Essentials of Orthopedic Surgery

Third Edition

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This text is dedicated to
Star Conway Wiesel and Elizabeth Jane Delahay
as they begin their nursing careers.
These two very capable, caring young people
represent the bright future of health care.

Sam W. Wiesel, MD
John N. Delahay, MD

Preface

The third edition of the *Essentials of Orthopedic Surgery* provides a concise overview of orthopedic surgery directed toward third- and fourth-year medical students. In this edition, physical diagnosis is a subsection in each chapter, which we believe gives better continuity. Additionally, at the end of each chapter we have created a number of multiple-choice questions considered appropriate for medical students to be able to answer.

Each chapter has been revised to reflect updated material and, as in previous editions, we have kept to a standardized format as much as possible. The topics are presented from a straightforward practical point-of-view, with the material being condensed to its most salient features.

Algorithms are at the heart of each chapter, with the decision points being based on practice standards and guidelines. This format allows the student, when confronted with a specific clinical problem, to formulate both a diagnostic plan and a treatment plan.

Also, we have enjoyed working with our new publisher—Springer—and with Robert Albano as well as Sadie Forrester, who have guided this text to publication.

Finally, and most importantly, it has been again a very exciting and stimulating experience to work with all the members of the Department of Orthopaedics of Georgetown University Medical Center. Since the last edition we have welcomed seven new members to the faculty, each a subspecialist. Everyone has given very generously of their time. We are most appreciative of each contribution and are proud of the final text.

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1
Basic Science of Bone and Cartilage Metabolism

John N. Delahay

Normal Bone Growth and Development

Bone is a biphasic connective tissue consisting of an inorganic mineral phase and an organic matrix phase. The hardness of bone allows it to provide several specialized mechanical functions: the protection of internal organs, the scaffold that provides points of attachment for other structural elements, and the levers needed to improve the efficiency of muscle action. In addition, bone serves two biologic functions: a site for hematopoietic activity and a reservoir of minerals needed for metabolic interchange.

Embryology

The major components of the musculoskeletal system originate from the mesoderm layer of the trilaminar embryo. This "middle layer" is populated by mesenchymal cells that are totipotent and capable of differentiating into a number of tissues. The sequence of events important in bone growth and development begins with the appearance of the limb bud around the fifth week of life. It is at that time that a tubular condensation of mesenchyme develops centrally in the limb bud. Discrete areas, called interzones, are seen between these condensations (Fig. 1-1) and represent the primitive joints.

During the sixth week, the mesenchyme differentiates into cartilage through the process of chondrification (Fig. 1-2). Interstitial and appositional growth occurs from within and from the surface, respectively. In the seventh week, the cartilage model is penetrated by a vascular spindle, which occurs coincidentally with the necrosis of the central cartilage cells. Once this vascular spindle is established, the central portion of the model is populated by osteoblasts. Matrix is secreted and this in turn is ossified, making immature (woven) bone.

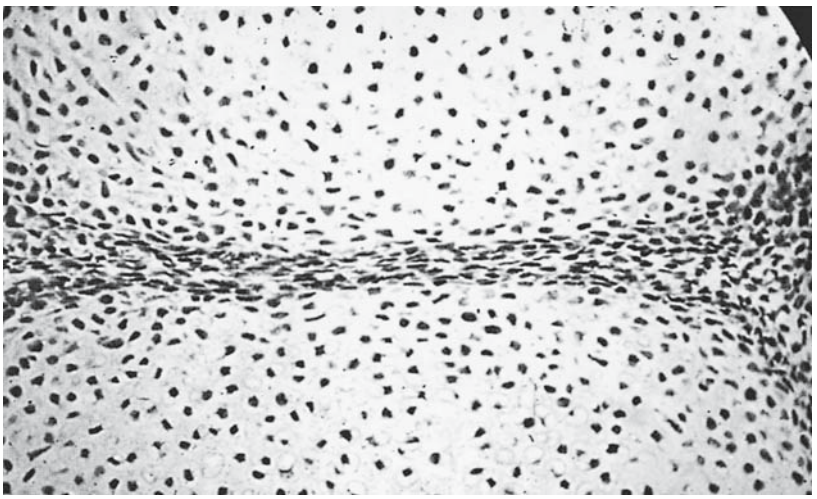


FIGURE 1-1. Histologic study of fetus, approximately 6 weeks gestation, depicting early joint formation. Note the identifiable cartilage and the condensed mesenchymal tissue of the interzone destined to become the joint. (From Bogumill GP. Orthopaedic Pathology: A Synopsis with Clinical Radiographic Correlation. Philadelphia: Saunders, 1984. Reprinted by permission.)

FIGURE 1-2. Histologic study of fetus, approximately 8 weeks gestation. Earliest ossification is depicted here. A sleeve, or collar, of bone is present on the outer surface of the cartilage model. (From Bogumill GP. Orthopaedic Pathology: A Synopsis with Clinical Radiographic Correlation. Philadelphia: Saunders, 1984. Reprinted by permission.)

Once the central portion of the model is ossified, it is referred to as a primary ossification center (Fig. 1-3). Further ossification of the skeleton occurs via one of two mechanisms: (1) enchondral ossification within a cartilage model (i.e., long bones), and (2) intramembranous ossification within a mesenchymal model (i.e., most flat bones and the clavicle).

From the second through the sixth embryonic months, progressive changes occur in the tubular bones. First, the central (medullary) canal cavitates, leaving a hollow tube of bone with a large mass of cartilage persisting at each end (Fig. 1-4). Within these masses of cartilage, the secondary ossification center, or epiphysis, will form (Fig. 1-5). A cartilage plate, the physis or growth plate (Fig. 1-6), persists between the developing epiphysis and metaphysis. This structure is responsible for growth in length, whereas the covering of the bone, the periosteum, is primarily responsible for growth in girth.

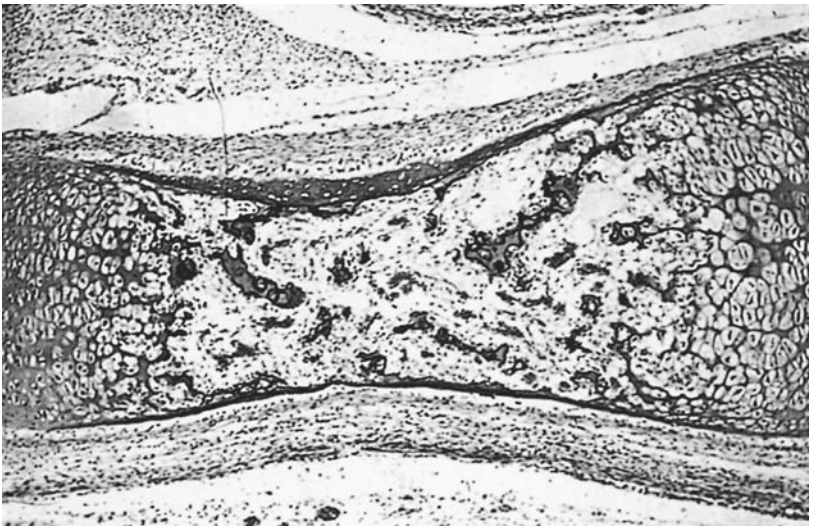


FIGURE 1-3. Primary ossification center of fetus, approximately 14 weeks gestation. The cartilage cells have been removed almost entirely from the center, leaving remnants of acellular cartilage matrix. Bone deposits on the cartilage remnants will form primary trabeculae. Note that the primary sleeve, or collar, of bone has extended along both margins and is located adjacent to the hypertrophied cartilage at each epiphyseal end. (From Bogumill GP. Orthopaedic Pathology: A Synopsis with Clinical Radiographic Correlation. Philadelphia: Saunders, 1984. Reprinted by permission.)

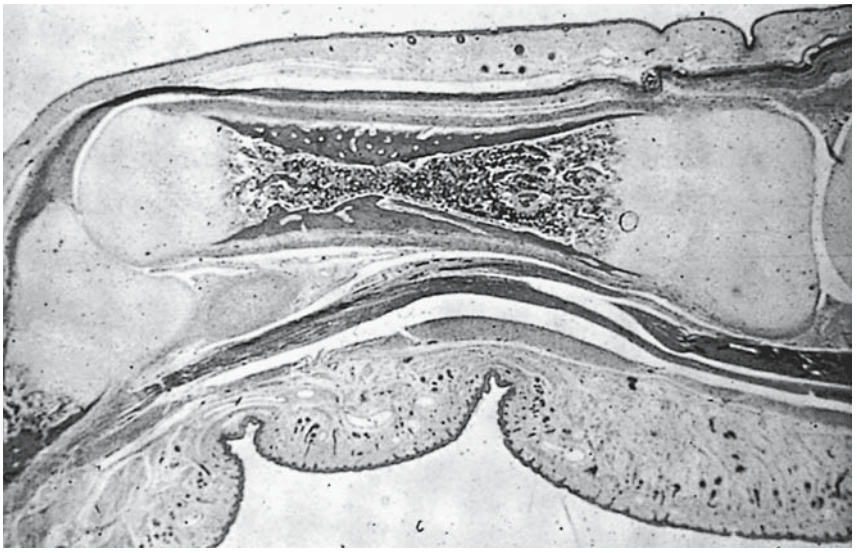
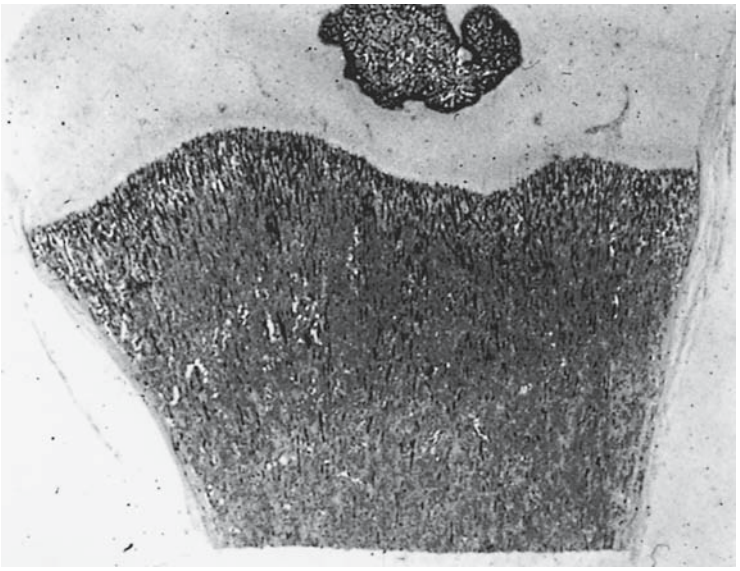


FIGURE 1-4. Primary ossification center, near term. There is complete replacement of cartilage in the diaphyseal portion of the cartilage model. The remaining cartilage is confined to both epiphyseal ends of the model. Note the increasing thickness of the cortical portion of bone, which is a result of conversion of periosteum to bone. A light-staining cambium layer is identifiable. The narrowest portion of the shaft is the site of initial vascular invasion and remains identifiable throughout life in many bones, especially in hands and feet. The eccentric position of this narrowed area indicates the disproportionate contribution to growth in length from each epiphysis. (From Bogumill GP. Orthopaedic Pathology: A Synopsis with Clinical Radiographic Correlation. Philadelphia: Saunders, 1984. Reprinted by permission.)



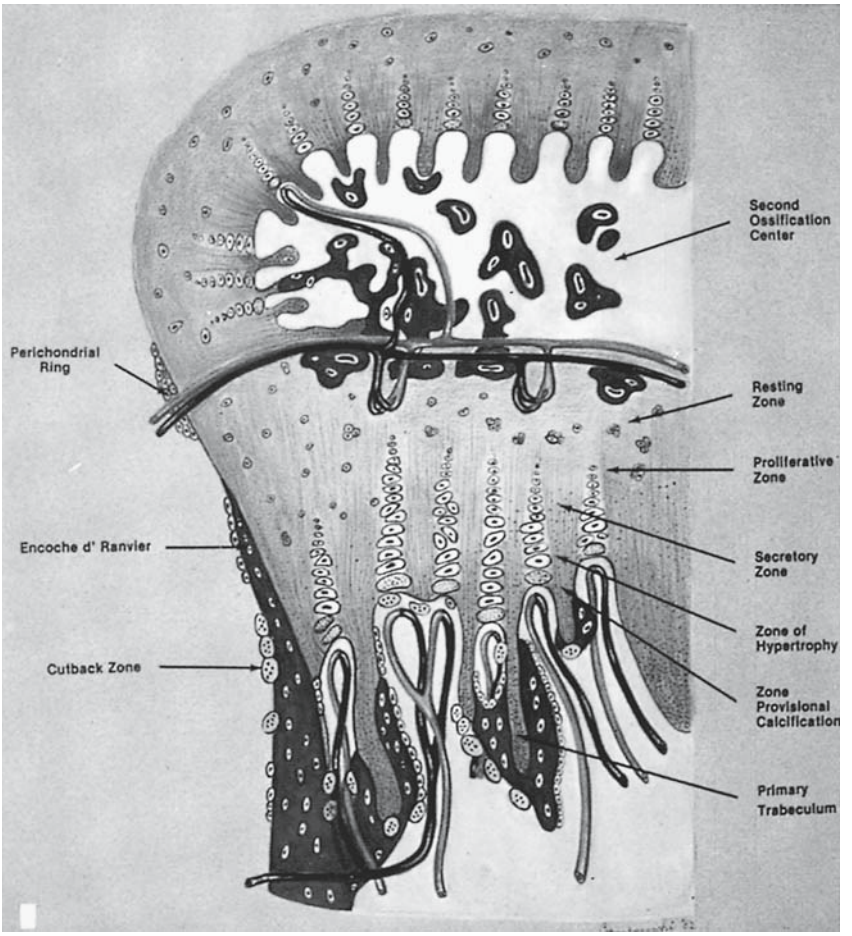


FIGURE 1-6. Schematic diagram of growth plate, consisting of resting zone, proliferative zone, secretory zone, zone of hypertrophy, and zone of calcification. The cross-sectional view helps place events at the growth plate in three-dimensional perspective. (From Bogumill GP. Orthopaedic Pathology: A Synopsis with Clinical Radiographic Correlation. Philadelphia: Saunders, 1984. Reprinted by permission.)

FIGURE 1-5. Early secondary ossification center of mature fetus. The formation of the secondary ossification centers in the lower tibia and upper femur coincide with fetal maturity. The secondary center begins not in the center of the epiphysis but nearer the growth plate. Expansion, therefore, is eccentric. (From Bogumill GP. Orthopaedic Pathology: A Synopsis with Clinical Radiographic Correlation. Philadelphia: Saunders, 1984. Reprinted by permission.)

Postnatal Development

The physis and the periosteum continue to function postnatally in the growth and development of the infantile skeleton. Numerous local and systemic factors impact on their activity; vascular, hormonal, and genetic effects all play important roles. In essence, the reworking or remodeling of bone that is already present occurs so that the bone can meet the mechanical and biologic demands placed on it.

Bone: The Tissue

Bone, whether it is immature or mature, consists of cells and a biphasic blend of mineral and matrix that coexist in a very exact relationship. The matrix phase consists of collagen and glycosaminoglycans, which are dimeric disaccharides. Both are products of the osteoblast. Calcium hydroxyapatite is the basic mineral crystal in bone. Despite the presence of some less structured amorphous calcium phosphate, the bulk of calcium in the skeletal reservoir is bound in the crystals of hydroxyapatite.

Osteoblasts are bone-forming cells that secrete the matrix components described. As ossification progresses, the osteoblasts become trapped in the matrix they produce and are then referred to as osteocytes. These cells are rather inert but are capable of a small degree of bone resorption. Osteoclasts are those cells whose primary function is the degradation and removal of mineralized bone. It is important to remember that the osteoclasts can remove only mineralized bone, and not unmineralized matrix.

Bone Organization

Microscopically, bone is generally described as mature or immature. Mature bone (Fig. 1-7) has an ordered lamellar arrangement of Haversian systems and canalicular communications that give it its classic histologic appearance. Immature bone (Fig. 1-8), in contrast, has a much more random appearance of collagen fibers dispersed in a matrix of irregularly spaced cells. It is produced rapidly by osteoblasts and "remodeled" by the local cell population, until the mature lamellar pattern is achieved. Immature bone is seen in the adult skeleton only under pathologic conditions (i.e., fracture callus, osteogenic sarcoma, myositis, etc.). Macroscopically (Fig. 1-9), the lamellar bone is configured either as dense cortical bone or as delicate spicules called trabeculae. In both areas, the cortex and the trabecular metaphysis, the bone is histologically the same (i.e., mature lamellar bone).

Turnover and Remodeling

Although the tendency is to think of adult bone as an inert tissue, nothing could be further from the truth. Throughout adult life there is a constant ebb and flow of bone formation and bone resorption. These two processes

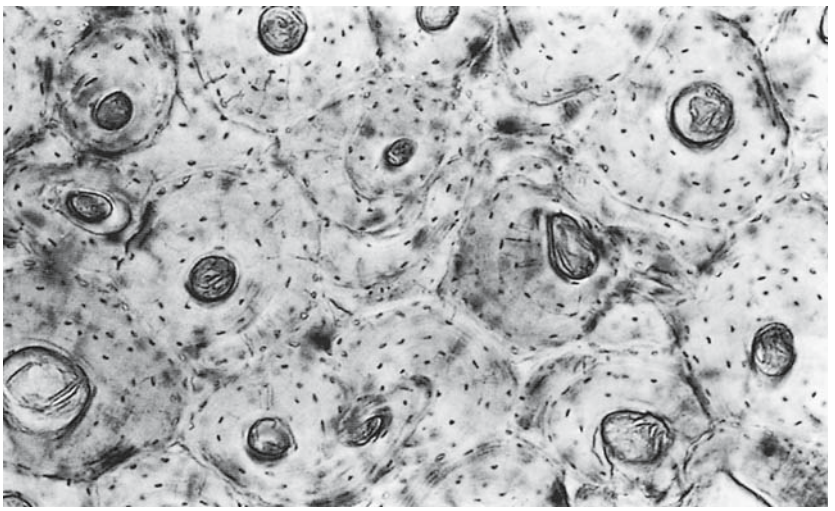


FIGURE 1-7. Mature bone: osteonal structure as seen in undecalcified material. Numerous interstitial fragments (osteonal fragments without an associated Haversian canal) are readily observed. (From Bogumill GP. Orthopaedic Pathology: A Synopsis with Clinical Radiographic Correlation. Philadelphia: Saunders, 1984. Reprinted by permission.)

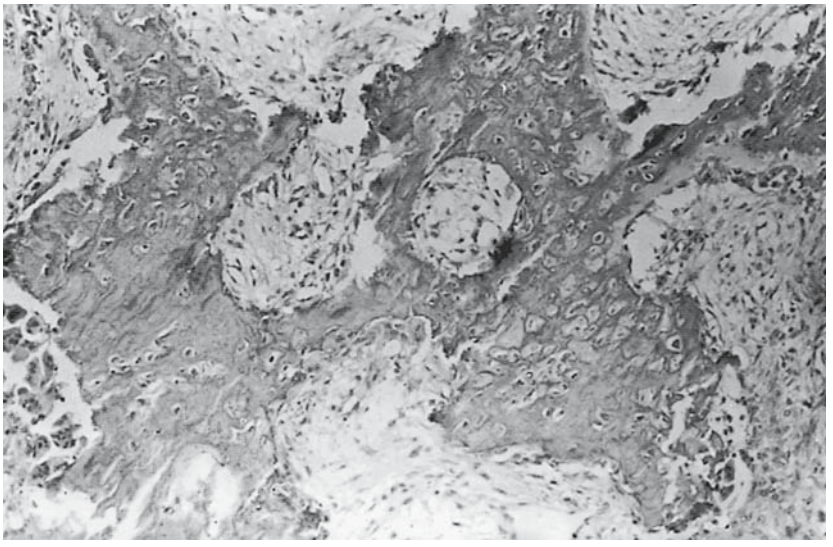


FIGURE 1-8. Immature bone (early callus). Note the large number of osteoblasts and osteocytes. (From Bogumill GP. Orthopaedic Pathology: A Synopsis with Clinical Radiographic Correlation. Philadelphia: Saunders, 1984. Reprinted by permission.)

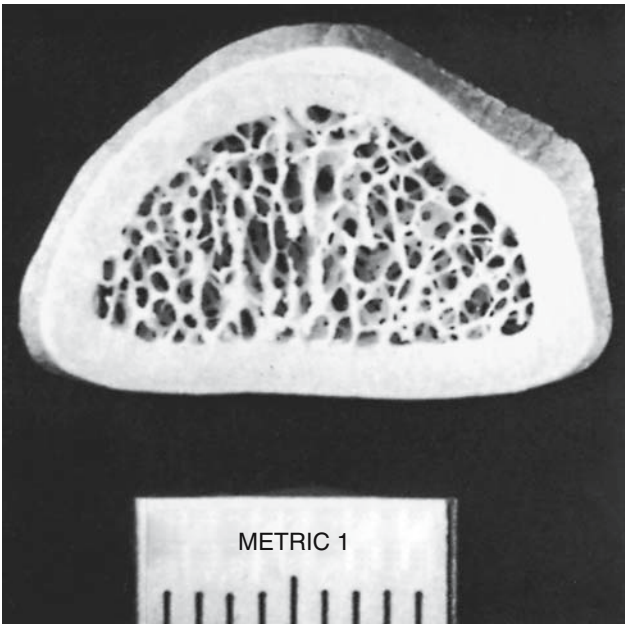


FIGURE 1-9. Cross section of the radius at the distal metaphysis. The majority of bone is cortical bone, in which the annual rate of turnover is only 2%.

are delicately balanced and keep the skeletal mass in a state of equilibrium. A number of authors have popularized the concept of "coupling"; bone formation and bone resorption generally increase or decrease in the same direction. When one process increases, so does the other, and vice versa. It is important, however, to consider the net effect of the rate changes in these two processes. For example, in osteoporosis, both formation and bone resorption increase, but resorption increases at a much greater rate, so that despite a coupled increase in bone formation the net effect is an overall decrease in bone mass. A number of factors, systemic and local, affect these processes and hence impact bone turnover and remodeling. Perhaps the most well defined factor is mechanical stress, which forms the basis for the classic Wolff's law. Simply stated, trabecular, and to a lesser degree cortical, bone remodels along lines of mechanical stress. Bone forms where it is needed to meet mechanical demands, and it is resorbed where the need is less. Current research suggests that bone functions as a transducer, converting mechanical energy from the applied load into electrical energy and a voltage gradient. In turn, this voltage gradient that is generated modulates cellular differentiation. Osteoblastic activity is thus seen in regions where the mechanical demands are the greatest. Osteoclas-

tic activity predominates the pattern when those mechanical demands decrease and less bone is required. This phenomenon has been called the "piezoelectric effect." Specifically, the deformation of bone apatite crystals by superimposed load generates the voltage gradient, which in turn alters the cell population to respond to that load.

Cartilage: The Tissue

Cartilage, like bone, is a connective tissue. Its histologic organization, however, is far less structured. There are three histologic types of cartilage, each serving a different function:

1. *Hyaline cartilage* covers the ends of long bones and provides a smooth, frictionless surface for articulation in a diarthrodial (synovial lined) joint.
2. *Fibrocartilage* is typically found in certain nondiarthrodial joints such as the pubic symphysis. It is also located at the margins of certain diarthrodial joints, forming structures such as the glenoid labrum and acetabular labrum. Following injury to hyaline cartilage, repair of the chondral defect is typically accomplished in the form of fibrocartilage.
3. *Elastic cartilage* is found in certain areas where resiliency is important. Examples include the tip of the nose and the ear lobe.

The most important of the three, hyaline cartilage, is a relatively aneural, avascular, and relatively hypocellular connective tissue. By weight, it is 70% water and 30% ground substance and cells. The ground substance of hyaline cartilage is composed primarily of type II collagen and GAG proteins (glycosaminoglycans). The collagen endows the cartilage with tensile strength, and the GAGs are critical for resiliency.

The cells are called chondrocytes and are dispersed throughout the chondral layers in four zones: tangential (most superficial), transitional, radial, and calcified. These chondrocytes are found in individual lacunae, where they maintain healthy cartilage by actively synthesizing new ground substance components.

The chondral layer receives the bulk of its nutrition by diffusion from the synovial fluid above and from the vasculature at the subchondral plate below. Normal diarthrodial (synovial lined) joint function depends on the presence of normal hyaline cartilage. In its fully hydrated state, hyaline cartilage provides an almost frictionless bearing, hence minimizing wear on the articular surface.

Abnormal Bone Development and Metabolism

Most skeletal diseases are the result of disruption of normal bone growth and development, breakdown of bone once it has been normally formed, or alteration of the normal mechanisms of bone formation or bone resorp-

tion. The etiologies of the pathologic states, as one would expect, are quite varied; but the final manifestations within the musculoskeletal system frequently show striking similarities.

Despite the etiology, damage to the growing skeleton will alter the overall shape of one or more bones, depending on whether the adverse process is localized or generalized. Similarly, disruption of osteoblast function will decrease the amount or the quality of the bone formed. Multiple factors are known to stimulate osteoclast activity, such as parathyroid hormone, the presence of particulate polyethylene, and certain neoplasms, resulting in localized or generalized bone resorption.

As one considers the etiology of skeletal disease, it is helpful to first group the possible differential diagnoses by disease category, which permits one to develop a comprehensive list of possible diagnoses that may explain the findings manifested by the skeleton. The seven disease categories are best remembered using the acronym "VITAMIN":

V, vascular disease
I, infection
T, tumor
A, arthritis
M, metabolic bone disease
I, injury
N, neurodevelopmental causes

The remainder of this chapter focuses on these diagnostic groups and the way in which they affect the skeleton. Specific emphasis is placed on generalized afflictions of the skeleton. In that light, certain disease categories are more likely to adversely affect the skeleton in a generalized fashion, specifically vascular, metabolic, systemic arthritis, and neurodevelopmental etiologies. The other etiologies—infection, injury, and tumor—are more likely to produce localized changes and, therefore, are considered in individual subsequent chapters.

Last, as a reminder, a differential diagnosis is a listing of plausible specific diagnoses that may explain observed findings such as physical or radiographic. It is not adequate to simply list a disease category because appropriate treatment of a given condition depends on identifying a specific etiology.

Metabolic Bone Disease

General Concepts

Disease processes affecting bone often can be understood as a change in the relationship of bone formation and bone resorption. It is therefore important to understand this relationship. Only by doing so can the net effect on the skeleton be appreciated.

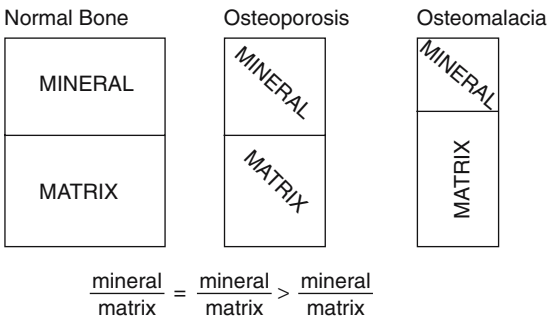


FIGURE 1-10. Ratio of mineral to matrix in certain disease states. In osteoporosis, the ratio remains constant despite an overall decrease in bone mass. However, in osteomalacia there is a decrease in the ratio of mineral to matrix as a result of skeletal demineralization; in addition, there is an overall decrease in bone mass.

The relationship (ratio) of mineral to matrix may be affected in abnormal metabolic states (Fig. 1-10). For example, osteoporosis is a loss of bone mass, but there is an equivalent loss of matrix and mineral; therefore, the ratio remains normal. In contrast, osteomalacia is a relative loss of mineral resulting in a predominance of matrix, hence decreasing the ratio of mineral to matrix. Serum calcium is rarely representative of skeletal activity. Considering that more than 95% of the body's calcium is stored in bone apatite, it is understandable that the 180 mg of ionized plasma calcium represents literally the "tip of the iceberg." Peripheral sampling of the serum calcium provides only a remote clue to the true content of skeletal apatite. It does, however, provide a convenient way to think about and classify metabolic bone disease.

Eucalcemic States: Osteoporosis

As mentioned, osteoporosis is a predominance of bone resorption over bone formation, with the net effect being bone loss (Fig. 1-11). There is a parallel loss of mineral and matrix, so their ratio remains normal. Essentially, osteoporosis is a decrease in bone mass with an increase in cortical porosity and in diaphyseal bone diameter. This latter phenomenon is an attempt by the organism to use what limited bone there is and to disperse it as far as possible from the neutral axis of the long bone. Mechanically, this increases the torsional rigidity of the bone. Numerous etiologies of osteoporosis have been identified (Table 1-1), but clinically most significant is the postmenopausal type, which occurs shortly after the withdrawal of estrogen (naturally or surgically) from the predisposed

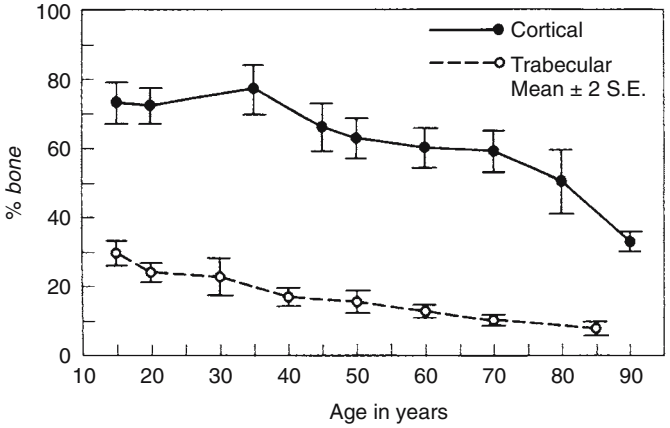


FIGURE 1-11. The relative decrease in cortical and trabecular bone with age in apparently normal persons. Note the relatively rapid loss early in life in trabecular bone and comparatively little loss at this age in cortical bone. The situation is reversed after age 55. (From Jowsey J. Metabolic Diseases of Bone. Philadelphia: Saunders, 1977. Reprinted by permission.)

female (Table 1-2). The yearly cost in dollars, as well as pain and suffering, is overwhelming. Women with this affliction frequently sustain classic osteoporotic fractures. These fractures typically involve the vertebrae, the wrist, the proximal femur, and/or the proximal humerus. In addition to the pathologic fractures, there is frequently a loss of height as a result of the cumulative effect of multiple vertebral fractures, as well as the progressive development of a kyphotic deformity in the thoracic spine, which is referred to as a "dowager's hump" (Fig. 1-12).

Patients present with a history of pain and/or repeated fractures. Occasionally they complain of early satiety because of some abdominal compression resulting from loss of height of the vertebral column. Similarly, the increasing kyphosis in the thoracic region may be responsible for some shortness of breath. On examination, typically one finds the prominent dowager's hump, a barrel chest, a protuberant abdomen, and generalized bone pain with percussion tenderness.

One of the most difficult problems in the past has been to determine bone mass. Typically, a crude estimate of bone density determined by plain radiograph has been used to extrapolate to the amount of bone previously lost. Classically, once osteopenia is noticeable radiographically, it has been estimated that the bone density is decreased by 30% to 50%.

Recently, additional diagnostic techniques have become available to more carefully estimate the amount of bone loss and, therefore, the amount of bone that remains. Isotope measurements, specifically single-photon

absorptiometry, using an iodine compound, or dual-photon absorptiometry, using a gadolinium compound, have been developed. They have significant technical limitations. The single-photon technique, measuring peripheral sites, such as the forearm and heel, is rarely an adequate reflection of the true bone mineral density in the axial skeleton. The dual-photon study, although providing more reliable information about the bone mineral density of the axial skeleton, continues to have some technical limitations.

TABLE 1-1. Causes of osteoporosis.

Primary
- Involutional (postmenopausal or senile)
- Idiopathic (juvenile or adult)

Secondary

Endocrine
- Hypogonadism
- Adrenocortical hormone excess (primary or iatrogenic)
- Hyperthyroidism
- Hyperparathyroidism
- Diabetes mellitus
- Growth hormone deficiency

Nutritional
- Calcium deficiency
- Phosphate deficiency
- Phosphate excess
- Vitamin D deficiency
- Protein deficiency
- Vitamin C deficiency
- Intestinal malabsorption

Drug
- Heparin
- Anticonvulsants
- Ethanol
- Methotrexate

Genetic
- Osteogenesis imperfecta
- Homocystinuria

Miscellaneous
- Rheumatoid arthritis
- Chronic liver disease
- Chronic renal disease

Immobilization
- Malignancy (multiple myeloma)
- Metabolic acidosis
- Cigarette smoking

Source: From Borenstein D, Wiesel SW. Low Back Pain: Medical Diagnosis and Comprehensive Management. Philadelphia: Saunders, 1989:329. Reprinted with permission.

TABLE 1-2. Types of involutional osteoporosis.

	Type 1 (Postmenopausal)	Type 2 (Senile)
Age (years)	51–75	Over 70
Sex ratio (M/F)	1:6	1:2
Type of bone loss	Trabecular	Trabecular and cortical
Fracture site	Vertebrae (crush) Distal radius	Vertebrae (multiple wedge) Hip
Main causes	Menopause	Aging
Calcium absorption	Decreased	Decreased
1,25-$(OH)_2$-vitamin D synthesis from 25-(OH) vitamin D	Secondary decrease	Primary decrease
Parathyroid function	Decreased	Increased

Source: Modified from Riggs BL, Melton LJ III. Involutional osteoporosis. N Engl J Med 1986;314:1676.

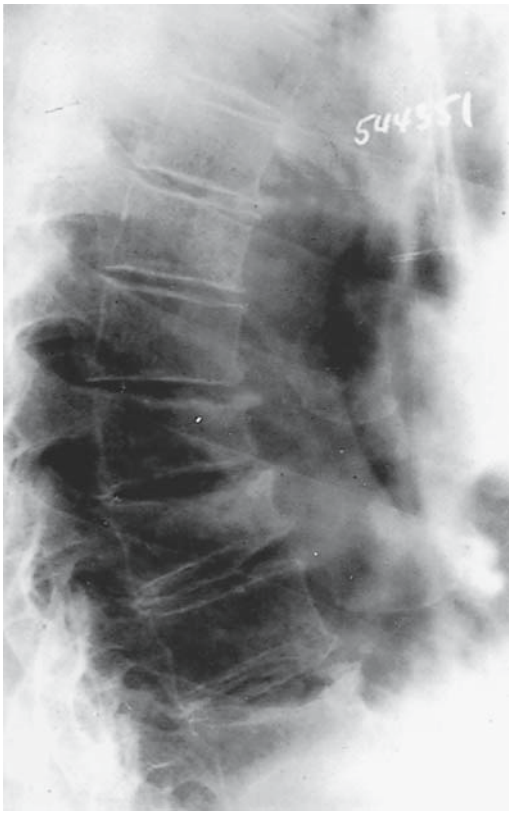


FIGURE 1-12. Radiograph of spine showing osteoporosis. Cortical bone appears accentuated by contrast with osteopenic marrow. Longitudinal trabeculae also appear accentuated because smaller transverse trabeculae are absent. Anterior wedging and endplate compression are present. (From Bogumill GP. Orthopaedic Pathology: A Synopsis with Clinical Radiographic Correlation. Philadelphia: Saunders, 1984. Reprinted by permission.)

As of this writing, it is probably fair to say that both these techniques have been replaced by dual-energy X-ray absorptiometry (DEXA) scanning. The DEXA technique is currently the standard, used in the evaluation of bone mineral density (BMD) in women approaching or following their menopause. This technique allows accurate and reproducible measures of density of the spine and the hip and does so with a minimal amount of radiation exposure. Current guidelines as recommended by the National Osteoporosis Foundation and the World Health Organization allow comparison of an individual's bone density to that of healthy normals. The difference is expressed as a T-score, which essentially represents one standard deviation above or below ideal bone mass. The definitions based on T-scores are as follows:

Normal	0 to –1
Osteopenia	–1 to –2.5
Osteoporosis	Less than –2.5

The unfortunate result of DEXA scanning, however, has been to adulterate the use of the term osteopenia. For many years, this term was defined as a generalized decrease in radiographic bone density. As such, it was nonpejorative and did not speak to a specific metabolic bone disease. In its present accepted context, the implication of using the term osteopenia is to imply a mild form of postmenopausal osteoporosis, which was certainly not the original connotation of the term. Diseases other than osteoporosis, such as hyperthyroidism and multiple myeloma, are characterized by observed decreases in radiographic bone density, hence osteopenia.

Without question, the most definitive diagnostic technique is direct bone biopsy with or without tetracycline labeling. It can clearly give the most reliable information regarding the presence of osteoporosis, its degree, and whether a superimposed osteomalacic state exists. Once the diagnosis has been confirmed and the risk analysis carried out, a treatment protocol can be tailored for the individual patient.

Most treatment regimens are considered either prophylactic or therapeutic. Prophylactic regimens include regular weight-bearing exercise, such as walking or jogging, supplemental calcium administration, and vitamin D administration with or without the administration of postmenopausal estrogen substitutes. The complications of oral estrogen administration, such as its relation to breast and cervical cancer and to heart disease and the incidence of deep venous thrombosis (DVT), make its general use controversial; however, its efficacy in maintaining skeletal mass is beyond question.

Therapeutic regimens, in contrast, are much more debatable. Current therapeutic regimens include the use of any or all of several different pharmacologic agents. Selective estrogen receptor modulators (SERMs) are drugs that behave either as an agonist or antagonist of estrogen. They have been shown, in selective populations, to decrease or minimize bone loss.

These drugs theoretically have an estrogen-like protective effect on bone. It has also been suggested that they have inhibitory (protective) effects on the breast and the endometrium.

Bisphosphonates are structurally similar to naturally occurring pyrophosphates. Because they have a strong chemical affinity for hydroxyapatite, they are potent inhibitors of bone resorption. They, therefore, are able to decrease the rate at which bone remodeling occurs and, as a result, to reduce the amount of bone resorption. It has been said that bisphosphonates are able to "freeze the skeleton." It is hoped that the consequence of decreasing bony resorption will be a coincident increase in bone mass. At the present time, the most popular bisphosphonate in current use is Fosamax, which has been approved for both the prevention and treatment of osteoporosis.

Calcitonin, a naturally occurring polypeptide hormone, is currently being administered in an effort to also decrease the rate of bony resorption by decreasing the number and activity of osteoclasts. The drug is currently being administered in the form of a nasal spray.

The current regimens used for the therapeutic management of osteoporosis include one or more of these drugs in addition to the standard prophylactic measures. Not infrequently, these agents are used cyclically or in an alternating fashion. Because the true measure of any therapeutic regimen for osteoporosis is an increase in bone density and a reduction in fracture risk or in the number of fractures, the true efficacy of these agents and various therapeutic regimens must be evaluated over the long term. As of this writing, the use of SERMs, bisphosphonates, and calcitonin all have shown early promise in this context.

Hypercalcemic States: Hyperparathyroidism

The effect of parathormone on bone is the same whether it is released as a result of a parathyroid adenoma (primary hyperparathyroidism) or by one of several secondary causes. In essence, parathormone stimulates osteoclastic activity, causing an intense resorption of bone (Fig. 1-13). The cavities resulting from this clastic activity fill with vascular fibrous tissue, resulting in the classic "osteitis fibrosa cystica." As the cavities coalesce, they form a single large cyst called a "brown tumor" because of the hemosiderin staining one sees within them. Clinical and radiographic changes result from this cavitation as well as from the erosive changes occurring under the periosteum.

Hypocalcemic States: Rickets and Osteomalacia

The same underlying mechanism accounts for rickets and osteomalacia: there is a general failure to mineralize bony matrix, resulting in the presence of unmineralized osteoid about bony trabeculae. This lack of mineral

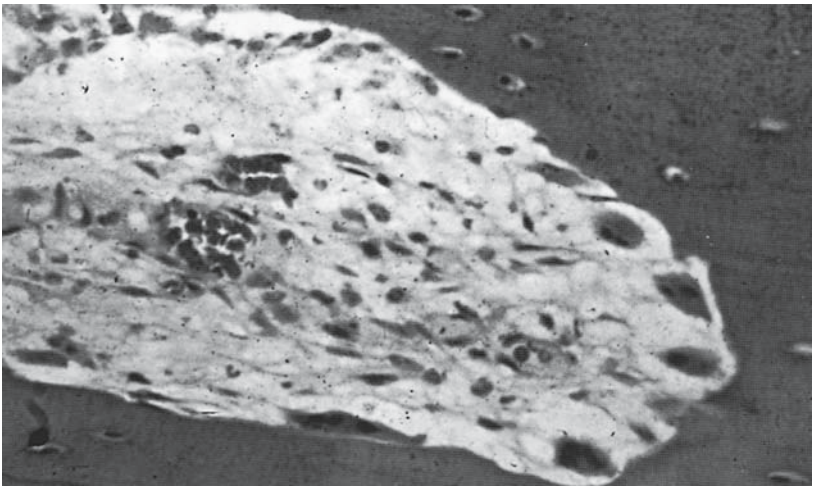


FIGURE 1-13. "Cutting cone." Successive relays of osteoclasts on the right resorb a tunnel of bone, making it longer and wider with each relay. Behind the cutting cone is a "filling cone" of successive relays of osteoblasts secreting osteoid. Resorption is facilitated by high-speed flow of well-oxygenated blood in small vessels, whereas refill is accompanied by dilated sinusoidal vessels with sluggish flow and low oxygen content. (From Bogumill GP. Orthopaedic Pathology: A Synopsis with Clinical Radiographic Correlation. Philadelphia: Saunders, 1984. Reprinted by permission.)

for adequate mineralization can be caused by a number of different etiologies: nutritional deficiency, malabsorption states, or renal disease (Table 1-3) are some of the more common. Despite the etiology, the metabolic effects on the skeleton are similar.

If the failure of mineralization impacts the skeleton before physeal closure, the result is rickets. The affected child will demonstrate the characteristic hallmarks of the disease: bowlegs, frontal bossing, ricketic rosary, and knobby joints (Fig. 1-14). All these findings are due to the presence of large masses of unmineralized osteoid. In addition, abnormalities of the physis and abnormal physeal growth can be anticipated.

If the process impacts the skeleton after physeal closure, the disease that results is osteomalacia. As noted earlier, the ratio of mineral to matrix decreases as a result of the paucity of mineral available to the skeleton. In the adult, these areas of unmineralized osteoid present as radiographically lucent areas in the bone, frequently referred to Looser's lines (Fig. 1-15). In addition, the bones themselves tend to be somewhat malleable and can bow under load; this is in contradistinction to osteoporotic bone, which is very brittle.

TABLE 1-3. Diseases associated with osteomalacia.

Disorder	Metabolic defect
Vitamin D:	Decreased generation of vitamin D_3
Deficiency	
Dietary	
Ultraviolet light exposure	
Malabsorption	Decreased absorption of vitamins D_2 and D_3
Small intestine	
Inadequate bile salts	
Pancreatic insufficiency	
Abnormal metabolism	
Hereditary enzyme deficiency	Decreased 1-alpha-hydroxylation of 25-(OH)-vitamin D
D-dependent rickets (type I)	
Chronic renal failure	Decreased 25-hydroxylation of vitamin D
Mesenchymal tumors	
Systemic acidosis	
Hepatic failure	
Anticonvulsant drugs	
Peripheral resistance	Absent or abnormal 1,25-$(OH)_2$-Vitamin D receptors
Vitamin D-dependent rickets (type II)	
Phosphate depletion:	
Dietary	Inadequate bone mineralization secondary to low serum concentrations
Malnutrition (rare)?	
Aluminum hydroxide ingestion	
Renal tubular wasting	
Hereditary	Decreased serum phosphate concentrations
X-linked hypophosphatemic osteomalacia	
Acquired	
Hypophosphatemic osteomalacia	
Renal disorders	
Fanconi's syndrome	
Mesenchymal tumors	
Fibrous dysplasia	
Mineralization defects:	
Hereditary	Abnormal alkaline phosphatase activity
Hypophosphatasia	
Acquired	
Sodium fluoride	Inhibition of bone mineralization
Disodium etidronate	
Miscellaneous:	
Osteopetrosis	Abnormal osteoclast activity
Fibrogenesis imperfecta	Unknown
Axial osteomalacia	Unknown
Calcium deficiency	Inadequate bone mineralization Secondary to low serum calcium concentration

Source: From Borenstein D, Wiesel SW. Low Back Pain: Medical Diagnosis and Comprehensive Management. Philadelphia: Saunders, 1989:339. Reprinted with permission.

FIGURE 1-14. Radiograph of wrist of child with active rickets exhibiting the irregular widened zone of provisional calcification that is replaced by abnormal osteoid. The cartilage masses are not visible, but the widened epiphyseal growth plate and irregular calcification are readily seen. Note pathologic fracture of radial shaft. (From Bogumill GP. Orthopaedic Pathology: A Synopsis with Clinical Radiographic Correlation. Philadelphia: Saunders, 1984. Reprinted by permission.)

FIGURE 1-15. Radiograph of osteomalacia showing a Looser's transformation zone. These lines appear at sites in which stress fractures would occur. Stress of normal use incites remodeling with removal of bone. In normal individuals, the removed bone is replaced by normal osteons. In persons with osteomalacia, the removed bone is replaced with abnormal osteoid, which fails to mineralize and leaves a linear radiolucency that may persist for years. (From Bogumill GP. Orthopaedic Pathology: A Synopsis with Clinical Radiographic Correlation. Philadelphia: Saunders, 1984. Reprinted by permission.)

Miscellaneous Metabolic Bone Disease: Renal Osteodystrophy

Renal osteodystrophy encompasses the skeletal changes that result from chronic, acquired renal disease. These changes are truly a "collage" of the other metabolic bone diseases. To understand the pathogenesis of renal osteodystrophy is to understand the basis of all the metabolic afflictions of the skeleton (Fig. 1-16). Chronic uremia allows a twofold drive to depress the serum calcium. First, the kidney is unable to excrete phosphate, hence the serum phosphate level rises. The serum calcium level is then of necessity driven down to maintain the fixed solubility product. Coincidentally, because the absence of a functional renal parenchyma stops the output of significant amounts of activated vitamin D, intestinal absorption of calcium is retarded, further depressing serum calcium. This dual mechanism profoundly depresses serum calcium and thus in turn mandates a parathormone response. The changes in the bone reflect the metabolic drives. The vitamin D deficiency is demonstrated by the presence of unmineralized osteoid (Fig. 1-17). The elevated levels of parathormone cause osteitis fibrosis cystica. Unique to this syndrome, the hyperphosphatemia results in a diffuse osteosclerosis. The latter finding causes one of the most pathognomic radiographic findings (Fig. 1-18), the "rugger jersey" spine.

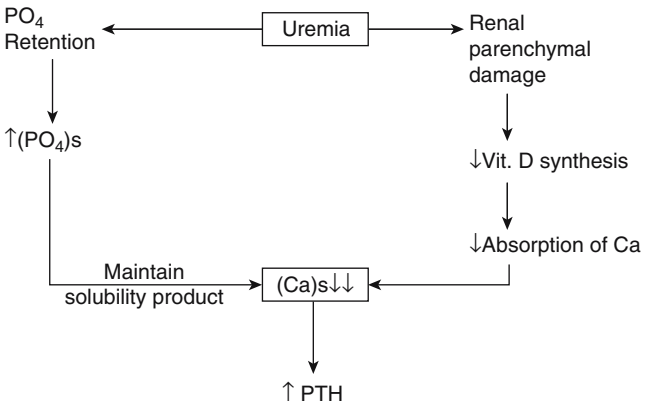


FIGURE 1-16. Pathogenesis of renal osteodystrophy.

FIGURE 1-18. Radiograph of patient with long-standing renal osteodystrophy. Marked osteoporosis attributable to secondary hyperparathyroidism is evident. There is bowing of the proximal femurs, marked lordosis, and pelvic tilt. The deformity of the pelvis is commonly seen in osteomalacia, but it does not usually occur in primary hyperparathyroidism. (From Bogumill GP. Orthopaedic Pathology: A Synopsis with Clinical Radiographic Correlation. Philadelphia: Saunders, 1984. Reprinted by permission.)

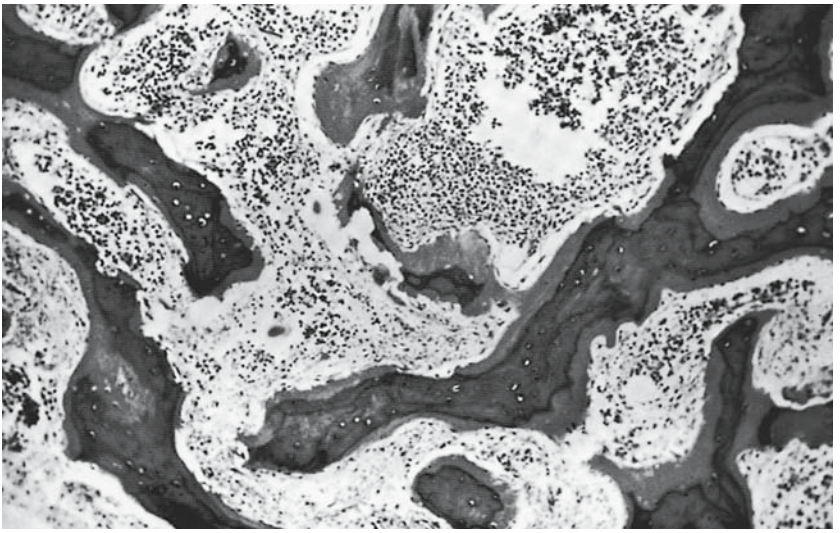
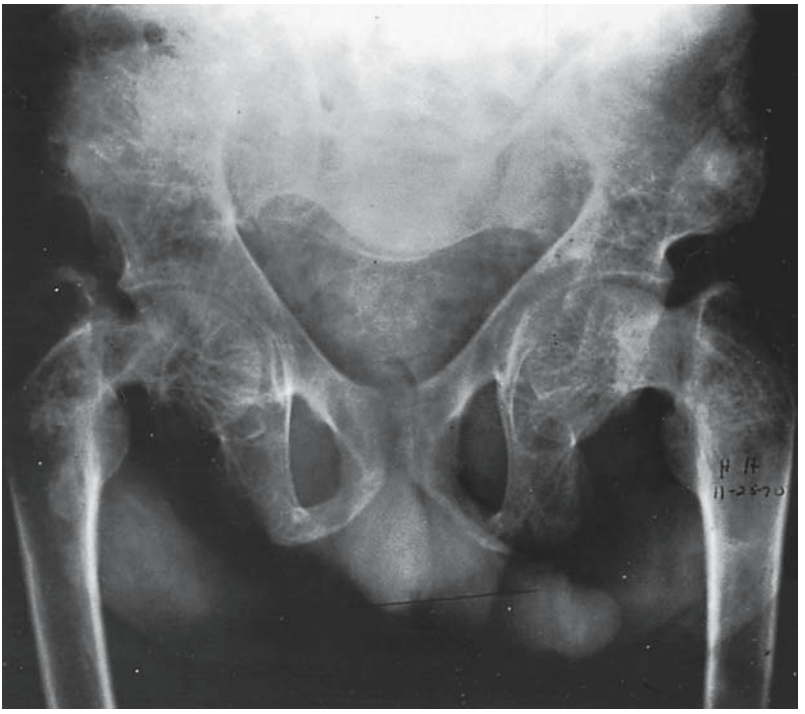


FIGURE 1-17. Renal osteodystrophy. Histologic section of bone exhibiting wide osteoid seams. These seams are seen in patients with primary renal disease, but they are not present in patients with primary hyperparathyroidism because the osteoid produced in primary hyperparathyroidism is normal. (From Bogumill GP. Orthopaedic Pathology: A Synopsis with Clinical Radiographic Correlation. Philadelphia: Saunders, 1984. Reprinted by permission.)



Sick Cell Syndromes: Osteogenesis Imperfecta and Osteopetrosis

The underlying mechanism seen in these conditions is a qualitative, functional deficit in a specific cell population, despite the fact that the population is quantitatively normal.

Osteogenesis imperfecta (Fig. 1-19) is typified by the impotence of the osteoblasts; they are unable to manufacture and secrete normal collagen. Ossification is, therefore, abnormal and results in inferior-quality bone. Clinically and radiographically, there is marked cortical thinning and attenuation of the diaphyseal caliber. The long bones, because of their altered anatomy, are at very high risk for fracture (Fig. 1-20). This bone fragility is the hallmark feature of osteogenesis imperfecta.

Because osteogenesis imperfecta is caused by a genetic mutation in the normal coding for type I collagen, there is significant phenotypic heterogeneity. In an effort to accommodate the variations in phenotype, the Sillence classification has been adopted by most authors. Four specific types are described in this classification:

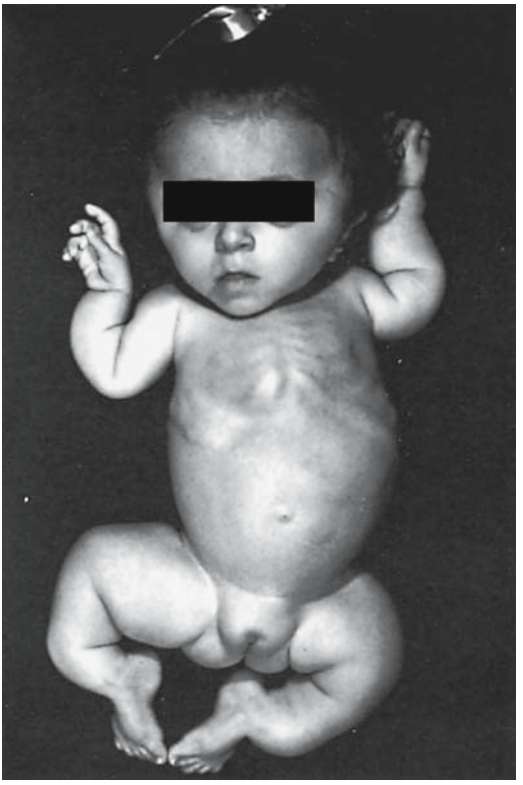


FIGURE 1-19. Deformity in a child with severe osteogenesis imperfecta. Note the prominence of the ribs in the abnormally shaped thoracic cage, the flattening of the skull with frontal bulging, and the malformed ribs. (From Gertner JM, Root L. Osteogenesis imperfecta. Orthop Clin North Am 1990;21(1):153. Reprinted by permission.)