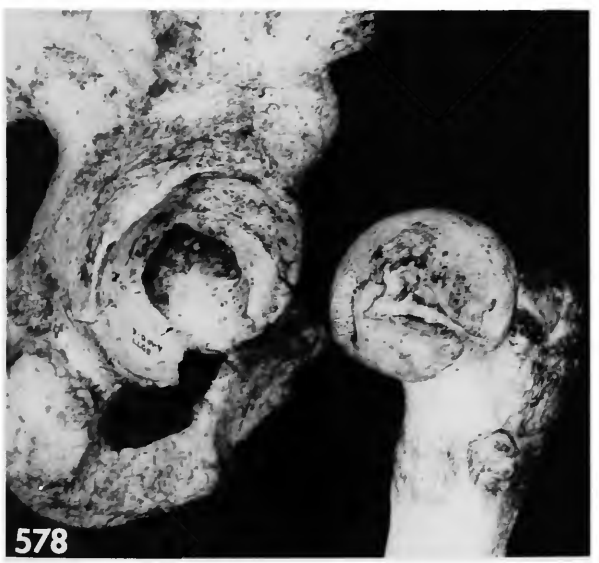
PALEOPATHOLOGY

In addition to the formation of a secondary acetabulum some abnormalities of the hip may result in only chronic slippage of the femoral head within the shallow actabulum, with no permanent dislocation and no formation of a new joint. It will not always be possible to distinguish between dislocation induced by trauma and congenital dislocation.

Baudouin (1906) found that the incidence of congenital dislocation in prehistoric European skeletal samples was rare. Moodie (1923) reported an example of hip dislocation in an ancient Peruvian skeleton in which a new joint was formed. Pales (1930, pl. 2: figs. 1, 2) published a photograph and a roentgen film of a pathological femur from the Neolithic period, which he attributed to congenital dislocation. However, the morphology is more compatible with slipped epiphysis. Brothwell (1967b) reviews several reports and cases of congenital hip dislocation. Morse (1969:33, 92, pl. 6B) includes a brief description of congenital hip dislocation in a specimen (Burial 10) from the Morse site in Illinois, USA. The abnormal bones include the left innominate and both femora. The acetabulum is abnormally shallow and distorted and the femoral heads irregular and small. A diagnosis of congenital hip dislocation would appear correct, although the reported age of 10 years appears too young.

An adult male skeleton from tomb A100E at the Early Bronze Age cemetery of Bab edh-Dhra in Jordan has an abnormally shallow acetabulum of the right hip. This is the same skeleton that had a separate neural arch. The acetabulum is also much larger in diameter than the corresponding acetabulum of the left innominate (Figure 577). There is a moderate amount of arthritic lipping on the margin of the abnormal acetabulum, but there is no secondary joint. The clearest evidence of dislocation is seen on the head of the right femur (Figure 578). The head itself is enlarged with two obvious defects on the inferior surface. The most anterior of these defects is a shallow curved depression about 1 centimeter wide by 2 centimeters long. The margins are

sharply defined and the base of the lesion is porous but well remodeled. The posterior defect is a narrow deep groove beginning with the pit for the ligamentum teres and continuing posteriorly through the boundary of the joint surface.



FIGURES 577, 578.—Chronic subluxation of the right hip, perhaps of congenital origin, in a skeleton from the Early Bronze Age site of Bab edh-Dhra, Jordan: 577, Comparative views of the right (left portion of photograph) and left acetabula showing the shallow and enlarged diameter of the right. 578, Bony components of the right hip; note the grooves from pressure erosion on the femoral head. (NMNH uncataloged, from tomb A100E.)

The curvature of the anterior defect corresponds to the curvature of the anterior margin of the acetabulum. There is little doubt that chronic and abnormal abrasion of the femoral head during repeated episodes of partial, anterosuperior subluxation produced this defect. The posterior defect is due to abnormal pressure on the joint surface by the ligamentum teres perhaps during dislocation. Another possibility is that part of an abnormally elongated ligamentum teres might have lain across the joint surface when the head was in the normal anatomical position. In view of the lack of a secondary joint and any evidence of fracture, congenital dislocation would appear to be the most appropriate diagnosis.

An example of hip dislocation with secondary joint formation is seen in an adult female from the ancient Pueblo site of Kwasteyerkiva in New Mexico, USA (NMNH 271828). The specimen consists of the right innominate and femur, although only the innominate was available for study. The most noticeable feature is the presence of a large secondary joint projecting well above the acetabular rim (Figure 579). The articular

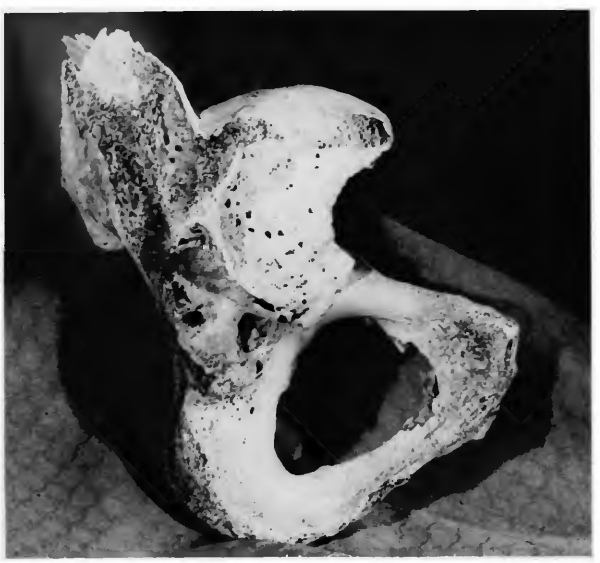


FIGURE 579.—Hip dislocation with secondary joint formation in the right innominate of an adult female from an ancient Pueblo site in New Mexico, USA. Note the bony remodeling in the original acetabulum and the porosity of the new joint. (NMNH 271828.)

surface is rough with many pores penetrating the surface. Considerable remodeling has taken place in the acetabulum. The original surface is very coarse and is almost completely covered over by a concave layer of bone, which may have been a temporary shallow joint before the femur slipped again and stimulated the formation of the final joint. Trauma cannot be ruled out, in part due to postmortem damage. The acetabulum appears to be abnormally shallow, which would support a diagnosis of congenital hip malformation leading to dislocation and secondary development of another joint.

Extremities

Pathology

Complete failure of development of one or several limb buds results in amelia; partial suppression results in peromelia. At times, only the proximal portions of the limb are rudimentary, while hands or feet are developed (phocomelia). As far as individual simple long bone defects are concerned, the radius and the tibia (Figure 580) are more often missing than the ulna or the fibula. Congenital bony ankylosis between the proximal radius and ulna (Figure 581) and between the distal tibia and fibula are typical anomalies, preventing, in the former, pronation of the forearm but not affecting the function of the ankle significantly in the latter. There are many, frequently hereditary and familial, syndromes concerning the number, length, and position of digits of fingers and toes (polydactyly, brachydactyly, clinodactyly). The number may be more or less than normal (Figure 582). Single rays may be fused through their entire length or only distally (various forms of syndactyly). Clubfoot deformity (pes equino-varus), often bilateral, can also occur

Figure 580.—Congenital absence of left tibia with hypertrophic fibula and equinovarus deformity of foot. (18-year-old female, WM S 16.2 from 1892.)

Figure 581.—Proximal congenital radioulnar synostosis (left) in pronation, radial head hypoplastic. (Adult, WM S 8.1.)

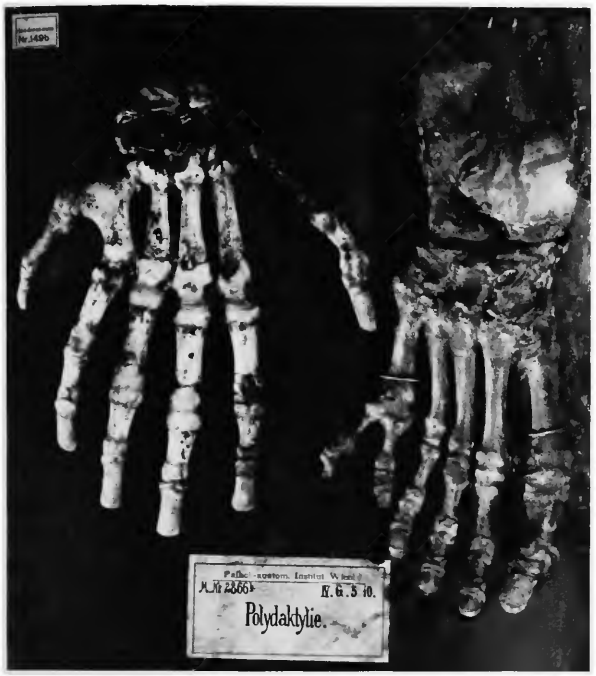
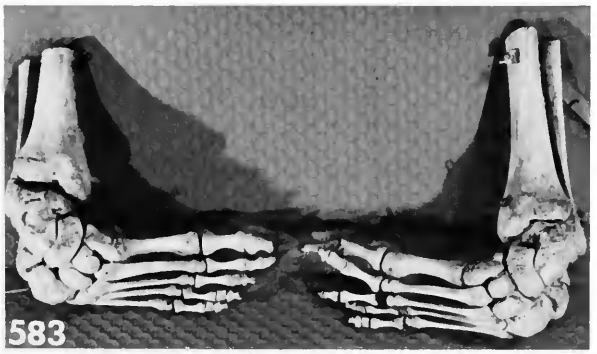
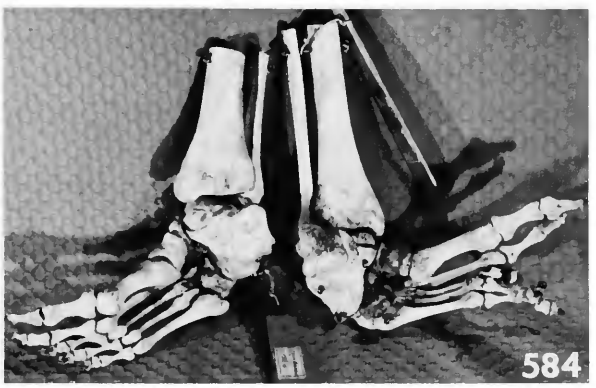


FIGURE 582.—Polydactyly of right hand and right foot. The supernumerary finger branches off from the fifth metacarpal and has only two phalanges; the supernumerary toe articulates with the fifth metatarsal and has three phalanges. (FPAM 2866b)





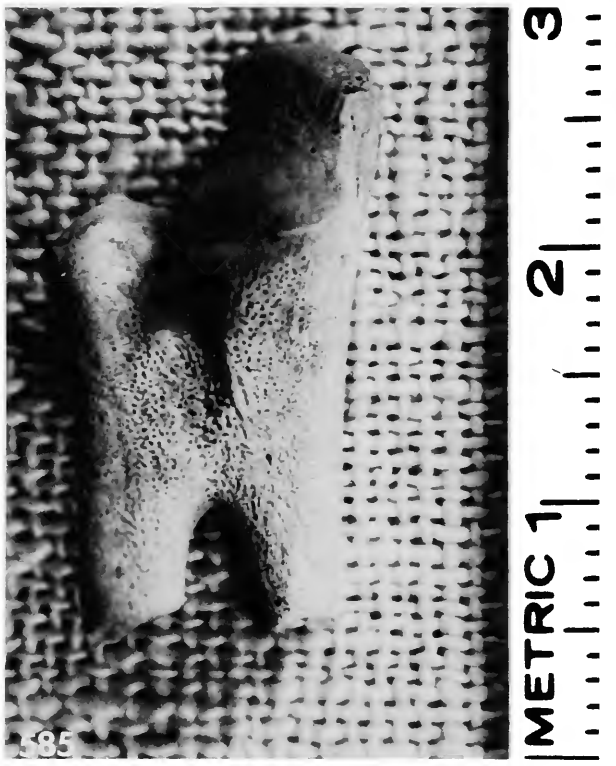
FIGURES 583, 584.—Congenital clubfoot deformity, bilateral (maximal pes equinovarus): 583, Dorsal view. 584, Plantar view. (About 16 years of age, FPAM 152.6 and 147.6.)

on a congenital basis with or without malformation of the spine (Figure 583, 584).

PALEOPATHOLOGY

Identification of congenital absence of one or more limbs in archeological specimens is difficult, due to the many postmortem conditions that affect the recovery of skeletons. A case has been reported by Hawkes and Wells (1976) in which such a diagnosis appears appropriate. The specimen was excavated from an archeological site in Hampshire, England, probably dated to the late sixth or early seventh century A.D. The skeleton (burial 38) is that of a man about 28 to 30 years of age. The preservation was good and the excavation was carefully done. The entire left extremity, scapula and clavicle were missing at the time of excavation. The authors conclude that this absence of the limb is a congenital condition. They point to the presence of other abnormalities and compensatory development of other bones in support of the conclusion that the abnormality was a long-standing antemortem condition.

Congenital fusion of the proximal radius and ulna has been noted in archeological specimens. Morse (1969:33, fig. 4) reports two cases of this abnormality from the Crable site in Illinois, USA. Both cases involve the left arm. Vyhnánek, Hanáková, Stloukal and Kolář (1965:2190) report the presence of congenital radioulnar synostosis in ancient Slavic skeletal material. The congenital nature of radioulnar synostosis is demonstrated in two specimens from archeological sites in North America. The first of these specimens (NMNH 384347) is the fused right radius and ulna from the skeleton of a near-term fetus or young infant (Figure 585). The specimen is from Ossuary II, Juhle site in Nanjemoy, Maryland, USA. No European trade goods were found, although the



FIGURES 585, 586.—Synostosis of the proximal right radius and ulna: 585, Near-term fetus or young infant from the late prehistoric Juhle site at Nanjemoy, Maryland, USA (NMNH 384347). 586, Adolescent skeleton from the historic period, Mobridge site in South Dakota, USA (NMNH 382993).

date is Late Woodland and thus could be post-Columbian. Even at the young age of this specimen, the radioulnar fusion is well established. The second example of radioulnar synostosis is from an adolescent skeleton from the Mobridge site in South Dakota, USA (NMNH 382993). This site contained trade goods and is thought to date around A.D. 1750. The right ulna and radius are fused in a slightly pronated position (Figure 586). The fusion has taken place in the radial tuberosity and the supinator fossa of the ulna. The radial head is not involved. There is no evidence of trauma and the young age of the specimen, as well as the nature of the fusion, make congenital radioulnar synostosis the appropriate diagnosis.

Deformities attributed to clubfoot have been described in the literature on paleopathology. Pales (1930:36–38) reviews some of the earlier reports on this condition. His discussion highlights the problems in distinguishing between congenital clubfoot and postparalytic deformities, such as those produced by poliomyelitis. Brothwell (1967b:424) has reviewed one reported case of clubfoot in a Twelfth Dynasty Egyptian mummy and finds the evidence inadequate to support this diagnosis. Brothwell (1967b:425–428) describes a fragmentary Neolithic skeleton from Gloustershire, England, with foot deformities attributable to clubfoot. Johnson and Kerley (1974) found evidence of clubfoot deformity in four skeletons from the pre-European site at Mokapu in Hawaii, USA. They also note the problem in distinguishing between congenital and postparalytic deformities. Johnson and Kerley make the useful conceptual distinction between malformation, which could be associated with congenital abnormalities, and deformation, which would be more likely in postparalytic changes.

Tumors

General Discussion

Pathology

Tumors and tumor-like cysts arising primarily in the skeleton are uncommon. They are the result of uncontrolled proliferation of any one of the tissue components of the osteogenic mesenchyme (bone, cartilage, fibrous tissue, or blood vessels). If the growth consists of well-differentiated (mature) tissue and remains localized, the tumor is called benign. If the tumor consists of poorly differentiated (immature) tissue, and continues to grow unchecked and can spread to other parts of the body through blood vessels and/or lymphatics, the neoplasm is designated as malignant. In contrast to those in other parts of the body, primary benign and malignant tumors of the skeleton mostly arise in young, actively growing individuals.

Malignant tumors arising in various organs and tissues of the body (carcinomas and sarcomas) may spread, mainly through the bloodstream, to bones, resulting in metastatic tumors. In contrast to the primary tumors of the skeleton, metastatic tumors are usually multiple and predilect the older age group. In present populations, metastatic tumors in bone are much more common than primary ones.

The identification of specific tumor types in dry bone is not always possible; however, location and age as well as alteration of the surrounding bone offer helpful clues. Obviously, tumors producing benign or malignant neoplastic bone offer the best chance of identification.

The following discussions concentrate on those lesions and features that offer some chance of recognition in archeological material. (For a review of all the aspects of skeletal neoplasms consult Jaffe, 1958.)

Paleopathology

With respect to the paleopathology of human tumors one needs to keep in mind the fundamental difference between modern medical experience and conditions associated with ancient populations. Most important are differences in average age at death. People in populations on which much of modern medical experience with tumors is based, live about twice as long as people in populations studied by paleopathologists. Since primary malignant tumors usually occur or arise

Figure 587.—Unicameral bone cyst of proximal left humerus; bisected wet preparation. Notice the thin expanded new cortex and the contact of the cyst with the growth plate. (7-year-old male, WM S 61a 44 from 1905.)

during the growth period, their frequency in ancient skeletons might be expected to be similar to modern skeletal samples. However, secondary tumors of bone are associated with the older age categories and are thus less likely to be found in archeological skeletons. Individuals who lived in antiquity usually died of other causes before secondary tumors could become a serious factor in morbidity and mortality.

Another factor is the well known environmental effect on the type and incidence of cancer. Carcinoma of the lung, for example, is associated with smoking and air pollution. Metastases to the skeleton from this type of cancer may be minimal where such environmental problems did not exist. It is important to emphasize, however, that exposure to other conditions, such as smoke from a wood fire, may be carcinogenic. Wells (1964b: 1611) has noted the relatively high incidence of nasopharyngeal tumors of bone in paleopathological specimens. Today, this condition is rare in the Western world but rather common in Africa. The condition is also rather common among the Chinese.

Primary Benign Tumors

Pathology

Cysts

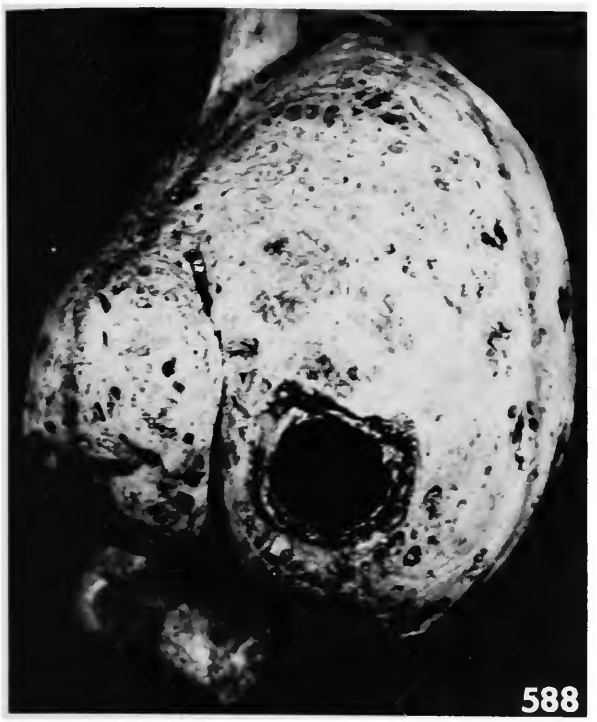
A cyst, in the anatomical sense, is a lesion characterized by a fluid-filled cavity surrounded by a distinct wall. Several types of lesions fulfilling these criteria occur in bone in different characteristic locations.

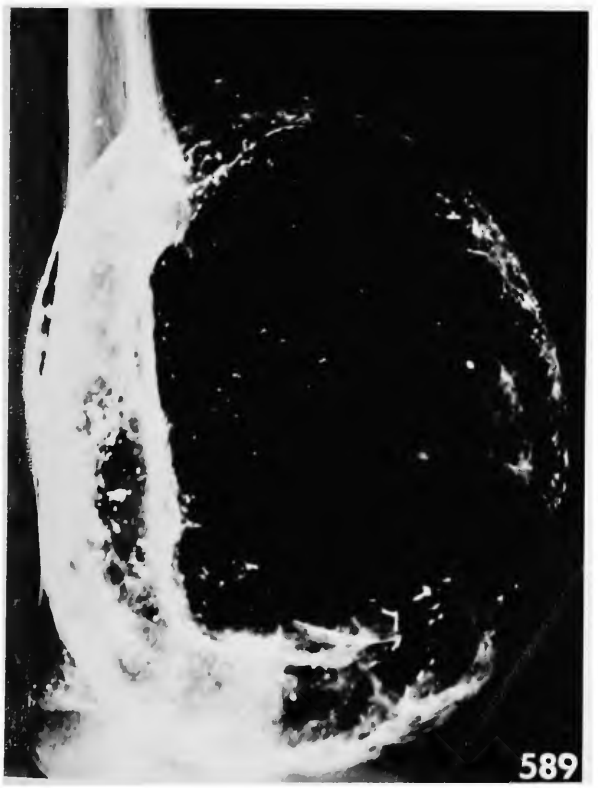
Unicameral Bone Cyst.—This tumor-like lesion occurs about as frequently as giant cell tumor. It consists, in the fully developed state, of a round or oval fluid-filled single cavity of several centimeters in diameter that is lined by a thin membrane of poorly vascularized osteogenic mesenchyme. The lesion occurs most often in young children or adolescents but may be carried into adult age. It is most common in long bones and starts in the metaphysis in close proximity to the growth plate (Figure 587). Its location is central in regard to the axis of the involved bone. It always spares the epiphysis. In the course of longitudinal growth of the individual, it usually seems to move away, towards the diaphysis, by interposition of newly grown bone between it and the growth plate. The most frequent locations are proximal humerus, proximal and distal femur, proximal and distal tibia, and proximal fibula. All other locations are unusual, especially in small tubular and in flat bones. In Jaffe's (1958:63) material of 75 cases, the male–female sex ratio was 2:1.

The enlargement of the cyst leads to smooth, tapering resorption of the overlying cortex, due to increasing fluid pressure. Ultimately the cortex may be replaced with a newly formed shell of cortical bone, which may show reinforcing ridges on the inner surface. Usually the actively enlarging cyst shows osteoclastic resorption on the inside and osteoblastic bone deposition on the outside of the shell. Pathological fractures, crushing the thin wall, are common and often heal readily.

Aneurysmal Bone Cyst.—This is a much less common lesion of older children and adolescents. It is always eccentric in location as regards the axis of the involved bone. It shows a distinct bulge, consisting of a thin newly formed shell over the eroded cortex, which accounts for its name (Figures 588, 589). The contralateral cortex of the involved bone and the adjacent cancellous bone are usually spared. The lesion occurs with equal frequency in long bones and in the spine. In long bones, the location is usually the metaphysis, seldom the diaphysis. The epiphysis is spared. In the spine, vertebral bodies, transverse processes and neural arches may be involved. If the lesion is large, pressure defects on adjacent vertebrae can occur. In 35 cases studied by Jaffe (1958:56), the lesion was more common in females.

The aneurysmal bone cyst is usually multilocular, but the different compartments are separated only by soft tissue septa, which are devoid of bone. The wall consists of highly vascular connective tissue with numerous giant cells. The





multilocular character of the lesion is reflected in the bosselated outline, especially of the wall facing the axis of the bone.

Traumatic Epidermal Inclusion Cyst.—Crushing trauma to the terminal phalanx, more often of fingers than toes, may displace some squamous epithelium of the nail bed into the fractured bone. This displaced epithelium continues to grow, forming a cystic space in the bone which fills with keratin material. In the dry bone this would present as a central cavity, usually not more than one centimeter in diameter, with or without evidence of a healed fracture. It would be difficult or impossible to distinguish the lesion from the much more common enchondroma.

Congenital Epidermal Inclusion Cyst.—This lesion is fairly rare and occurs only in the bones of the calvarium. In the course of the closure of the cranial cavity during embryonic development, ectodermal skin epithelium may become trapped in the developing bone of the cranial vault. The cyst resulting from the proliferation of such epithelium may be located in any part of the vault but occurs most frequently in the parietal bone. Usually the lesion is single. The cyst is lined by squamous epithelium and filled with keratin. Mostly the cyst is small, confined to the diploë, appearing as a round or anteroposteriorly elongated radiolucent area on X-ray. Occasionally such cysts can become rather large, 10 centimeters or more in diameter. In these cases the overlying tables bulge, the inner one more than the outer. Occasionally the cyst lining may show various skin appendages (hair follicles, sebaceous glands and sweat glands) representing all skin elements (dermoid cyst). These occur particularly in the vicinity of the orbit (Figure 590). The small round inclusion cysts may not be distinguishable from the more common eosinophilic granulomas of the skull in children.

Figures 588, 589.—Aneurysmal bone cyst of left distal femoral metaphysis: 588, Showing a big cystic mass with thin reticulated bony shell. 589, X-ray of specimen, showing the slight mineralization of the shell and some sclerosis at the deep border. (12-year-old male, PMES 1 TF 14(2) from before 1842.)

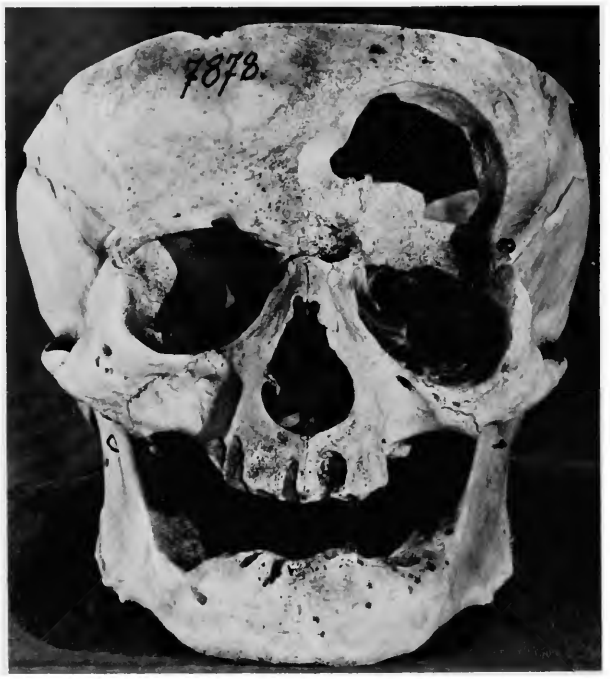


FIGURE 590.—Cranial defect from dermoid cyst. Notice the smooth destruction of the left frontal bone and supraorbital ridge with inward sloping, slightly sclerotic edges. (61-year-old male, DPUS 7878 from 1905.)

Osteoblastic Tumors

OSTEOMA.—This condition is a benign borderline-neoplastic lesion consisting of mostly dense lamellar bone with vascular channels but practically without marrow spaces. In several typical locations and appearances, it occurs almost exclusively in the skull. The mineral density of the lesion permits excellent preservation in archeological material.

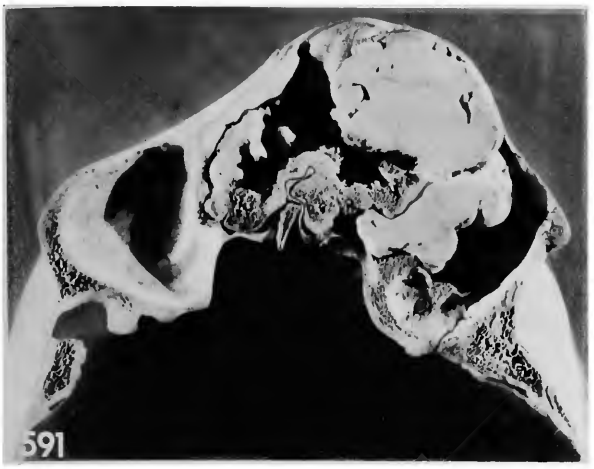
The most common lesion is the so-called button osteoma of the cranial vault, usually located on the outer table and consisting of an ivory-hard smooth lump of not more than 2 centimeters in maximal diameter. Sometimes it shows a peripheral circular constriction at the junction with the outer table. The lesion is found in at least one percent of all autopsies. Usually the lesion is single, but multiple lesions occur. The most frequent locations are the frontal and parietal bones. Similar lesions on the internal table of the cranial vault are much rarer.

The second most common osteoma is a horseshoe-shaped or circular overgrowth of cortical bone on the inner aspect of the bony auditory canal. The maximal thickness usually does not exceed 5 millimeters but narrows the canal considerably. Both of these lesions are well documented in various races and in archeological material.

The third type of osteoma is a more formidable lesion, most commonly consisting of a tumorous bony proliferation projecting into the frontal or other paranasal sinuses (Figures 591, 592). This lesion begins with a fibro-osseous proliferation made up of fibrous tissue and woven bone trabeculae. As the lesion matures it becomes more solidly bony with lamellar bone predominating. These tumors may occasionally reach large sizes, disfiguring the facial bony contours, projecting into the orbit, and/or the nasal cavity. Occasionally a sizeable nodular projection into the anterior cranial fossa with compression of the frontal lobe of the brain occurs. This lesion is much less common than the two other cranial osteomas described above. Osteomas should be easily identifiable even on archeological specimens.

OSTEOID OSTEOMA.—This type of osteoma is a not infrequent, small, tumor-like lesion consisting of poorly mineralized woven bone developing in the cortex or in the spongiosa of a bone. The lesion rarely exceeds one centimeter in diameter. It occurs most commonly in older children and young adults. The location is most often on a long bone of an extremity. The localization in 150 cases of Jaffe's (1958:94) series was 25 percent in the femur, 25 percent in the tibia, 35 percent in the fibula, humerus, vertebrae (transverse process and arch), and tarsal bones (talus and calcaneus) combined. Most other bones may be affected occasionally, but the cranium seems to be largely spared.

The lesion produces a small radiolucent focus, with or without central calcification, which, if the lesion involves the cortex, elicits an osteosclerotic response of the surrounding bone, out of propor-



FIGURES 591, 592.—Osteoma of frontal sinus: 591, Cut surface view from below. Notice the ivory-like tumor masses bulging and penetrating the sinus wall externally and endocranially. 592, Frontal view, showing multiple tumor perforations through outer table. (Adult, HM P809 from before 1799.)

tion to the size of the lesion. This reactive buildup of dense lamellar bone may appear as an external or internal cortical thickening of several centimeters axial length, reaching its maximum over the small lesion (Figure 593). If located in the spongiosa, the reactive perifocal bone formation is usually much less marked. The density of the reactive bone should preserve the appearance of the lesion in dry bone, although the osteoid portion would disintegrate. The differentiation from a small intracortical abscess may not be possible.

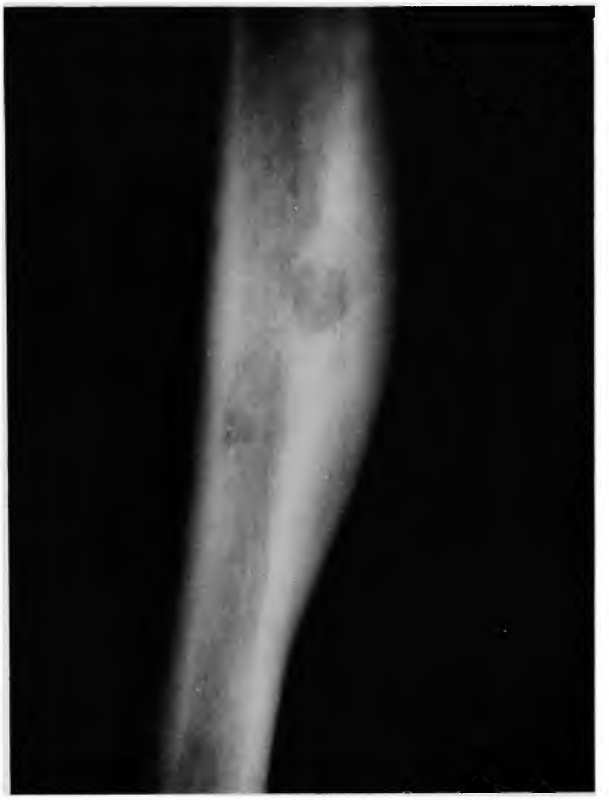


FIGURE 593.—X-ray of osteoid osteoma of femur. Notice the lytic corticomedullary lesion with a central nidus of increased density and pronounced cortical hyperostosis. (Adult, AFIP 116267.)

OSTEOBLASTOMA.—This is a rare benign tumor of bone consisting of various components of osteogenic mesenchyme producing poorly mineralized woven bone trabeculae. To a great extent, the histologic pattern resembles that of osteoid osteoma. However, this tumor may reach considerable size. The lesion appears most often in the spine (vertebral bodies and/or arches) and small tubular bones of hands and feet (Jaffe, 1958:108). The tumor occurs mostly in adolescents and young adults with no predilection to sex.

In long bones, the tumor is usually in the metaphysis and is seen as a moderately radiolucent lesion with occasional perifocal osteosclerosis. Identification in dry bone cannot be made with assurance.

Chondroblastic Tumors

CHONDROMA.—This is a benign tumor consisting of hyaline cartilage. It is one of the more common bone tumors, usually arising within the bone (enchondroma), much less often in the periosteum (juxtacortical chondroma). The lesion is most frequently found from later childhood through middle age. There is no sex predilection. It occurs usually in the metaphysial area of a tubular bone of the extremities with special predilection of the small tubular bones of hands and feet. The localization in Jaffe's (1958:169) material, arranged in order of decreasing frequency, was as follows: finger phalanges, metacarpals, humerus, femur, toe phalanges, metatarsals, tibia, fibula, and ulna. The tumor does not occur in bones developing through intramembranous ossification. At least in part, enchondromas develop from residual portions of growth cartilage that have become detached from the growth plate. This used to be common in the healing of rickets. Scherer (1928) found metaphysial cartilage rests in 1.7 percent of over 1000 right femora of individuals over 25 years of age examined at autopsy. This explains that chondromas are located in the proximity of the area of the growth plate: in either metaphysis of long bones, in the distal metaphysis of metacarpals and metatarsals, and in the proximal metaphysis of phalanges. The tumor partly destroys the cancellous bone and, if large, scallops the inner cortex of long bones. The outer contour of large long bones may not be altered. In the small bones of hands and feet the outer contour may be distorted and expanded, revealing a thin, new cortical shell, which may be perforated. The joint surfaces are always spared. The center of the tumor is usually, and often heavily, calcified and sometimes ossified.

The juxtacortical chondroma is rare. It usually is located over the metadiaphysial area of a large long bone of an extremity. In its development it causes a cup-shaped depression of the underlying cortex with an elevated cortical lip surrounding the defect. However, it usually remains separated from the interior of the bone by a distinct sclerotic border. The central portion of the tumor may calcify and/or ossify.

CHONDROMATOSIS (Ollier's Disease).—This is a rare condition in which multiple and often numerous chondromas manifest in early childhood in various parts of the enchondrally ossified skeleton. Again, the most frequent manifestations concern the bones of the extremities, including hands and feet. In the small bones these cartilage tumors cause ballooning deformities bordered by a thin cortical shell (Figure 594). The cranial base and facial skeleton, although enchondrally ossified, is usually spared (Jaffe, 1958:185). There is a high incidence of development of a malignant cartilage tumor (chondrosarcoma) in later life. Ollier's disease produces changes which are, in several ways, different from those of solitary enchondroma. Speiser (1925) has shown that, in addition to the large metaphysial enchondromas

FIGURE 594.—Multiple enchondromas of right fifth metacarpal and basal phalanx. Notice the ballooning projections on both bones covered by a thin shell of new cortex. (Adolescent, PMES ITD 120(1).)

derived from the growth plate, numerous smaller chondromas are formed by faulty differentiation of the inner layer of the periosteum. In the process of growth some of these assume an elongated, streak-like, radiolucent appearance. The presence of large cartilage masses during the growing period results in alteration of the normal process consisting of diminished growth, bowing deformities due to uneven growth, and widening of the metaphyses of involved bones due to delayed and incomplete remodeling.

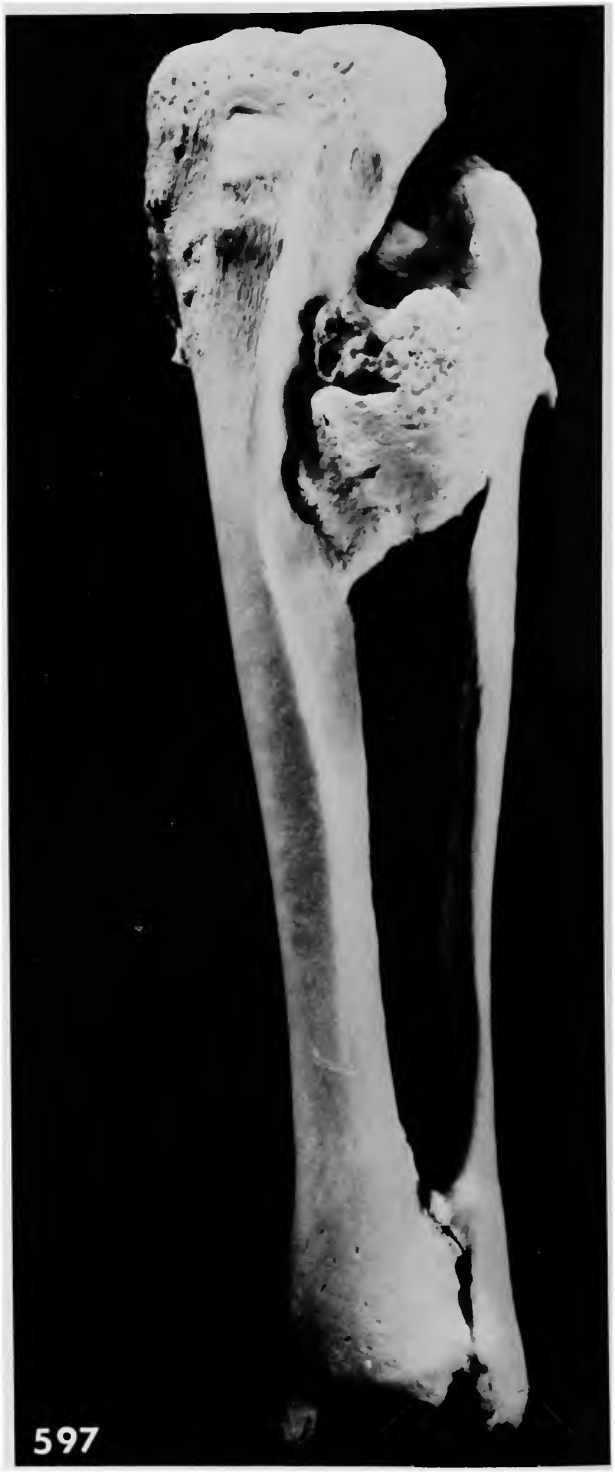
Cartilaginous Exostosis (Osteochondroma).—Cartilaginous exostosis, appearing as a solitary lesion, is one of the most common benign bone tumors. Its initiation is limited to the growing period of the skeleton beginning most often in childhood. There is no sex predilection. The lesion may occur on any bone that develops by enchondral ossification, but the cranial base and the facial bones are rarely involved. The tumor most commonly occurs in proximity to the growth plate on the metaphysial surface of long bones. The distal metaphysis of the femur and the proximal metaphysis of the tibia are the site of the majority of the lesions. Other long bones of the extremities, including small tubular bones of hands and feet, ribs, pelvis, and scapula are less commonly involved. The spine and sternum are usually spared.

The lesion begins close to the growth cartilage with a faulty differentiation of the inner layer of the periosteum into cartilage. This cartilage undergoes enchondral ossification from the underlying osteogenic mesenchyme, mimicking to a great extent the enchondral growth of the growth plate. Since, in the involved area, the periosteum produces cartilage instead of bone, there is never a cortex separating the medullary spaces of the exostosis from those of the affected bone. Thus, the exostosis enlarges continuously due to the growth of the basal layer of the cartilage cap. Usually the growth of the exostosis stops when the nearby growth plate terminates its growth.

The lesion begins as a rounded outgrowth on the periosteal surface. The final shape is greatly modified by mechanical stresses (muscle pull and tendon insertions) in the affected area. Thus, the common exostoses around the knee area become elongated polypoid structures with bulbous tips pointed away from the joint; upward on the femur and downward on the tibia. This is an effect of secondary remodeling. Fractures of slender, elongated exostosis in this area are not uncommon. The flaring remodeling of the metaphysis is often inhibited on the side of the bone bearing the osteochondroma. On the proximal humerus, osteochondromas tend to remain broad-based and rather bulky without pedunculation. Osteochondromas of the pelvis appear as cauliflower-like osteochondrous masses projecting outward or toward the pelvic canal (Figure 595).

In dry bone and on X-ray, the tumor shows easily recognized diagnostic characteristics. If transsected, the continuity of the cancellous marrow spaces of the lesion and of the involved bone are readily apparent as is the reflection of the cortex onto the lesion. The inner structure consists of more or less regular cancellous bony trabeculae

Figure 595.—Osteochondroma of left ilium. (59-year-old female, IPAZ autopsy 1776 from 1962.)



FIGURES 596, 597.—Multiple cartilaginous exostoses: 596, Right femur; notice lack of metaphysial remodeling in area of exostoses. 597, Left tibia and fibula with multiple exostoses. (54-year-old male, IPAZ S.1201 from 1965.)

mingled with areas of densely calcified cartilage or sclerotic bone.

HEREDITARY MULTIPLE EXOSTOSES (Diaphysial Aclasia).—This dominant hereditary condition, which affects multiple bones of the skeleton, is rare. It usually manifests itself in several siblings and several generations of one family. There is a marked predominance in the male sex. Basically, the individual lesion is identical with that of solitary cartilaginous exostosis. The predominant location is again the knee area, often involved symmetrically (Figures 596, 597). In addition, the proximal humeral metaphysis, the distal tibial metaphysis, and the small tubular bones of the hands are frequently involved. Other locations in decreasing order of frequency are: proximal fibula, iliac bone, metatarsals and toes, radius, ulna, ribs, and scapula. The spine is rarely affected; neither are the facial bones. The skull base occasionally shows a lesion on the endocranial aspect. The number of lesions may vary from a few to several hundred in different cases.

If numerous lesions are present in a metaphysial area, remodeling of the metaphysis is lacking, leaving a broad metaphysis without flare (Figure 596). Growth is retarded and, at times, irregular, leading to shortening and axial deviation of the affected bone. If the distal ulnar metaphysis is severely involved, the ulna is shortened and the radius, due to its intimate attachment to the ulna, is abnormally curved, giving the appearance of a Madelung's deformity of the forearm. In a moderate number of cases in the older age group, a chondrosarcoma develops from the residual cartilage cap in one or several of the lesions (Figure 598).

CHONDROBLASTOMA.—This fairly rare benign tumor is made up of primitive chondroblasts and giant cells. It occurs mainly in adolescents and young adults, with a marked preponderance of males over females. The tumor occurs most commonly in the epiphyses of long bones (Figure 599). In fact, this is the only bone tumor that

FIGURE 598.—Secondary chondrosarcoma on cartilaginous exostosis of proximal right femur (center); the distal right femur (left), and the proximal right fibula and tibia (right) show smaller benign exostoses. (43-year-old male, GHPM 4041 from 1841.)

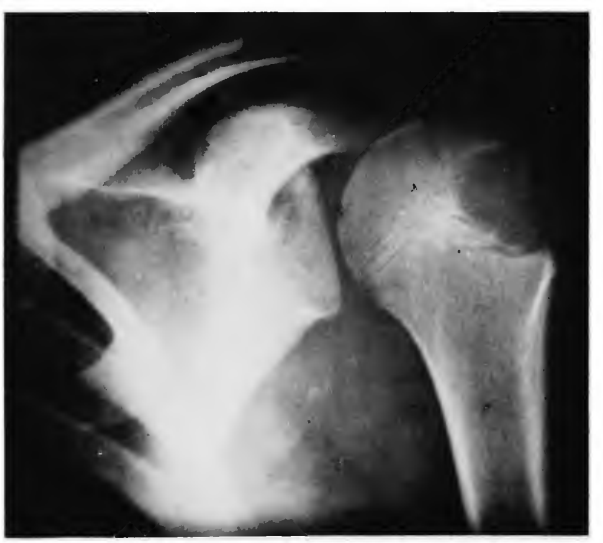


FIGURE 599.—X-ray of chondroblastoma of proximal left humeral epiphysis. Notice the eccentric lucency with a thin bony shell and slight sclerotic reaction. (Adolescent, courtesy Dr. S. Werthammer, Huntington, West Virginia, USA, 1952.)

always arises and often is confined to an epiphysis or apophysis. This is helpful in identification in dry bone. The location in 30 cases of Jaffe's (1958: 44) material, in decreasing order of frequency, was in the following epiphyses: distal femur, proximal tibia, proximal humerus, distal tibia, proximal femur, calcaneus, talus, ilium, and ischium.

The tumor usually produces a purely lytic lesion of several centimeters in diameter. After closure of the adjacent growth plate the lesion may extend into the adjacent metaphysis. There is often some degree of calcification in the center, which, however, probably would be lost in dry bone. Occasionally, bony sclerosis is found adjacent to the lesion. In the humerus and femur the separate apophyses for the greater tubercle and for the greater trochanter are favored locations (Jaffe, 1958:45).

Chondromyxoid Fibroma.—This rare benign bone tumor occurs in adolescents and young adults. The lesion is most commonly found in the metaphysial area of long and short tubular bones, mainly of the lower extremity. The most common bone involved is the tibia. The lesion is located eccentrically but starts within the cancellous bone. It consists of neoplastic proliferation of fibroblastic cells producing large amounts of mucoid matrix. There are also giant cells scattered through the lesion. The radiologic picture is similar to that of nonossifying fibroma, but the lesion tends to be more rounded and there is often marked bony sclerosis in the wider vicinity of the lesion. In small tubular bones, after closure of the growth plate, the lesion may extend into the epiphysis.

Fibroblastic Tumors

Fibrous Cortical Defect.—This is a very common lesion of the skeleton, present in about one-third of all children with a male–female ratio of 2:1 (Jaffe, 1958:78). It seldom appears before 2 years of age. The location is always on the metaphysial cortex of the long bones. The lesion begins in the periosteum, close to the growth plate but may be moved away from it in the process of subsequent growth. It is always longer than wide and may be multiloculated. The adjacent portion of the cortex may be sclerotic, and there is always a layer of dense bone separating it from the medullary space (Figure 600). This is the area where, in the normal process of growth, remodeling resorption of the metaphysial cortex occurs. The lesions are often multiple and symmetrical. The most commonly involved area is the distal metaphysis of the femur. Other locations, in order of decreasing frequency, are: proximal tibia, proximal and distal fibula, proximal femur, distal tibia, humerus, radius, and ulna (Jaffe, 1958:80).

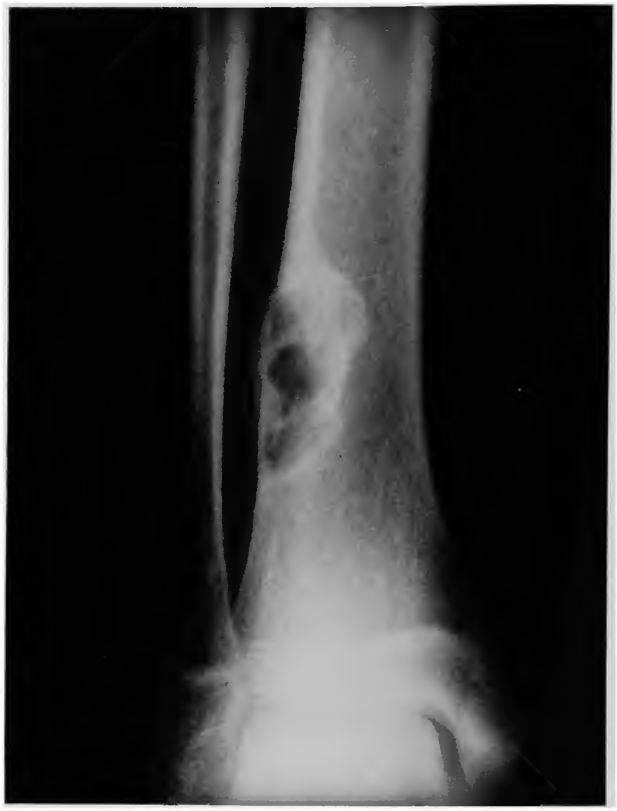


Figure 600.—X-ray of fibrous cortical defect of distal right tibia. Notice the eccentric lobulated lesion with slightly bulging cortex and pronounced deep sclerotic margin. (Adolescent, MGH surgical specimen 14645 from 1969.)

The lesion consists of spindle-shaped mesenchymal cells and giant cells, probably representing a faulty development of the remodeling inner layer of the metaphysial periosteum. On the dry

bone the lesion would present as a streak-like or multiloculated defect beneath the cortex, bordered by smooth surfaced sclerotic bone in the depth. These lesions may reach several centimeters in length but usually not more than one centimeter in diameter. They may fill in spontaneously or remain stationary. In few instances they behave agressively and become truly neoplastic (nonossifying fibroma).

In Caffey's (1955) series of 1000 healthy children examined radiologically, the most common location of the defect was found on the distal femur and with equal frequency as a unilateral or bilateral symmetrical lesion. The solitary unilateral lesions were, in both sexes, twice as often located in the right than in the left femur. Of 254 femoral defects, 224 were present on the medial cortex of the metaphysis and only 30 on the lateral cortex. The anterior cortex was involved only once. Multiple defects within a single femur were found only 19 times. No defects were demonstrated in the first two years of life, but in the third, almost 8 percent of the children showed defects. In the fourth year, the number of affected children rose sharply to 15 percent, maintaining a frequency of 14 to 17 percent through the twelfth year. In the thirteenth year, the frequency of cortical defects dropped sharply to 10 percent and reached 8 percent in the fourteenth year. The defects in the distal femoral metaphysis were 25 times more frequently encountered than those in the proximal tibial metaphysis and 60 times more common than those in the proximal fibular metaphysis. Cortical defects in the proximal femoral metaphysis were rare.

Nonossifying Fibroma.—As already indicated (p. 374), this neoplastic lesion derives from a fibrous cortical defect that behaves aggressively and continues to grow. The location is the same as that of the latter. The age of the individuals affected is older (later childhood and adolescence). The occurrence is much less common than the fibrous cortical defect and is usually unifocal. The lesion always starts eccentrically in the metaphysis of long bones and, even after penetration into the medullary portion of the bone, is separated by a distinct, frequently scalloped, bony shell from the medullary tissue. Only in a thin long bone, like the fibula, may the tumor ultimately transect the entire diameter, but even then the contralateral cortex is usually less involved. However, since large portions of cancellous bone and cortex are progressively destroyed, pathological, usually diagonal, fracture occurs commonly, especially in weight-bearing bones. The tumors may reach up to 10 centimeters in length.

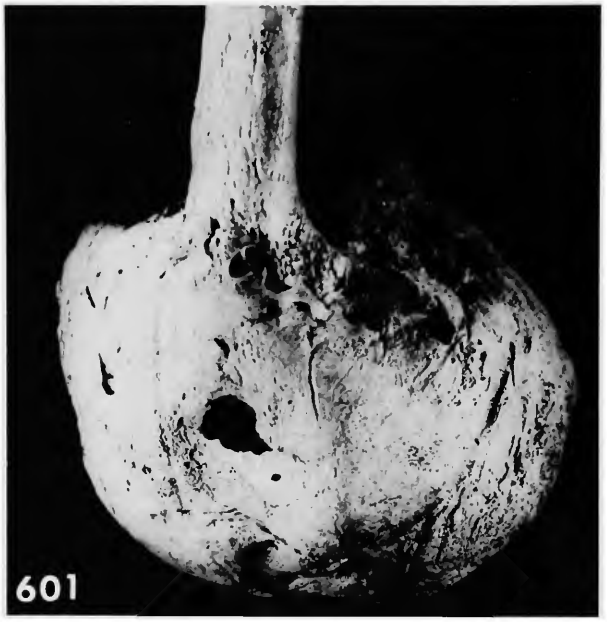
Cortical Desmoid.—A lesion, similar to fibrous cortical defect, is occasionally seen on long and short tubular bones in which a similarly scalloped cortical defect is filled with tough mature collagen. There is also a separating cortical shell at the deep margin of the lesion. The individuals affected are often adult. On the dry bone, the defect would appear very similar to that of a fibrous cortical defect.

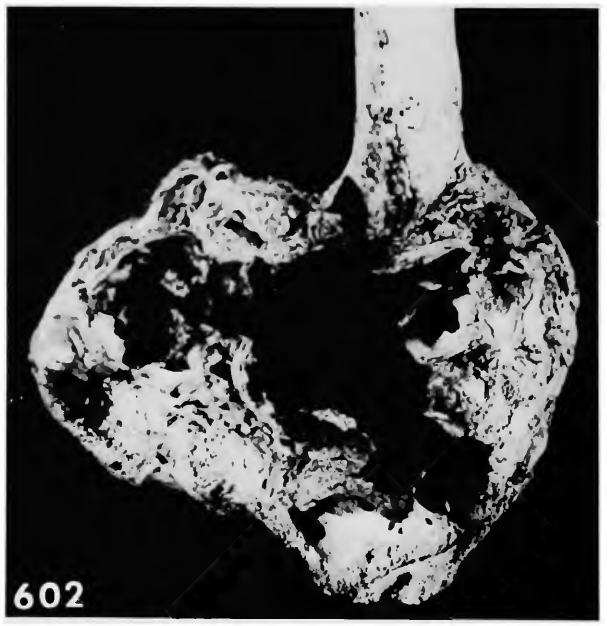
Giant Cell Tumor

One of the most common primary bone tumors is the giant cell tumor (osteoclastoma), a borderline-benign neoplasm made up of stromal cells and multinucleated giant cells similar to osteoclasts. The lesion most commonly occurs in adolescents and young adults, mostly less than 40 years of age. The tumor is eccentric, epimetaphysial in location, and most often found in long bones. The predilected areas are distal femur, proximal tibia, distal radius, and proximal humerus. All other locations are rare, but it can occur in vertebrae, pelvis, cranial vault, or small bones of the extremities.

The most characteristic lesion, and the only one that could be identified with a reasonable degree of certainty in dry bone, is the epimetaphysial lesion of long bones. The tumor usually completely destroys the preexisting spongiosa and the original cortex, allowing the formation of a thin, new, periosteal cortical shell in an expanded position, which often has small rounded perforations and reinforcing ridges on the inner surface (Figures 601, 602). This accounts for the radiolog-

ical "soap bubble" appearance of the cortical shell. The lesion often excavates the epiphysis and closely approaches the articular surface, without penetrating into the joint. The zone of transition into the intact cortex on the diaphysial end of the lesion is usually sharp and narrow. Pathological fractures through advanced lesions, especially in weight-bearing bones, are not uncommon. The normal consistency and appearance of the rest of the bone and of other bones of the same skeleton should permit differentiation from reactive giant cell tumors (brown tumors) in severe hyperparathyroidism.





FIGURES 601, 602.—Giant cell tumor of distal femur: 601, Outside view, showing expanded cortical shell with small perforations. 602, Cut surface, showing lytic center and ridged cortical shell. (HM P837.)

Hemangioma

True hemangioma is a neoplasm formed by proliferating blood vessels. This type of lesion is very rare in any part of the skeleton. However, lesions showing marked dilatation of preexisting bone marrow sinusoids that are not true neoplasms are commonly found in the vertebrae (Putschar, 1929). These vascular lesions are often combined with proliferation of fatty marrow (angiolipoma).

This vertebral lesion is rather common in older individuals. Schmorl and Junghanns (1971:325–327) report an incidence of 12.2 percent in males and 15.9 percent in females over 60 years of age in their series of close to 4000 complete spinal columns studied at autopsy. Of these, two-thirds were solitary and one-third multiple lesions affecting different vertebrae. In their large series, close to 70 percent of the lesions were found in the thoracic vertebrae, especially the three lowest ones, 20 percent in the lumbar vertebrae, and the rest were evenly divided between lower cervical and upper sacral vertebrae.

This lesion does not alter the contour of the vertebral body. The hemopoietic marrow is replaced by dilated blood vessels with a varying admixture of fat cells. Characteristically, the vertical trabeculae are reduced in number and increased in diameter, giving the picture of coarse, vertical striation on X-ray (Figure 603). Small lesions may not show any reduction or alteration of trabeculae.

There are rare instances of true vascular neoplasms of the spine. They mostly occur in the younger age group and destroy the bone, ballooning the outer configuration of the vertebra and

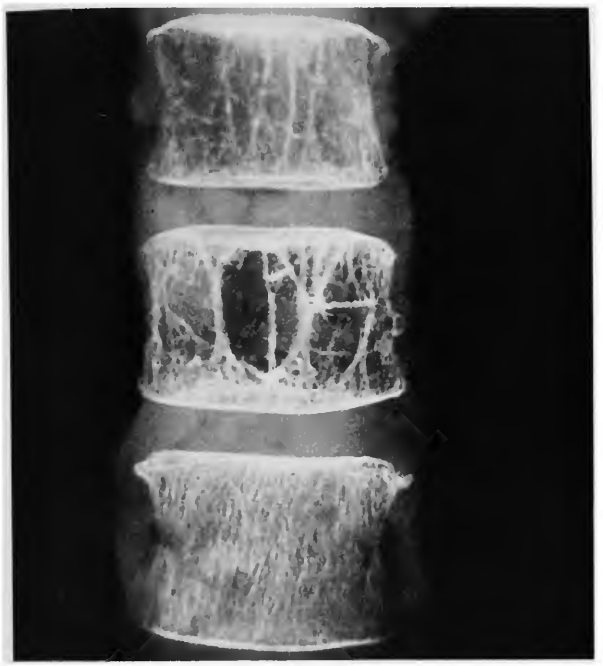


FIGURE 603.—X-ray of angiolipoma of two vertebrae. Notice reduced number of thickened trabeculae in top and center vertebra; bottom vertebra normal. (Adult, studied by Putschar.)

encroaching upon the lumen of the spinal canal. Pathological fractures occur in these tumors but are absent in the common older age vascular ectasias.

True hemangiomas occur in the cranial vault

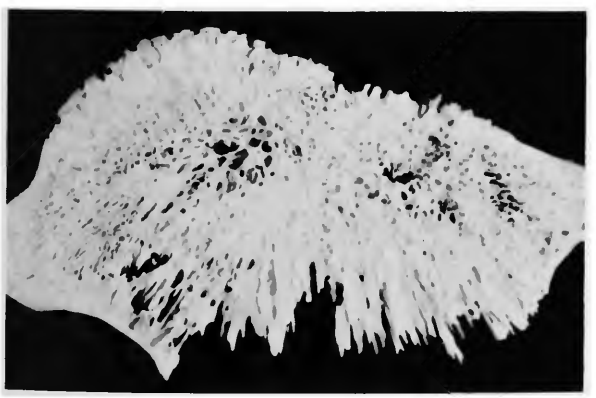
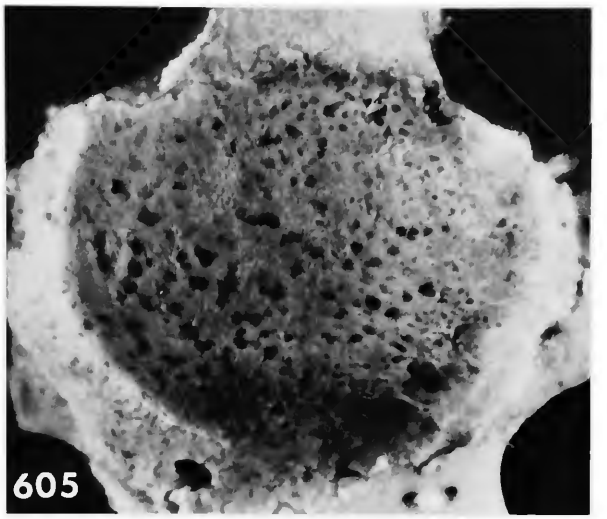
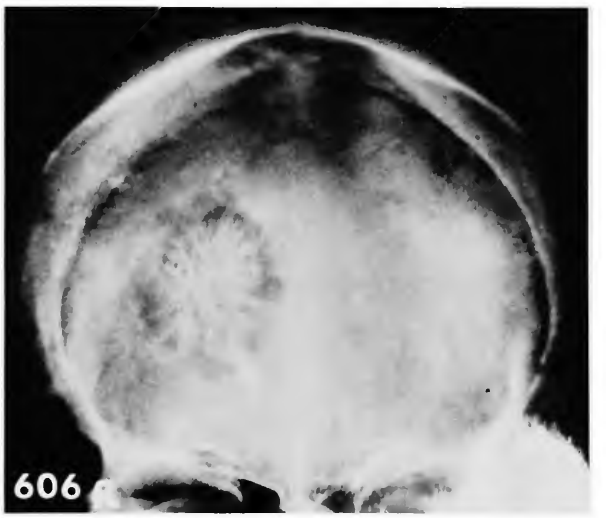


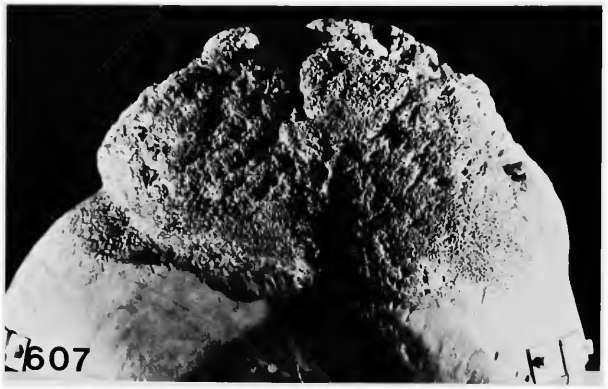
FIGURE 604.—Hemangioma of cranial vault. Transverse cut showing radiant bone trabeculae between distended marrow spaces, protruding bluntly outward and sharply inward. (35-year-old female, IPAZ surgical specimen 16132 from 1966.)

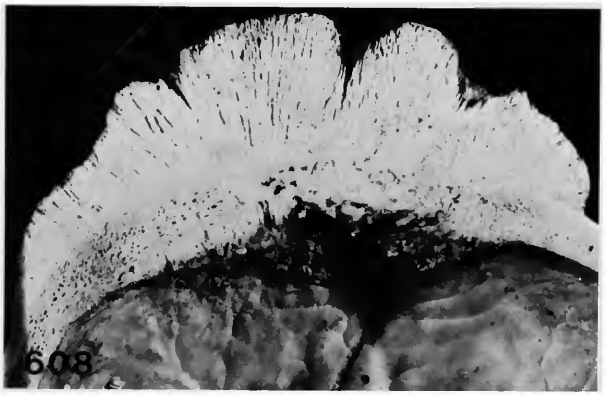




FIGURES 605, 606.—Hemangioma of frontal bone: 605, Wet specimen, endocranial view, showing spongy bone with large vascular channels. 606, X-ray, showing the typical wheel-spoke pattern of cranial hemangioma. (47-year-old female, 2 years duration, PMSG 9/1.741a, surgical specimen 2369 from 1955.)

showing a characteristic radial arrangement of coarse diploic trabeculae around large vascular channels, giving a "sunburst" effect on X-ray (Figures 604–606). These lesions are usually round, several centimeters in diameter, and destroy the inner and outer table. The tumor expands mostly outward and shows a circular lytic margin in the periphery. Hemangiomas in long





FIGURES 607, 608.—Meningioma of cranial vault of 6 years duration: 607, External view; notice the massive bone formation outward. 608, Cut surface; notice the radiant arrangement of the new bone, sclerosis of diploë, destruction of both tables and only slight endocranial build-up. (20-year-old Sinhalese male, WM N9.2 from 1884.)

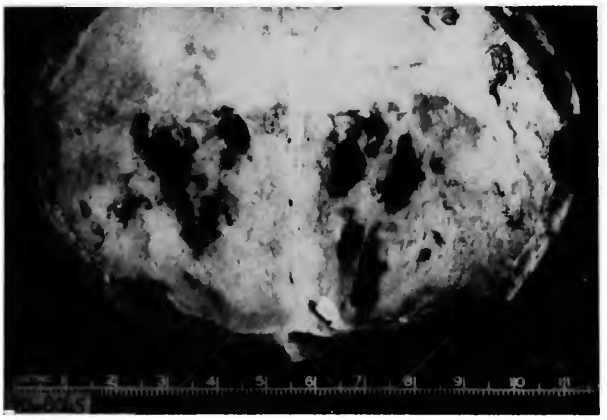


FIGURE 609.—Unusually deep niches for pacchionian granulations of frontal bone. Notice the bilateral deep (dark) defects of the inner table. (63-year-old female, MGH surgical specimen 8045 from 1970.)

bones are extremely rare. They produce loculated lytic and sometimes sclerotic lesions, which are not characteristic enough for identification in dry bone.

Meningioma

Meningioma is not primarily a bone tumor. It arises from the mesothelial lining cells of the dura mater of the brain and spinal cord. Not uncommonly the cranial meningiomas invade the cranium through the internal table often eliciting a massive response of reactive bone in form of radiant spicules, which may project outward from the destroyed outer table (Figures 607, 608). This finding may be indistinguishable from osteosarcoma in dry bone, but a destructive defect of the inner table without internal spiculation favors the diagnosis of meningioma. Meningioma invading the cranial base tends to be more osteosclerotic with less tendency of spicule formation. Unusually large arachnoid granulation may occasionally create deep, smooth-walled defects in the cranial vault. However, they are usually lined by a thin internal table (Figure 609).

PALEOPATHOLOGY

Osteoblastic Tumors

Of the several primary benign tumors of bone known in modern medical practice few have been identified in archeological specimens. This may be due partly to the lack of knowledge by paleopathologists of the gross morphological features associated with these tumors but also may be due to the problems in identifying a tumor in a dry bone specimen. Osteomas are fairly common in archeological specimens and have been described by several authors. Small button osteomas occur primarily on the outer table of the skull vault. Typically this osteoma is no larger than one centimeter in diameter. It is solitary, dense, with little relief but with a slight degree of undercutting at the boundary with normal bone (Figure 610). Large osteomas also occur as seen on a skull from near Lima, Peru (NMNH 242462). This tumor measures 2.0 centimeters in diameter and

FIGURE 610.—Button osteoma of the frontal bone in an adult female skull from Pachacamac, Peru. (NMNH 267377.)

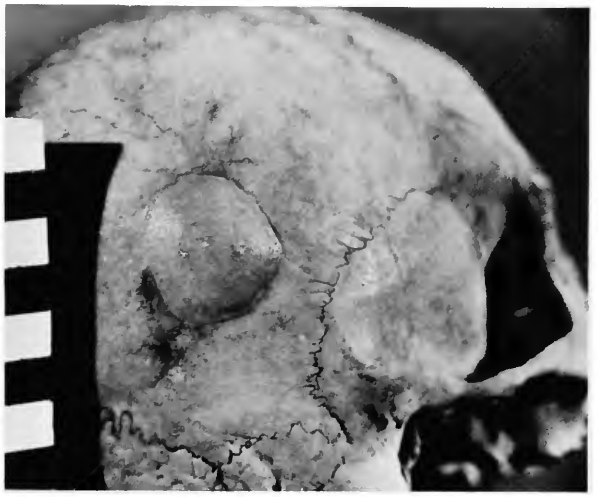


FIGURE 611.—Large osteoma of the right parietal in an adult skull from a site near Lima, Peru. (NMNH 242462.)

almost 4 millimeters above the plane of normal bone at the center of the lesion (Figure 611).

Another common type of tumor seen in archeological specimens is the small bony growth, which may partially to completely fill the external auditory meatus. Hrdlička (1935) was unable to reach any firm conclusion regarding the cause of these ear exostoses. He did not feel that they were the result of infectious or malignant diseases and suggested chemical or mechanical irritation as a possibility. Hrdlička (1935:80) observed that males are far more subject to the disease. His report of racial variation suggests a genetic component but environmental or cultural conditions cannot be ruled out. Hrdlička (1935:80) did find that the incidence increased with the age of the individual. Other scholars, including Adis-Castro and Neumann (1948) and Gregg and McGrew (1970), have described cases in archeological populations. Stewart (1979b:268) has called attention to lytic lesions of the bony external auditory canal, which he suggests may be the bony response to a soft-tissue tumor (ear cholesteatoma). The circumscribed enlargement of the canal does

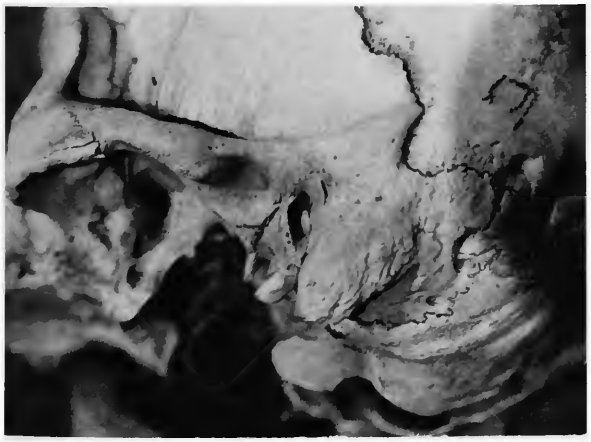


FIGURE 612.—Bony tumor of the left external auditory meatus of an adolescent skull from Chicama, Peru. (NMNH 264344.)

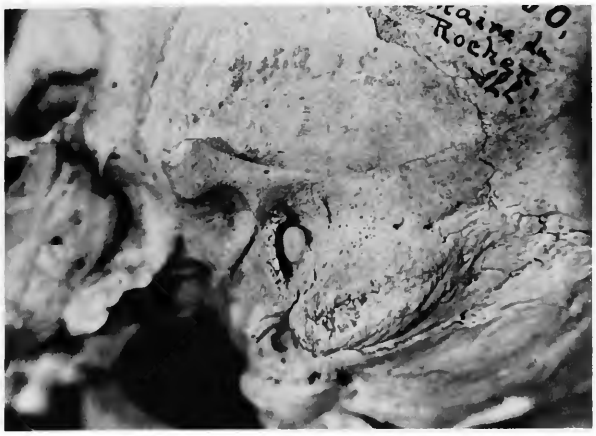


FIGURE 613.—Large bony tumor of the left external auditory meatus of an adult male from an archeological site in Illinois, USA. (NMNH 243180.)

not exceed the size of an acorn. The condition appears to be fairly common among the Eskimos and Aleuts of Alaska, USA.

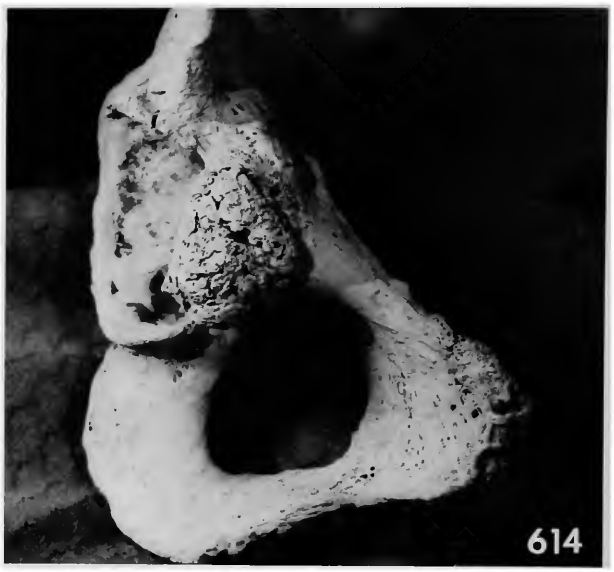
There are numerous examples of ear exostoses in the collections of the National Museum of Natural History, USA. Only two will be described here. The first of these is an adolescent skull from Chicama, Peru (NMNH 264344) illustrating what appears to be the incipient stage of this condition (Figure 612). All secondary teeth have erupted. The basioccipital synchondrosis is not fused indicating an age of around 18 years at the time of death. Sex is thought to be female. Both auditory canals exhibit narrowing; the right auditory meatus has a slight, hypertrophic ridge that may represent the early stage of tumor development.

The second skull is from Illinois, USA (NMNH 243180). It is from a fully adult male. The mandible is missing and the maxilla has a slight development of bony tumors in the buccal region of both sides. Like the previous specimen the auditory canals are abnormally narrow. In addition there are large bony tumors in both external meatus with the most pronounced development on the left side (Figure 613). Both tumors arise from the posterior portion of the canal.

Beraud, Morel and Boyer (1961) describe an osteoma of the ethmofrontal region in a skull from France. The specimen is from the medieval cemetery of St. Hermentaire and is dated to the 11th century A.D. The authors estimate the age at death to be 22–25. The tumor encroaches on the endocranium.

Chondroblastic Tumors

CARTILAGINOUS EXOSTOSIS.—A probable example of cartilaginous exostoses is seen in a right innominate in a Twelfth Dynasty Rock Tomb at Lisht in Upper Egypt (NMNH 256474). There are no other bones associated with this specimen. The morphology of the innominate indicates that it is from an adult female skeleton. The tumor occurs at the junction between the fused pubic and iliac bones (Figures 614, 615). The lesion measures 25 by 33 millimeters and extends 18 millimeters above the plane of the normal adjacent surface. The surface of the tumor is irregular and has a rounded, lumpy appearance. The joint surface of the acetabulum is normal; however,



FIGURES 614, 615.—Cartilaginous exostosis in the area of fusion between the pubic and iliac bones of a right innominate bone: 614, Anterolateral view, showing the irregular surface of the tumor. 615, Superior view, showing projection of the tumor from the bone surface. (Adult female from the Rock Tombs, Lisht, Egypt, Twelfth Dynasty, NMNH 256474.)

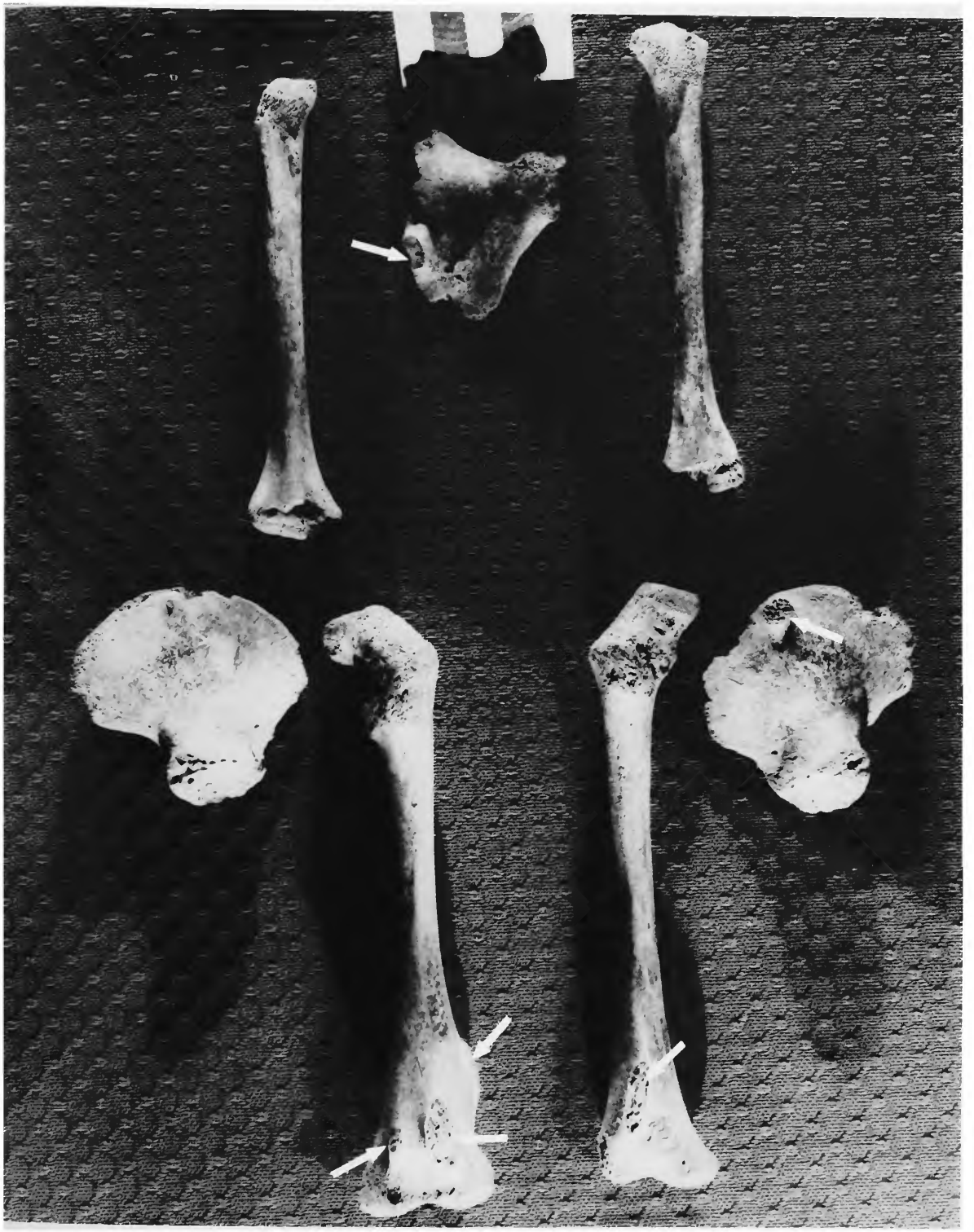


FIGURE 616.—Hereditary multiple exostoses in a child's skeleton from a Saxon cemetery, Winchester, England. Arrows indicate the location of some of the exostoses; scale in cm. (BMNH G932, CG69, TR XL 1085.)

the tumor bone has extended the anterosuperior margin of the acetabulum. The X-ray film of the tumor reveals that the medullary space of the exostosis is continuous with the innominate.

HEREDITARY MULTIPLE EXOSTOSES (Diaphysial Aclasia).—Singer (1962) describes a case of diaphysial aclasia in a young adult female skeleton from a cave in South Africa. The archeological age is reported to be post-European. There are multiple exostoses on all the major long bones. Gladykowska-Rzeczycka and Urbanowicz (1970) report a case of this condition in a skeleton of an adolescent from a cemetery in Pruszcy Gdanski, Poland. The site is dated to between the first century B.C. and the fourth century A.D. There are multiple bony projections primarily of the metaphysial areas of the major long bones. Sjøvold, Swedborg, and Diener (1974) describe a most interesting burial from a cemetery on the island of Gotland off the southeastern coast of the Swedish mainland. The burial is dated to about A.D. 1250 and includes the skeleton of a young female between 17 and 20 years of age and a near-term fetus. Both skeletons exhibit multiple exostoses. The authors suggest that exostoses protruding from the pelvis prevented the birth of the infant and led to the death of both the mother and fetus.

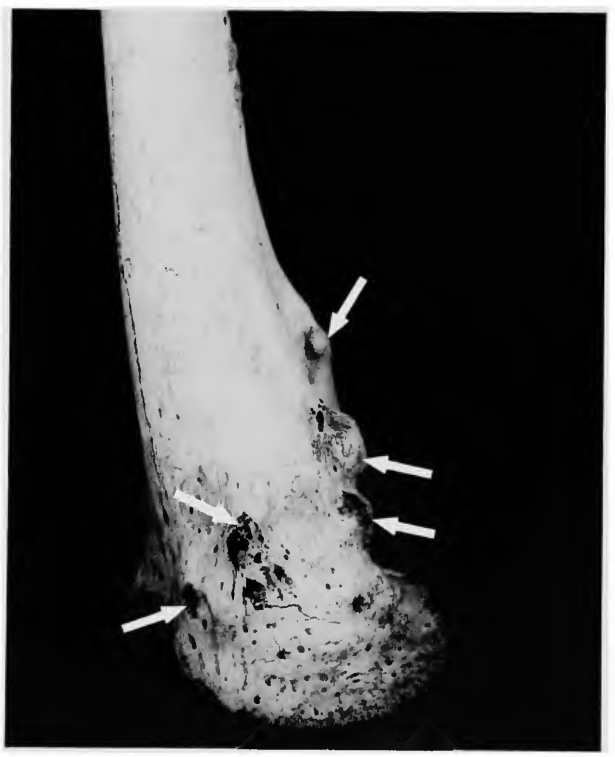


FIGURE 617.—A probable case of hereditary multiple exostoses in an isolated right femur of an adult from an archeological site in Chicama, Peru. Medial view of the distal femur, arrows indicate the location of some of the exostoses. (NMNH, uncataloged.)

Another archeological example of multiple cartilaginous exostoses is located in the Winchester Saxon skeletal-pathology collections of the British Museum (Natural History) in London. This specimen (BMNH G932, CG69, TR XL 1085) includes most of the major postcranial bones. The age estimate based on femur length is around 4 years, sex is unknown. Prominent bony projections can be seen on the distal metaphyses of the left humerus and of both femora (Figure 616). The left scapula and both iliac bones are affected as well. Other bones, including the ribs, vertebrae, and tarsals, are normal. Since this tumor is usually benign, it is unlikely to have been the cause of death.

A New World example of multiple cartilaginous exostoses is seen in a right femur of an adult from Chicama, Peru (NMNH, uncataloged). There are no other bones associated with the specimen and the archeological age is unknown. There is a large bony exostosis, which developed between the greater trochanter and the superior surface of the femoral neck. The remaining lesions are in the distal metaphysial region (Figure 617) and consist of multiple bony projections.

Meningioma

Abbot and Courville (1939) attribute a parosteal osteoblastic lesion seen in two skulls located in the Museum of Man, San Diego, California, USA, to periosteal reactions to underlying tumors of the dura (meningiomas). One skull was from Peru and had a large lesion of the skull vault consisting of fused spicules of bone, which radiate out perpendicular to the skull surface. The abnormal bone is attached to the underlying outer table, which is largely intact. The second skull is

FIGURES 618, 619.—Multiple lytic lesions of the skull possibly due to meningioma. Specimen an adult male from an archeological site on St. Lawrence Island, Alaska, USA: 618, Posterolateral view, showing large lesion on the left parietal. 619, Detailed view of large, parietal lesion; note the partial bony fill-in of the exposed diploë in the larger lesion. (NMNH 280091.)

from San Nicolas Island off the coast of southern California. The lesion is similar to the Peruvian specimen. Meningiomas are known to produce lytic lesions in early stages of the tumor development (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952:1628), but blastic lesions are common (Cushing, 1922:150).

Rogers (1949) briefly describes two skulls from Egypt having lesions, which he attributes to meningiomas. The first of these is from the First Dynasty and shows a hyperostotic lesion of the right parietal involving both the inner and outer table. The second specimen is from the Twentieth Dynasty and exhibits a large honeycomb lesion with a focus in the right parietal but involving the left parietal and the frontal bone. The lesion is large including most of the right parietal. It is not clear whether the inner table was involved. The first of these two cases differs from the two cases of Abbot and Courville (1939) in involving both tables of the skull.

MacCurdy (1923:264, pl. 39) describes an adult male skull from Paucarcancha, Peru, with a large tumor of the left parietal and frontal bones (probably the same Peruvian skull described later by Abbot and Courville (1939)). The coalescing osteophytes extend outward from the skull about 4.5 centimeters. The tables of the skull have been destroyed beneath the tumor. MacCurdy attributes this tumor to osteosarcoma, but a meningioma would seem to be more likely.

An Eskimo skull from St. Lawrence Island, Alaska, USA (NMNH 280091) exhibits lesions that can be attributed to meningioma of the lytic type. The archeological provenience is obscure. The skull is male and fully adult, probably in the 45 to 65 year age range. The mandible is missing as are all the postcranial bones. On the external table of the vault, there is a diffuse porosity, which is also seen to a more limited degree on the nasal bones and maxilla. There has been some postmortem erosion that obscures the surface texture somewhat, but the remaining surface has a pumice-like quality. On the posterior portion of the left parietal the most dramatic feature is a large lytic lesion, which is confluent with a smaller lytic lesion encroaching on the right parietal (Figures 618, 619). The larger of the two

lytic lesions has a maximal diameter of 4 centimeters and a slightly scalloped but well-circumscribed appearance, with the inner table somewhat more affected than the outer. The smaller lytic lesion is similar in appearance but less well circumscribed, suggesting lytic activity at the time of death. Other smaller lesions occur primarily on the left side of the skull with only one moderately large lesion, which measures 1.5 centimeters in diameter in the right temporal bone, and which also involves the sphenoid. The skull is somewhat thicker than normal, measuring 13 millimeters near the edge of the major lytic focus.

There are several conditions that could give rise to the pattern seen in the skull. Eosinophilic granuloma produces lesions similar to that seen in this skull. Metastatic carcinoma and angiosarcoma are also possibilities.

Primary Malignant Bone Tumors

Pathology

Primary malignant bone tumors are comparatively rare lesions that, in contrast to other malignant lesions, most frequently occur during the growing period. These sarcomas arise from osteogenic mesenchyme and are classified according to their matrix production as osteosarcomas, chondrosarcomas, fibrosarcomas, reticulum cell sarcomas, and angiosarcomas. The most common primary malignant bone tumor is the osteosarcoma followed by chondrosarcoma and Ewing's sarcoma in decreasing frequency. All other types are rare.

Osteosarcoma

This tumor occurs most often in adolescents and young adults. Males are about twice as often affected as females. The appearance of this tumor is closely related to the areas and periods of greatest enchondral growth. The most active growth plates in the skeleton are those of the distal femur, the proximal tibia, and the proximal humerus. The most common location of this tumor is in the distal femoral metaphysis, followed in frequency by the proximal tibial metaphysis and proximal humeral metaphysis. The epiphyses are practically never the primary site and often remain uninvolved, being protected by the open growth plate. The tumor predilects the long bones of the extremities, rarely involving the cancellous bones, the small tubular bones of the extremities, or the skull. Of the skull bones, the mandible is most often involved. Osteosarcomas arising past middle age may develop in abnormal bone, particularly in Paget's disease of bone.

Osteosarcoma arises within the bone at or near the metaphysial side of the growth plate. The tumor destroys varying amounts of the normal bone and extends through the cortex into the surrounding soft tissue. The appearance of the tumor, especially in dry bone, depends on the amount of mineralized bone formation. This ranges from almost purely lytic forms (Figure 620) to those with massive production of sclerotic tumor bone (Figures 621, 622). The bone produced by the tumor is woven bone, occurring in sheets and masses without architectural arrangement of trabeculae. The extracortical portion of the tumor may exhibit a radiant alignment of tumor bone, so-called sunburst (Figure 623). The elevation of the periosteum at the margin of the cortical breakthrough leads to some reactive, nonneoplastic bone formation decreasing in thickness away from the tumor. In profile, this area appears triangular on X-ray (Codman's triangle) (Figures 624, 625). The epiphysis adjacent to the tumor may ultimately become involved, but the joint surface usually remains intact. The tumor extends through the medullary cavity but practically never involves the epiphysis on the end remote from the tumor. Occasionally osteosarcoma metastasizes to other bones (Figures 626, 627).

Parosteal osteosarcoma arises on the outside of the bone in or near the periosteum. This tumor is less frequent than endosteal osteosarcoma and shows no particular sex predilection. Its occurrence is more evenly spread throughout adult age. It, too, occurs most commonly on the distal metaphysis of the femur (Figure 628). This tumor is slow growing and may produce large extraosseous masses of dense bone (Figure 629). Penetration

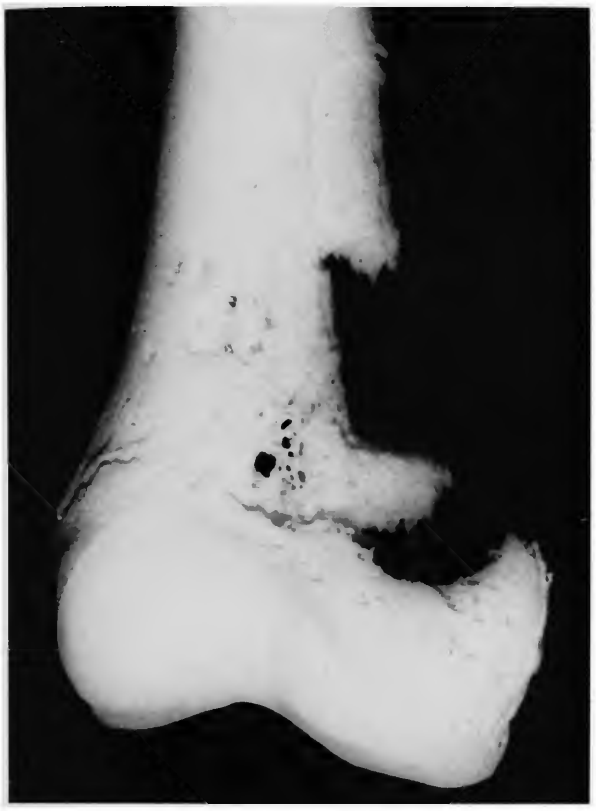
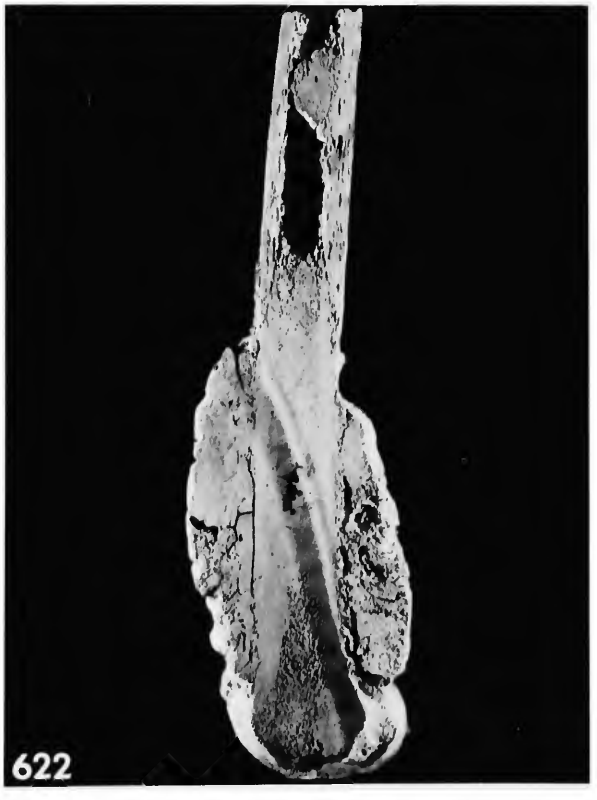


FIGURE 620.—Osteolytic osteosarcoma of distal right femur, anterior view. Notice cortical destruction and hypervascularity. The epiphysis is spared. (13 year-old female, CGH surgical specimen 2229 from 1957.)

into the marrow cavity is late or absent (Figure 630). The differentiation from parosteal traumatic myositis ossificans is not always possible on dry bone.

Chondrosarcoma

This tumor produces malignant cartilage of varying degrees of maturity. It also arises most commonly in the metaphysis of long bones of the extremities, either directly or on the basis of a previously benign cartilage tumor or developmental cartilage rest. The age distribution is more



FIGURES 621, 622.—Sclerosing osteosarcoma of distal femur: 621, Outside view, showing extraosseous portion of tumor extending to knee joint. 622, Cut surface, showing sclerotic tumor bone filling the medullary cavity. (Young adult male, died with pulmonary metastases, HM P826 from 1786.)

FIGURE 623.—Osteosarcoma of left frontoparietal area. The irregular tumor bone projecting outward is similarly projected endocranially. Specimen recovered from a grave. (Adult, ANM 2042.)

widely spread from late adolescence through adult age. There is no marked sex predilection. The most common locations are the proximal and distal metaphysis of the femur, the proximal metaphysis of the humerus and the pelvis. Although the small tubular bones of hands and feet are frequently the site of single or multiple enchondromas, chondrosarcomas are uncommon in these bones. The tumor masses tend to be nodular, destroying the cancellous bone and causing endosteal scalloping of the cortex by pressure erosion (Figures 631, 632). The more mature forms of chondrosarcoma tend to undergo spotty calcification and, in part, enchondral ossification. Only such calcified or ossified chondrosarcomas would show characteristic features in dry bone. Although the enchondral ossification of a chondrosarcoma tends to be more mature and orderly than the tumor bone of an osteosarcoma, the

FIGURES 624, 625.—Sclerosing osteosarcoma of distal left femur: 624, Surface. 625, Cut surface. Notice extensions of tumor bone through the cortex into soft tissue and shelf of reactive bone at proximal tumor margin (Codman's triangle). (12-year-old male, MGH surgical specimen 11078 from 1972.)

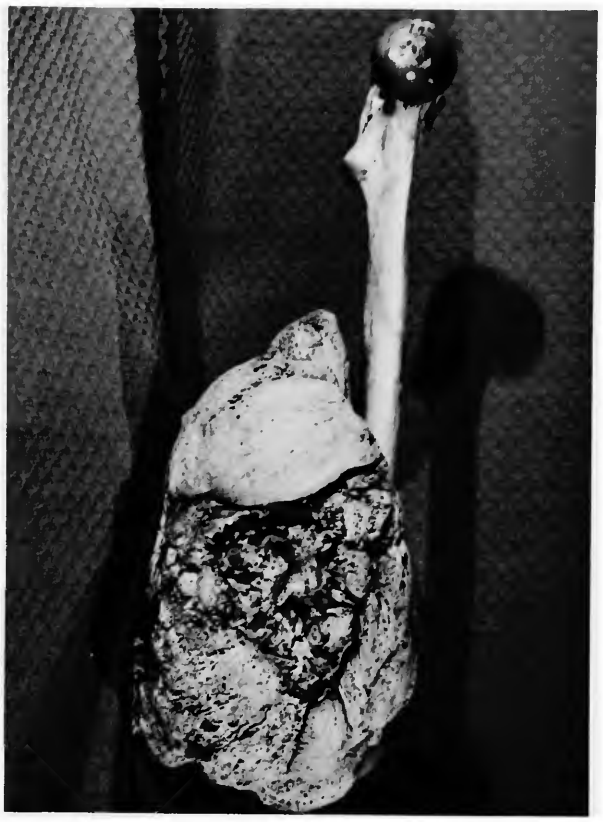


FIGURE 628.—Parosteal osteosarcoma of distal left femur. (Young adult, FPAM 2102 from before 1848.)

differentiation between the two may be dubious in dry bone.

Ewing's Sarcoma

This is the third most frequent primary malignant tumor of bone. It occurs most frequently in childhood and adolescence, rarely in adults. The tumor is formed by very primitive mesenchymal round cells, which do not produce matrix of any kind. The most common location for this tumor

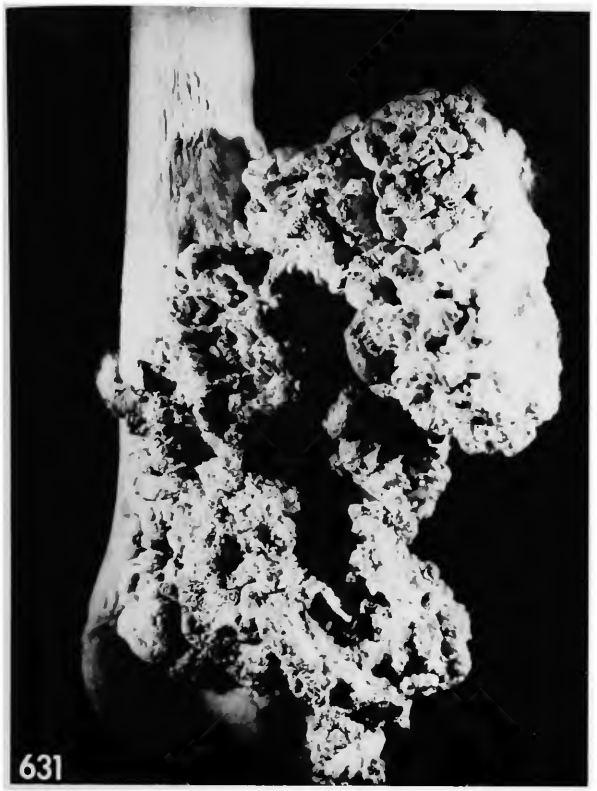
FIGURES 626, 627.—Metastatic sclerosing osteosarcoma of ninth thoracic vertebra: 626, Lateral view, showing tumor bone replacing the vertebral body. 627, Cut surface, showing ivory-like tumor bone. There was a smaller second focus in the twelth thoracic vertebra. (20-year-old male, FPAM 3171, autopsy 56719 from 1870–71; scales in cm.)

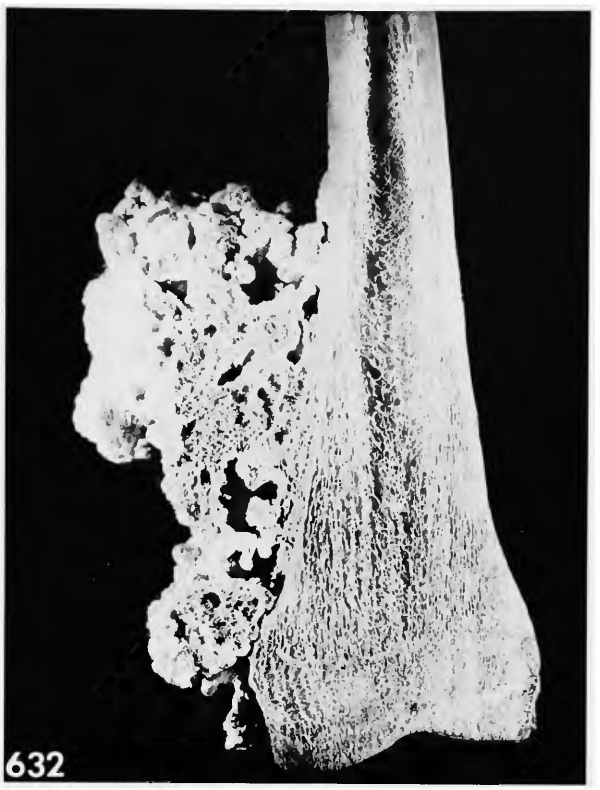
FIGURES 629, 630.—Parosteal osteosarcoma of proximal right tibial metaphysis: 629, External view, showing layer of reactive bone at junction of cortex and tumor. 630, Cut surface, showing dense radiating tumor bone. Notice slight involvement of underlying cortex. (Young adult, FPAM 3639 from 1878; scales in cm.)

is a long bone of an extremity or a pelvic bone. The structure of the tumor, consisting of loose round cells not held together by matrix, is responsible for the permeative pattern of this lesion. The tumor cells can easily push through intertrabecular marrow spaces and invade Haversian canals of the cortex. The result of this is that a large segment of the bone, including the diaphysis, is involved. The destruction of trabecular bone may be less marked, but resorption along Haversian canals leads to lamination of the cortex. The tumor spreads into and through the periosteum, eliciting some reactive bone formation of surface parallel ("onion skin") or radiant ("sunburst") type. Of all the bone tumors, this is the one most often metastasizing to other bones. The presence of reactive bone may make the differentiation between Ewing's and osteosarcoma impossible on dry bone. The absence of massive destruction and the long axial involvement, together with permeative intracortical resorption, would arouse suspicion that the lesion may have been a Ewing's sarcoma or a reticulum cell sarcoma, which usually affects adults, especially past 50 years of age.

Chordoma

Chordoma is a rare tumor arising from notochordal rests. The tumor mostly affects individuals above 40 years of age and somewhat predilects males. During embryonic development the notochord extends from the skull base to the tip of the coccyx, through the entire spine. In the





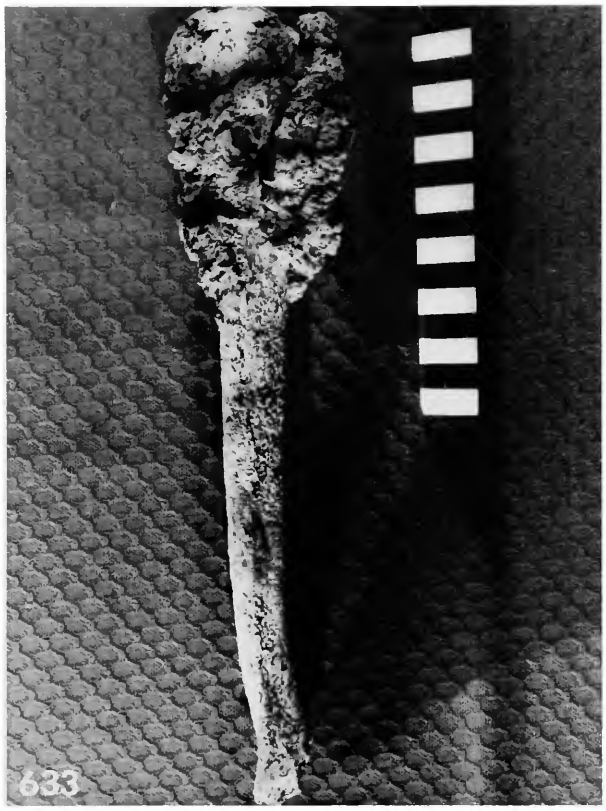
FIGURES 631, 632.—Parosteal chondrosarcoma of distal right femur: 631, Outside view; notice the nodular character of the mineralized portion of the tumor. 632, Cut surface; notice the mature trabecular pattern of the bone replacing the core of the tumor. (31-year-old male, 10 years duration, WM S.72a.2 from 1894.)

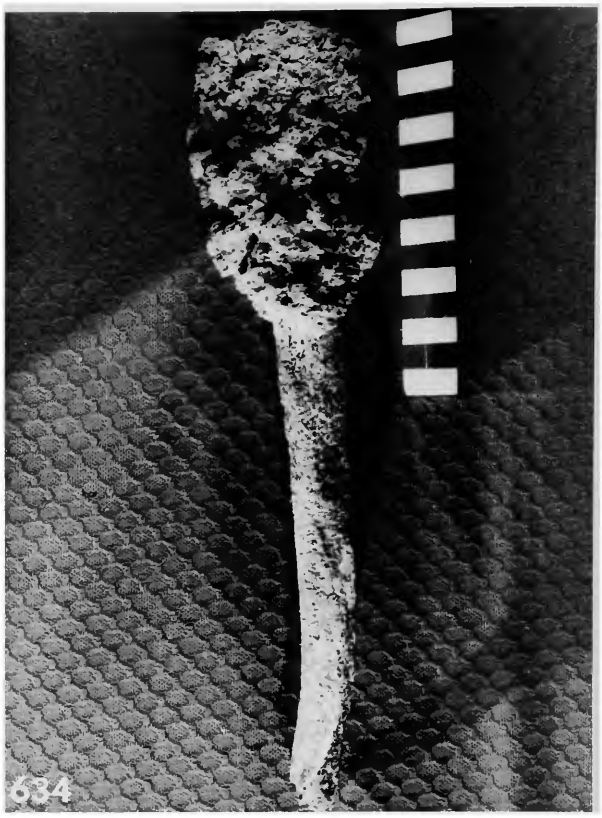
process of vertebral development the notochordal tissue is extruded into the nucleus pulposus of the intervertebral disc. In the movable part of the spine, it is mostly destroyed by mechanical attrition. Only in mechanically sheltered areas of the axis do notochordal remnants survive into later life. These areas are the skull base, the sacrum, and the second cervical vertebra, where the odontoid process is united with the body of C2 by fusion of an intervening disc. These facts explain the localizations of chordomas: 55 percent sacral, 35 percent skull base, and only 10 percent vertebral, especially cervical.

Small notochordal rests are common in the clivus of the skull base and may show a small, pitted defect in dry bone in this area. The tumor is purely destructive producing large, lytic defects in the spheno-occipital portion of the skull base or the sacrococcygeal area centered on the midline. Since no mineralized matrix is formed, nothing of the tumor itself could remain in dry bone.

PALEOPATHOLOGY

Primary malignant tumors of bone are rare in modern medical practice and are equally rare in archeological specimens. Ruffer and Willmore (1914) report a tumor of the pelvis from Egypt dated about A.D. 250. The tumor affects the right innominate, particularly the ischium and inferior ilium. It appears to have started in the cancellous tissue of the pelvis and is slightly expansive in nature, producing a deformation of the obdurator foramen and encroaching on the acetabulum. The authors rule out carcinoma and infection and suggest osteosarcoma adding that they can-





FIGURES 633–635.—Osteosarcoma or chondrosarcoma of the proximal left humerus of a Celtic warrior (approximately 800–600 B.C.) found near Muensingen, Canton Bern, Switzerland: 633, Medial view; note that the articular surface is spared. 634, Lateral view. 635, X-ray. (NHMB A95; scales in cm.)

not determine if the tumor was primary or secondary. The illustration of the lesion is of little value but does not show the active irregular growth of bone usually seen in osteoblastic osteosarcoma. The lesion appears to be chronic rather than acute, which would tend to rule out osteosarcoma. However, a tumor of some type is possible as is infection.

Dastugue (1965) describes a tumor of the right maxilla and zygomatic bone on a skull from a Middle Age site associated with the town of Caen in France. The tumor measures 60 millimeters wide by 40 millimeters high and encroaches both on the nasal passage and the right orbit. The surface of the tumor is very irregular with relatively large coalescing spicules. There are three inferior projections. Curiously, there is antemortem damage to the right zygomatic bone adjacent to the tumor and Dastugue suggests the possibility of surgical intervention to explain this defect. The tumor appeared malignant to Dastugue and he expresses the opinion that it caused the death of the individual. The morphology of the lesion is compatible with a primary malignant tumor of bone. However, other possibilities, such as callus following trauma to the face, seem more probable. In my opinion, particularly in view of the injury to the zygomatic bone, trauma rather than surgical intervention is more probable.

A tumor of the mandible was found in an American Indian skeleton excavated in the state of West Virginia (Kelln, McMichael, and Zimmermann, 1967). The authors date the specimen to the mid-seventeenth century. The lesion is rather large, measuring about 24 millimeters in diameter and 19 millimeters in depth. The mandible is probably from a female of about 20 years of age. The authors suggest several possible neoplastic conditions including osteoma and osteosarcoma as the cause of the lesion. The position of the tumor near the symphysis and the young age of the individual are compatible with a diagnosis of primary malignant tumor. However, the chin is prone to trauma and infection and bony reaction to these morbid conditions is more likely.

An unambiguous example of a primary malignant tumor of bone is found in the skeletal collections of the Natural History Museum in Bern, Switzerland. This specimen (NHMB A95) is the left humerus from a Celtic warrior tomb dated to approximately 800–600 B.C. and found near the town of Muensingen in Canton Bern. Brothwell (1967a:331) provides a brief description of the specimen. Although damaged by postmortem erosion, the humerus is complete except for the distal epiphysis, which was lost postmortem. The lesion extends completely around the proximal humerus but is least developed on the medial aspect. The bone associated with the lesion is approximately 7 centimeters in maximum diameter and abnormal tissue extends slightly more than one-third the length of the humerus (Figures 633, 634). Tumor bone overlies the insertion of the joint capsule but does not involve the joint surface itself. The gross morphology of the tumor consists of large coalescing bony projections, which have a coarse coral-like appearance. While the age at death of the individual cannot be determined, the humeral head appears to be fused indicating an age in excess of 15 years.

X-ray films of this specimen were provided through the courtesy of Dr. Walter Huber, the Director of the Natural History Museum in Bern. The films reveal a lytic process extending 2 to 3 centimeters into the externally normal cortex distal to the lesion (Figure 635). The location of the tumor, as well as its gross and X-ray film appearance, is compatible with the diagnosis of primary osteosarcoma or chondrosarcoma.

Metastatic Tumors

Pathology

Tumors metastatic to the skeleton are a common occurrence in carcinoma but rare in sarcoma. The tumor cells reach the skeleton, usually the bone marrow, by the blood stream. The frequency of skeletal metastases of different tumors depends, to a considerable extent, on the location of the primary tumor. Larger tumor particles

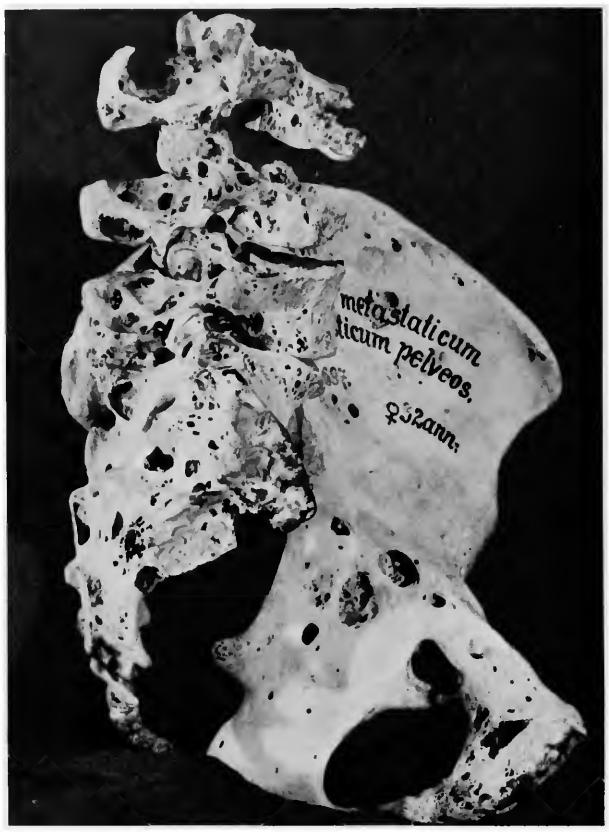


FIGURE 636.—Osteolytic metastatic carcinoma of spine and pelvis. Notice disseminated round lytic lesions exposing spongiosa and the involvement of the neural arches and spinous processes. (32-year-old female with breast cancer, FPAM 5697, autopsy 182394 from 1929.)

(tumor emboli) are usually retained in the primary filter, but individual tumor cells, as in round cell tumors especially, pass readily through a capillary filter. The primary filter for tumors drained by the caval system is the lung, for those in the portal drainage area the liver, while tumors arising in the lung have direct access to the systemic circulation through the pulmonary veins.

The frequency of skeletal metastases varies with different observers. The largest autopsy statistics based on two decades of cases of the Zurich Pathological Institute preceding 1948, are as follows: Of over 3000 malignant tumors, 12 percent metastasized to the skeleton (Walther, 1948:256). Jaffe (1958:599–600) feels that this figure is abnormally low due to early postoperative deaths in previous decades. He states that presently at least 76 percent of tumor cases with metastases that come to autopsy show involvement of the skeleton. In anthropological material, in the absence of radical surgery, with the malignancy having taken its natural course, a high figure for skeletal metastases should be expected.

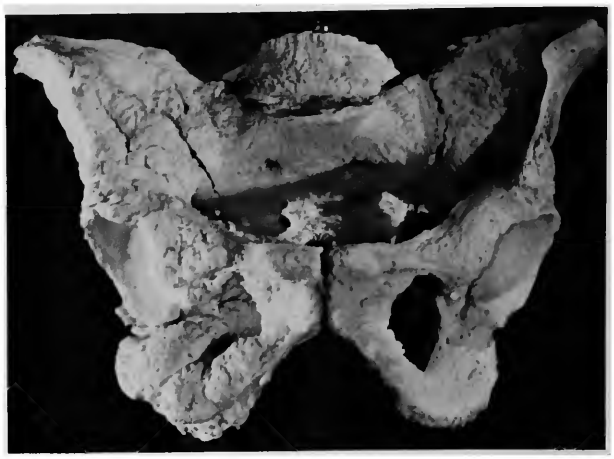
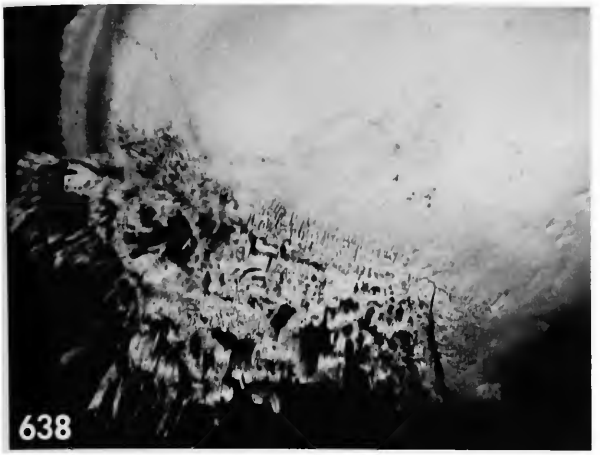
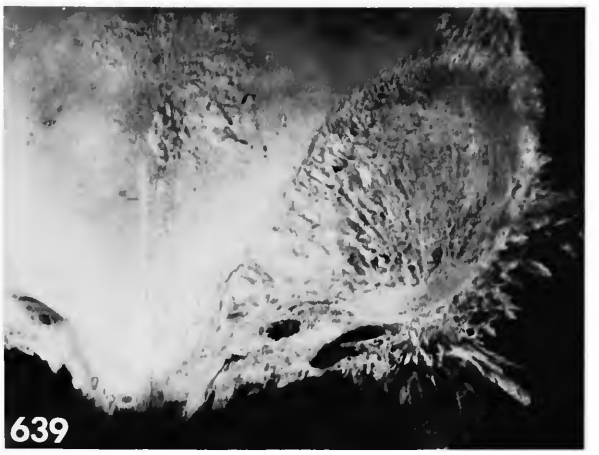


FIGURE 637.—Massive osteoblastic metastatic carcinoma of pelvis and spine. Notice the diffuse nodular hyperostosis. (66-year-old male with cancer of the prostate, IPAZ autopsy 497 from 1946.)

Everybody agrees that the carcinomas most commonly metastasizing to the skeleton are those of the female breast and of the male prostate. The figures in Walther's (1948:277) series are as follows: breast 47.2 percent, prostate 42.4 percent, thyroid 30.8 percent, lung 29.8 percent.

The bones most commonly involved in metastatic carcinoma are the spine (including the sacrum), the proximal metaphysis and epiphysis of the femur, ribs, sternum, skull, pelvis, and proximal humerus, in decreasing order of frequency. This coincides with the location of hemopoietic marrow in the older adult. The figures of Walther (1948:279) concerning only autopsy cases with skeletal metastases are as follows: spine 80 percent, femur 40 percent, ribs and sternum 25 percent, skull and pelvis 25 percent, humerus and shoulder girdle 7 percent, other extremity bones 1 to 2 percent. Skeletal metastases distal to the elbow and knee area are distinctly uncommon.





FIGURES 638–640.—Osteoblastic metastases of adrenal neuroblastoma: 638, External view of frontal bone, showing radiating reactive bone spicules in supraorbital area. 639, Endocranial view, showing radiating trabeculae replacing the inner table. 640, Basal external view of sphenoid, showing involvement of both wings. (14-month-old male, PMES 1VUH(1 and 1a) from before 1929.)

The tumor cells reaching the bone marrow grow and first replace the marrow without change of the bone architecture. The majority of metastatic deposits discovered at autopsy are not demonstrated by external inspection or radiography. In the Zurich series, only 36 percent of the skeletal metastases were discovered during life (Walther, 1948:189). Continued growth of the tumor cells usually leads to destruction and osteoclastic resorption of cancellous bone and internal scalloping or complete destruction of cortical bone (osteolytic metastases). At least microscopically, however, some reactive bone formation is often seen in some of the lesions and/or in their vicinity. In other cases, a strong osteoblastic response to the presence of the tumor cells is observed, consisting of new bone deposits replacing old trabeculae or coating remaining old trabeculae. This results in blotchy or diffuse densities on X-ray. It is not entirely clear what causes this difference. In general, however, fast growing tumors are mainly osteolytic, while slow growing ones elicit an osteoblastic response.

Carcinoma of the breast mostly shows osteolytic metastases, while carcinoma of the prostate usually shows osteoblastic deposits. The great majority of metastatic bone tumors are multiple and involve more than one bone. Solitary and relatively slow growing metastases are most often caused by renal cortical or thyroid carcinomas. These solitary lesions occur with predilection in the shaft of femur or humerus, less commonly in the cranial vault. They may appear on X-ray as a cystic lesion, with an expanded shell of new cortex showing soap bubble appearance. They may mimic benign lesions, except for the fact that diaphysial benign lesions are very uncommon.

Lytic metastases progressively destroy cancellous bone and erode the cortex of long bones from within. Disseminated lytic metastases may be indistinguishable from multiple myeloma in dry bone. In the spine, the most frequently involved

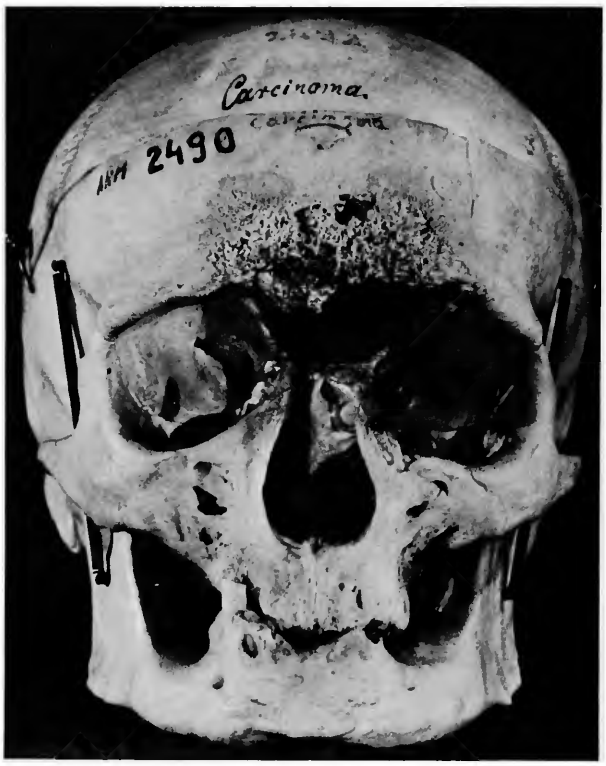


FIGURE 641.—Probable carcinoma of the ethmoid with destruction of ethmoid, medial orbital wall, and adjacent frontal bone. Notice the almost complete absence of bony reactivity. (Elderly individual, ANM 2490.)

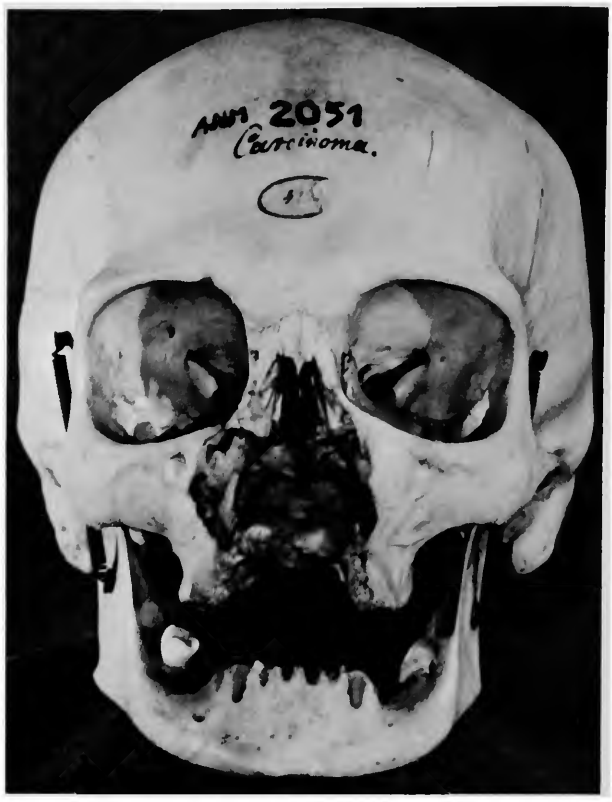


FIGURE 642.—Carcinoma of the nasal cavity with destruction of palate and interior portion of maxilla. Notice the complete absence of reactive bone. (Adult, ANM 2051.)

vertebrae are the lower thoracic, followed in decreasing frequency by the lumbar, sacral, and cervical vertebrae. The metastases are prevailingly located in the vertebral bodies, but neural arches and spinous processes may also be affected (Figure 636). Compression fractures are common, secondary to lytic metastases. In the cranial vault, the most marked destruction is in the diploë, while the outer and inner tables are long spared. Metastases in the long bones predilect the intertrochanteric area of the femur or proximal portion of the humerus. Pathological fractures secondary to lytic deposits in long bones are common. The frequency in Copeland's (1931) radiological series is as follows: renal cortical carcinoma 45 percent, thyroid carcinoma 33 percent, malignant melanoma 33 percent, carcinoma of the colon 33 percent, carcinoma of the lung 25 percent, carcinoma of the breast 15 percent, carcinoma of the stomach 14 percent, carcinoma of the prostate 0.2 percent and no pathological fractures in cancers of other primary sites.

Subperiosteal metastases are uncommon. If present they are usually lytic, causing deep scalloping of the underlying cortex. This is seen most commonly in renal cortical carcinoma (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952(2): 1008). Osteoblastic metastases are most commonly observed in carcinoma of the prostate. The distinct lesions may become confluent and extremely sclerotic. There is usually little if any bone destruction, and the outer contours of the involved bones are not significantly altered except for those advanced cases where massive subperiosteal bone is deposited in the form of mossy osteophytes (Figure 637). The distribution of metastatic prostate carcinoma is different from that of the usual hematogenous metastases. The ear-

liest and most massive involvement is in the pelvis, sacrum, and lumbar vertebrae. This is probably due to the fact that the pelvis and vertebral venous plexuses freely communicate and thus permit direct colonization of the adjacent bones without passage through the pulmonary filter (Batson, 1940). In most instances blastic or lytic pelvic metastases begin in the floor of the acetabulum, spread into the pubic rami, and lastly involve the iliac wings (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952(2):1007). The differentiation from Paget's disease can be difficult on gross inspection or X-ray. However, microscopic sections should reveal the typical mosaic structure if the lesion is Paget's.

Special mention should be made of the appearance of skeletal metastasis in neuroblastoma, a malignant tumor of the adrenal or of the sympathetic chain in infants and children, usually under 3 years of age. This tumor, a round cell lesion, freely seeds to the bone marrow in wide distribution, including trunk, skull, and extremities. The lesions often elicit osteoblastic reaction leading to "hair-on-end" appearance in areas of the cranial vault and periosteal layered-bone formation of varying thickness on the long bone shafts of the actively growing childhood skeleton (Figures 638–640). The periosteal deposis differentiate it from similar skull lesions in severe anemias.

Craniofacial involvement may occur by direct extension of paranasal carcinomas. Craniopharyngeal carcinoma may erode the cranial base from without. Carcinoma of the ethmoid may destroy the adjacent frontal bone and the orbital wall (Figure 641). Carcinoma of the nasal cavity may destroy part of the maxilla and of the hard palate (Figure 642). In these cases the absence of reactive bone and the strictly localized area of destruction usually permits, in dry bone, differentiation from lesions of tertiary syphilis, leprosy, and lupus vulgaris.

Paleopathology

Secondary tumors of bone are by definition malignant. These types of tumors are presently much more common then primary malignant tumors of bone. Thus, even though they are associated with the older age categories, they might be more common in archeological skeletons. However, like primary malignant tumors, they pose problems in differential diagnosis in dry bone specimens, particularly from infectious diseases.

A skeletal specimen from a cemetery near Dubendorf, Canton Zurich, Switzerland, dated between the eleventh and fifteenth centuries exhibits a diffuse osteoblastic reaction. The specimen is an adult male probably in excess of 50 years of age at death and is currently stored in the Anthropological Institute of the University of Zurich, Switzerland (AIUZ 7757). The right innominate contains a lesion largely limited to the periosteal surfaces of the ilium (Figure 643). The lesion consists of extensive fine porous bony buildup. There is no obvious destruction of underlying bone nor evidence of abscess or cloacas. There is a similar bony reaction on the right anterior proximal femur. Two of the ribs in this specimen also show periosteal lesions (Figure 644). The disseminated nature of the disease and the morphology of the lesions are compatible with metastatic tumor. The extensive involvement of the innominate and the sex of the case suggest prostate cancer.

Osteolytic metastatic cancer of the skeleton is probably the most common type of malignant lesion seen in archeological specimens. The major problem in diagnosis is to distinguish between metastatic carcinoma and multiple myeloma. In the skeleton these two types of cancer reflect a morphological gradient making differential diagnosis impossible in many cases. A presentation of three problem cases is provided in the discussion on reticuloendothelial and hemopoietic disorders (pp. 267–269).

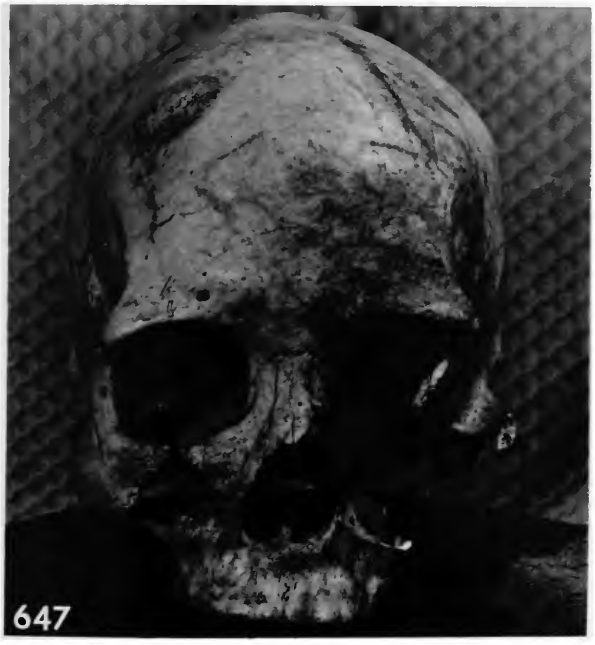
Bony responses to neoplasms may produce mixed blastic and lytic lesions. An archeological example of this condition is seen in a skeleton from Kachemak Bay, Alaska, USA (FM SEL30 AMU3 Burial 11). The skeleton was excavated in 1974 and dated circa A.D. 350 by carbon-14 analysis. The skeleton was shown to me through the

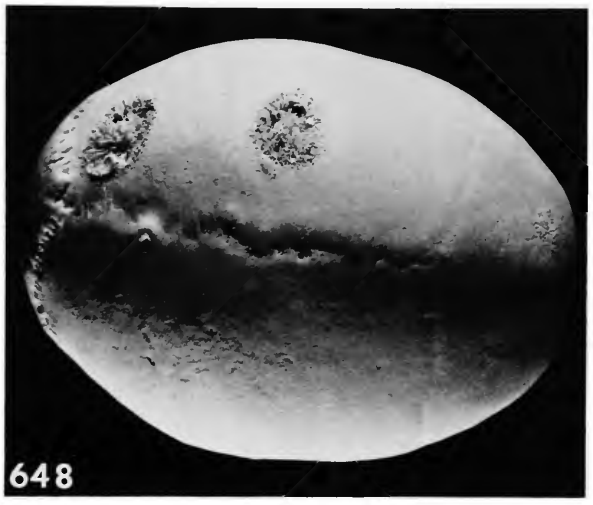
FIGURES 643, 644.—Osteoblastic lesions of the right innominate possibly due to metastatic carcinoma of the prostate. Specimen from an adult male skeleton from a cemetery near Dubendorf, Switzerland, dated between the 11th and 15th centuries AD. 643, Right innominate. 644, Right rib from lower chest region. (AIUZ 7757.)

courtesy of John Lobdell, currently with the Department of Anthropology at the University of Alaska, Anchorage, Alaska, USA. The specimen consists of the skull and the incomplete portions of the postcranial skeleton. The sex is female and the pitting on the posterior portion of the pubic symphysis suggests multiple pregnancies. Skeletal age is in the 50-year-plus range.

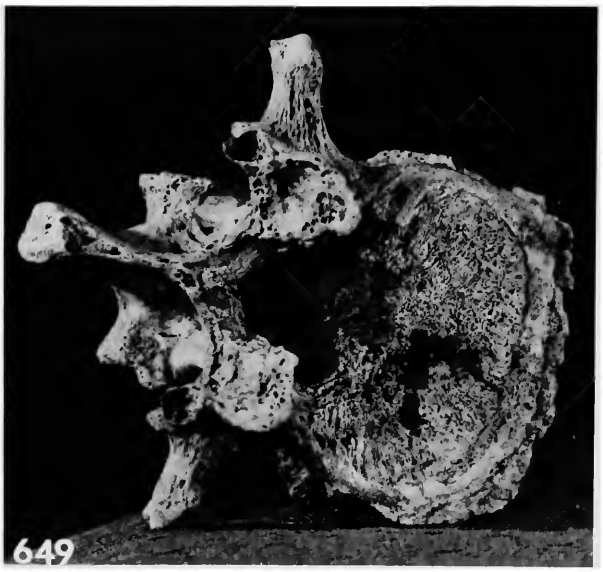
The disease process is multifocal with lesions on the skull, ribs, vertebrae, left humerus, both innominates, the right femur and both tibiae. The skull lesions are circular and fairly large (Figures 645–647), the smallest is about 25 millimeters in diameter and the largest about 45 millimeters in diameter. One lesion is on the frontal, two are entirely on the left parietal, and a third is partially on the left parietal but crosses the sagittal suture into the right parietal. The lesions show an active lytic front at the boundary with normal bone. The lytic process results in the bone having the appearance of a fine sponge. There is no osteoblastic circumscription of the lytic process. The residual porous, spongy bone provides the scaffolding for the formation of coarse fiber bone in the more central portions of the lesion. The central osteoblastic activity produces bone that rises above the plane of the normal bone. The lesions in this skull are similar to lesions occurring in the skull of a modern case of metastasis from lung carcinoma. The latter case is from the Institute of Pathological Anatomy at the University of Zurich, Switzerland (IPAZ autopsy 1850/67). In the skull lesions there is a lytic front adjacent to normal bone with fiber bone formation in the more central portion of the lesion (Figure 648).

The postcranial lesions in the Alaskan specimen appear to be more variable morphologically than those of the skull, although the picture is complicated by considerable postmortem breakage. The cervical vertebrae are missing postmortem. The upper thoracic vertebrae are somewhat osteoporotic but are normal, except for slight degenerative-arthritic changes. However, T10 and T12 both have lytic lesions of the lamina. Similar destructive lesions occur on the laminae of the first two lumbar vertebrae. The vertebral bodies of L1, L2, L3, and L5 show evidence of





FIGURES 645–648.—Mixed blastic and lytic lesions of the skull: 645, Left lateral view of a skull showing two large porous lesions on the parietal bone. Specimen from an archeological site dated about A.D 350 located near Kachemak Bay, Alaska, USA. 646, Detailed view of parietal lesion adjacent to the sagittal suture. 647, Frontal view, showing lesion of right portion of frontal bone. (FM SEL 30 AMU3 Burial 11.) 648, Metastatic skull lesions from lung carcinoma in a modern case. (IPAZ autopsy 1850/67.)



FIGURES 649–651.—Postcranial lesions in archeological skeleton from Kachemak Bay, Alaska: 649, Superior surface of the first lumbar vertebrae; note the lytic lesion of the posterior portion of the vertebral body. 650, Blastic and lytic lesions of the diaphysis of the right femur. 651, Lytic lesion of the proximal tibia. (FM SEL30 AMU 3 Burial 11.)

erosion and disk herniation (Figure 649). The first through the third vertebral bodies are wedged due to partial collapse. The wedging defect of the vertebrae is well consolidated, suggesting a long-standing condition that does not appear to be related to the active lytic process in the vertebrae. The few rib fragments show active lytic involvement.

On both innominates there is a large lytic area in the region of the acetabulum in which the bone is entirely destroyed. Both these lesions appear active. The left femur appears to be normal, although the distal end is missing postmortem. Only the diaphysis of the right femur is preserved. It shows multiple lesions, all of which combine lytic and blastic processes (Figure 650). The proximal ends of both tibiae have large, purely lytic defects in the metaphysial cortex (Figure 651). Both lesions are oblong, the left measuring about 40 by 20 millimeters on the X-ray film. Postmortem damage has destroyed part of the lesion on the right tibia. What remains suggests a lytic lesion that is even larger than the one seen on the left.

The morphological similarity of the skull lesions to those seen in known metastatic carcinoma of bone make carcinoma a probable cause of the lesions. The lesions of the long bones would be compatible with this diagnosis. Another possible consideration is a mycotic infection.

Lesions of the Joints

In order to avoid duplication, the joint manifestation of the specific infections have been discussed in connection with the respective bone lesions in the discussion on infectious disease (pp. 141, 198).

Septic Arthritis

Pathology

Septic arthritis is caused by bacteria entering the synovium and the joint cavity by the blood stream, extension of infection from the bone (osteomyelitis) or soft tissue, or by direct introduction through a wound. Occasionally any joint may be involved, but about one-third of the cases involve the knee and one-third the hip joint, leaving one-third for all other locations. About 10 percent show osteomyelitis. The causative organisms identified are staphylococci in about one-third and streptococci in about one-fourth of the cases (Heberling, 1941). Other organisms causing septic arthritis are gonococci, pneumococci, meningococci, and various Gram-negative rods from enteric infections (*Salmonella*, *Shigella*). Gonococcal arthritis is more often multiarticular than other septic joint infections. Harkness (1942) observed multiarticular involvement in over 80 percent of instances of gonococcal arthritis.

The acute phase of septic arthritis is limited to the synovial membrane and the articular cartilage and will not be recognizable in dry bone

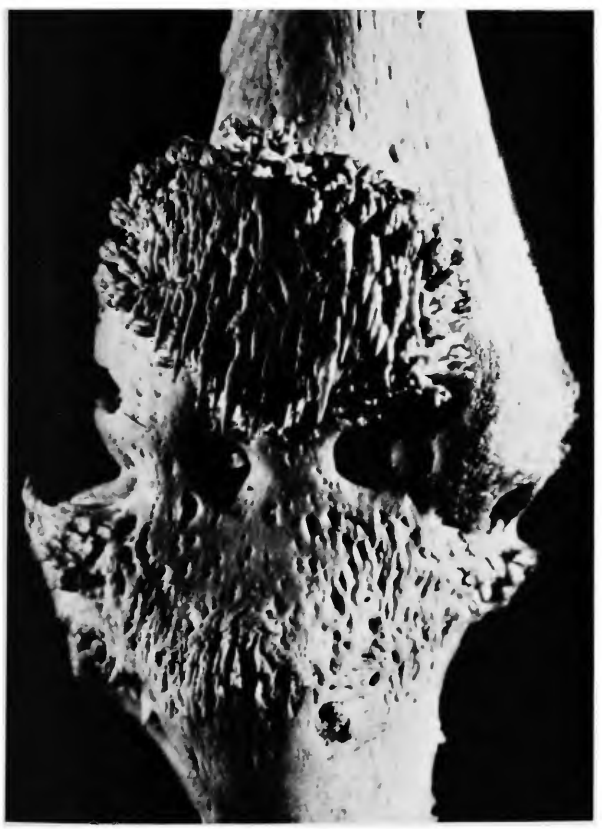


Figure 652.—Septic arthritis of right knee with bony ankylosis following violent trauma to the knee; anterior view. Notice reactive bone formation and hypervascularity around the joint and on the patella. (45-year-old male, WM HS 84.7 from 1842.)

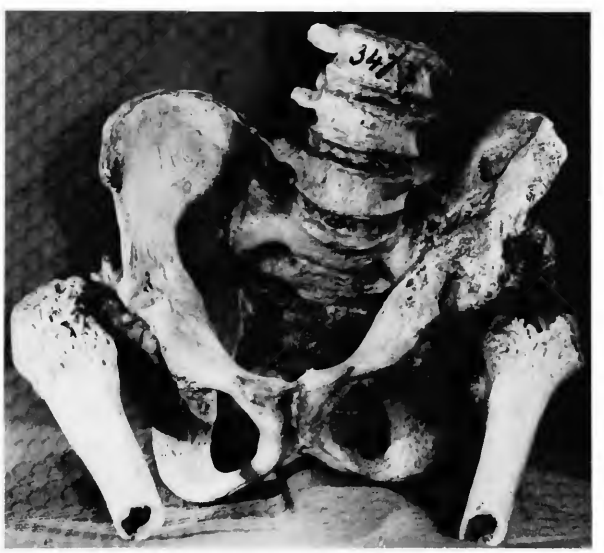
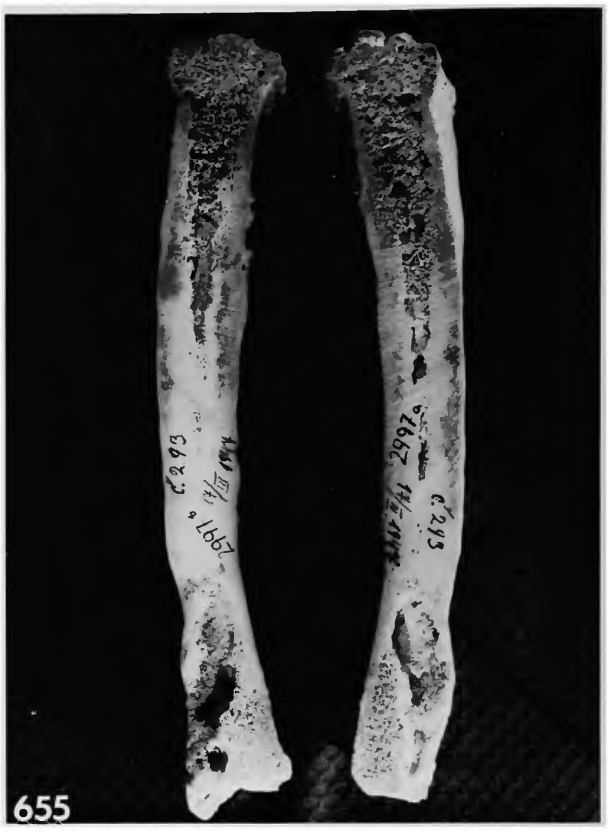
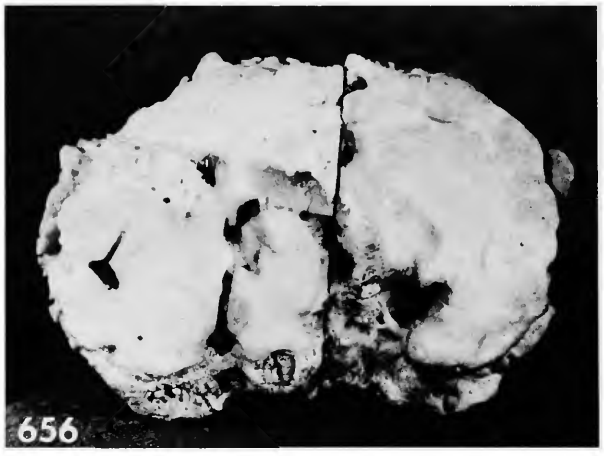


Figure 653.—Chronic osteomyelitis of left ilium with left sacroiliac ankylosis and septic arthritis of congenitally dislocated left hip. (Underdeveloped 24-year-old male, PMUG 3477, autopsy 9928 from 1879.)





FIGURES 654–656.—Chronic sclerosing osteomyelitis of left tibia with involvement of the proximal epiphysis and joint surface: 654, External view, showing periosteal hyperostosis. 655, The bisected specimen shows sclerotic obliteration of the medullary cavity and a recent surgical defect in the distal metaphysis. 656, The proximal articular surface of tibia shows multiple perforations. (ANM 2997.)

unless the subchondral bone has become involved and eroded. Unrelieved septic arthritis will often terminate in bony ankylosis (Figure 652). The acute and final stages are very similar to tuberculous arthritis in dry bone, although there is usually less bone destruction and concomitant shortening in septic arthritis than in tuberculosis.

In infants, before separation of the blood supply of the femoral head from that of the proximal metaphysis, direct extension of osteomyelitis to the hip joint is common. In such cases, destruction of the femoral head and of the growth cartilage

FIGURE 657.—Septic arthritis of left elbow secondary to partly healed comminuted fracture. Notice destruction of joint constituents, massive reactive bone formation and ulnarhumeral ankylosis; posterior view. (Adult, WM S 81.1 from 1851.)

may result in bony ankylosis between the femoral metaphysis and the acetabulum, followed by marked growth deficit of the proximal femur.

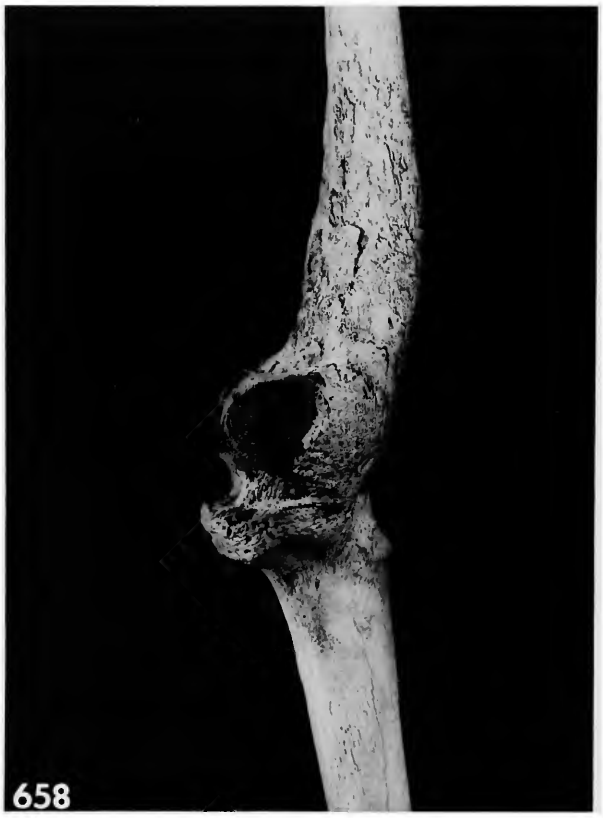
The most marked destructive changes in dry bone are seen in arthritis complicating chronic osteomyelitis (Figures 653–656) or infected fractures (Figure 657).

PALEOPATHOLOGY

References to arthritic change resulting from infection of one or more joints are uncommon in the literature on paleopathology. Wells (1962b) has attributed the degeneration of the left humeral head in a young female skeleton to septic arthritis. This case is from an Early Saxon cemetery at Caister-on-Sea, England. Jarcho, Simon, and Bick (1963) have described a fused hip from a pre-Columbian archeological site at Wupatki Pueblo, Arizona, USA. The authors conclude that the fusion was the sequela of infection of the hip. Brothwell (1965a, pl. 7E) illustrates a pair of humeri from a medieval adult skeleton from Scarborough, England. The head of the left humerus is abnormal with a flattened, irregular joint surface. Brothwell suggests a septic condition, such as osteomyelitis, as the cause for the arthritic change. The humerus is unusually short. This could be the result of diminished growth due to infection during childhood. However, the combination of reduced length and rather robust diaphysis also suggests the possibility of a congenital abnormality, such as achondroplasia.

Moodie (1928) suggested an association between degenerative arthritis and dental disease. He based this observation on degenerative arthritis of the right temporomandibular fossa and the presence of dental abscess in a female skull from an archeological site at Cinco Cerros, Peru. The skull (No. 348) is part of the skeletal collection at the San Diego Museum of Man, California, USA. However, it is apparent in Moodie's (1928) figure 1 that there is a fracture through the temporomandibular fossa with some refill of the defect indicating healing. Thus, a more probable diagnosis would be degenerative arthritis following trauma.

Inglemark, Møller-Christensen, and Brinch (1959) report a correlation between arthritic change in the spine and the presence of dental infection in a cemetery population from Aebelholt Monastery in Northern Zealand, Denmark, dating from approximately A.D. 1185 to 1559. The authors controlled for one variable, age, which would be a major factor in this correlation. However, this does not mean that a causal relationship between the two pathological conditions can be assumed, because there may be other underlying factors to which the pathological conditions may be related. Morse (1969, pl. 11E) briefly describes an arthritic right hip joint from an adult male skeleton. The acetabulum is shallow and enlarged. The head of the femur is



FIGURES 658, 659.—Septic arthritis of the knee resulting in fusion of the left femur and tibia: 658, Medial view; note the periosteal reactive bone on the distal femur. 659, X-ray of mediolateral view. (Adult, surface find from Chancay, Peru, NMNH uncataloged.)

deformed. Two isolated pieces of bone appear to be sequestra from the head of the femur. The specimen is from the Klunk site in Illinois, USA, and is probably pre-Columbian (Hopewell Culture). Morse attributes the arthritic changes to infection.

Clear evidence of septic arthritis is seen in the fused left femur and tibia from an isolated surface find near Chancay, Peru (Figures 658–661). There are no data on cultural association or date. The specimen is part of the skeletal collections of the National Museum of Natural History, Washington, D.C., USA (NMNH, uncataloged). Although the bones are somewhat short, they are robust. Arthritic change is limited to the knee. The X-ray film indicates normal cortical bone thickness and density indicating that the limb was used throughout the life of the individual (Figure 659, 661). The knee has fused in the extended position with some rotation of the axis of the tibia relative to the femoral condyles. Because of this rotation the medial condyle of the tibia projects posteriorly. There is extensive periosteal bone reaction on the distal diaphysis and metaphysis of the femur. Periosteal reactive bone on the tibia is minimal and limited to the bone adjacent to the proximal articular surface. There is a cloaca in the posterior cortex of the distal femur just superior to the lateral condyle (Figure 660). The X-ray film reveals considerable sclerosis in the distal femur. The periosteal reactive bone, sclerosis, and cloaca are indicative of infection and inflammation, making a diagnosis of septic arthritis highly probable.

FIGURES 660, 661.—Septic arthritis of the knee resulting in fusion of the left femur and tibia: 660, Posterior view; note the cloaca (arrow) in the femur. 661, X-ray of anteroposterior view. (Adult, surface find from Chancay, Peru, NMNH uncataloged.)

Rheumatoid Arthritis

PATHOLOGY

Rheumatoid arthritis is a generalized connective tissue disease of unknown etiology with most significant involvement of joints. Genetic factors and climatic conditions are thought to be important. Generally, the disease is thought to be more common in cold than in hot climates. However, a statistical study carried out on the Blackfoot Indians of northern Montana and the Pima Indians of southern Arizona confirmed neither of these viewpoints (Bunim, Burch, and O'Brien, 1964). The disease manifests itself in the adult and in the juvenile forms. Although the pathology in both conditions is essentially the same, the areas of involvement are sufficiently different to necessitate a separate discussion. In either situation I am here only concerned with the late stages exhibiting significant bone changes.

Adult Rheumatoid Arthritis

This disease usually commences in the fourth and fifth decades and continues through the lifetime of the patient. There is a great predilection of females over males with a ratio of about 3:1. The disease is not rare. In northern Europe, about 4 percent of the males and 16 percent of the females over 65 years of age showed some presumptive or definite evidence of this disease (Lawrence, Laine, and de Graaff, 1961).

Rheumatoid arthritis usually involves multiple joints and is frequently symmetrical. Joints of the hands are most frequently, most characteristically, and often first involved. In the hands, the

predilected locations are the metacarpophalangeal joints and the proximal interphalangeal joints. The second interphalangeal joints are usually spared. The knee is the most frequently involved large joint. The temporomandibular joint is affected in about 25 percent of the cases (Garrod, 1890:250). Other commonly involved joints are wrist, carpal joints, shoulder, and elbow. Tarsal, tarsometatarsal, metatarsophalangeal joints, and toe joints are less often involved than the corresponding joints of the hands. The hip joint is the least often affected of the large joints. The joints of the trunk are much less commonly involved. If the spine is affected, the changes are usually most marked in the diarthrodial joints of the cervical spine, including the joint between axis and atlas.

The disease process consists of an inflammatory infiltration and proliferation of the synovium, which is slowly progressive over many years and ultimately destroys and disfigures the joint. The process in untreated cases often terminates in either loosening of the joint with subluxation or ankylosis (Figure 662) with bony continuity. In the active phase the joint changes are accompanied by marked osteoporosis in the adjacent bones. If joint mobility is maintained, degenerative arthritis is superimposed on the rheumatoid lesion. However, in contrast to primary degenerative arthritis, osteoporosis still prevails and marginal lipping is less prominent.

The bone changes on large joints consist of osteoporosis of the articular ends, thinning and resorption of the subchondral bone plate, subchondral bone resorption, and lytic foci due to subchondral cysts (Figure 663). The contour of the articular surface is deformed and irregular.

Subluxation is most commonly observed on metacarpophalangeal joints with axial deviation towards the ulnar side. On dry bone, evidence of subluxation would consist of extension of the convex articular surface on the side of deviation. Corresponding unilateral marginal lipping may be present on the concave articular facet.

Ankylosis may fuse carpals and tarsals into solid blocks of extremely porotic bone showing sparse trabeculae. Carpometacarpal and tarsometatarsal joints may also be found ankylosed.

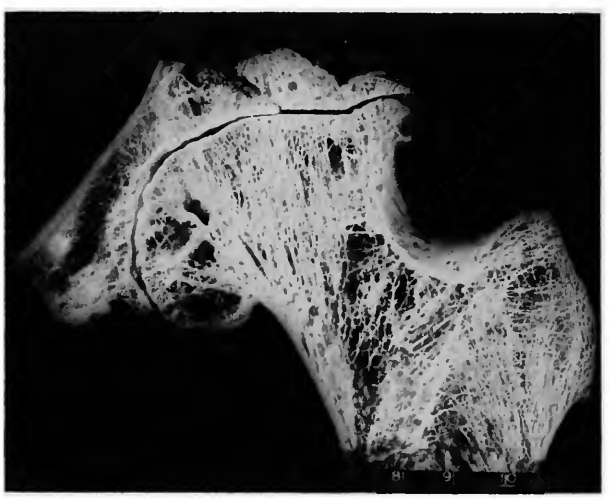


FIGURE 662.—Rheumatoid arthritis of hip with cartilaginous ankylosis. Notice minimal separation between acetabulum and femur and severe osteoporosis, especially of the unburdened portion of the femoral head. (73-year-old male, MGH autopsy 34283.)

FIGURE 663.—Rheumatoid arthritis of shoulder. Bisected proximal humerus shows large cyst and severe osteoporosis; 24 years duration, most joints involved. (59-year-old male, IPAZ autopsy 303 from 1960.)

The large joints and interphalangeal joints may undergo bony ankylosis, usually in flexed position, secondary to contracture and muscular imbalance.

Excessive bone resorption (mutilating arthritis), probably on a basis of neurovascular disturbance, is occasionally observed. It occurs most frequently on hands or feet, resulting in concentric atrophy of metacarpals, metatarsals, and phalanages. These extreme cases of bone resorption in themselves may be indistinguishable from those seen in leprosy.

Juvenile Rheumatoid Arthritis (Still's Disease)

Juvenile rheumatoid arthritis (Still's disease) usually begins in early childhood with a second peak before puberty. The predilection of the females over males for getting the juvenile form is less marked than in the adult form. The pathology is identical with that of the adult form, but there are some differences in location and type of involvement. The disease often begins in a knee or other large joint and occasionally may be mono-articular. In contrast to the adult form, the cervical spine is involved in about two-thirds of the cases (Ansell and Bywaters, 1963). Intervertebral diarthrodial joints and vertebral bodies may become fused. The atlantoaxial joint occasionally may become subluxed or fused.

Although wrists and ankles, carpal and tarsal joints are frequently involved, the other joints of hands and feet are more often free. If the interphalangeal joints are involved, the distal interphalangeal joints also participate in about 15 percent of the cases (Ansell and Bywaters, 1963). In contrast to adult rheumatoid arthritis, the hips are frequently diseased and the temporomandibular joint is usually spared. Subluxation of metacarpophalangeal joints is towards the radial side, but usually less marked than in the adult form. Ankylosis occurs more often in the carpal and tarsal joints than in large joints (Figure 664). Only the advanced and severe cases will show significant bone changes.

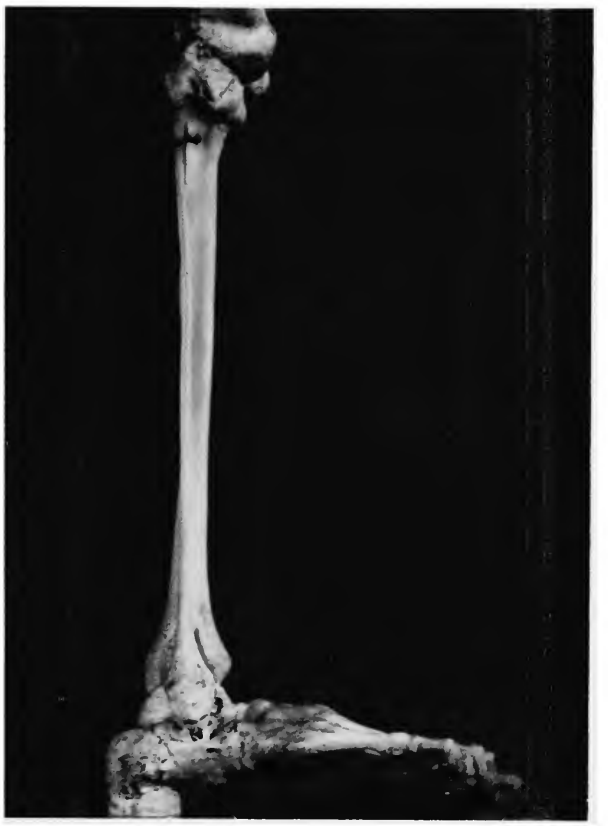


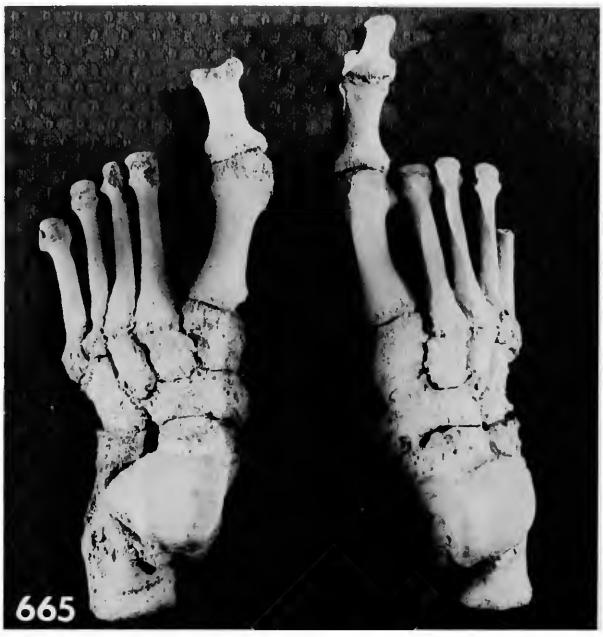
FIGURE 664.—Juvenile rheumatoid arthritis of lower right leg, with ankylosis of ankle, tarsal, and tarsometatarsal joints and extreme osteoporosis of disuse. Notice the translucency of the tibial cortex. Eight years duration, most joints affected. (18-year-old male, PMES 1SUT16(1) from 1838.)

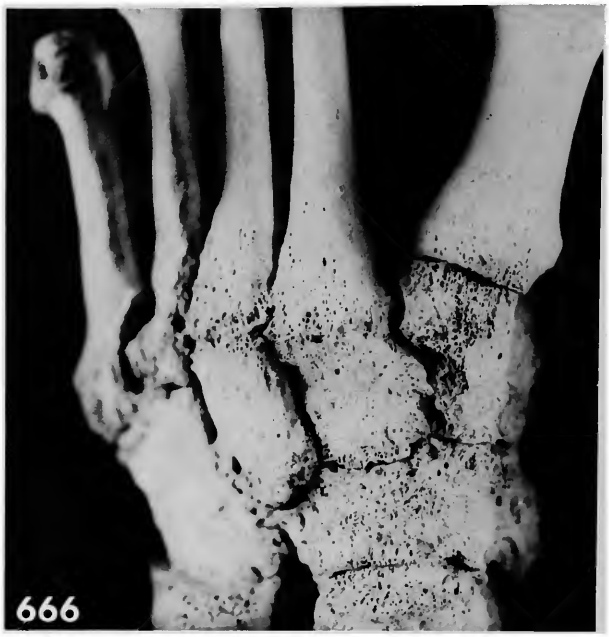
Psoriatic Arthritis

In psoriasis, a chronic disease of unknown etiology, arthritis similar to rheumatoid arthritis occurs. The disease involves and sometimes is limited to the distal interphalangeal joints of fingers and toes. In this condition osteoporosis is often absent, but mutilating arthritis of hands and feet is observed (Avila, Pugh, Slocumb, and Winkelmann, 1960).

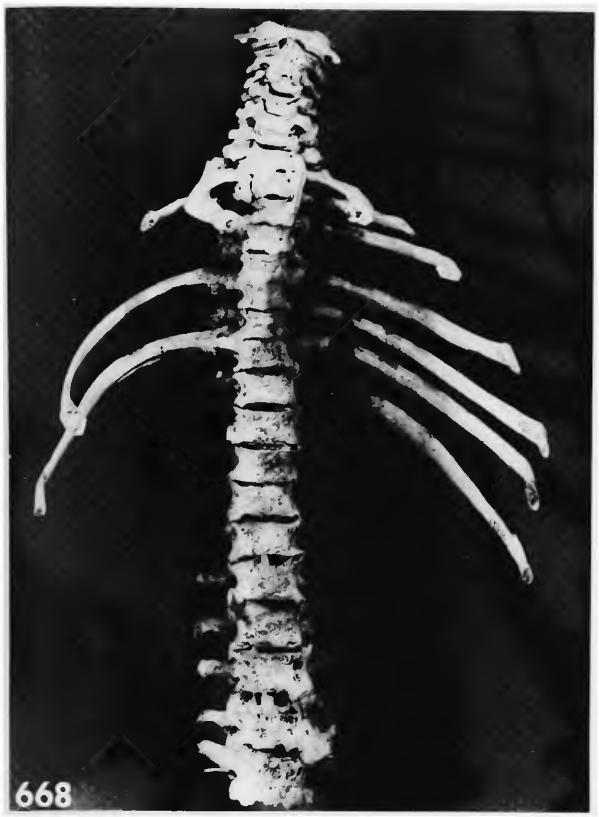
PALEOPATHOLOGY

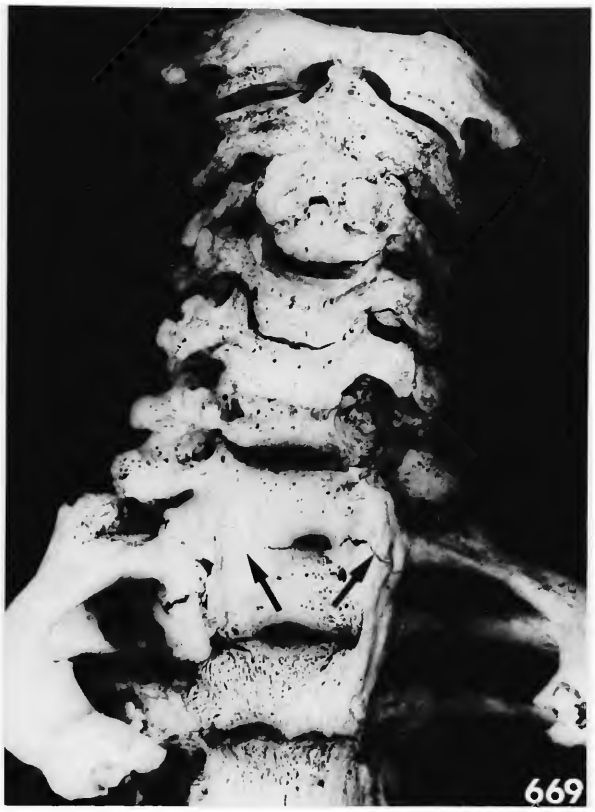
Because of the problems in differentiating rheumatoid arthritis from other types of arthritic con-





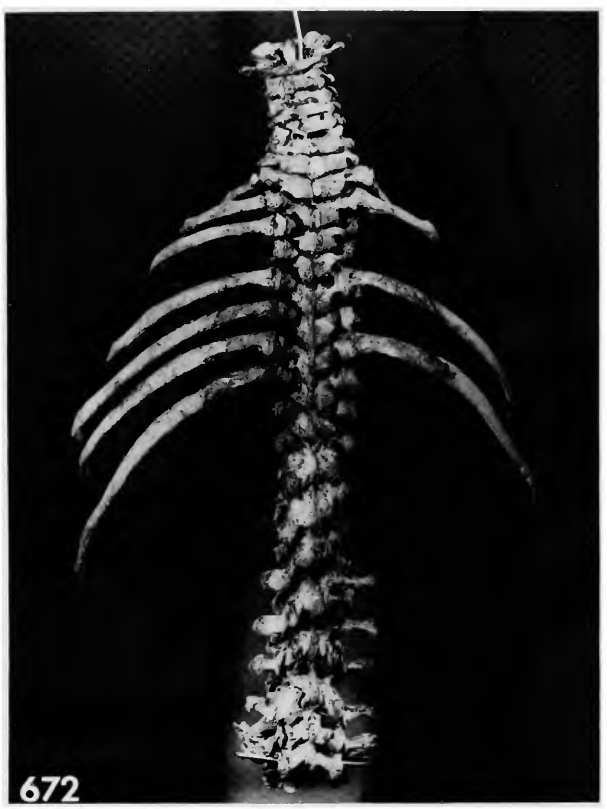
FIGURES 665-667.—Rheumatoid arthritis of the feet: 665, Superior view; note the porosity apparent in the tarsal bones. 666, Detail of the fusion between the tarsal and metatarsal bones of the left foot. 667, Detail of the bony fusion of the right great toe. (Adult specimen from the medieval cemetery at Winchester, England, BMNH XXIII, Burial 2.)

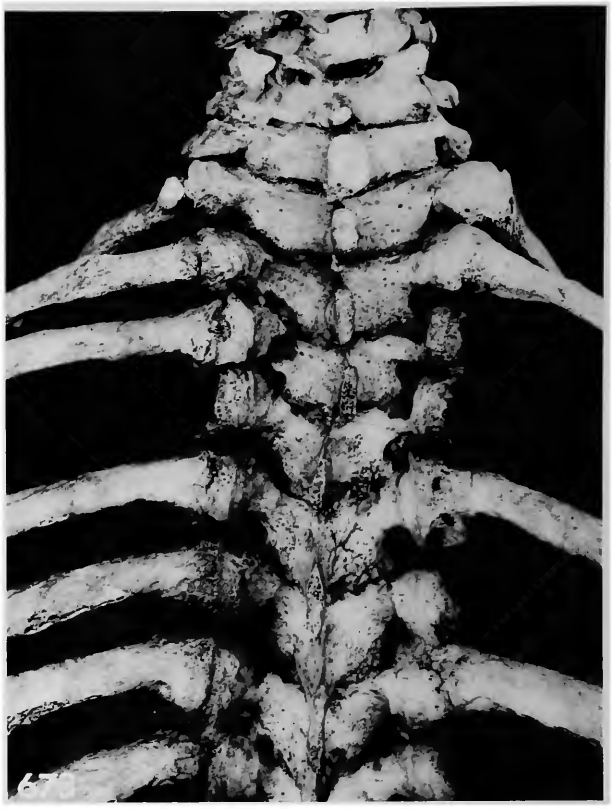




FIGURES 668, 669.—Probable rheumatoid arthritis: 668, Anterior view of the spine with fused ribs. 669, Detail of cervical and first two thoracic vertebrae; note the fusion of the first rib and the fusion of the seventh cervical and first thoracic vertebral bodies (arrows). (Adult male skeleton from Golovnin Bay, Alaska, USA, NMNH 346117.)

FIGURES 670, 671.—Probable rheumatoid arthritis: 670, Inferior aspect of third (right) and fourth (left) lumbar vertebrae; note the severe arthritic change of the diarthrodial joints. 671, Detail of rib fusion to thoracic vertebrae. (Adult male skeleton from Golovnin Bay, Alaska, USA, NMNH 346117.)





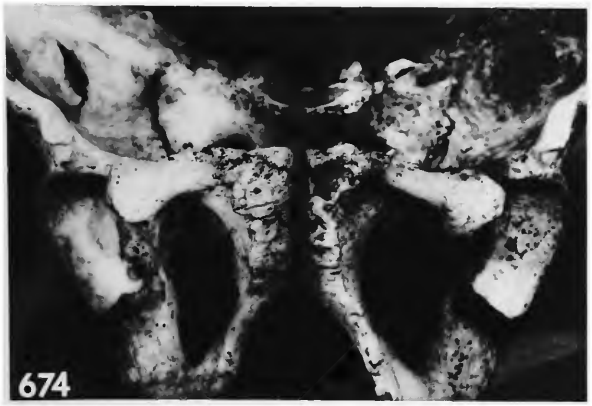
FIGURES 672, 673.—Probable rheumatoid arthritis: 672, Posterior view of spine. 673, Detail of joint fusion of ribs and thoracic vertebrae. (Adult male skeleton from Golovnin Bay, Alaska, USA, NMNH 346117.)

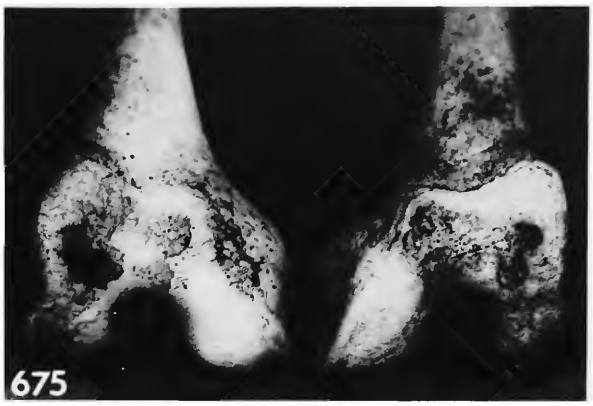
ditions in dry bone specimens, there are very few case descriptions of this type of arthritis in the literature on paleopathology. Indeed the evidence for rheumatoid arthritis in antiquity is so limited that Short (1974) suggests that this disease did not occur until fairly recent times with no convincing evidence before A.D. 1676.

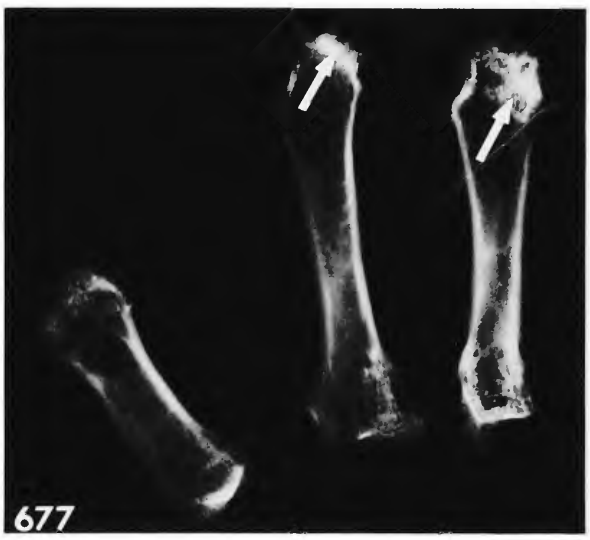
May (1897) describes a skeleton excavated by Sir Flinders Petrie from a tomb at Deshasheb, Egypt, dated to the Fifth Dynasty. The skeleton is from a male probably 50 to 60 years of age. There is considerable arthritic degeneration of the spine. The arthritic change affects with decreasing severity the cervical, thoracic, and lumbar vertebrae. The vertebral bodies are flattened, but the anterior margins bulge out with irregular lip-like projections. The plate published as part of May's report shows a deformity of the hands and feet compatible with a diagnosis of rheumatoid arthritis.

Hormell (1940) attributes arthritic changes in a skeleton from Lower Egypt, dated about 1300 B.C., to rheumatoid arthritis. Morse (1969:15, pls. 6E, 11A) reports a case of possible rheumatoid arthritis from the Dickson Mound in Illinois, USA. The skeleton is from an old male whose knees were buried in a partly flexed position. Although evidence of arthritic change in the skeleton is minimal, Morse argues that the flexed knee is the only example of this burial position at the site and that this may be evidence of rheumatoid deformity without marked bone changes.

A possible case of rheumatoid arthritis is found in the skeletal material excavated in Cathedral Green at Winchester, England (BMNH XXIII, Burial 2). This specimen is medieval in date and







FIGURES 674–677.—Probable rheumatoid arthritis: 674, Anterior view of pelvis; note the marginal osteophyte development and erosion in the region of the pubic symphysis. 675, Erosion and porosity of the distal joint of the femur. 676, Erosion and porosity of the distal joint of the second and third, right metacarpal bones. 677, X-ray of first, second, and third right metacarpals; note the subchondral joint sclerosis of the distal, second and third metacarpals (arrows). (Adult male skeleton from Golovnin Bay, Alaska, USA, NMNH 346117.)

consists of most of the bones of both feet. There is bilateral ankylosis of most tarsometatarsal joints accompanied by marked osteoporosis (Figures 665–666). The porosity associated with the joints is indicative of a chronic inflammatory condition. The interphalangeal joint of the right great toe is fused as well (Figure 667).

Another possible case of rheumatoid arthritis is a male Eskimo skeleton about 34 years of age (Figures 668–677) from Golovnin Bay, Alaska, USA (NMNH 346117). The specimen consists of most of the bones from the postcranial skeleton. The skull and mandible were not recovered. The archeological age is not known. The articular surfaces of the shoulder joints are intact. However, there has been some arthritic lipping and marginal erosion and porosity. There is some degenerative change on the articular surfaces of

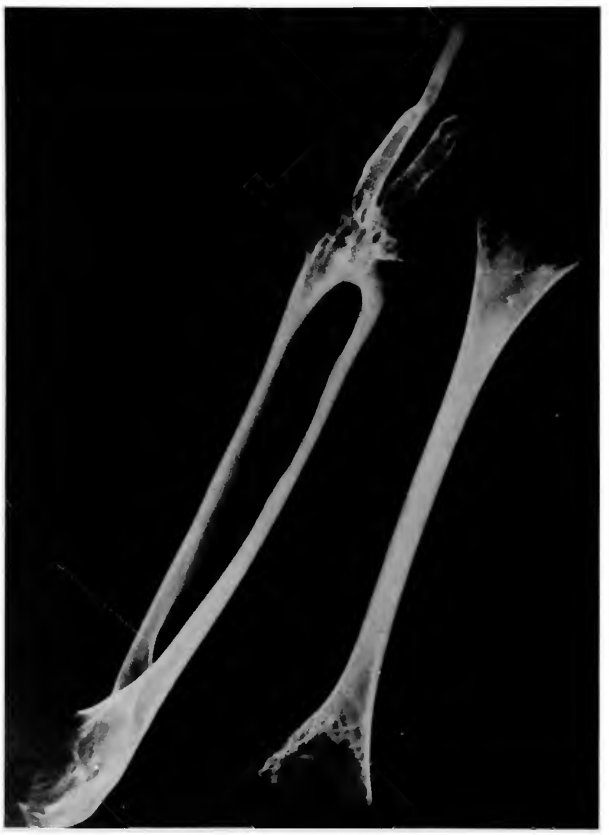


FIGURE 678.—X-ray of rheumatoid arthritis of the arm in an Indian skeleton from Puye, New Mexico, USA; skeleton is that of an adult male and is from the historic period. Note the flexion contracture of the hand and the lytic, expansive nature of the metaphyseal areas of the humerus. (X-ray courtesy Dr. Christy Turner,. Arizona State University, GQ 391.)

both elbows. This includes some porosity on the left side and considerable disruption of the articular surface of the right ulna. Marginal porosity is minimal. The radioulnar joint surfaces of the wrist exhibit some marginal lipping but are otherwise normal. The carpal bones are missing except the right and left capitate, left scaphoid and lunate, and the right triquetrum. Joint surfaces appear normal, but there is periarticular erosion on the left scaphoid. The first, second, third, and fifth metacarpals of the right hand were recovered as was the fifth metacarpal of the left hand. Only the distal joints of the second and third metacarpals show arthritic change with both having joint surface degeneration with porosity and development of marked peripheral osteoarthritis (Figures 676, 677). The clavicles are normal except for erosion of the right acromioclavicular joint surface. The normal complement of vertebrae are present, and all exhibit arthritic change of both the amphiarthrodial and diarthrodial joints. Arthritis is minimal on the cervicals and very marked on the thoracic and lumbar vertebrae (Figures 668–673). This pattern is atypical for rheumatoid arthritis. Many of the diarthrodial joints of the vertebrae and between the vertebrae and ribs are fused (Figures 669, 671). The vertebral bodies have remained intact although there is considerable development of marginal osteophytes and evidence of periosteal reactive bone particularly on the anterior surface of the body (Figure 670). In some vertebrae, adjacent osteophytes have fused. In those diarthrodial joints that can be inspected, there is considerable erosion and degeneration of the joint surface with extensive marginal lipping.

The bones of the pelvis are not fused. The sacroiliac joint surface has sparsely distributed pores about 1 millimeter in diameter. There is periarticular reactive bone, particularly at the superior margins of the joint. The pubic symphysis is normal, but there is evidence of some erosion and extensive osteophyte development on the anterior margin (Figure 674).

The joint surfaces of the hip show minimal arthritic change primarily in the right acetabulum where there is porosity on the anterosuperior surface. There is very little marginal lipping. Arthritic degeneration of the knee joint surfaces is limited to the femur and patella, although all joint components exhibit slight marginal lipping. The joint surface degeneration on the femur is largely restricted to the areas of articulation with the patella (Figure 675). Both surfaces show erosion and porosity with isolated areas of hypertrophic bone development. The joints of the ankles and feet are incomplete, but those which are present do not exhibit any evidence of articular surface degeneration and there is minimal marginal lipping.

Differential diagnosis of this case involves, in

addition to rheumatoid arthritis, ankylosing spondylitis and degenerative arthritis. The fusion of the diarthrodial joints of the spine would be very unlikely for degenerative arthritis. Differentiation from ankylosing spondylitis is much less certain. Favoring a diagnosis of rheumatoid arthritis would be peripheral joint involvement, lack of fusion of the sacroiliac joint, and the absence of any squaring off of the vertebral bodies. The distribution of arthritic change in the spine with minimal arthritis of the cervical vertebrae is more typical of ankylosing spondylitis. However, the overall pattern of arthritic change favors a diagnosis of rheumatoid arthritis.

From a Pueblo Indian site dated around A.D. 1559–1672 Steinbock (1976:305–309) describes a skeleton, which has partial ankylosis of the spine and many other joints, including some bones of the hands and fingers (Figure 678). Steinbock attributes this condition to ankylosing spondylitis, and this is certainly a possible diagnosis. However, the involvement of peripheral joints, including severe flexion contractures of the hands, is more typical of rheumatoid arthritis.

Ankylosing Spondylitis (Marie-Strümpell's Disease)

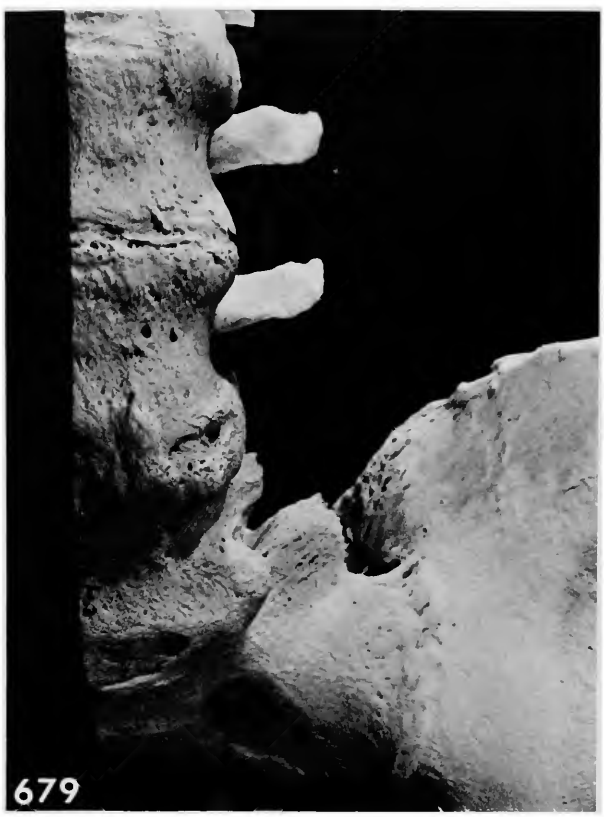
Pathology

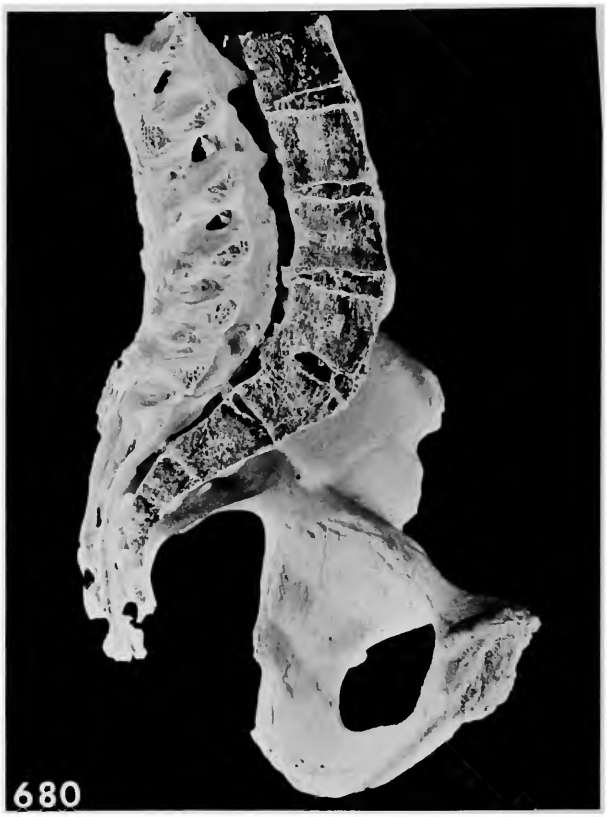
Ankylosing spondylitis is a progressive inflammatory disease of unknown etiology primarily affecting the diarthrodial joints of the spine, the costovertebral joints, and the sacroiliac joints. The pathologic changes in the active phase are practically identical with those of rheumatoid arthritis. The question of the relationship of the two diseases is still open but does not concern us here. The manifestations are sufficiently different to justify a separate discussion.

Ankylosing spondylitis prevailingly involves males, in contrast to rheumatoid arthritis. The male to female ratio is about 9:1. The disease usually begins in the second or third decade of life. In about half of the cases other joints, particularly hips or shoulders, are affected (Polley and Slocumb, 1947). Involvement of peripheral joints, if present, is indistinguishable from rheumatoid arthritis anatomically. The disease usually begins in the lumbar spine and in the sacroiliac joints and progressively ascends until the entire spine and all costovertebral joints are affected (Figure 679).

The disease develops as an inflammatory arthritis of the diarthrodial joints of the spine, which leads to early bony ankylosis, either limited to the periphery of the joints or across the entire facet. In the same type of process, the joints between the necks of the ribs and the transverse processes and those between the heads of the ribs and the vertebral bodies become ankylosed. In addition, the peripheral portion of the annulus fibrosus becomes ossified, bridging the inner portions of the intervertebral discs, which at first are not affected, but later bony replacement of the nucleus pulposus does occur. The vertebral bodies lose their concave flare on the outer circumference, resulting in the so-called squaring of their contour in lateral view. This process of fusion of the spinal segments is accompanied by pronounced osteoporosis of the vertebral bodies (Figure 680). The result of this ankylosing process is the rigid, so-called bamboo spine with loss of its physiological curvatures. In late stages, also, the various ligaments attached to the spine ossify.

The changes at the sacroiliac joints are usually bilateral and are never absent if the lumbar spine is involved. They commence with irregular erosion of the articular surfaces accompanied by sclerotic condensation of the subchondral bone on both the sacral and the iliac sides of the joint. This process ultimately establishes trabecular continuity between ilium and sacrum and completely effaces the joint. After establishment of complete ankylosis the subchondral sclerosis disappears (Schinz, Baensch, Friedl, and Uehlinger, 1951–1952(2):1514). Erosive changes also occur on the symphysial facet of the pubis, but ankylosis of the symphysis pubis is uncommon. If peripheral joints are involved in this disease, early bony ankylosis without marked bone destruction commonly occurs (Rutishauser and Jacqueline, 1959: 10–11). The atlantoaxial, the atlantooccipital, and occasionally the temporomandibular joint can also be ankylosed.

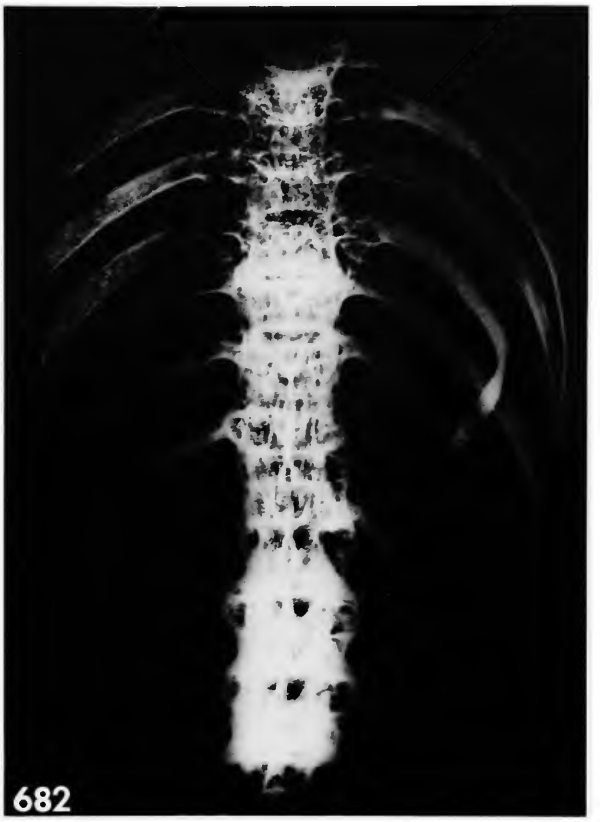




FIGURES 679, 680.—Ankylosing spondylitis: 679, Frontal view, showing smooth ankylosis of left sacroiliac joint and "bamboo spine." 680, Cut surface, showing ankylosis of the diarthrodial joints, ossification of interspinal ligaments and the anulus fibrosus, and severe osteoporosis of vertebral bodies. (67-year-old male, IPAZ autopsy 572 from 1963.)

PALEOPATHOLOGY

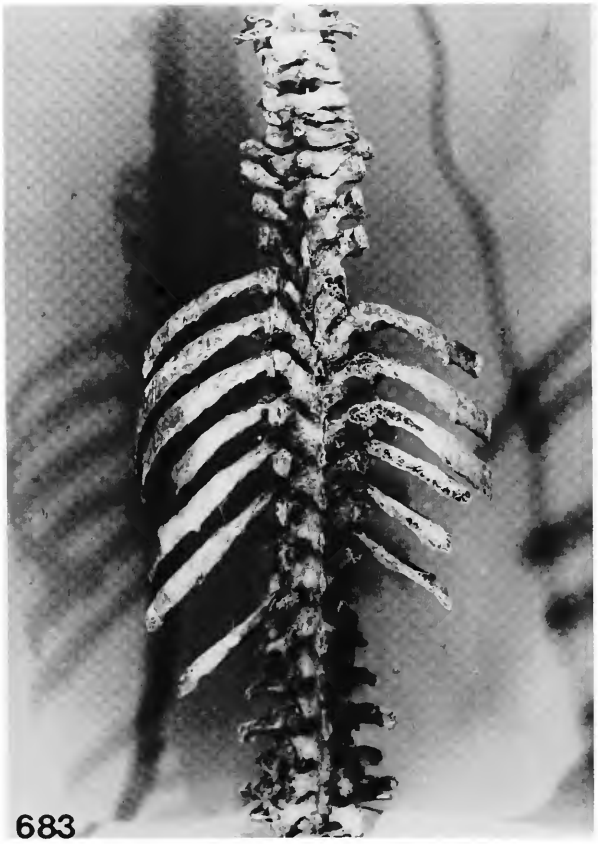
Ruffer and Rietti (1912) and Ruffer (1918b) describe a case of "spondylitis deformans" in an adult male from the Third Dynasty in Egypt. In this case the vertebrae are fused from the fourth cervical through the coccyx. Both the anterior and posterior longitudinal ligments were ossified. Wood-Jones (1910b:278) reports many cases of fusion of several vertebrae in ancient Nubian skeletons. Elliot-Smith (in Wood-Jones, 1908a:59) comments on an adult male from Nubia that showed complete bony ankylosis of the entire vertebral column. Pales (1930:117, pl. 24: figs. 1, 2) describes from the Neolithic period two isolated, fused thoracic vertebrae, which he feels were due to ankylosing spondylitis. Shore (1936) reviews several specimens from the pre-Dynastic site of Hierakonpolis, Egypt. All the specimens had at least partial ankylosis of the vertebral column. He attributes the abnormality to infection. Bourke (1967:357–360) describes three possible cases of ankylosing spondylitis from Egypt. Two of these are from the cemetery at Hou. One specimen from the B cemetery included the second cervical through the lumbar vertebrae. The diarthrodial joints and the spinal ligaments were ossified with smoothing of the anterior vertebral bodies. The second specimen is from cemetery Y and dated to the Twelfth Dynasty. It consists of six fused thoracic vertebrae. Ossification had occurred in the diarthrodial joints as well as spinal ligaments. The third case included all lumbar vertebrae and is from the cemetery at Nagada

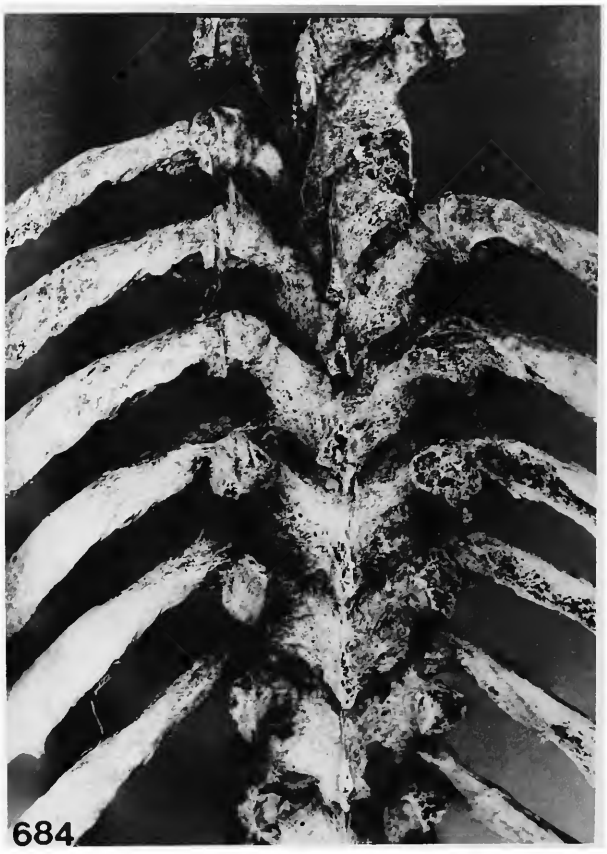


FIGURES 681, 682.—Ankylosing spondylitis of spine: 681, Anterior view of spine. 682, X-ray of anteroposterior view of fused vertebrae and ribs. (Adult male skeleton from Kuskokwim River, Alaska, USA, NMNH 351296.)

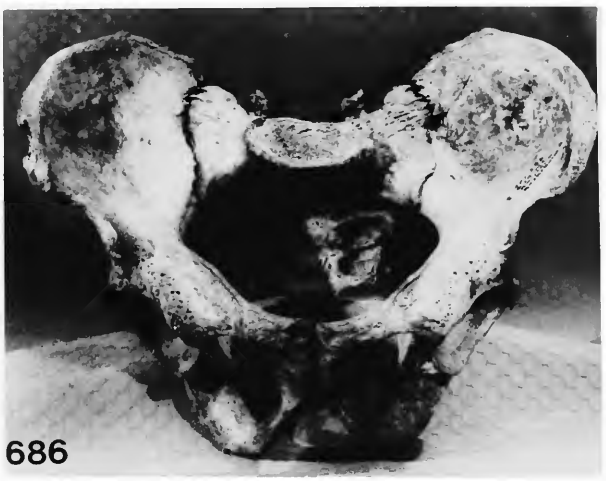
but currently stored at the British Museum (Natural History). Both the diarthrodial joints and amphiarthrodial joints were fused. Kidd (1954) reports the presence of ankylosing spondylitis in two skeletons from a post-Columbian, Huron cemetery in Ontario, Canada. Morse (1969, pl. 2 B, C) briefly describes two possible cases of ankylosing spondylitis from archeological sites in Illinois, USA. Bass, Gregg, and Provost (1974) report one case of ankylosing spondylitis in 2600 skeletons of Plains Indians from North and South Dakota, USA. Steinbock (1976:304–309) attributed the fusion of the spine in a Zuni skeleton from New Mexico, USA (NMNH 239208) to ankylosing spondylitis.

A probable example of ankylosing spondylitis is seen in the postcranial skeleton of an adult male Eskimo from Alaska, USA (NMNH 351296). On the skull there is some arthritic degeneration of the occipital condyles with marginal lipping and porosity of the left surface. The temporomandibular joints are normal. The sixth cervical vertebra is missing. The diarthrodial joints of the second and third cervical are fused. There is some erosive degeneration of the atlantooccipital joint surface and some marginal lipping of all the cervical diarthrodial joint surfaces, but this is minimal. The first thoracic vertebra (T1) is unfused. T2 through T4 are fused and may have been fused to T5 although postmortem damage makes this uncertain. T5 through the fourth lumbar vertebrae were fused during life. Interlocking osteophytes between L4 and L5 would have limited the movement of L5. The





FIGURES 683, 684.—Ankylosing spondylitis of spine: 683, Posterior view of spine. 684, Detail of diarthrodial joint fusion of thoracic vertebrae and ribs. (Adult male skeleton from Kuskokwim River, Alaska, USA, NMNH 351296.)



FIGURES 685, 686.—Ankylosing spondylitis of spine and pelvis: 685, Right lateral view of spine; note smooth anterior surface of fused vertebral bodies. 686, Anterior view of pelvis; note the fusion and osteophyte development of the sacroiliac joint. (Adult male skeleton from Kuskokwim River, Alaska, USA, NMNH 351296.)

FIGURES 687, 688.—Anklylosing spondylitis of spine: 687, Detail of rib and vertebral body fusion; note smooth anterior surface of vertebral bodies. 688, X-ray of lateral view of fused vertebrae; note the "squaring off" of the vertebral bodies. (Adult male skeleton from Kuskokwim River, Alaska, USA, NMNH 351296.)

fifth through twelfth ribs appear to have been fused to the vertebrae during life, although postmortem damage makes this uncertain in some vertebral joints. Generally the diarthrodial and amphiarthrodial joints were fused (Figures 681–684). There has been remodeling of the thoracic vertebral bodies producing the square profile characteristic of ankylosing spondylitis (Figures 685, 687, 688). The sacroiliac joints are fused across the joint surfaces (Figure 686); there is also marginal lipping which, on the left anterosuperior margin, has fused. In this specimen the characteristics of the fusion of the pelvis, spinal column, and ribs and the lack of peripheral joint involvement make the diagnosis of ankylosing spondylitis highly probable.

Metabolic Arthritis

PATHOLOGY

Gouty Arthritis

Gout is a disease due to a disturbance of the purine metabolism characterized by accumulation of its end product—sodium urate—in the body. Gout seldom manifests itself before 40 years of age and shows such a strong sex predilection that 9 out of 10 patients are male. Gout is rare in African Negroes, which may be due to a dietary rather than racial difference (Shepherd-Wilson and Gelfand, 1962). Of the many manifestations of this disease, I am here only concerned with

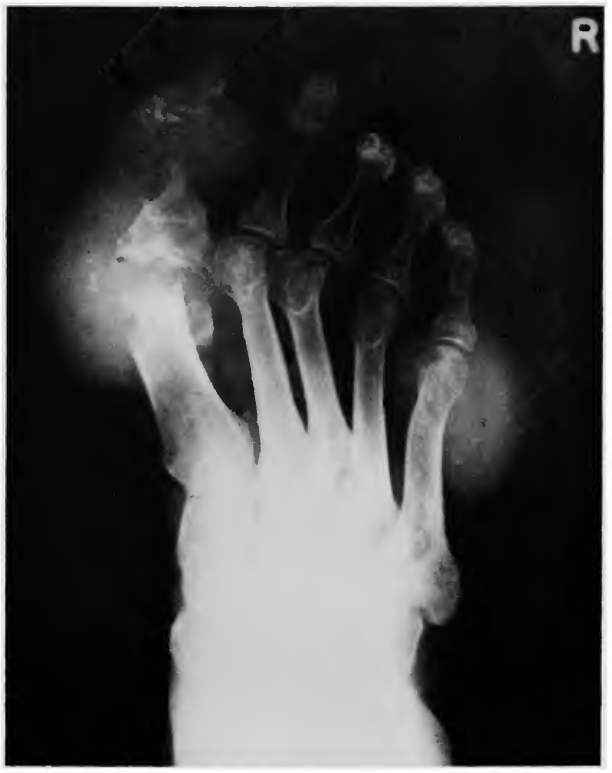


FIGURE 689.—Gout. X-ray of right foot showing severe involvement of the first metatarsophalangeal and interphalangeal joint. The scooped-out subperiosteal defects on both sides of the joint are characteristic. (Studied by Putschar; no data.)

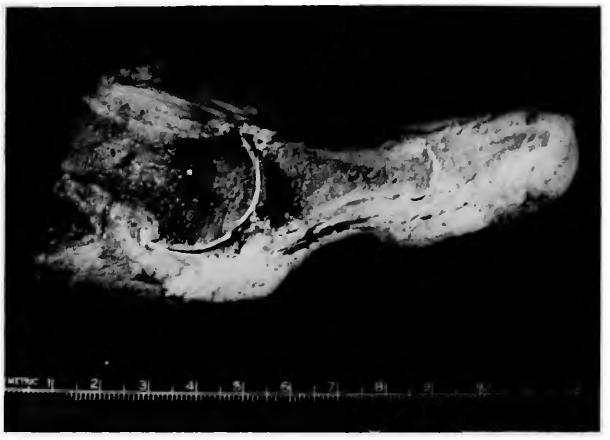


FIGURE 690.—Gout. Longitudinal cut of first toe, showing para-articular deposits of urates (white) extending into metatarsal and phalanges. (83-year-old male, MGH autopsy 33972 from 1970.)

chronic gouty arthritis in its advanced stages, mostly affecting old individuals.

In the course of chronic gout, sodium urate crystals are deposited in various tissues, particularly cartilage, para-articular connective tissues, and bone marrow. These gross deposits elicit an inflammatory foreign body reaction, resulting in the formation of circumscribed nodules filled with the crystals (tophi). The crystal deposition in the cartilage leads to early degeneration and ushers in secondary degenerative arthritis. Gouty arthritis affects mostly joints of the extremities rather than those of the trunk. The most common location is the metatarsophalangeal joint of the great toe. Generally, small joints are more often involved than big ones and the lower extremities more often than the upper. With the loss of soft tissue, the crystal deposits are also lost so that only bone changes of advanced gouty arthritis in typical skeletal locations could suggest the diagnosis in dry bone material. The most characteristic change is the pressure erosion due to para-articular tophi on metatarsophalangeal, metacarpophalangeal, and interphalangeal joints of toes and fingers. These scooped-out defects are located near the margin of the articular surface and often involve both bones forming the joint (Figure 689). They bulge into the bone but usually remain separated from the marrow cavity by a thin layer of bone (Figure 690). Intraosseous tophi may manifest as lytic lesions, expanding the small bones of hands and feet. These lesions are usually multiple and may simulate, in dry bone, the findings of enchondromatosis, predilecting the same skeletal areas. The main difference would be that, over large intraosseous tophi, the subchondral plate would be at least partly destroyed, while over an enchondroma it would be intact.

Ochronosis (Alkaptonuria)

Ochronosis is due to an inborn gene defect resulting in absence or deficiency of the enzyme homogentisic acid oxidase. Homogentisic acid is an intermediate product in the metabolism of the amino acids phenylalanine and tyrosine. In members of affected families homogentisic acid, form-

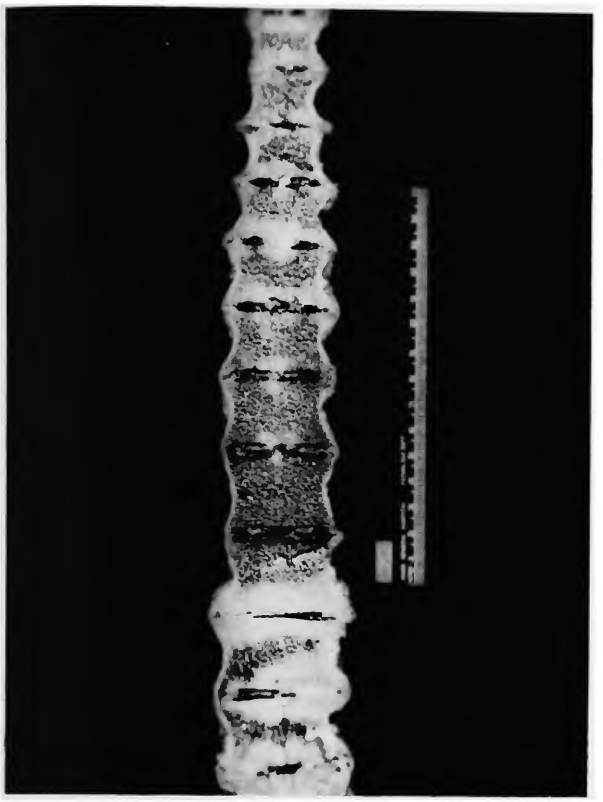


FIGURE 691.—Ochronosis of vertebral column. The affected intervertebral discs are narrowed and discolored black. There is marked osteosclerosis and bony bridging of some segments. (MGH autopsy 29047 from 1965.)

ing a black melanin-like pigment, accumulates in various tissues of the body, especially in cartilage. The lifelong accumulation of homogentisic acid damages the cartilage of the intervertebral discs and of the diarthrodial joints. The cartilage becomes brittle and undergoes mechanical attrition and degeneration. In the spine, the intervertebral disc spaces are narrowed and peripheral osteophytic bridging is common (Figure 691). Not unusually, ossification of the nucleus pulposus with bony union of the two adjacent vertebrae occurs. Complete ankylosis across the destroyed intervertebral disc is sometimes also seen. In the affected cases, all intervertebral discs are involved, resulting, finally, in a straight or kyphotic rigid spine, as in the late phase of ankylosing spondylitis. The radiologic finding of focally increased density is mainly due to ossification. In the large diarthrodial joints (knee, hip, and shoulder) ochronosis leads to earlier and more severe degenerative arthritis. On dry skeletal material, the diagnosis of ochronosis could only be suspected. The main reason for discussing this entity here is that purported cases of ochronosis have been observed in Egyptian mummies where cartilage tissue was, at least in part, preserved.

Hemophilic Arthropathy

Hemophilia is a heritable sex-linked disorder of the blood-clotting mechanism, which is transmitted as a recessive Mendelian trait and manifests itself only in males. Until recently, in the great majority of cases, repeated intra-articular hemorrhages, precipitated by minor trauma, led ultimately to severe joint disease. Although intra-articular hemorrhages begin in childhood, demonstrable bone changes do not develop until adolescence or adult life and increase with the age of the individual.

Thomas (1936) reported that in a series of 98 hemophiliac males 61 percent showed permanent joint deformity. The most commonly affected joint was the knee (68 percent) followed by the ankle (56 percent), elbow (53 percent) and hip (16 percent), with less frequency of other and especially of small joints. The changes often involve more than one joint and, especially in the knee, may be bilateral.

Repeated articular hematomas and their subsequent resorption and organization lead to marked synovial proliferation and hemosiderotic fibrosis and to damage of the cartilage. The underlying bone is particularly eroded at the margins of the articular cartilage by undermining ingrowth of fibrous tissue. In this manner erosion and widening of the intercondylar notch of the distal femur occurs, which is fairly characteristic for this condition (Schinz, Baensch, Friedl and Uehlinger 1951–1952(2):1427). In the elbow, the trochlea of the ulna is first and often most severely affected. In advanced cases extensive destruction of subchondral bone with alternating porotic and sclerotic areas ensues. Multiple and large sub-

chondral cysts are often present, especially in the distal femur and proximal tibia. Femoral condyles and corresponding portions of the tibial plateau can be extensively destroyed. Large pressure erosions through the cortex can occur near the capsular attachment. Severe grooving and eburnation of secondary osteoarthritis is often absent due to limited motion in the severely altered joint. For the same reason the irregularities in the level of the joint surface are more marked than in ordinary degenerative arthritis. In some cases limited mobility leads to bony ankylosis, especially between patella and femur and between femur and tibia. Early changes may be indistinguishable from tuberculous arthritis in dry bone because of the undermining marginal erosion common to both.

Hemophilic Pseudotumor

In some cases of hemophilia, massive subperiosteal or medullary hemorrhages occur. These subperiosteal or intraosseous hematomas may create large pressure defects and reactive bony shells simulating neoplasm. Such pseudotumors may be found on the diaphysis or near the joint. Pathological fractures through such lesions do occur (Ghormley and Clegg, 1948). Medullary hematomas may also result in formation of large metaphysial and diaphysial cysts (Stiris, 1958).

Paleopathology

Gouty Arthritis

A possible case of gout has been reported by Elliot-Smith and Dawson (1924:157–158) in an old male mummy from a Christian cemetery near the temple of Philae, Egypt. Many of the joints of the extremities have white concretions. The most noticeable of these is seen on the metatarsal bones of the great toe. One of these concretions measured 23 by 10 by 5 millimeters. The upper extremity was less affected than the lower. Analysis of the concretion produced a reaction typical of uric acid. Rowling (1961:414) has reconfirmed this analysis.

More recently Wells (1973) has described a male skeleton from a Roman cemetery (A.D. 150) at Gloucester, England. There were lytic lesions in the bone adjacent to the joints of the ankles and feet, suggestive of the type of bone reaction that could occur in long-standing chronic gout.

Ochronosis (Alkaptonuria)

Simon and Zorab (1961) examined one young female Egyptian mummy from the Roman Period before 30 B.C. The X-ray films of the mummy show radiodensities of the intervertebral discs and end-plate cartilage between all vertebrae. Corresponding X-ray films of other mummies did not show this feature. On the basis of similar radiologic findings in known cases of ochronosis, the authors suggest that the mummy may have had ochronosis during life.

Wells and Maxwell (1962) describe a possible case of ochronosis in another female Egyptian mummy. This specimen is dated to the Twenty-sixth Dynasty and exhibits dense shadows in the region of the lumbar discs with narrow, translucent zones adjacent to the dense zones. These roentgen features are seen in modern clinical cases of ochronosis, and the authors conclude that this disease was present in the mummy specimen. They also prepared histological sections of affected tissue but found no satisfactory microscopic support for their conclusion.

The major unresolved problem in these reports is that shrinkage of cartilage occurs during desiccation of the mummy, and as cartilage discs shrink they become more radiodense. The variation in radiodensity between mummies reported by Simon and Zorab (1961) may reflect different conditions of desiccation and preservation.

Stenn, Milgram, Lee, Weigand, and Veis (1977) report another possible case of ochronosis in Egyptian mummy material. The radiologic appearance of the spine suggests mineralization of the intervertebral discs. Furthermore, samples of the discs had zones of black pigment suggestive of local concentrations of homogentisic acid. Chemical analysis of this dark substance supports this suggestion.

Cartilage in naturally desiccated human mummies is typically very dark or black. This indicates that chemical changes in cartilage through time naturally produce dark-pigmented substances. Whether such cartilage would produce an analytical pattern similar to cartilage containing homogentisic acid has not been demonstrated. Until this type of experiment has been conducted and the effect of other variables on mummy tissue considered, it would appear prudent to defer final judgment on the presence of ochronosis in antiquity.

Degenerative Arthritis

Pathology

Degenerative arthritis is the most common of all articular diseases. As the term "degenerative" indicates, it is not an inflammatory disease but develops on the basis of aging changes and degeneration of articular cartilage. There is no sharp borderline between aging phenomena and incipient degenerative arthritis. The condition is slowly progressive, and demonstrable bone changes are preceded by many years of alterations of the articular cartilage alone. In addition to this primary degenerative arthritis, intimately associated with aging and physiological wear and tear, secondary degenerative arthritis develops early in structurally or functionally abnormal joints. Thus, degenerative arthritis becomes, in part, the end-stage of various inflammatory, traumatic, metabolic, and congenital or acquired joint diseases, provided that joint function continues under altered conditions for a long time.

The Appendicular Skeleton

The joint earliest and ultimately most often involved is the knee, followed in decreasing order of frequency by the first metatarsophalangeal joint, the hip, the shoulder, the elbow, the acromioclavicular joint, and the sternoclavicular joint. These statistical data, presented in Table 12, are taken from a study of Heine (1926), in which the above-listed joints were examined in 1000 autopsies of individuals above the age of 15 years. This does not mean that other joints not accessible at autopsy or not included in this study do not also considerably contribute to the total picture of degenerative arthritis. Heine's data indicate that a significant degree of degenerative arthritis, which may be accompanied by bone changes, does not appear before the fourth decade and increases with age after that time. Since, for our purpose here, only the advanced lesions showing significant bone changes are of interest, the life span of an archeological population will greatly influence the recognition and frequency of this condition in skeletal material.

Table 12.—Incidence of moderate to severe degenerative arthritis, stated in percentage of incidence (sample: 1000 autopsies of individuals above 15 years of age; modified after Heine, 1926)

Age by decade	*Knee*	*Great toe*	*Hip*	*Shoulder*	*Elbow*	*Acromioclavicular*	*Sternoclavicular*
4th	1.0	0	0	0	0	0	0
5th	0.8	0	0.8	0	0	0.8	0
6th	2.6	7.9	0.7	0.7	1.3	1.3	0
7th	12.0	13.7	2.7	2.6	5.2	9.6	1.1
8th	33.3	18.2	12.2	9.7	10.5	12.3	1.5
9th	39.4	24.6	16.7	15.7	15.5	11.4	7.0

In the knee joint, the articular changes begin in the patellar cartilage followed by lesions on the patellar facet of the femur, then on the tibial condyles not protected by the menisci, on the anterior portion of the medial condyle and, lastly, on the anterior portion of the lateral condyle of the femur. The posterior portions of the condyles are usually spared (Bennett, Waine, and Bauer, 1942). This pattern follows the locations of maximal pressure and friction, stressing the mechanical factors in the causation of degenerative arthritis.

Significant bone changes usually develop many years after the initiation of degenerative and proliferative cartilage changes. These usually begin at the margin of the articular cartilage and manifest themselves as marginal lipping and ex-

ostoses. It is realized that marginal lipping can represent a feature of joint remodeling without degenerative cartilage changes. However, for our purpose in the study of dry bone, it has to be included in the manifestations of degenerative joint disease. Admittedly there is no sharp line of separation between aging changes and degenerative joint disease, but severe and irregular marginal lipping definitely represents a manifestation of the latter.

Focal or complete attrition of articular cartilage leads to a sclerotic osteoblastic response in the subchondral bone and to mechanical attrition and polishing of the exposed bony articular surface (eburnation). These polished areas are absolutely diagnostic of severe degenerative arthritis. Within and around eburnated areas smaller and larger pits occur on the joint surface leading into subchondral bone. In part, such defects connect with smaller and larger cystic spaces, which may be sizeable in the articulating bone ends. In ball and socket joints, such as the hip and shoulder, these processes lead to enlargement, altered curvature, and deformation, especially of the convex joint surface. The eburnation in these joints has a smoothly polished appearance resembling porcelain. In joints with a hinge-like action, such as the knee and elbow, eburnated areas may exhibit deep parallel grooves. In joints with little mobility, such as the diarthrodial joints of the spine, eburnation may be minimal or absent, while marginal lipping dominates the picture.

In addition to changes on and around the articular surface, subsynovial osteophytes, as on the neck of the femur or humerus, and osteophytes at the insertion of capsule and ligaments occur in advanced cases. Regardless of the severity of changes, ankylosis does not occur in degenerative arthritis. A brief discussion of the local pecularities of degenerative arthritis in different joints follows.

Hip.—The most severe changes are found on the femoral head. Eburnation and erosion are most marked on the upper surface. Marginal exostoses may completely overhang the femoral neck (mushroom deformity). Osteophytes occur on the intracapsular portion of the neck. Large lytic cavities are not uncommon in the head. Acetabular changes mostly consist of marginal lipping and deepening, leading occasionally to protrusion into the pelvis. The acetabular roof is often sclerosed and may also contain cystic cavities. Degenerative arthritis of the hip often is preceded by other disease processes in this joint, particularly Perthes' disease and slipped epiphysis.

Shoulder.—The changes are similar to those in the hip but usually less severe because of lack of weight-bearing.

Knee.—The dominant feature is marginal lipping, often most pronounced on the patella. Eburnation is usually less extensive than in the hip. Exaggerated medial condylar lipping is seen in genu varum deformity and on the lateral condyle in genu valgum. The tibial surface is usually less affected because of the protective effect of the menisci.

Elbow.—Eburnation occurs on all joint surfaces. Excavation is observed on the trochlea of the ulna. The radial head shows more overhanging lipping.

Big Toe.—The frequent severe involvement of the first metatarsophalangeal joint in Heine's (1926) series may, in part, reflect the effect of heeled shoes, particularly in women. However, even in a barefoot population the pressure transmission through the first metatarsal is great, resulting in eburnation and sclerosis, especially of the metatarsal head. Lateral deviation of the big toe (hallux valgus) leads to formation of a new joint facet of the lateral slope of the metatarsal head.

Fingers.—Degenerative arthritis usually involves the distal interphalangeal joints, characterized by marked marginal osteophytes (Heberden's nodes). This condition is usually multiarticular and bilateral. It is much more common in females than in males.

Spondylitis Deformans (Spinal Osteophytosis)

Degenerative changes leading to bony spur formations in the vicinity of intervertebral discs are exceedingly common in skeletal material past

middle age. Although these changes around synchondroses are not arthritic in the strict sense, they represent an equivalent to the changes on the diarthrodial joints and are best discussed in this connection. In the study of Schmorl and Junghanns (1971:186–187) on more than 4000 autopsied spines, some degree of spinal osteophytosis was present in 60 percent of females and 80 percent of males at the end of the fifth decade, increasing to almost 100 percent in the ninth decade. Any segment of the spine may be affected and the lesions usually involve several or many vertebrae. Minimal lesions begin to appear as early as the third decade.

The lesion typically consists of shelf-like bony protrusions on the cortex of the vertebral body. Arising at the junction of the body and the fused marginal epiphysis, the lesion points toward the adjacent intervertebral space. The pathogenesis of this lesion is not entirely clear. Schmorl (in Schmorl and Junghans, 1971:188–189) thinks that disruption of annulus fibrosus bundles inserting on the marginal epiphysis, followed by protrusion of disc material, are the causative factors. We need not concern ourselves here with the various other theories. Suffice it to say that degeneration of the intervertebral disc is not necessarily a precursor of osteophytic formation. These shelf-like osteophytes of two adjacent vertebrae approach each other and pinch off a peripheral portion of annulus fibrosus. Complete fusion of the osteophytes is not uncommon. On dry bone it becomes obvious that the shelf of newly formed bone is buttressed by ossification within the anterior spinal ligament and extends over the entire height of the vertebral body in severe cases (Figure 692). These osteophytes are quite characteristic and easily recognized on skeletal material. If the fusion of osteophytes is complete and of long standing, remodeling may give these bony bridges a smooth surface, resembling the changes observed in ankylosing spondylitis. However, the diarthrodial vertebral joints and the costovertebral joints are not fused in spondylitis deformans.

Degenerative and traumatic changes of the intervertebral disc may lead to herniation of disc cartilage through the annulus fibrosus anteriorly,

FIGURE 692.—Deforming spondylitis with massive bony bridges across intervertebral discs. (71-year-old male, IPAZ autopsy 1485 from 1957.)

laterally, or posteriorly or through the vertebral end-plate into the subchondral spaces of the vertebral body. Of these the latter are the most common and are the only ones that can be identified on dry bone.

Such extruded disc cartilage can proliferate to a limited extent, forming round or lobulated nodules up to one centimeter in diameter. Such disc protrusions are present in the great majority of spines of individuals past 40 years of age and increase in size and frequency with age. They do not occur in children and adolescents. These cartilage nodules are more frequently seen in the lower thoracic and lumbar vertebrae and occur most often in the center of the end-plate where the closure of the notochord canal leaves a weak spot (Schmorl, 1927; Putschar, 1927a, 1927b). In

dry bone they appear as round or oval defects in the bony end-plate bordered by a basket-like arrangement of reactive bone within the vertebral body. Corresponding lesions in two adjacent vertebrae bordering the same disc are common.

Paleopathology

The expression of degenerative arthritis in archeological material affects both joint surfaces and the articular margins of bone. Degenerative arthritis of joint surfaces may result in bone destruction and/or bony hypertrophy. Arthritic destruction is expressed as porosity, often with eburnation (polishing) of the disturbed bone surface. Arthritic hypertrophy is less common and is seen as an irregular thickening of the joint surface, which may be somewhat porous. Marginal degenerative arthritis is also expressed as a destructive and/or hypertrophic process although the latter, in contrast with joint-surface degenerative arthritis, is more common. In diarthrodial joints, hypertrophic, marginal, degenerative arthritis is usually seen as a fairly smooth extension of the joint surface. In amphiarthrodial joints of the vertebral bodies, hypertrophic arthritic change gives rise to osteophytes or finger-like projections from the bony periphery of the joint.

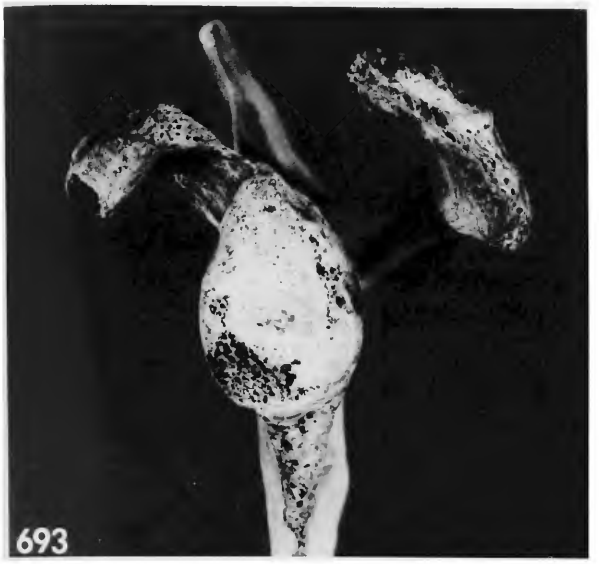
While several factors may contribute to the development of degenerative arthritis, stress undoubtedly is the most significant of these factors. Thus, as Ortner (1968:146) suggested, the distribution and severity of degenerative arthritis in the skeleton are likely to vary between societies and with different roles (e.g., male–female) within a society. Hrdlička's data (1914:62–69) indicates that the elbow joint was the most frequent site for degenerative arthritis in ancient Peruvian skeletal remains. The knee was next in severity (Table 13). This finding differs from modern populations where the knee is most frequently affected.

In a study comparing modern American Whites, Blacks, pre-Columbian Indians from New Mexico, and protohistoric Eskimos, Jurmain (1977) found the Eskimos to be most severely affected by degenerative arthritis. He concludes that their vigorous life style is the major causative factor in this difference. In an earlier study, Stewart (1947) found differences in the degree and location of vertebral degenerative arthritis between Eskimo, Pueblo Indian, and modern White skeletons. He suggested that cultural differences were important, but he also noted that racial variation in the structure of the vertebral column may affect patterns of movement of the vertebrae and thus affect the expression of spinal degenerative arthritis.

Table 13.—Frequency of degenerative arthritis in major long bones of adult archeological skeletons from the Chicama Valley, Peru (from Hrdlička, 1914:62–69)

Bones	*No. examined*	*No. arthritic*	*% of total*
Humeri	593		
Proximal		1	0.2
Distal		12	2.0
Ulnae	301		
Proximal		16	5.3
Distal		0	0
Radii	255		
Proximal		6	2.4
Distal		1	0.4
Femora	1210		
Proximal		16	1.3
Distal		36	3.0
Tibiae	781		
Proximal		12	1.5
Distal		0	0
Fibulae	266		
Proximal		1	0.4
Distal		2	0.8

Although stress is a major factor in degenerative arthritis, it is likely that different types of stress will affect the development of arthritis in different ways. It is here that the paleopathologist with access to the entire skeleton and joint surfaces, could contribute to the understanding of degenerative arthritis. For example, what is the effect of relatively continuous but moderate stress on a joint, as contrasted with intermittant but high stress? In such a problem comparisons of degenerative arthritis in joints of the left and right side and male versus female joints or skele-



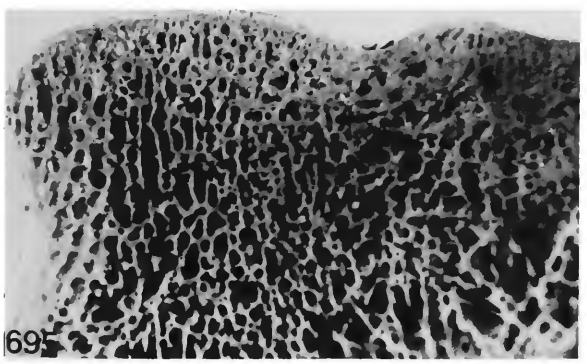
FIGURES 693, 694.—Degenerative arthritis of the shoulder and hip: 693, Lateral view of the left scapula showing porosity of the glenoid cavity. 694, Porous degeneration of the right hip joint. (Adult female Eskimo skeleton from Hooper Bay, Alaska, USA, NMNH 339115.)

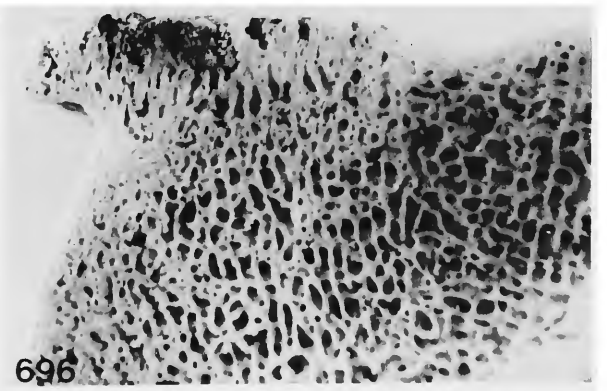
tons from different cultures where characteristics of body use are known or can be inferred might be used to clarify this and other problems.

Perhaps the biggest problem in the study of degenerative arthritis is the methodology for qualifying and, ideally, quantifying the observations. The degenerative conditions of bone destruction and hypertrophy need to be described in terms of location in the skeleton and in the joint and also with respect to the extent of involvement (Jurmain, 1975, 1977). In this context an important methodological consideration is the percentage of the total joint surface area affected by degenerative change.

The Appendicular Skeleton

Severe degenerative arthritis is seen in both the shoulder and hip joints (Figures 693, 694); however, its occurrence in the shoulder joint is uncommon in archeological skeletons. An example of what, in my experience, is moderate degenerative change is seen in the scapulae of an adult female Eskimo from Hooper Bay, Alaska, USA (NMNH 339115). The right scapula has a small area of porosity on the posterior margin of the glenoid fossa with a slight amount of marginal lipping.

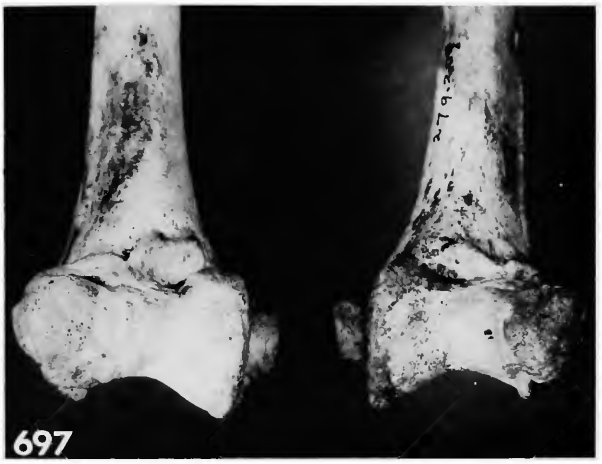


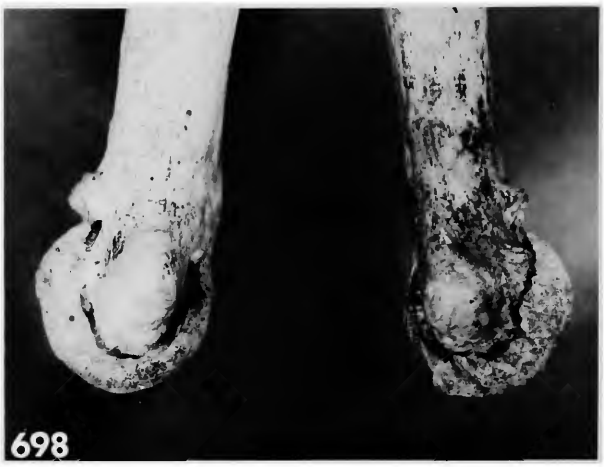


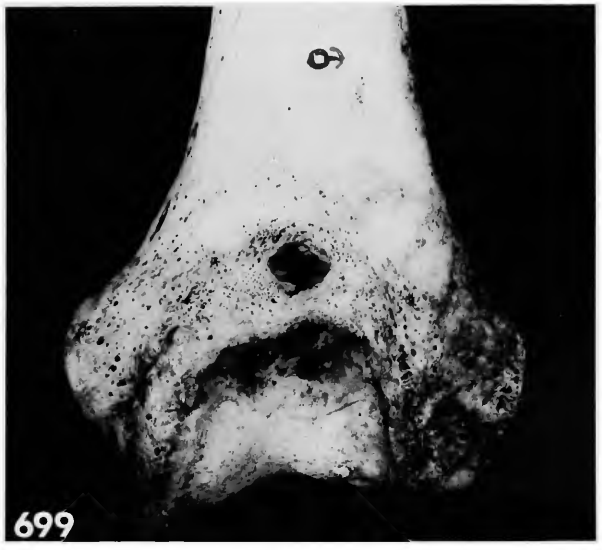
FIGURES 695, 696.—Subchondral bone in a section through the capitulum: 695, Normal joint surface and subchondral trabeculae. 696, Sclerosis of trabeculae underlying porous degenerative change of the capitular joint surface. (NMNH, unidentified humeri from the Huntington collection.)

The left scapula has considerable porosity on the anteroinferior surface of the glenoid fossa with a small area of porosity on the posterior margin (Figure 693). Marginal lipping is minimal.

In archeological skeletal remains the elbow is a common site of degenerative arthritis. Ortner (1968) discussed the anatomical and functional aspects of arthritic change in the elbow. Briefly, of the three related but distinct changes that occur, the first is a degenerative change of the joint surface, which is most common between the head of the radius and the capitulum. This is probably related to the fact that both flexion-extension and rotation are movements associated







FIGURES 697–700.—Degenerative arthritis of the elbow and wrist: 697, Anterior view of the distal humerus; note the porosity of the capitular surface and the bone hypertrophy of the capitular and trochlear fossae. 698, Marginal lipping adjacent to the trochlear joint surface. 699, Hypertrophic bone filling in the olecranon fossa. 700, Degenerative arthritis of the distal joints of the radius and ulna. (Adult male Eskimo skeleton from Golovnin Bay, Alaska, USA, NMNH 279209.)

with this joint surface. Porous degeneration of the joint stimulates sclerosis of subchondral trabeculae (Figures 695, 696). Second, there is marginal lipping primarily adjacent to the trochlear joint surface. Third, there is bony fill-in of the three fossae of the humeral component of the elbow.

An adult male Eskimo from a site on Golovnin Bay, Alaska, USA (NMNH 279209) exhibits the major features associated with degenerative arthritis of the humerus. Nearly the entire capitular joint surface is porous with slight traces of eburnation (Figure 697). Marginal lipping is most pronounced on the margins of the trochlear surface (Figure 698). There is considerable fill-in of both the capitular and trochlear fossae (Figure 697). This condition is more severe on the right side. Similarly the olecranon fossa has a large bar of hypertrophic bone filling in most of the depression (Figure 699). The hypertrophic bone in the fossae would have limited the amount of flexion and extension of the elbow.

The distal joint of the radius and ulna of the same specimen also shows marked degenerative arthritis (Figure 700). The joint between the ulna and radius exhibits marked marginal lipping as does the joint surface of the wrist. There is coarse porous degeneration, particularly on the ulnar surface, with some evidence of eburnation. It is possible that fracture may have stimulated some of the arthritic degeneration in the wrist. The bones of the hands are often lost in archeological skeletons. However, a common site for arthritic change in Eskimo skeletal specimens is the joint between the trapezium and the first metacarpal bone.

Arthritic degeneration of the hip is a fairly common condition in archeological specimens. Wells (1963a) and Pitt-Rivers (1965) are among many who have described typical cases. An example of moderate arthritic change is seen in the Eskimo skeleton also discussed below in the context of spinal degenerative arthritis (NMNH 339115). The right hip joint of this specimen exhibits marked porosity on the superior joint surface of the acetabulum with a trace of porosity on the peripheral margin of the remaining joint

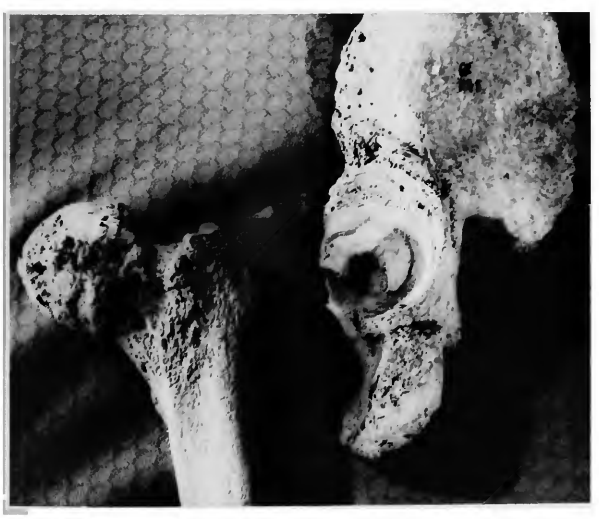


FIGURE 701.—Severe degenerative arthritis of the left hip. Note the abnormal contour of the femoral head. (HMCS GR 1248.)

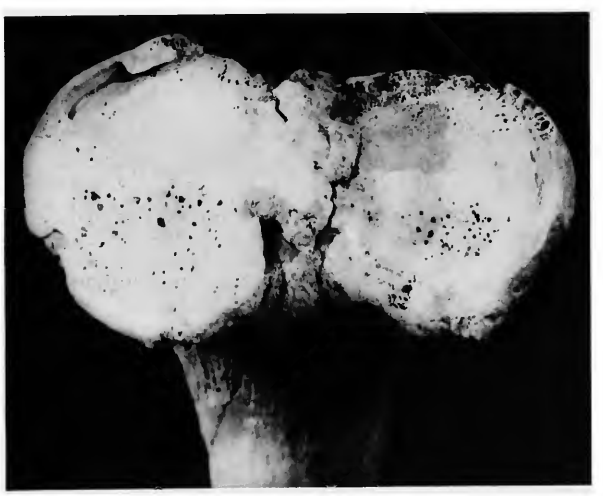
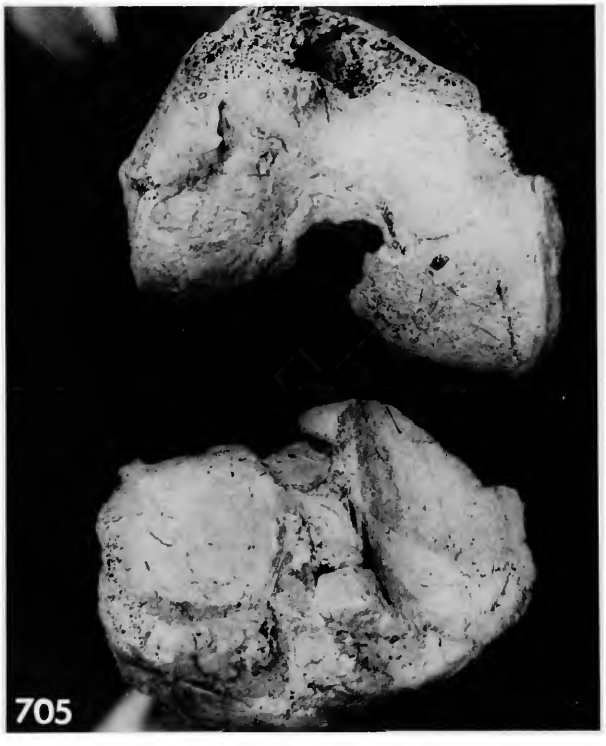
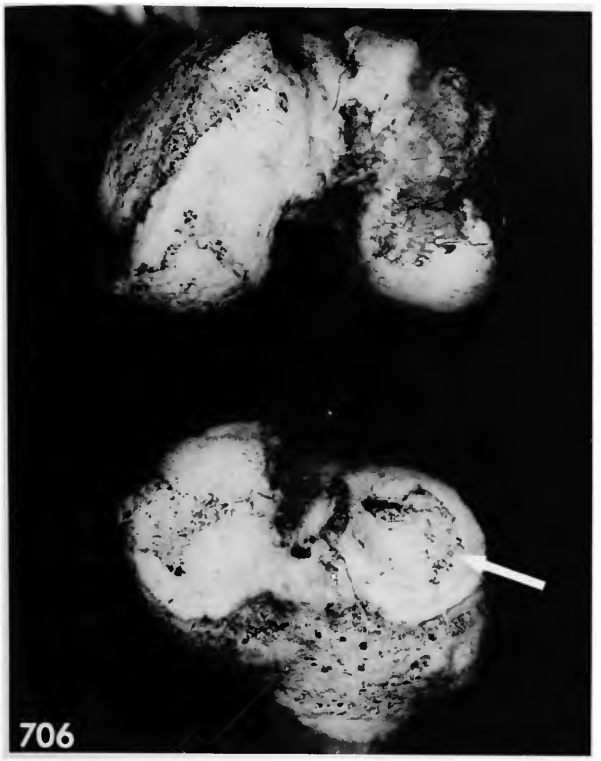
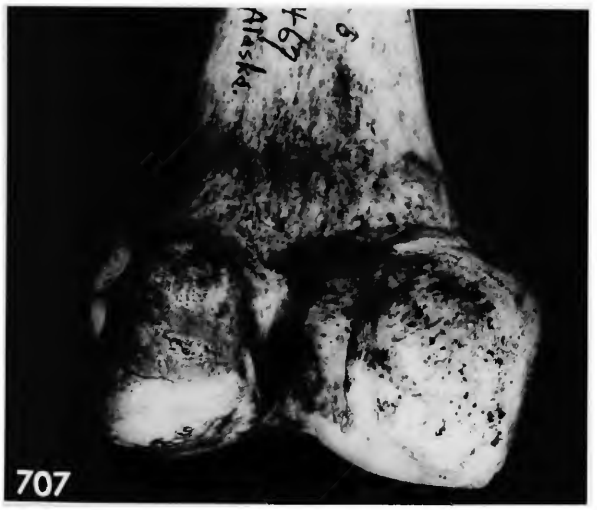


FIGURE 702.—Marginal lipping, porosity, and eburnation of the proximal joint of a right tibia. (Specimen from an archeological site in Paragonah, Utah, USA, NMNH 292012.)



FIGURES 703–705.—Degenerative arthritis of the spine and knee: 703, Anterior view of fused tenth thoracic through fifth lumbar vertebrae. Note the extreme development of hypertrophic bone on the right side. The diarthrodial joints are not fused. Both features are not associated with ankylosing spondylitis. The absence of hypertrophic bone on the left side is due to the overlying, descending aorta. 704, Right lateral view of fused vertebrae. 705, Bone surfaces of the right knee joint. Note erosion of the medial surface and the marginal lipping. Not apparent in the figure is extensive eburnation of the worn surfaces. (Specimen from an archeological site in Jersey County, Illinois, USA, NMNH 380071.)





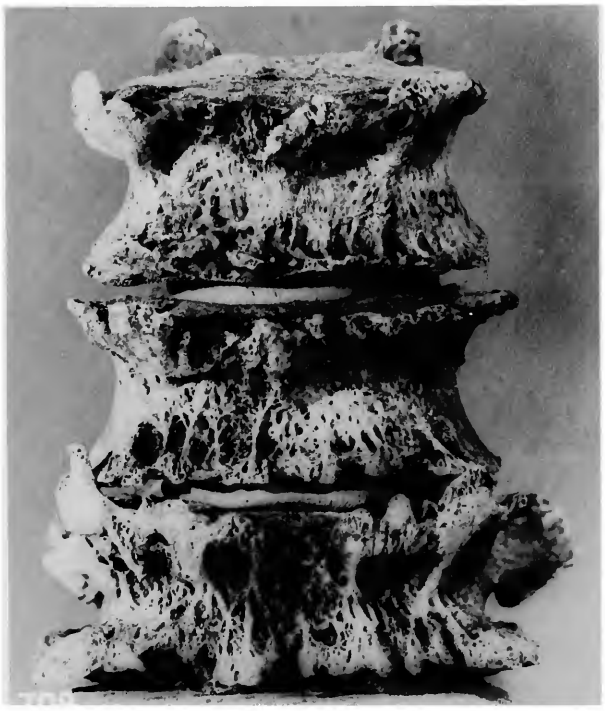
FIGURES 706, 707.—Porous, hypertrophic degenerative arthritis of the left knee: 706, Joint surfaces of the femur and tibia; note particularly the hypertrophic build-up of irregular bone on the lateral surface of the tibia (arrow). 707, Posterior view of femur, showing porous hypertrophic bone on the medial condyle. (Adult male Eskimo skeleton from an archeological site in Wales, Alaska, USA, NMNH 333467.)

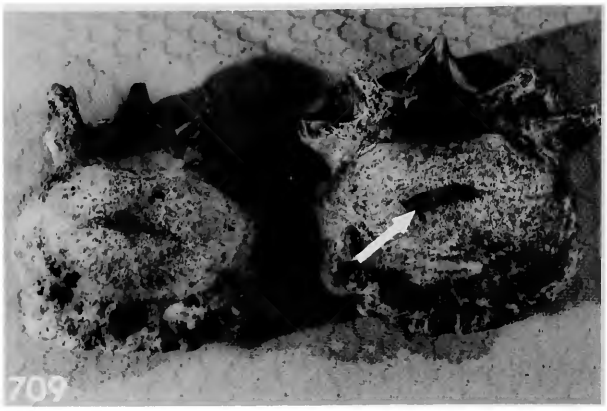
surface. There is a slight amount of marginal lipping. The posteroinferior surface of the right femoral head has an area of porosity about 1 by 1½ centimeters (Figure 694). A much more severe case of degenerative arthritis of the hip is seen in archeological specimen from the Historical Museum in Chur, Switzerland (HMCS GR 1248). The specimen is from the ancient cemetery at Bonaduz. The severe arthritic degeneration is associated with the left hip (Figure 701). The acetabulum shows severe porosity and eburnation with enlargement of the joint. There is marked marginal lipping superiorly and posteriorly. The left femur shows marked degeneration of the joint surface with eburnation and marginal lipping. The appearance of this case is very similar to a Peruvian specimen described by Hrdlička (1914: 60–61). He used the term "caput penis" to describe the rather distinctive morphology of this severe type of degenerative change of the femoral head.

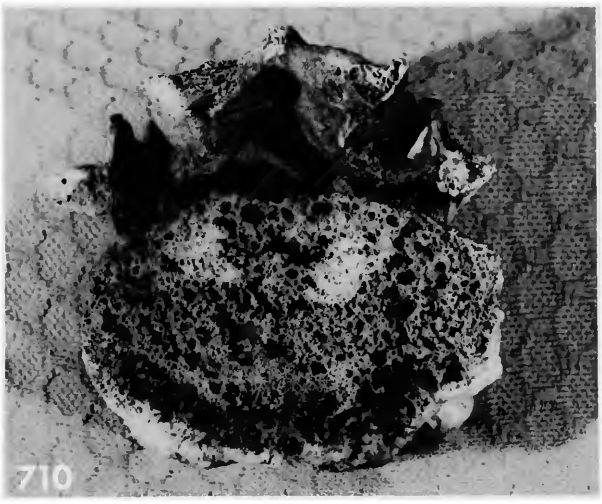
The incidence of degenerative arthritis of the knee is only slightly less than that of the elbow in archeological material. The condition is well known in the literature on paleopathology (e.g., Wells, 1965a). Three specimens serve to illustrate the significant features of degenerative change of the knee. The first of these is an isolated right tibia from Paragonah, Utah, USA (NMNH 292012). The posterior joint surfaces of both tibial condyles are eroded, porous, and eburnated (Figure 702). The degenerated surface is also grooved, which is indicative of long-standing bone-to-bone contact. There is well-developed marginal lipping, which has extended the joint surfaces particularly on the medial and posterior margins.

The second case is the right knee of a specimen from Jersey County, Illinois, USA. This skeleton (NMNH 380071) has a fused spine (p. 433). The medial condylar surface of the knee is eroded, porous, and eburnated with slight grooving (Figure 705). The lateral condyles are intact. There is very marked marginal lipping. The left knee is similarly affected by degenerative change.

The third specimen is from an adult male Eskimo from an archeological site in Wales,







FIGURES 708–711.—Degenerative arthritis of the third through fifth lumbar vertebrae and left and right first metatarsals: 708, Osteophyte development of vertebral bodies. 709, Superior aspect of the fourth (left) and fifth (right) lumbar vertebrae; note the erosive lesion suggestive of the presence of Schmorl's node in the fifth vertebral body (arrow). 710, Inferior view of the fifth lumbar vertebra; note the marked porosity of the vertebral body surface. 711, The distal articular surface of the right and left first metatarsal bones; note the porosity and eburnation associated with the central ridge of the joint surface. (Adult male Eskimo skeleton from an archeological site in Pastolik, Alaska, USA, NMNH 332556.)

Alaska, USA (NMNH 333467). Both knees are affected. The right knee exhibits erosion, porosity, and eburnation of the lateral condylar surfaces. The medial posterior portion of the femoral portion of the knee has a lytic porous focus with a small area of hypertrophic bone adjacent to the porosity. There is some marginal lipping. The left knee exhibits degenerative changes, which are somewhat different from those described earlier (p. 427). On the medial condyle of the femur, the arthritic change is characterized by a large area of irregular hypertrophic bone on the posterior portion (Figures 706, 707). There is some porosity, eburnation, and marginal lipping. The lateral condyle has a small area of irregular hypertrophic bone on the posterior portion. Marginal lipping is minimal. There is a defect between the condylar surfaces suggestive of an old well-healed fracture. On the tibia the medial condyle is eroded, porous, and eburnated with slight marginal lipping. On the lateral condyle, arthritic change is primarily a focus of irregular hypertrophic bone rising above the joint surface. On both condyles there is slight marginal lipping. The tibial portions of the ankle joints of this specimen also exhibit slight, porous degeneration of the joint surface. There is minimal marginal lipping. Arthritic degeneration of the ankle is uncommon in archeological specimens, even when there is marked degenerative change in other joints.

Bones of the feet commonly exhibit degenerative arthritis. The bones most often affected are those of the great toe. An example of this condition is seen on the distal joint surface of the right and left first metatarsal (Figure 711) from an Eskimo burial from Pastolik, Alaska, USA (NMNH 332556). The left metatarsal exhibits considerable porous erosion on the central ridge of the joint surface. There is minimal marginal lipping, although there is an erosive depression on the medial margin of the joint extending onto the adjacent cortex. On the right first metatarsal there is some porosity, but the major degenerative change is eburnation of the central ridge. There is also an erosive depression on the medial margin. The other metatarsals are normal.

The Axial Skeleton

SKULL AND MANDIBLE.—In the adult the skull has only two joints—the temporomandibular and atlantooccipital. The latter is most appropriately considered with the vertebral column. Degenerative arthritis of the temporomandibular joint has received little attention in the literature on paleopathology (see reviews of by Pales, 1930: 165–173, and Alexandersen, 1967:586–590). Given the hard use of the jaw often thought to be associated with archeological skeletal populations it is surprising that the frequency of arthritic change in the temporomandibular joint is not greater. As with other joints, degenerative change in this joint may involve porosity, eburnation, and hypertrophy of the joint surface. Marginal lipping is not common.

A fairly typical expression of temporomandibular degenerative arthritis is seen in an adult female skull from the Nushagak Region in Alaska, USA (NMNH 363530). Both the glenoid fossa and the mandibular condyles are involved (Figure 712) but the most severe change is on the glenoid fossa. There the porous lesions are restricted to the anterior edge of the fossa and are

FIGURE 712.—Degenerative arthritis of the left temporomandibular joint. (Adult female Eskimo skull from an archeological site in the Nushagak Region, Alaska, USA, NMNH 363530.)

characterized by erosion and porosity of the surface.

VERTEBRAL COLUMN.—Wood-Jones (1910b: 277) observed that virtually all adult ancient Nubian skeletons showed some evidence of degenerative arthritis of the spine. A similar generalization would apply to virtually any archeological skeletal sample. However, the frequency, age of onset, severity, and distribution within the spine varies in different populations (Snorrason, 1942; Stewart, 1947; Zorab, 1961; Chapman, 1965).

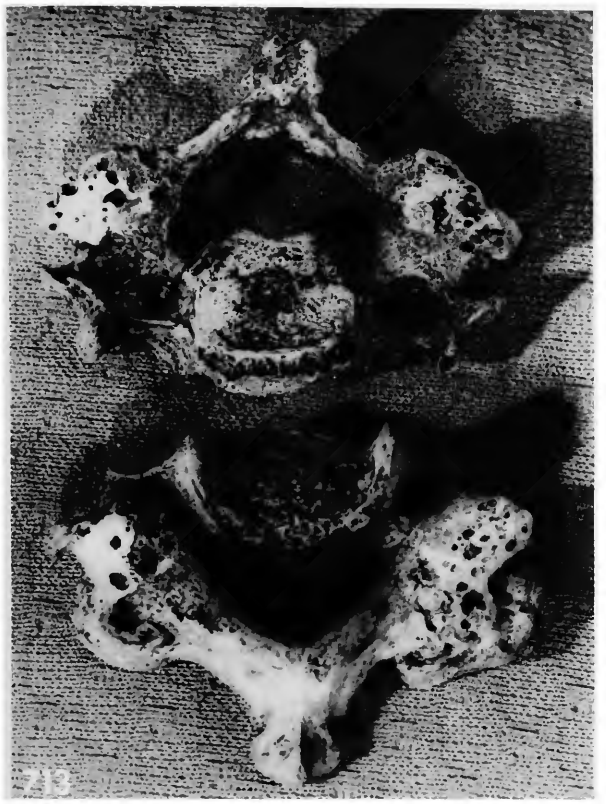
All vertebrae have diarthrodial joints, which permit movement of adjacent joint surfaces. These joints are between the facets of the articular processes of adjacent vertebrae. In addition the vertebral bodies are connected by cartilage discs in which gliding does not occur. All movement is due to the compression and expansion of the disc. The thoracic vertebrae have additional joints for the articulation of the ribs on the bodies and transverse processes. Porosity, eburnation, and marginal lipping are associated with degenerative arthritis of the diarthrodial joints. Erosion and marginal osteophyte development are associated with degenerative change of the vertebral bodies; porosity and eburnation occur rarely. The joints between ribs and vertebrae (costovertebral joints) exhibit marginal lipping and porosity but rarely eburnation.

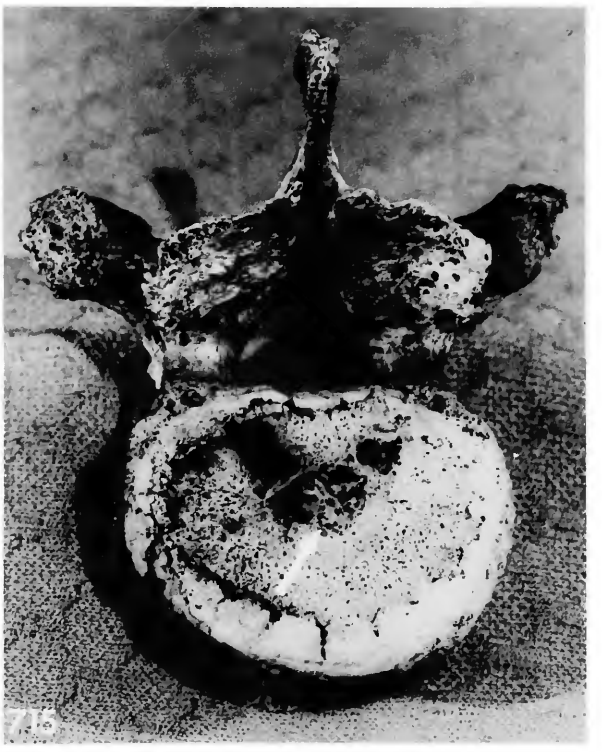
Two cases serve to illustrate degenerative arthritis of diarthrodial joints of the vertebrae. The first of these is an adult female skeleton from Hooper Bay, Alaska, USA (NMNH 339115). The archeological age is uncertain. The diarthrodial joints between the vertebrae all exhibit arthritic degeneration (Figures 713, 714). However, the cervicals have the most extensive involvement including porosity, eburnation, and marginal lipping (Figure 713). The lumbar vertebrae are somewhat less affected and the thoracic vertebrae least affected. The vertebral bodies of the lower thoracic and lumbar vertebrae exhibit the greatest osteophyte development. The inferior surface of the tenth vertebral body shows the typical bony erosion associated with Schmorl's nodes (Figure 715).

The second example is from an archeological site on the Seward Peninsula, Alaska, USA (NMNH 342482). The specimen is from an adult male Eskimo of uncertain archeological age. Diarthrodial joints between the vertebrae exhibit minimal evidence of abnormality except for the fusion of C2 and C3 and degenerative arthritis of the joints from the seventh cervical through the sixth thoracic (Figures 717–719). The spine of T1 was broken off (Figure 716) and the degenerative change of the diarthrodial joints may be a sequela of the trauma. This conclusion is supported by the relative lack of degeneration in the remaining vertebrae.

Degenerative arthritis of the amphiarthrodial vertebral joints is more easily noticed and has received greater attention in the literature on paleopathology. Two degenerative conditions are associated with these joints. In the first of these, degeneration of the disc may result in herniation of the disc and pressure erosion of one or more foci on the bony articular surface (Schmorl's nodes) of the vertebral body. With disc degeneration there may also be a generalized porosity of virtually the entire surface of the vertebral body. The second type of degenerative arthritis involves marginal development of osteophytes. In some cases the antecedent cause of osteophyte development appears to be anterior herniation of the disc. This condition is suggested in a specimen from an Eskimo skeleton from Pastolik, Alaska, USA (NMNH 332553). The specimen is from a young adult male of uncertain archeological age. The vertebral column is incomplete but the degenerative change is limited to the joint between the fourth and fifth vertebral bodies (Figures 720, 721). The lesion consists of anterior periarticular lipping with adjacent periosteal reactive bone on the anterior cortex of the bodies with rather marked erosion of the anterior joint surfaces particularly on the fifth vertebral body. The lesions of the two bodies are suggestive of an anterior herniation of the disc resulting in focal pressure erosion and peripheral periosteal reactive bone.

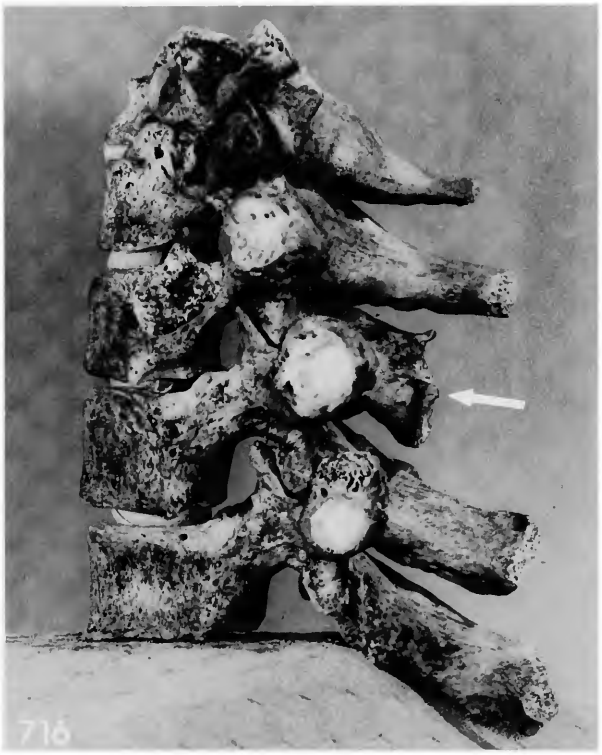
Another example of degenerative arthritis of the vertebral bodies is seen in the spinal column





FIGURES 713–715.—Degenerative arthritis of vertebrae: 713, Diarthrodial joint eburnation and porosity of the third and fourth cervical vertebrae. 714, Porous degeneration of the diathrodial joint between the tenth and eleventh thoracic vertebrae. 715, Depression in the inferior vertebral body due to Schmorl's node (arrow). (Adult female Eskimo skeleton from an archeological site in the Hooper Bay area of Alaska, USA, NMNH 339115.)

of another specimen from Pastolik, Alaska, USA (NMNH 332556). The skeleton is an adult male of uncertain archeological age. The vertebral column is complete, although there is some postmortem damage. All of the vertebral joints, both diarthrodial and amphiarthrodial, show evidence of degenerative arthritis. However, the most severe changes are seen in association with the amphiarthrodial joints of the lumbar vertebrae (Figures 708–710). The superior joint surfaces of L1, L2, and L5 exhibit slight to moderate lytic activity suggestive of pressure erosion from a Schmorl's node. The inferior surface of L5 (Figure 710) and the corresponding surface of the first sacral body are porous. There is extensive development of marginal osteophytes, some of which were in contact with osteophytes of adjacent vertebral bodies. Within the periarticular lipping



FIGURES 716–719.—Localized degenerative arthritis of the spine probably initiated by trauma: 716, Left lateral view of sixth cervical through third thoracic vertebrae; note truncated spine of first thoracic vertebra (arrow). 717, Superior aspect of the sixth (left) and seventh (right) cervical vertebrae, showing porous degeneration of the vertebral body. There is some hypertrophic bone formation on the body surfaces. 718, Porosity and eburnation of the left diarthrodial joint between the seventh cervical (left) and the first thoracic (right) vertebrae. 719, Bilateral diarthrodial joint porosity between the second (left) and third (right) thoracic vertebrae. (Adult male Eskimo skeleton from an archeological site on the Seward Peninsula, Alaska, USA, NMNH 342482.)

there is a concave depression, often with cyst-like cavities, between the outer edge of the original body and the outer edge of the degenerative lipping (Figure 709). The severity of the periarticular lipping increases toward the fifth lumbar body. These vertebrae exhibit a typical, although rather severe, pattern of degenerative arthritis of the spine.

FIGURES 720, 721.—Erosion and osteophyte development of the fourth and fifth lumbar vertebral bodies: 720, Left lateral view; note the anterior erosion of the adjacent edges of the vertebral bodies with evidence of bony reaction. 721, Anterior view; the appearance of the lesion is suggestive of the bony reaction to an anterior herniation of the disk. (Adult male Eskimo skeleton from an archeological site in Pastolik, Alaska, USA, NMNH 332553.)

Very extensive development of hypertrophic bone from the vertebral bodies may result in bony ankylosis. A possible example of this condition is seen in an archeological skeleton from Jersey County, Illinois, USA (NMNH 380071). The burial appears to be associated with the pre-Columbian period (Hopewellian). The skeleton is incomplete, but all the major long bones, pelvis, and the fused tenth thoracic through fifth lumbar vertebrae are present. The joints of the long bones all exhibit degenerative arthritis, particularly of the knee in which there is joint surface degeneration as well as extensive periarticular lipping. The most obvious arthritic changes are seen on the fused lower vertebrae. The extensive bony hypertrophy is largely limited to the right side and consists of massive bony overgrowths of marginal bone (Figures 703, 704). The disc spaces are normal and fusion does not appear to have occurred between the diarthrodial joints. The massive bone hypertrophy and lack of diarthrodial joint involvement contrasts with conditions seen in either rheumatoid arthritis or ankylosing spondylitis.

Other Lesions of the Joints

PATHOLOGY

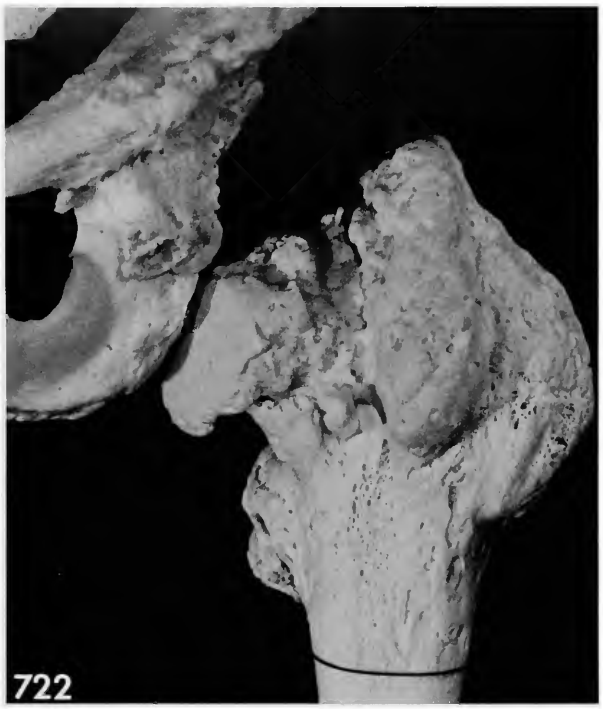
Neuropathic Arthropathy (Charcot's Joint)

Neuropathic arthropathy develops secondary to disturbance of the sensory and neurovascular innervation. The most common causes of Charcot's joint are (1) tabes dorsalis, a late manifestation of neurosyphilis, (2) syringomyelia, which is an intrinsic disturbance of the spinal cord, (3) diabetic neuropathy, and (4) traumatic nerve damage.

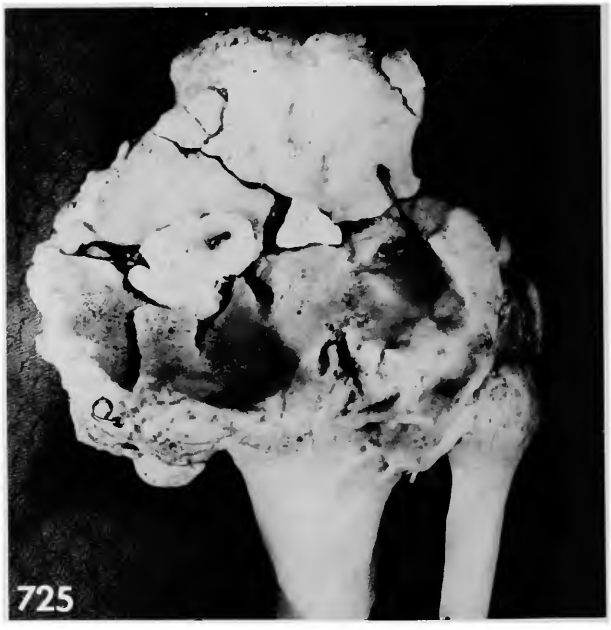
About 5 to 10 percent of tabetic patients develop a Charcot's joint. Tabetic arthropathy mostly affects large joints and localizes in the lower extremity in 75 percent of these cases (Figures 722, 723). The most commonly involved joint is the knee (about 50 percent), followed in descending order by the hip, shoulder, ankle, tarsus, and elbow. Only occasionally the dorsolumbar region of the spine, the temporomandibular, sternoclavicular, and finger or toe joints are involved. The affection is frequently bilateral.

Charcot's joint is the result of uncontrolled mechanical abuse in the absence of normal pain and position sensations, combined with abnormal neurovascular control of circulation.

In syringomyelia, a rare disease mainly located in the cervicodorsal portion of the spinal cord,



FIGURES 722, 723.—Charcot's joints of hip and knee in a patient with tabes doralis: 722, Left hip, showing joint destruction and massive periarticular bony build-up. 723, Left knee, showing massive osteocartilaginous loose bodies and subluxation. (73-year-old female, IPAZ 6650, autopsy 1817 from 1961.)



FIGURES 724, 725.—Charcot's joint of right knee: 724, Anterior view, showing large bone fragments between tibia and patella. 725, Posterosuperior view, showing deep excavation and eburnation of tibial articular surface and polished bone fragments. (PMUG 4942/60.)

destructive joint disease develops in about 25 percent of the patients. About 80 percent of these cases involve joints of the upper extremity, particularly the shoulder, and often are unilateral.

In diabetic neuropathy, mostly concerning elderly patients with long-standing diabetes, mostly the joints of the feet and, less commonly, of the ankle are affected (all statistical data from Jaffe, 1972:847–874). A similar distribution of neuropathic arthropathy occurs in late stages of neural leprosy. The end result is severe destruction, attrition, and fragmentation of the joint ends of the bones and formation of many loose bodies containing bone fragments. In addition, focal osteoporosis or osteosclerosis and transmetatarsal pathological fracture may be observed. Severe eburnation and articular grooving, as well as reactive exostoses of superimposed degenerative arthritis, are found (Figures 724, 725). In archeological material the fragments may not be recovered but the irregular destruction of the convex and the deep excavation of the concave joint end, as, for instance, the femoral condyles and the tibial plateau, should be recognizable.

Pigmented Villonodular Synovitis

Pigmented villonodular synovitis is a rare condition characterized by tumor-like proliferation of the synovial stroma with accumulation of blood pigment. Usually the disease is limited to the soft tissues of the joint. The reason to discuss it here is that the joint ends of the articulating bones occasionally become involved. The most common location is the knee joint, but any other diarthrodial joint may be involved. The disease is usually monoarticular and affects young adults. In joints with a tight synovial compartment, as interphalangeal joints, undermining erosion of the bone next to the cartilaginous articular surface can occur on both sides of the joint. This can look about the same on dry bone as the corresponding lesion in gouty arthritis, although in the latter the lesion is rarely limited to only one joint. In exceptional cases extensive involvement of epiphyses occurs, producing large lytic lesions in the affected bone ends. A solitary lesion of this type could, in the dry bone, be indistinguishable from an epiphysial tumor (chondroblastoma or enchondroma).

Synovial Osteochondromatosis

Synovial osteochondromatosis is a rare condition characterized by formation of multiple foci of metaplastic cartilage in the synovium. These cartilaginous foci frequently undergo enchondral ossification. Some of them separate from the synovium and become loose bodies in the joint cavity. The condition occurs more commonly in young adult males than in females. Only large joints are affected and the knee is more frequently involved than the hip or the shoulder. Usually only one joint is involved. The osteocartilaginous nodules, which can be very numerous, usually appear rounded and are of about equal size. Since the nodules arise from the synovium, the articular bone ends appear normal. This fact and the smooth contour, even size, and large number of these ossified nodules would permit differentiation from loose bodies of Charcot's joint and of osteochondritis dissecans in dry bone material. Of course, if the condition did not proceed beyond the cartilage stage, nothing recognizable would be present in the dry bone material.

Lesions of Jaws and Teeth

The Jaws

Pathology

The jaws participate in a great variety of lesions, which also may affect other parts of the skeleton. Here I will discuss only the lesions that are found exclusively in the jaws, because of their intimate relationship to the teeth and to the complicated development of the facial skeleton. These lesions fall into two groups: odontogenic and nonodontogenic.

Odontogenic Cysts

The complicated dental development resulting from the coordinated interplay of the ectodermal enamel epithelium and of the mesodermal tooth bud lends itself to a variety of malformations. Since the third molar is regressing in the human and may be not developed, or at least not erupted, anomalies affecting its development are not too uncommon.

Primordial Cyst.—This cyst is the result of an abortive development of a dental follicle without formation of a tooth. The cyst has a fibrous wall and is lined by odontogenic epithelium. It predilects the area of the third molar, more often in the mandible than in the maxilla. As the cyst enlarges it may occupy part of the ascending mandibular ramus. In dry bone it would appear as a cystic cavity that may bulge the overlying cortex but is not intimately associated with a dental root and does not contain a tooth.

Dentigerous Cyst.—This type of cyst is the product of a later stage of deranged dental development. After the crown of a tooth is already developed the enamel epithelium fails to involute but remains, forming a cyst into the base of which the incomplete tooth projects. Again it is the area of the third molar that is predilected. This type of cyst usually develops from a tooth bud of the second dentition or from a supernumerary dental follicle. In dry bone it would be similar to the primordial cyst except for the presence of an incomplete tooth projecting into the cavity if it were preserved intact.

Radicular Cyst.—This third type of odontogenic cyst is the most common of all. It is not the result of maldevelopment but of infection of the pulp cavity and the root canal of an erupted tooth. It starts as an apical abscess or granuloma and secondarily acquires an epithelial lining from remnants of the epithelial sheath of the dental root. It may be located on any erupted tooth of the mandible or maxilla and is always intimately associated to the root of an erupted tooth. This tooth may have fallen out but its alveolus would still connect with the cyst cavity. In contrast to the dentigerous cyst, where the tooth is below the cyst, in the radicular cyst the tooth is above.

Odontogenic Tumors

A variety of tumors can arise from epithelial or mesenchymal components of the dental bud. The most important of these is the ameloblastoma arising from enamel epithelium. This lesion is more common in the mandible than in the maxilla, predilecting the area of the third molar. Males are more often affected than females. The lesion is unusually frequent in certain populations of African Negroes, but it can occur in all races. The lesion begins in childhood or adolescence and slowly progresses. It is basically a benign

lesion. The fully developed tumor presents as a cystic mass, often multiloculated, which expands the bone but maintains a thin, ridged, bony shell. Enamel is not formed by the tumor tissue and a tooth is not included in the lesion.

All the other odontogenic tumors are of mesenchymal origin. The odontoma is a tumor prevailingly consisting of dentin, but cementum and enamel can be intermixed. This tumor may be a shapeless mass of tissue of increased radiodensity, but in its most mature form it may show a conglomeration of fairly well-developed teeth. Thin, polished, undecalcified sections should readily reveal the finely tubular structure of dentin microscopically. The cementoma is usually a tumor of moderate size, intimately attached to one or several adjacent erupted teeth. Cementum is a mineralized tissue resembling bone, which covers the dental root in a thin layer and serves as attachment of the alveolar ligament. Tumor-like proliferations of cementum are not uncommon in the jaws of persons with Paget's disease.

The myxoid mesenchyme normally forming the tooth pulp may give rise to an odontogenic fibroma. This would appear in the dry bone merely as a smooth-walled, more or less expanded, lytic defect without any identifiable characteristics.

Nonodontogenic Cysts

These cysts are related to the closure of various fissures in the development of the facial skeleton or remnants of the nasopalatine duct. Most of them are located in the maxilla, and all but the unicameral bone cyst arise on the basis of minor developmental disturbances.

The median cysts occur between the central incisors of the maxilla or of the mandible, due to trapping of epithelium between the two halves of the maxilla or mandible. This is the only non-odontogenic epithelial cyst found in the mandible.

The globomaxillary cyst results from epithelial inclusions between the maxillary and globular processes of the maxilla. It is always located between the lateral upper incisor and the canine tooth in the alveolar process.

The nasoalveolar cyst is due to epithelial remnants between the lateral nasal and the maxillary process. It is not located between the teeth and tends to bulge into the nasal cavity when it enlarges.

The nasopalatine cyst is based on persistence and continued secretion of the epithelial lining of the vestigial nasopalatine duct. It may form unilaterally or bilaterally in the incisive canal and destroy or partly destroy the anterior portion of the hard palate.

Unicameral bone cysts occur in the jaws as in other bones. In the mandible they tend to be located below the nerve canal rather than primarily in the alveolar process. In the maxilla they tend to bulge the cortex forward and towards the antrum.

Nonodontogenic Tumors

Various fibro-osseous tumors occur in the jaws, more often in the maxilla than in the mandible. They are characterized by production of finely or coarsely trabecular, mostly woven, bone. The relationship of such lesions to fibrous dysplasia is open to question. Bilateral disfiguring fibro-osseous maxillary tumors are seen in young individuals (cherubism). Recognition of such lesions in dry bone would depend on perfect preservation of frail trabecular bone.

A variety of lytic lesions, such as eosinophilic granuloma, "brown tumor" of hyperparathyroidism, giant cell reparative granuloma or metastatic carcinoma, occur with some frequency in the jaws. In dry bone such lesions are devoid of identifying characteristics in themselves but may be interpretable in the context of findings elsewhere in the skeleton.

Carcinoma of the oral or nasal cavity, the paranasal sinuses, and even the facial skin may cause extensive destruction of a jaw or other facial bone by direct invasion. As a rule the lesion would show a frayed margination in dry bone with little, if any, osteosclerotic reaction.

The Teeth

There are many parallels between the diseases that affect bone and those that affect teeth. Trauma, infection, metabolic abnormalities, congenital malformations, and tumor are among morbid conditions that affect both. However, there are fundamental differences in the biology of teeth that affect the expression of disease. Dental enamel lacks cells, vascular and nerve supply. For this reason enamel can be affected by morbid conditions only during the development of the dental crown and by extrinsic lytic processes (primarily dental caries) after eruption of the teeth. Dentin also has a very limited potential response to morbid conditions. Cells (odontoblasts) line the inner surface of dentin and cellular extensions pass through the dentin. Blood and nerve supply, as the major components of the pulp cavity, are in close proximity to the dentin. Odontoblasts have limited potential to form secondary dentin and this is often seen in worn teeth. Dentin may be affected by morbid conditions present during development, and like enamel, may be affected by lytic processes after eruption.

There is a vast literature on dental pathology. Pindborg (1970) and Gorlin and Goldman (1970) are the major sources for the discussion of dental disease. Much has been published on dental paleopathology as well. However, this aspect of paleopathology has not received the same attention as skeletal paleopathology. Much of the following discussion of dental paleopathology is the result of my own experience and the observations made on specimens from the human skeletal collections of the National Museum of Natural History, Washington, D.C., USA.

Dental Caries

Pathology

Pindborg (1970:256) defines dental caries as an infectious and transmissible disease in which progressive destruction of tooth structure is initiated by microbial activity on the tooth surface. Unlike destructive processes of bone, destruction of dental hard tissue is the direct result of lytic activity by bacteria. Bacterial organisms associated with dental caries include lactobacilli and streptococci, although the causative mechanisms by which organisms produce caries is still not clear (Darling, 1970:273).

Dental surfaces exposed to potential caries include: (1) the cutting or chewing (occlusal) surfaces, (2) the smooth surfaces of the crown including the mesiodistal surfaces (interproximal) in contact with other teeth and buccolingual surfaces, and (3) the root, which may be directly affected by caries after the recession of the gingiva. Each of these surfaces has varying cariogenic potentials. These potentials, in combination with varying mouth bacteria and diet, appear to produce different patterns of dental caries (Keyes, 1968). Thus, the characteristic location of dental caries in an archeological skeletal sample may provide insight regarding the diet of the people.

It is important to emphasize that all cavity-like defects in teeth are not the result of caries. Pits and fissures are naturally occurring features of a normal tooth. They may, of course, become a site for caries. The distinction between a natural pit and a cavity induced by dental caries may not be apparent externally. However, when dental caries passes through the enamel and reaches the dentin, the destructive process spreads rapidly in the dentin, adjacent to the boundry between enamel and dentin, creating a funnel-shaped lytic lesion (Figure 726). Evidence of such a lesion would be indicative of caries and would be apparent in an X-ray film or could be demonstrated with a dental explorer or bent needle.

Pindborg (1970:256–258) notes the existence of acute and chronic dental caries. The former tends to be associated with young individuals and appears as a white chalky spot in early stages. Chronic caries is often seen on interproximal surfaces of teeth often opposed to a carious lesion on an adjacent tooth. Lesions in chronic caries tend to be darker in color varying from yellow to brown.

FIGURE 726.—Dental caries of the right mandibular first molar. Interproximal caries (white arrow) and occlusial surface caries (central lesion). On the central lesion note the spread of the decay process below the enamel (black arrows). (Mesiodistal section, NMNH unidentified anatomical specimen.)

While bacterial activity appears to be a necessary condition for the development of caries, factors intrinsic to the tooth structure may affect the development and location of caries. Aside from the anatomical features mentioned above, developmental defects in the quality of the enamel create conditions favorable to cariogenic activity.

Untreated caries may result in destruction of the entire crown and/or significant portions of the root. Exposure of the pulp chamber creates a high risk of infection with the almost inevitable sequela of abscess and destruction of the supporting tissues. Abscess of a tooth frequently leads to its exfoliation followed by alveolar repair involving bone resorption and partial refill of the alveolus. Hematogenous dissemination of the infection may also occur and results in serious complications including meningitis and hematogenous osteomyelitis (see p. 123 and Figures 165–168).

PALEOPATHOLOGY

Two basic approaches characterize the literature on the paleopathology of dental caries. The first of these is largely descriptive, in which an important focus is the history and antiquity of dental caries (e.g., Baudouin, 1923). In the more recent literature the major emphasis has been on the difference in caries frequency between populations (e.g., Angel, 1974b), with emphasis on caries as an indicator of varying food resources as in the nutritional differences between hunter-gatherers and agriculturists (e.g., Cassidy, 1974; Sciulli, 1977; Turner, 1979; Cook and Buikstra, 1979). Caries frequency is low among hunter-gatherers (approximately two to three lesions per mouth) and more than twice as high among agriculturists. Both malnutrition affecting tooth development and higher carbohydrates in the diet have been invoked to explain the higher caries frequency in agricultural populations. With the recent addition of refined sugar and flour to the diet, caries frequency has become even greater (Brothwell, 1963:274).

There are two major problems in studying dental caries in archeological skeletons. First, teeth are frequently lost or damaged postmortem. Postmortem tooth loss is most common with the anterior teeth, which are usually less affected by caries. Antemortem tooth loss also creates problems in evaluating caries incidence. This condition is often but not always the result of caries complicated by abscess. The second problem is that most archeological skeletons exhibit considerable dental attrition due to coarse particles in the food. The effect of this wear is to grind away occlusal surfaces before dental caries can develop. Occasionally this wear is so severe that the pulp cavity is exposed with the tooth root and supporting tissues often becoming infected. If wear is extreme the abscess is more likely to be due to attrition rather than caries, although it may not always be possible to make this distinction.

OCCLUSAL SURFACE CARIES.—This type of caries is primarily associated with the molars and premolars and begins in the crevices of the occlusal surface. In early stages there may be only a small penetrating hole through the enamel surface (Figure 727) as seen in a young adult mandible from an archeological site in Arizona

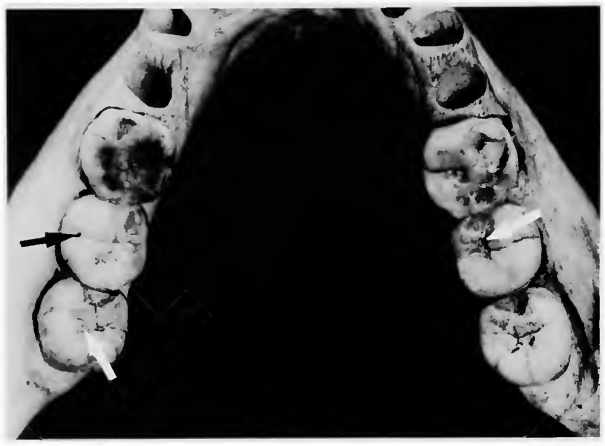


FIGURE 727.—Occlusal surface caries, early to moderate stages in the lower jaw. The lower right first molar exhibits moderate wear and a fairly large cavity in the mesial-buccal quadrant. Other molars exhibit early development of dental caries (white arrows) with only small holes apparent externally. Probing with a dental explorer reveals much greater destruction below the enamel. There is a pit on the lower left second molar (black arrow), which is not carious. (Young adult female mandible from an archeological site in Arizona, NMNH 239202.)

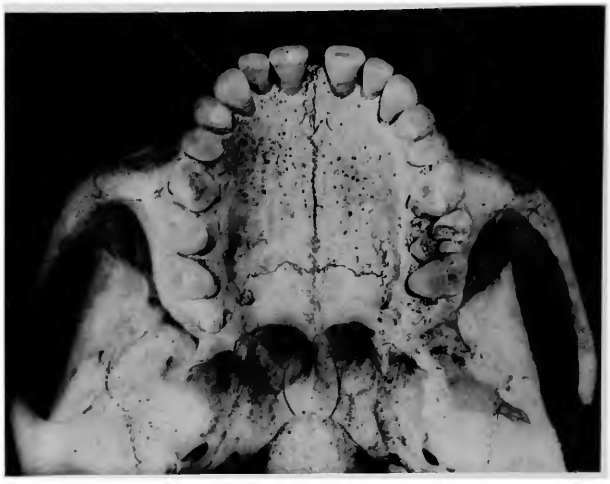


FIGURE 728.—Severe occlusal surface caries of the upper left second molar. Virtually the entire crown has been destroyed. The other teeth are worn but not carious (Adult male from Early Bronze Age site of Bab edh-Dhra, Jordan, NMNH A100N Burial 1.)

(NMNH 239202). A much more severe example of probable occlusal surface caries is seen in the upper left second molar (Figure 728) of an adult male from the Early Bronze Age site of Bab edh-Dhra in Jordan (NMNH A100N Burial 1). This is the only evidence of caries in the dentition. There is some alveolar resorption at the base of the carious tooth that is suggestive of infection.

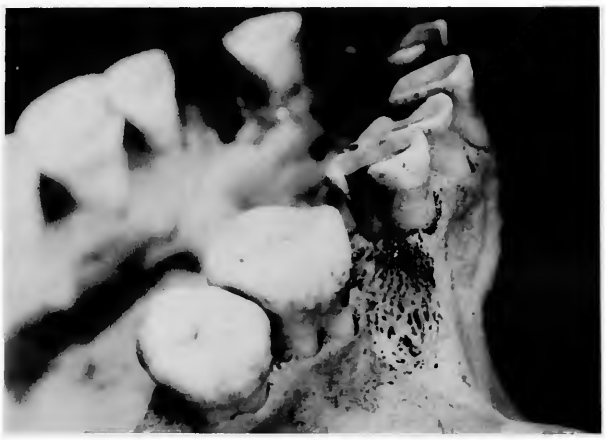


FIGURE 729.—Interproximal caries of the upper left second premolar. Adjacent first molar is missing antemortem, probably due to caries and abscess. (Adult female from Moorehead Cave, Texas, NMNH 372528.)

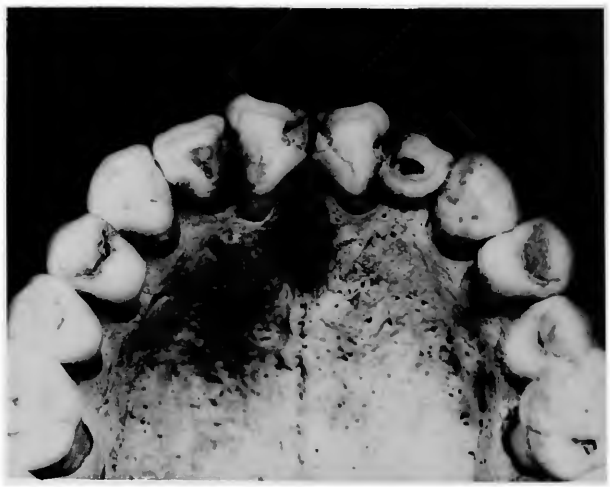
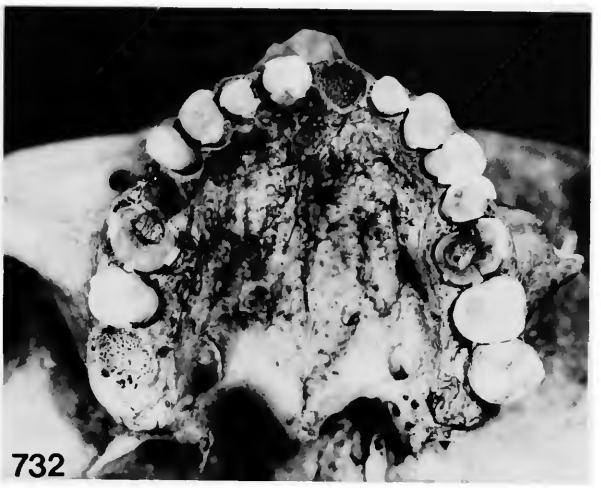
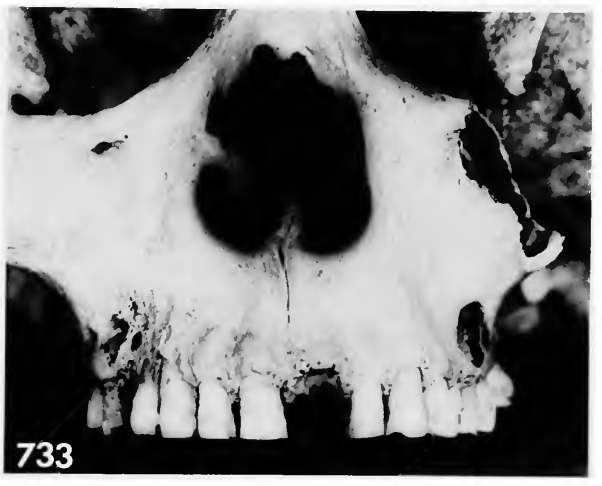


FIGURE 730.—Interproximal caries of the maxillary incisors. (Adult male from a Late Woodland site in Virginia, USA, NMNH 382419.)

FIGURE 731.—Root caries of the mesial, interproximal surface of the lower left first molar. Note the evidence of moderate alveolar resorption indicated by the distance between the cemento-enamel junction and the alveolar bone. The soft tissue would have receded with the bone, exposing the tooth root to caries; anterolateral view. (Adult female from archeological site of Puye, New Mexico, USA, NMNH 262944.)





FIGURES 732, 733.—Dental caries with destructive lesions penetrating the pulp cavity of both first molars: 732, Occlusal view; left central incisor missing postmortem. Right second premolar and third molar sockets show evidence of alveolar remodeling suggestive of antemortem tooth loss. There is an abscess associated with the premolar. 733, Anterior view of maxilla, showing lytic bony response to abscessed first molars and right second premolar. (Adult male from archeological site of Puye, New Mexico, USA, NMNH 262948.)

NONOCCLUSAL SURFACE CARIES ON THE CROWN.—An example of interproximal caries is seen in a skull of an adult female from Moorehead Cave, Texas, USA (NMNH 372528). The upper left second premolar is carious on the distal surface. The first molar appears to have been lost antemortem (Figure 729). The molar socket shows considerable resorption and evidence of abscess. The carious lesion on the premolar continues past the cemento-enamel junction into the root. The lesion did not penetrate to the pulp cavity.

Interproximal caries can also affect the incisors. An example of this condition is seen in an adult male skull from the Shannon site in Virginia, USA (NMNH 382419). This site is Late Woodland in date with no European trade goods. All of the interproximal surfaces between the maxillary incisors are carious (Figure 730).

ROOT CARIES.—An example of root caries is seen on the mesial interproximal surface of the lower first molar of an adult female skull from the pre-Columbian archeological site of Puye, New Mexico, USA (NMNH 262944). On the buccal side of this tooth the upper margin of the alveolar bone is almost 4 millimeters below the cemento-enamel junction (Figure 731). On the lingual side this dimension is almost 5 millimeters.

These distances are more than twice the normal distance between alveolar bone and the cementoenamel junction and are indicative of considerable alveolar resorption, which exposed the root to caries.

Secondary Effects of Dental Caries.—If tooth destruction from dental caries penetrates the pulp cavity, infection of the support tissues is an almost inevitable result. Bone tissue reaction to infection initially involves bone destruction providing a drain for pus. Chronic infection leads to at least partial bony circumscription around the infectious focus. Tooth exfoliation and hematogenous dissemination of the infection are additional potential secondary effects.

Most of these morbid processes, secondary to dental caries, are demonstrated in the maxillary dentition of an adult male skull from the pre-Columbian site of Puye, New Mexico, USA (NMNH 262948). The left central incisor is missing postmortem. The right second premolar and right third molar are missing antemortem. In the case of the premolar there is evidence of abscess at the base of alveolus. The alveolus of the third molar is shallow with evidence of remodeling suggestive of infection. However, there is no evidence of a draining sinus through the alveolar bone. The major periapical lesions are associated with the first molars. Both teeth have large cavities in the crown, which have penetrated the pulp cavity (Figure 732). The resulting infection has resulted in lytic lesions of the maxilla, which penetrate the external cortex of the maxilla on both sides and the inner cortex on the left (Figure 733). Whether this condition led to disseminated infection elsewhere in the body cannot be determined. Indeed evidence for such a complication would be rare in dry skeletal material.

Periodontal Disease

Pathology

Ruben, Goldman, and Schulman (1970:394) note that several morbid conditions result in periodontal disease. Identification of many of these conditions depends on good medical histories, soft tissue pathology, and bacteriology. It is thus unlikely that all the specific factors that induce periodontal disease will be identifiable in dry bone specimens. For this reason I shall limit my discussion to the more common causative factors and those which may be identifiable in archeological specimens.

As a general principle, periodontal disease involves an inflammatory response to one or more irritants. This inflammation often results in resorption of alveolar bone creating an abnormally large distance between bone and the cementoenamel junction. A common irritant is calculus and its precursor, bacterial plaque, which consists of a sticky coating including protein, food particles, living and dead microorganisms. When plaque mineralizes, it becomes calculus and in this form can be found on archeological skeletal specimens as relatively hard additions to the tooth surface. The role of calculus in periodontal disease is not clear. Ruben, Goldman, and Schulman (1970:397) express the opinion that mechanical irritation from calculus is secondary to inflammation directly induced by bacterial activity. In any case the presence of calculus on dental tissues is an important consideration in evaluating the cause of periodontal disease in archeological skeletal material.

Metabolic problems may result in inflammatory conditions that affect the periodontal tissues. In scurvy, for example, the quality of the connective tissue associated with teeth is weakened. Bleeding results from weakened blood vessel walls being traumatized by chewing. The inflammatory response in scurvy may lead to exfoliation of teeth, particularly those with single roots. Similarly, protein deficiency may lead to diminished quality of supporting tissue and periodontal disease.

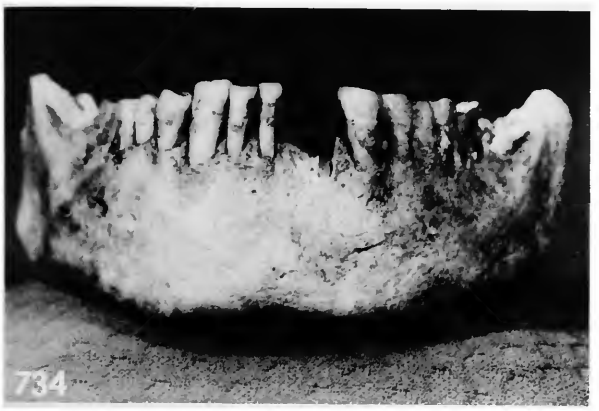
Peridontal abscess may result in relatively localized periodontal disease. The effect on supporting tissue may be similar to that produced by periapical abscess. However, the focus of infection in periodontal abscess is outside the pulp cavity and often between the roots of multirooted teeth

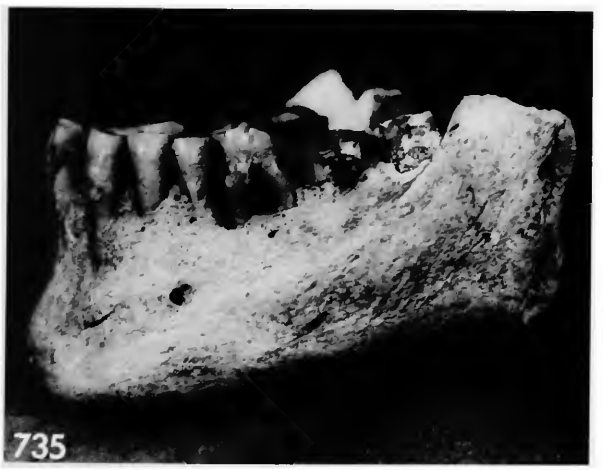
(Ruben, Goldman, and Schulman, 1970:423). Abscess of alveolar bone without evidence of caries involvement of the pulp cavity would be suggestive of periodontal abscess.

Paleopathology

In archeological skulls the major manifestation of periodontal disease will be local or general alveolar resorption. The distinction will need to be made between alveolar resorption from periodontal disease and resorption associated with periapical abscess and tooth loss. Clearly if there is alveolar resorption but little or no evidence of caries or tooth loss, a diagnosis of periodontal disease is appropriate. An example of this condition is seen in the mandible of a skeleton from an archeological site on Horr's Island, Florida, USA (NMNH 352156). The maxilla was broken postmortem and only fragments recovered. On the mandibular teeth there is considerable root exposure, which is indicative of alveolar resorption (Figures 734, 735). Dental calculus is adherent on the exposed buccal root surfaces of the incisors and canines. Alveolar resorption is particularly noticeable in the bone supporting the first molars. Although there is considerable wear, there is no evidence of pulp-cavity exposure or caries. The presence of calculus in association with alveolar resorption and lack of dental caries supports a diagnosis of periodontal disease.

Another example of marked tooth root exposure with no evidence of dental caries is found in the mandible of a skeleton from an archeological





Figures 734, 735.—Alveolar resorption indicative of periodontal disease: 734, Anterior view of lower jaw showing marked exposure of the roots of the anterior teeth; note the dental calculus adjacent to the exposed roots. 735, Left lateral view, showing root exposure of the premolars and molars. There is considerable dental attrition but no evidence of caries or exposure of the pulp chambers of any teeth. (Adult male from Horr's Island, Florida, USA, NMNH 352156.)

Figure 736.—Molar root exposure and reactive alveolar bone suggestive of periodontal disease. Note calculus on the distolingual portion of the third molar (arrow). Left medial view of mandible. (Adult male from Canaveral, Florida, USA, NMNH 377439.)

site at Canaveral, Florida, USA (NMNH 377439). The maxilla was broken postmortem and only fragmentary portions were recovered. These fragments exhibit considerable root exposure of the right incisors, canine, and first premolar. These teeth were extensively worn with evidence of secondary dentin formation in the incisors. The mandibular incisors appear to have been lost antemortem as was the left first premolar. The right canine, premolars, left second premolar and first molar were probably lost postmortem. However, in the case of the left first molar, alveolar resorption had resulted in very poor support for the tooth roots. There is considerable root exposure of the second and third molars and marked build up of calculus on the crown and exposed root (Figure 736). Rough porosity characterizes the bone associated with the left molar roots. This is indicative of an inflammatory condition, probably including the overlying soft tissues. The pattern of alveolar exposure and lack of caries supports a diagnosis of periodontal disease.

Disturbances in Dental Development

Pathology

Abnormal Quality of Teeth

Several morbid conditions may result in an abnormal quality of teeth. Some conditions affect dentin (see above), some affect enamel, and others affect both. The most common condition is a hypoplastic defect in the enamel. Pindborg (1970: 75–210) provides an extensive review of the various conditions that produce this defect, many of which are rare or caused by modern problems. The reader is referred to this source for a more comprehensive treatment of this subject. I shall limit my comments to those pathological conditions that are fairly common in paleopathological specimens.

Infectious diseases, primarily congenital syphilis and tuberculosis, as well as metabolic and endocrine disorders, are known to adversely affect the formation of dental tissue. Fairly typical multiple hypoplastic lines are seen in the dentition of a 26-year-old male from the Pathology Museum of the University of Graz in Austria (PMUG 3421, autopsy 9497 from 1879). Autopsy records state that the individual was a syphilitic. Transverse hypoplastic lines are most pronounced on the maxillary dentition, particularly the central incisors (Figure 737). In addition to the defective lines, there are numerous fine pores in the enamel.

Congenital syphilis may result in defective first molars (mulberry molars). The crown size is smaller than normal, tending to be smaller than that of the second molar; normally the crown of the first molar is the larger. The occlusal surface is very irregular. A skull from a modern female about 30 years of age exhibits dental defects fairly typical of congenital syphilis (NMNH 219398). The upper right central, lower right central, and lower left lateral incisor crowns have hypoplastic defects of the incisal edge (Figure 738). The first molars are smaller than the second and have a pitted, poorly formed, occlusal surface (Figure 739).

Dental hypoplasia is also thought to be associated with rickets. Figures 424 and 426 show hy-

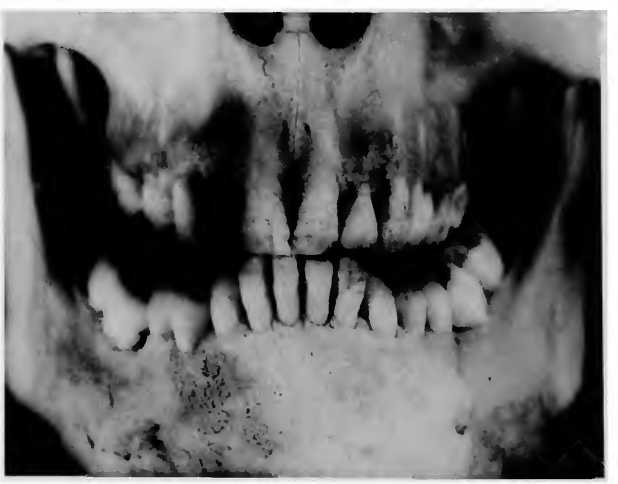
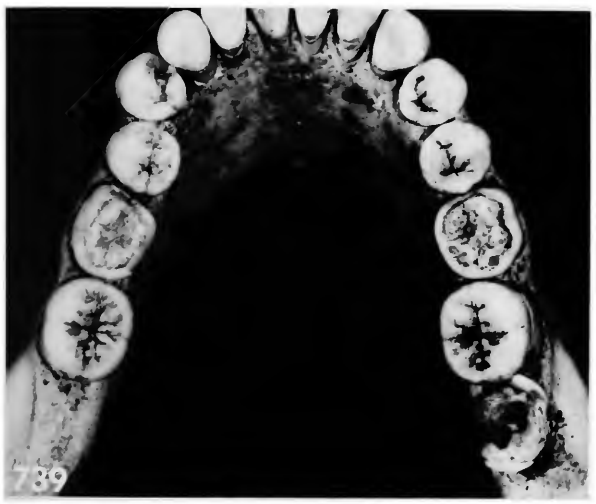
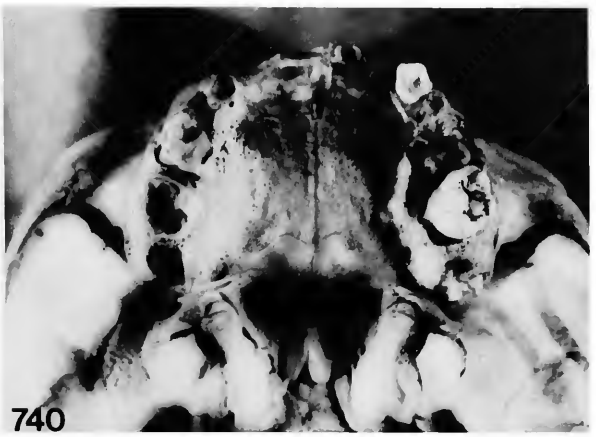
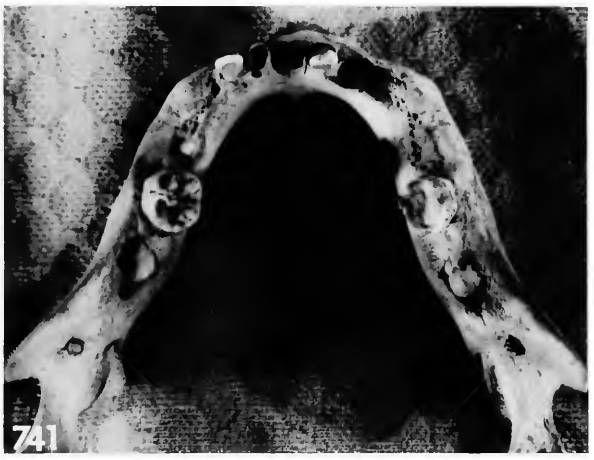


Figure 737.—Dental hypoplasia associated with syphilis. Note multiple transverse, hypoplastic lines and pitting of enamel between the lines. (26-year-old male, PMUG 3421, autopsy 9497 from 1879.)



FIGURES 738, 739.—Dental hypoplasia associated with probable syphilis: 738, Anterior view; note defect of incisal edges of incisors. 739, Occlusal view of mandibular molars. The first molar is smaller than the second and has an abnormal pitted occlusal surface typical of congenital syphilis. (Female about 30 years of age, NMNH 219398, dissecting-room specimen from before 1903.





FIGURES 740, 741.—Dental hypoplasia associated with rickets: 740, Occlusal view of maxillary dentition; note defective crowns of second deciduous molars and right first permanent molar. 741, Occlusal view of mandibular dentition. The crowns of both first permanent molars are defective. (Child about 6 years of age, FPAM 2694 from before 1858.)

poplastic defects of the teeth. A rachitic skull (FPAM 2694) from the Federal Pathologic-Anatomy Museum in Vienna, Austria, exhibits marked hypoplastic defects of the teeth (Figures 740, 741). The skull is from a child about 6 years of age. The most severe changes are seen in the crowns of the upper second deciduous molars, the right upper first permanent molar, and the first permanent molars of the mandible. Both the enamel and dentin appear to be affected.

Tuberculosis is most commonly a disease of childhood, so it is not surprising to find hypoplastic dental lesions associated with tuberculosis. A skull of a child, between 3 and 6 years of age, reported to have had tuberculosis, is part of the collections of the National Museum of Anthropology in Prague, Czechoslovakia (ANM 2028). All of the deciduous teeth have erupted and

FIGURE 742.—Dental hypoplasia associated with a case of tuberculosis. Hypoplastic lines or spots are visable in the crowns of all the deciduous teeth. (Child between 3 and 6 years of age, ANM 2028 from before 1895.)

FIGURE 743.—Dental hypoplasia associated with tuberculosis. Note multiple hypoplastic lines in the teeth. (17-year-old-male, FPAM 2016, autopsy 16648 from 1842.)

exhibit hypoplastic lines and spots (Figures 742). There are no autopsy data available for this case, so that the diagnosis of tuberculosis cannot be evaluated. The lytic lesion on the mandible is not typical of tuberculosis. The disease processes would have had to begun in late fetal life or early infancy to have produced these effects on the incisors.

An undoubted case of tuberculosis with multiple hypoplastic lines is associated with the skull of a 17-year-old boy, which is part of the Federal Pathologic-Anatomy Museum in Vienna, Austria (FPAM 2016, autopsy 16648 and 1842). There are multiple lytic lesions of the skull. The incisors, canines, and premolars that remain in the jaws show hypoplastic lines (Figure 743).

Abnormal Quantity of Teeth

Rather commonly, there is an increase in the normal number of teeth (hyperodontia). This typically affects the permanent dentition and most often results in more than the normal 32 teeth (hyperodontia). The extra teeth may be heterotopic, meaning that they develop outside of the alveolar region, or they may be normotopic, meaning that they develop in the alveolar region and erupt in the normal orientation. Supernumerary teeth may be the result of the retention of deciduous teeth or the development of extra permanent teeth.

Fewer than normal teeth (hypodontia) occurs when one or more teeth fail to develop. The frequency of agenesis varies with the tooth. In descending order of frequency the commonly affected teeth are: third molars, maxillary lateral incisors, second premolars, mandibular central incisors, and maxillary first premolars (Salzmann, 1957:256). Occasionally, often because there is insufficient room, one or more teeth will become impacted and fail to erupt. This is not a true case of hypodontia, since the dental element did develop. However, the external appearance of the dentition and jaw may be very similar and an X-ray film would be needed to differentiate dental agenesis from impaction.

Abnormal Size and Shape of Teeth

Teeth may be larger than normal (macrodontia). This condition may affect the entire dentition or just one tooth. It may be symmetrical or unilateral. In the latter case macrodontia is easily confused with the fusion of two teeth and differentiation may not be possible (Pindborg, 1970: 35). Teeth smaller than normal (microdontia) are more common. This condition is associated with a number of congenital morbid conditions, including heart disease, Down's syndrome, and cleft palate (Pindborg, 1970:36). Often the shape in microdontia will be abnormal as well.

Dental Anomalies

There is considerable normal, genetically based variation in the shape of the teeth. Variants include the shovel-shaped incisors of Asian populations and many variations in molar cusp pattern. Fairly common dental anomalies include fused teeth and enamel pearls. Teeth may be fused at the crowns, the roots or both. The factors contributing to fusion include both genetic abnormalities and inflammation. Pindborg (1970: 47–57) distinguishes between concrescence, in which tooth roots are joined only by hyperplastic cementum, and fusion, which occurs during the development of the teeth. Fusion affects the deciduous dentition more commonly than the permanent teeth. Incisors and canines are the most frequently affected teeth.

Enamel pearls are thought to be the result of abnormalities in embryological development. They may project from the surface of the tooth (extradental) or may be included within the dentin (intradental). The former are the ones encountered fairly commonly in archeological specimens. They occur only on multirooted teeth and vary in size from barely visible to about 2 millimeters in diameter. They are most frequent on the maxillary teeth where they tend to be on the interproximal surfaces. On the mandibular teeth they tend to be on the buccal or lingual surfaces (Pindborg, 1970:45–47).

Dental Crowding

The development of teeth appears to be primarily under the control of genetic fields and environmental conditions, which are relatively independent of those factors that affect the development of jaws. Because of this independence, the size of the two types of structure may not be congruent. Most often the incongruence is expressed as an inadequate size of the jaw relative to the size of the teeth. This is particularly true of the mandible. Inadequate space for the teeth results in tooth crowding. This problem may be particularly severe in conditions such as pituitary dwarfism, where the lack of growth hormone severely limits bone development but has minimal effect on tooth size.

Paleopathology

Abnormal Quality of Teeth

I have previously described (p. 207) a possible case of congenital syphilis in which the deciduous dentition had marked hypoplastic enamel defects (Figures 320, 322). Most of the examples of dental hypoplasia in archeological skeletons are not as noticeable. Two examples of hypoplasia in archeological specimens illustrate more subtle manifestations. The first of these is a specimen from the protohistoric archeological site of Mobridge in South Dakota, USA (NMNH 325416). The skull is from an adult female skeleton. The teeth are worn, with development of secondary dentin on the incisors and upper canines. The wear may have obliterated some of the hypoplastic lines. The third molars did not develop. All the teeth except the molars have at least one hypoplastic transverse line (Figure 744).

Multiple hypoplastic transverse lines are present on the canines (Figure 745) in an adult male skull from an archeological site in Fort Concho, Texas, USA (NMNH 243490). The upper incisors are missing and the lower incisors have been damaged postmortem. The lower lateral incisor, however, appears to have at least one hypoplastic line, suggesting that the incisors may have been defective. The premolars and molars are normal.

FIGURE 744.—Enamel hypoplasia expressed as hypoplastic transverse lines of the buccal crown surface of all teeth except the molars. Anterior tooth wear is marked with secondary dentin apparent on the occlusal surfaces. (Adult female from Mobridge site, South Dakota, USA, NMNH 325416.

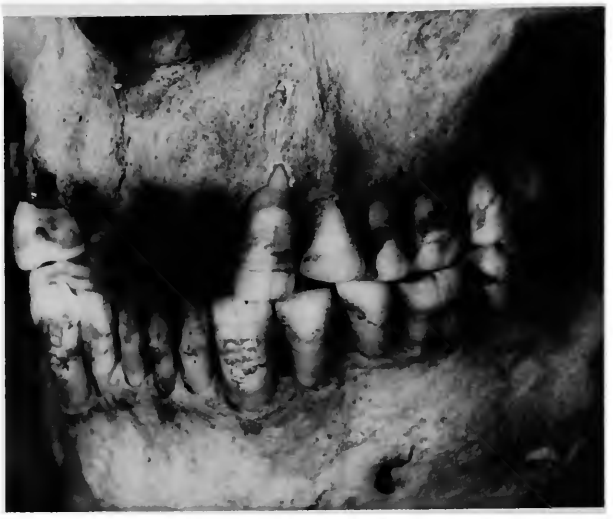


FIGURE 745.—Multiple hypoplastic transverse defects of the canine teeth. (Adult male from Fort Concho, Texas, USA, NMNH 243490.)

Abnormal Quantity of Teeth

Supernumerary teeth (hyperodontia) are rather common in archeological specimens (e.g., Brothwell, 1965a:114, 115). Two cases serve to illustrate the development of supernumerary teeth outside the alveolar region of the jaws (heterotopic). The first of these is the facial portion of a child's skull from Chicama, Peru (NMNH 293595). The mandible was not recovered. The archeological age is unknown. Both second permanent molars have erupted, although the third permanent molar is still deep in the maxilla. Most of the teeth have been lost postmortem. The alveoli indicate that the normal number of teeth

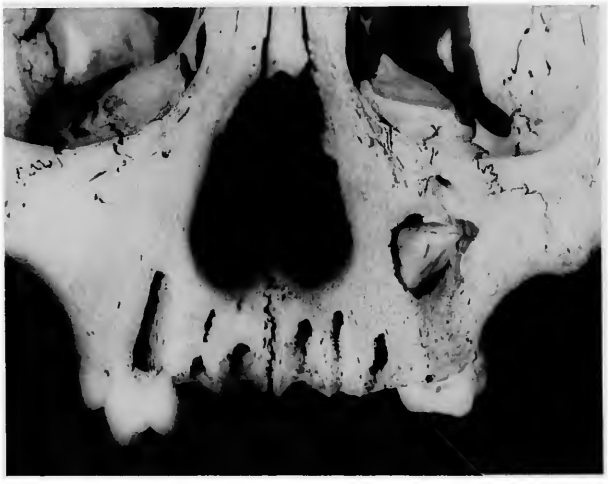


FIGURE 746.—Heterotopic supernumerary canine projecting through the left maxilla. (Child's skull about 12 years of age from Chicama, Peru, NMNH 293595.)

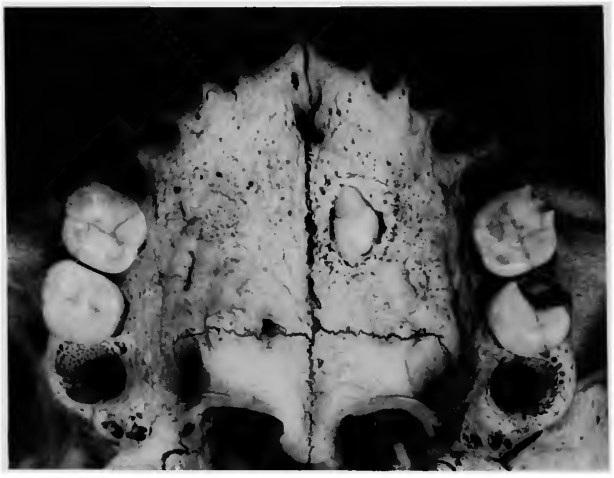


FIGURE 747.—Heterotopic supernumerary canine projecting through the left palate (Adult skull from Pachacamac, Peru, NMNH 267104.)

were present. However, the alveolus of the left canine indicates that this tooth was much smaller than normal. Projecting through the maxilla in the region over the first premolar is the crown of a normal sized canine (Figure 746). An X-ray film reveals the relatively normal size and shape of the canine root.

The second example of heterotopic hyperodontia is an adult skull from Pachacamac, Peru (NMNH 267104). The mandible was not recovered. Only the first and second molars are present, the remaining teeth were lost postmortem. The alveoli are intact and indicate a normal number and root morphology for the missing teeth. A dental element with the crown and root morphology of a canine projects partially through the left palate (Figure 747).

It should be emphasized that not all heterotopic teeth are supernumerary. Another adult skull from Pachacamac, Peru (NMNH 266005) illustrates this point. Except for a heterotopic central incisor, all the teeth are missing. Some alveoli are present indicating postmortem loss. However, there is also alveolar bone resorption, indicative of antemortem tooth loss as well. There is a dental element with the morphology of an upper central incisor embedded in the maxilla in the region above where the central incisor should have been. The crown of the tooth projects buccally (Figure 748). The tooth root is shortened but may project posteriorly. This is a case where the tooth erupted in the wrong direction, but there is no evidence that the tooth was supernumerary.

An example of a supernumerary tooth erupting in the normal orientation and in the alveolar region (normotopic) is seen in the mandible of an adult female skull from the Virgin Islands (NMNH 385695). The specimen is from the historic period. There is some antemortem tooth loss, but the only evidence of supernumerary teeth is a fourth molar of the left mandible (Figure 749). This condition is not present on the right side.

Retention of deciduous teeth may lead to extra teeth in the tooth row. This condition can be

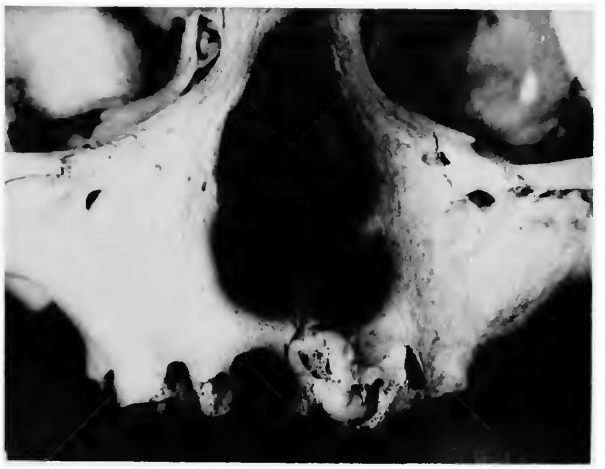


FIGURE 748.—Heterotopic maxillary central incisor, probably not supernumerary. Crown of tooth is in contact with anterior nasal spine. (Adult skull from Pachacamac, Peru, NMNH 266005.)

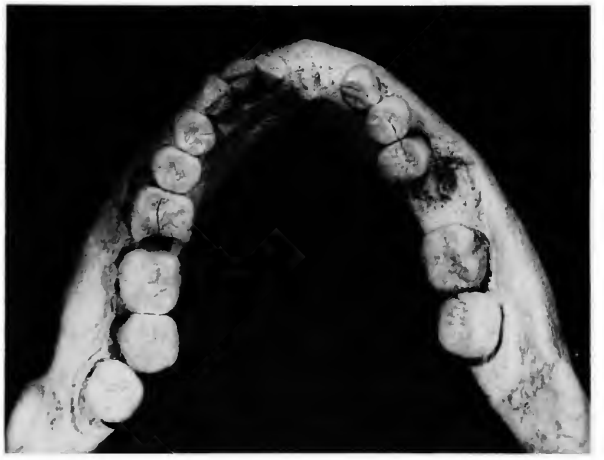
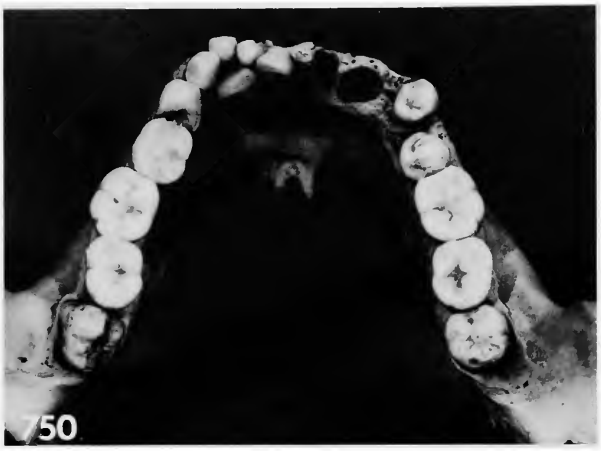
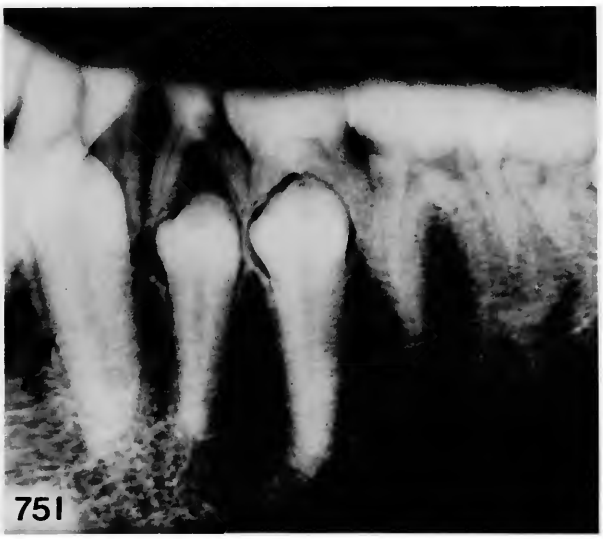


FIGURE 749.—Supernumerary molar on the left side of a mandible. (Adult female skeleton from an archeological site in the Virgin Islands, NMNH 385695.)

observed in the mandible of a young adult specimen from Chicama, Peru (NMNH 264521). There is no skull associated with the mandible. Age cannot be determined precisely, although the third permanent molars have erupted indicating a minimal age of 18 years. The entire primary dentition has been retained on the left side (Figure 750). The permanent incisors have erupted. The central incisor is in a relatively normal posi-

tion. The lateral incisor is displaced lingually. The tip of the impacted permanent canine is projecting through alveolar bone. The premolars are completely impacted beneath the primary molars (Figure 751). On the right side, the permanent dentition has erupted. However, remnants of some of the deciduous roots still project through alveolar bone. The left permanent incisors and canine are missing postmortem.





FIGURES 750, 751.—Retention of primary dentition into adulthood on the left side of the mandible. Although the permanent teeth have erupted in the right side, a few fragments of the deciduous roots remain embedded in alveolar bone: 750, Occlusal view. 751, X-ray of left side; note impacted permanent canine (arrow) and premolars. (Adult mandible from Chicama, Peru, NMNH 264521.)

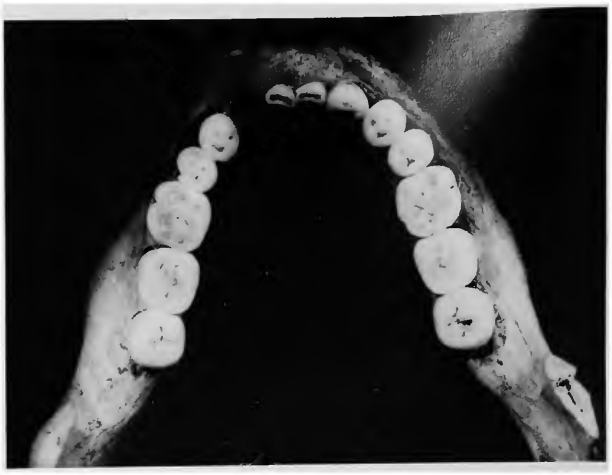


FIGURE 752.—Dental agenesis of the mandibular lateral incisors, occlusal view. (Adult female from Mobridge, South Dakota, USA, NMNH 325417.)

Dental agenesis (hypodontia) is rather common in archeological specimens. It is most commonly seen in the failure of the third permanent molar to develop (Brothwell, Carbonell, and Goose, in Brothwell, 1963:182). An example of dental agenesis of the lower lateral incisors is found in an adult female skull from Mobridge, South Dakota, USA (NMNH 325417). The maxillary dentition is normal. On the mandible, the canines are adjacent to the central incisors (Figure 752). An X-ray film of the anterior teeth shows no evidence

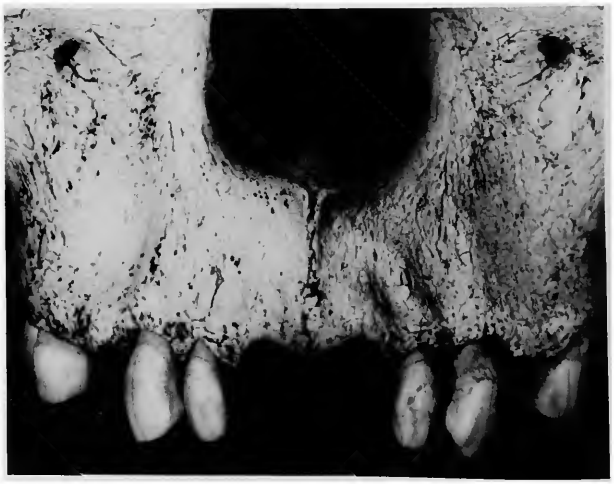


FIGURE 753.—Abnormally small maxillary lateral incisors in an adult skull from Chicama, Peru (NMNH 264518.)

of impacted teeth, indicating that the lateral incisors did not develop.

Abnormal Size and Shape of Teeth

Abnormal variations in size and shape of teeth should be readily recognizable to people familiar with skeletal specimens. One example is sufficient to illustrate this condition. This specimen is an adult skull, lacking the mandible, from Chicama, Peru (NMNH 264518). Many teeth are missing postmortem, although the alveoli indicate a normal number of teeth. The upper lateral incisors are smaller than normal (Figure 753).

Dental Anomalies

Three cases of dental fusion illustrate the basic features of this dental anomaly. The first of these is seen in the mandible of a young child about 1½ years of age. The specimen is from the historic period in Nebraska, USA (NMNH 243355). Fusion occurred between the central and lateral right incisors (Figure 754). The second case is from an archeological site in New Mexico, USA (NMNH 269221) and involves the deciduous mandibular right lateral incisor and canine (Figure 755). The child was about 8 years of age. An X-ray film indicates that the roots were fused. The third case is from an adult dentition in the mandible of a skeleton from an archeological site in New Jersey, USA (NMNH 285307). Fusion has taken place between the right lateral incisor and canine and has created a very large tooth, which might, on superficial inspection, be confused with macrodontia (Figure 756). The roots of the two teeth are fused.

Another dental anomaly involves the development of "enamel pearls" on the surface of the roots. An example of multiple enamel pearls is seen in the mandible of an adult female skeleton from South Dakota, USA (NMNH 325367). An enamel pearl developed on the distal surface of the roots of the left second and third molars (Figures 757, 758). The growth encroached on the alveolar bone creating a marked cavity adjacent to the second molar but only a slight depression adjacent to the third molar.

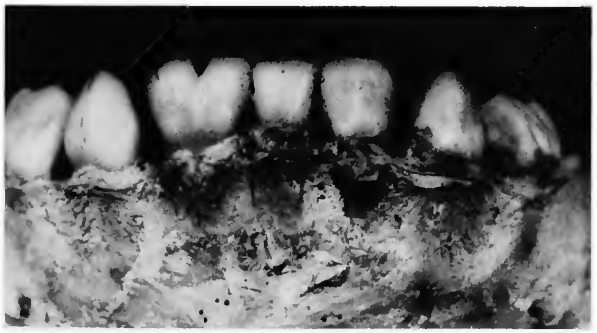


FIGURE 754.—Fusion of the mandibular right incisors. (American Indian child about 1½ years of age from a historic period site in Nebraska, USA, NMNH 243355.)

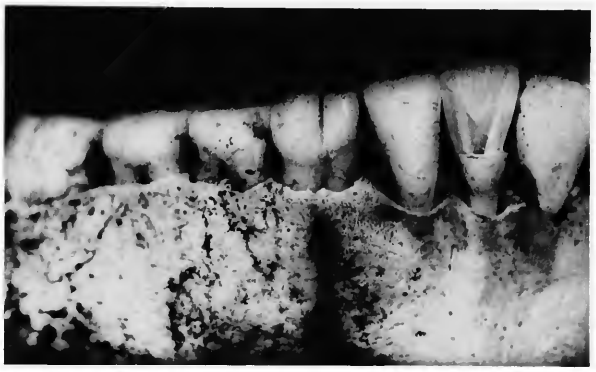


FIGURE 755.—Fusion of the mandibular deciduous right lateral incisor and canine. (Child, about 8 years of age, from an archeological site in New Mexico, USA, NMNH 269221.)

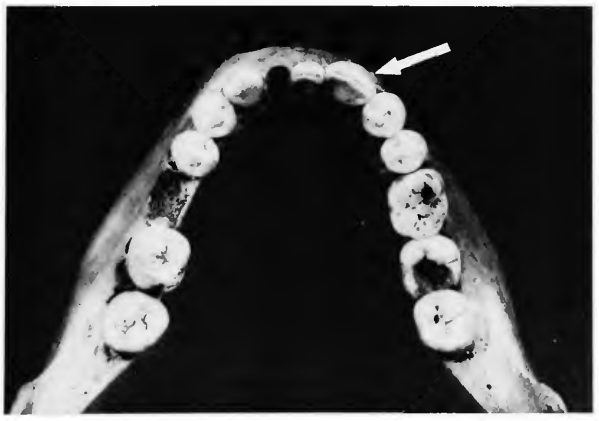
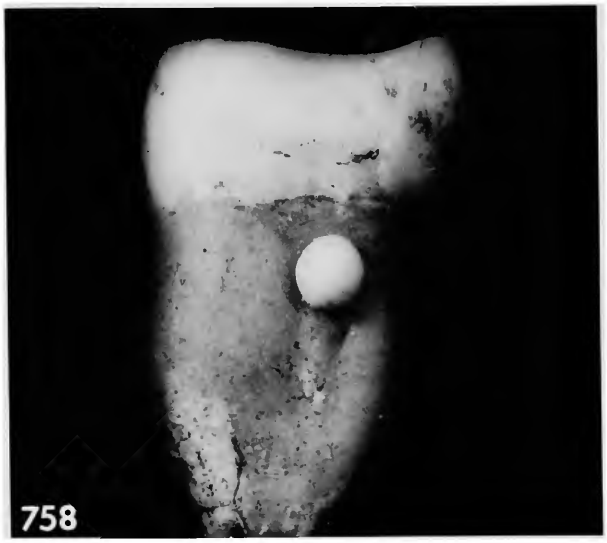


FIGURE 756.—Fusion of the permanent mandibular, right lateral incisor and canine (arrow). An X-ray reveals that the roots are fused. This fusion should be distinguished from abnormally large teeth (macrodontia). (Specimen is from an archeological site in New Jersey, USA, NMNH 285307.)



FIGURES 757, 758.—Enamel pearls on the distal root surface of the mandibular, left permanent second and third molars: 757, Molars, in situ; note that the enamel pearl encroaches on alveolar bone. 758, Detail of enamel pearl on second molar. (Specimen from archeological site in South Dakota, USA, NMNH 325367.)

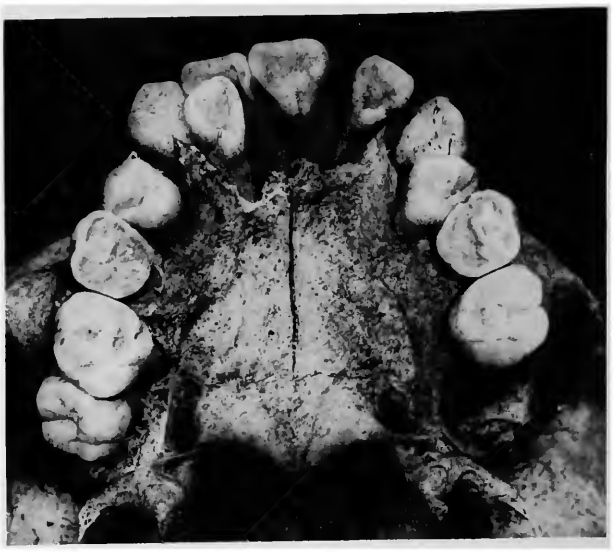


FIGURE 759.—Severe crowding of the maxillary dentition in a child's skull about 10 years of age. Note the displacement of the anterior teeth. (Specimen is from an archeological site in Canaveral, Florida, USA, NMNH 377496.)

Dental Crowding

A small skull, lacking the mandible, of a child from an archeological site in Canaveral, Florida, USA (NMNH 377496) illustrates the problems that develop when normal or large permanent teeth erupt in a small jaw (Figure 759). The dental eruption stage is somewhat anomalous due to the retention of the second deciduous molars; however, the estimated age would be about 10 years. The fragmentary long bones are small and gracile. The estimated age, based on femur length, is about 8 years suggesting subnormal growth. The skull is small with a maximum length of 149 millimeters and a maximum breadth of 131 millimeters. The right lateral incisor is displaced lingually due to inadequate space in the maxilla. Although the skeleton is small, there is no evidence of pituitary dysfunction. The probable pituitary dwarf described in the chapter on endocrine disorders did not exhibit serious crowding in the dentition, in part due to the prognathous jaws (see Figures 468, 469).

Dental Trauma

PATHOLOGY

In modern populations the major causes of dental fracture are automobile accidents and injury during participation in sports or the rough and tumble play of children. Pindborg (1970:277)

reports that the central maxillary incisors are most frequently fractured, and males fracture teeth twice as often as females. Enamel, dentin, pulp, and cementum may be involved in fracture. The crown is involved in about 98 percent of the cases of dental fracture, the root in about 2 percent (Pindborg, 1970:279). Crown fractures most frequently involve both the enamel and dentin, less commonly the enamel only. Exposure of the dentinal tubules may permit infection of the pulp and this may lead to periapical abscess.

Paleopathology

Antemortem dental fracture in dry bone specimens may be very difficult to differentiate from postmortem damage in the ground or in museum storage. Most often postmortem damage involves the enamel, which chips off easily. However, major portions of the tooth may split involving the dentin and pulp cavity. One possible criterion for making the distinction between ante- and postmortem fracture is the smoothing of the fractured edges of the tooth, which would occur in antemortem fracture if sufficient time had elapsed between the fracture and death.

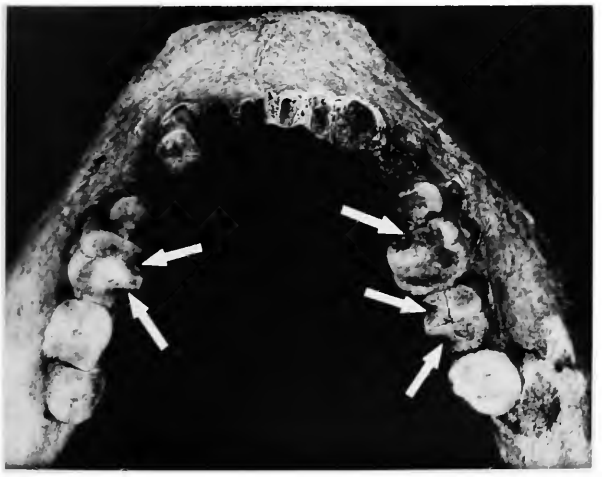


Figure 760.—Possible antemortem fracture of the dental crowns (arrows), occlusal view of mandible. Note the rounded edges of the remaining crowns suggesting wear of the teeth after the fracture. (Adult female from archeological site on Perico Island, Florida, USA, NMNH 373547.)

Figure 761.—Dental mutilation of the maxillary left incisors. (Adult female, probably a Negro burial, from Saint Croix, Virgin Islands, NMNH 382048.)

A possible example of antemortem tooth fracture is in the mandibular teeth of an archeological skeleton from Perico Island, Florida, USA (NMNH 373547). Fairly substantial portions of the crowns of the left and right second premolars, first molars, and right second molar have been chipped off primarily on the buccal side (Figure 760). The fractured edges of the affected crowns have been smoothed, suggesting attrition of the broken surface after the fracture. All the teeth are worn, indicating a fairly abrasive diet, so attrition of broken edges could be expected if antemortem.

Fracture of teeth may be induced intentionally as an expression of a cultural practice. Two basic cultural practices occur. The first of these is ritual ablation or removal of teeth (Baudouin, 1923). Typically this involves the incisors, which may be extracted or broken off leaving the roots. The second practice is mutilation of the teeth. This practice also, typically, involves the incisors. The teeth may be ground, filed, or broken to create a special shape or they may be inlayed with metals or mineral materials. This practice is primarily associated with the continents of Africa and South America with some evidence for its presence in the indigenous cultures of North America (e.g., Jhering, 1882; Starr, 1909; Stewart, 1944;

Romero, 1958, 1970). Tooth filing, associated with blackening with burnt oil, is common in Sumatra and probably throughout much of the Pacific.

The variation in the practice of dental mutilation is remarkable, although beyond the objectives of this book. One previously unpublished case will serve to illustrate this type of dental trauma. The specimen is from Saint Croix in the Virgin Islands and was discovered when it was washed out by a storm (NMNH 382048). The morphology of the skull is distinctively Negro and may have been the remains of one of the slaves in the slave camps located on the island. The archeological age is not known but is certainly in the historic period. The skull appears to be that of an adult female. The maxillary right incisors and third molar are missing postmortem, as is the mandibular left central and right incisors and the right third molar. The maxillary left incisors have been mutilated, creating a point with its apex between the two teeth (Figure 761).

Dental Attrition

Pathology

Pindborg (1970:294) makes the distinction between attrition resulting from natural mastication (physiologic attrition) and attrition, often localized, caused by abnormal use or position of teeth (pathologic attrition). The factors contributing to attrition are varied and include the occlusion of the opposing teeth, the quality of the teeth, abrasives in the diet, and the use of abrading instruments, such as a pipe or the use of teeth to hold objects.

Attrition of the occlusal surfaces may destroy the enamel, exposing the underlying dentin. Attrition continued beyond this point may threaten exposure of the pulp cavity. In response to attrition, the odontoblasts begin forming secondary dentin in the threatened areas of the pulp cavity, and attrition may expose the secondary dentin. If attrition occurs at a rate faster than the formation of secondary dentin, the pulp cavity will be exposed creating the potential for infection of the pulp and supporting alveolar bone.

Attrition also takes place on the opposing surfaces of adjacent teeth (interproximal attrition). This type of wear is due to the contact between teeth and the slight movement of the teeth that accompanies chewing. This type of attrition is relatively slight, rarely if ever exposing the dentin.

Paleopathology

Dental attrition is a well-known phenomenon in archeological skeletons. Because of its association with biological aging, the degree of wear has been used as a method for estimating the age of the skeleton (e.g., Brothwell 1965a:69). Brothwell notes (1965a:68) that methods of aging based on attrition and developed for one population cannot be applied to a population where the conditions contributing to wear are different.

The severity of attrition is much greater in archeological skeletons than most modern skeletons (Pindborg, 1970:296). Mehta and Evans (1966:254) studied attrition in archeological skeletons from Arkansas, USA. A majority of teeth had dentin exposure (62 percent). This finding is similar to attrition among modern primitive groups, such as the Australian Aborigines.

Three archeological cases illustrate the essential features of dental attrition. The first case illustrates extensive attrition with secondary dentin formation but no pulp cavity exposure (Figure 762). The specimen is the mandible of an adult male from an archeological site in South Dakota, USA (NMNH 325360). The third molars apparently did not develop. The left central incisor and second premolar are missing postmortem.

A rather unusual pattern of attrition occurs on the mandibular central incisors of an adult female skull from an archeological site in New Mexico, USA (NMNH 262915). While attrition has exposed secondary dentin in the maxillary and mandibular incisors, the lower central incisors are beveled anteriorly (Figure 763). This type of wear

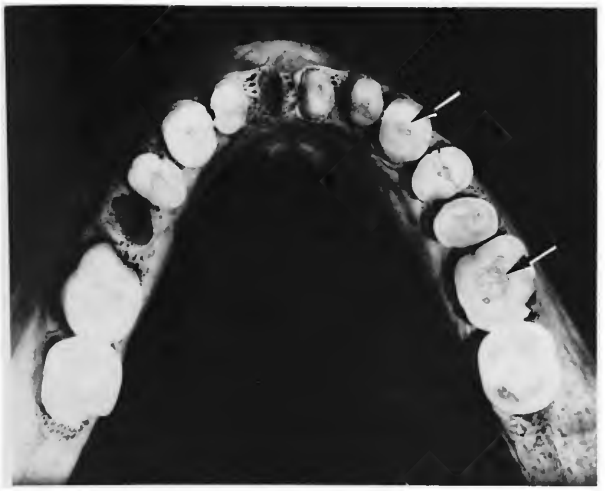


FIGURE 762.—Marked dental attrition and secondary dentin formation (arrows). Note large darkened area of secondary dentin in the center of the crowns of the first molars. (Specimen from an archeological site in South Dakota, USA, NMNH 325360.)

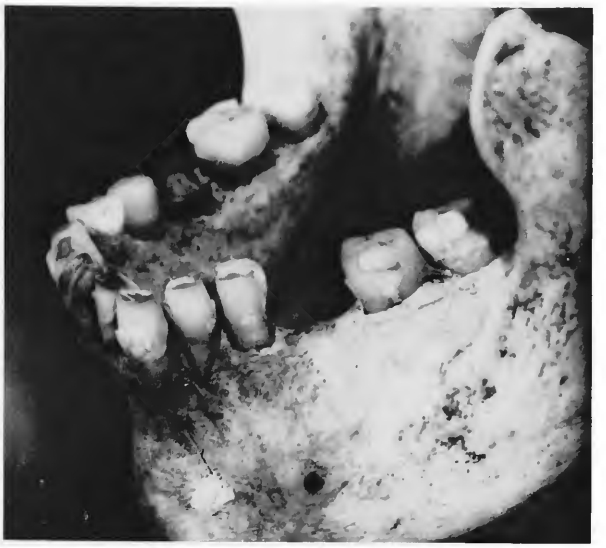


FIGURE 763.—Angular attrition of the mandibular central incisors, probably due to wear from causes other than chewing food. (Adult female, Pueblo Indian, New Mexico, USA, NMNH 262915.)

is suggestive of attrition due to the use of teeth for purposes other than chewing food.

Marked antemortem tooth loss, severe attrition complicated by pulp cavity exposure and periapical abscesses are seen in the dentition of an adult male skull from the pre-Columbian site at Puye, New Mexico (NMNH 262957). On the maxilla the incisors and canines are present but badly worn. The incisors have exposed secondary dentin and the canines have wear exposing the pulp cavity on the left. There is carious destruction of the crown of the right canine. There is a periapical abscess associated with the left canine. The remaining teeth are missing antemortem with considerable alveolar resorption. On the mandible the central incisors are missing antemortem as apparently are the third molars. The remaining incisor crowns are worn away with exposure of secondary dentin. The canine crowns are almost worn away. There is pulp exposure of the left lateral incisor and left canine with a periapical abscess associated with both teeth (Figure 764). The crown of the left first premolar has been destroyed by caries.

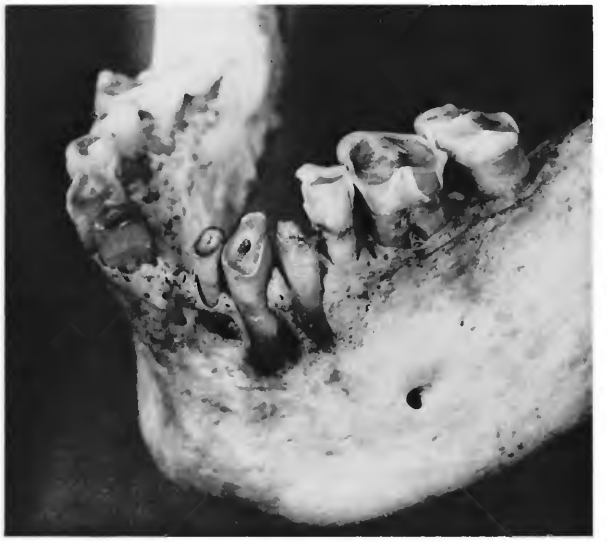


FIGURE 764.—Severe attrition of the mandibular incisors and canines with exposure of secondary dentin. The pulp cavities of the left lateral incisor and left canine are exposed; both teeth have a periapical abscess. (Adult male from the archeological site of Puye, New Mexico, NMNH 262957.)

Dental Discoloration

Pathology

Discoloration may occur in dental tissue during formation of the teeth or after the teeth have erupted. It may affect otherwise normal teeth or teeth with defective enamel and/or dentin. Some of the conditions that result in discoloration in modern cases are the effect of dental or medical intervention and are of no significance in archeological skeletons. Conditions that can result in color changes of teeth and that might be seen in archeological specimens include: fluorosis, congenital heart disease, erythroblastosis fetalis, neonatal hepatitis, congenital defects of the bile duct, porphyria, hemorrhage or necrosis of the pulp, the use of tobacco, and betel (Pindborg, 1970: 212, 221). A summary of pattern and color changes is presented in Table 14.

TABLE 14.—Abnormal conditions resulting in discoloration of the teeth, with typical pattern and color change (based primarily on Pindborg, 1970:211–224)

Cause of discoloration	*Pattern*	*Color*
Fluorosis	Mottled	Yellow to brown
Congenital heart disease	Diffuse	Bluish white
Erythroblastosis fetalis	Diffuse	Green to yellow, brown or gray
Neonatal hepatitis	Diffuse	Yellowish brown
Congenital defect of bile duct	Diffuse	Green (particularly the roots)
Porphyria	Striated	Pinkish brown (roots indigo)
Hemorrhage or necrosis of pulp	Diffuse	Light brown to gray
Tobacco	Diffuse	Brown
Betel	Diffuse	Dark brown (occlusal surface and roots tend not to be affected)

Paleopathology

Discolorations of teeth and their pathological implications have received little attention in the literature on paleopathology. With the exception of staining from betel chewing, I am unable to provide examples of paleopathological specimens that illustrate these conditions. Hopefully the brief review above will call attention to the potential of observing teeth for color changes as a supplemental diagnostic feature.

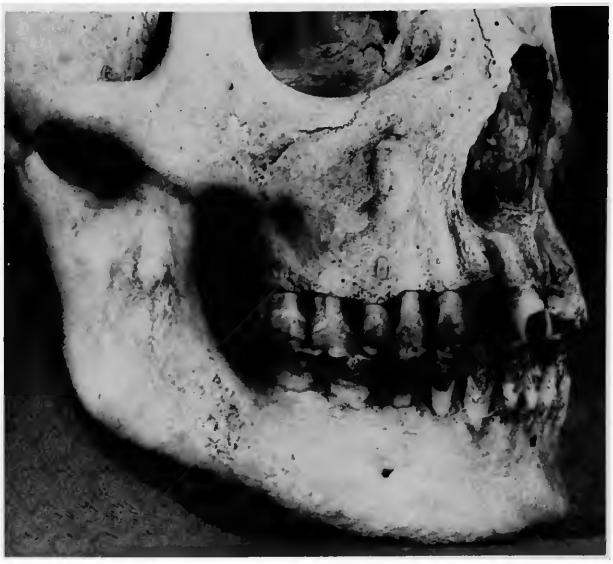


FIGURE 765.—Dental discoloration of the crowns from chewing betel nut; roots and occlusal surfaces tend not to be affected. (Adult male from Malay Archipelago, NMNH 225052.)

The example of discoloration from chewing betel nut or intentional staining with blackened coconut oil is seen in a skull and mandible of an adult male from the Malay Archipelago (NMNH 225052). The specimen was collected in the late nineteenth century and little is known about the cultural association. Accession records indicate the tribal association is the Buginese (Boeginees). Some of the teeth are missing postmortem. Those that remain show little evidence of wear or caries. There has been some alveolar resorption suggestive of periodontal disease. The maxillary right central incisor has an unusual wear pattern on the buccal side suggestive of intentional filing. Staining has resulted in a dark brown color of all crown surfaces (Figure 765) except most of the occlusal surfaces. An exception to this is the maxillary right third molar. However, the opposing molar on the mandible had not erupted indicating that the lack of staining on the occlusal surface was due to chewing. This conclusion is supported by the presence of slight staining deep in the crevices of the premolars and molars.

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