Textbook of Musculoskeletal Disorders

Umile Giuseppe Longo
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Editors

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Francisco Forriol and Alexander Jedrzejczak

Overview
Metabolic diseases affect bone as a regulating organ of mineral metabolism, mainly calcium and phosphorus. Mechanical integrity depends on a balance in the regulation of bone resorption and bone formation.

1.1 Bone and Its Function

Bones constitute a part of the musculoskeletal system that defines the shape of the body and protects vital organs. Bones are essential in movement, acting as a rigid lever for the action of muscles. In addition, they have a hematopoietic function and act as phosphorus and calcium (P/Ca) homeostasis regulators. Bone is a living structure that grows, develops, and is continually modified during life due to the coordinated functions of its cells—osteoblasts, osteocytes, and osteoclasts. The coordinated actions of osteoblasts (bone-forming cells) and osteoclasts (bone-absorbing cells) allow bone tissue to repair itself, after a fracture, without scarring. In addition, the skeleton is essential for the immune system and acts as a storehouse of electrolytes and other essential substances for nerve conduction, muscle and heart contraction, clot formation, and other metabolic patterns. Bone mass contributes to blood calcium homeostasis.

1.2 Structure of Bone Tissue

Bone is composed of an organic and inorganic component. The organic component is made up of cells and an organic matrix and accounts for 30% of the bone's dry weight. Bone cells compose only a small portion (2%) of the organic component, with the remainder consisting of osteoid produced by osteoblasts. The osteoid is mainly composed of collagen, which accounts for the majority of the proteins in the organic component. The collagen is deposited along mechanical stress lines and creates a template for mineral deposition. The non-collagen proteins, even though only constituting a minor portion of osteoid protein content, play important roles in regulating osteoblast proliferation, metabolism, matrix production, and mineralization. The inorganic or mineral component (70% of dry weight) is mostly made up of a carbonate-rich hydroxyapatite analog. The hydroxyapatite analog incor-

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porates carbonate as well as magnesium, sodium, potassium, chloride, fluoride, strontium, and other bone-seeking elements. The analog has crystalline imperfections, making it more soluble than pure hydroxyapatite and therefore more readily available for metabolic activity and for exchange with body fluids. The union of collagen and hydroxyapatite provides the viscoelastic and anisotropic qualities of bone. Hydroxyapatite crystals provide mechanical resistance against compression forces; however, their elasticity is low. Collagen fibers are not resistant to compression, but they withstand torsional forces and have excellent elasticity.

The adult skeleton is made up of two distinguishable histological types of bone tissue, cortical (compact) and cancellous (trabecular or spongy). Cortical bone is solid and dense, with low porosity (<30%), while cancellous bone is full of holes and highly porous (50–90%). All normal adult bone is lamellar bone, whether it has a cortical or a cancellous structure. Cortical bone accounts for 80% of skeletal bone mass and consists of osteons and Havers and Volkmann canals. In a typical long bone, cortical bone forms the wall of the shaft, and cancellous bone is located in the metaphysis and epiphysis of the long bones. Cancellous bone is also found in the body of short bones such as the vertebrae and contains hematopoietic tissue. Although only making up 20% of skeletal bone mass, cancellous bone has a larger surface area and its surface-to-volume ratio is approximately ten times that of cortical bone. Vascularization is more abundant in cancellous bone tissue than in cortical bone.

The fundamental functional unit of adult cortical bone is the osteon. The osteon is filled with osteocytes and consists of concentric layers of bone or *lamellae*. The osteon has extensions that flow into the cortical bone canaliculi (Havers and Volkmann channels), communicating with cells covering the endosteal and periosteal bone surface. The Haversian canals (canals of Havers) are found centrally in the osteons and contain blood vessels, nerve supplies, and a supportive extracellular matrix. The lateral Volkmann canals carry blood vessels from one osteon to another. These canaliculi deliver oxygen and other nutrients to osteocytes through diffusion or convection forces resulting from mechanical motions.

In contrast to cortical bone structure, cancellous bone is made up of a porous honeycomb-like matrix. This matrix is composed of mature lamellar bone trabecula, in which osteocytes are concentrically oriented and have a well-developed canalicular network. The bony trabeculae fill the cross section of the bone, occupying approximately 20% of its volume. Depending on the location, such as in the proximal end of the femur, the trabeculae are quite regularly arranged reflecting the direction of mechanical stress acting on the bone.

1.3 Bone Remodeling

There are two populations of bone stem cells: mesenchymal stem cells (MSCs) and hematopoietic stem cells (HSCs). Osteoblasts and bone-lining cells originate from MSCs, while osteoclasts derive from osteoclast progenitors (mononuclear cells).

Bone is formed through two mechanisms, intramembranous and endochondral, each resulting in the same chemical composition. Although the biochemical compositions are comparable in both cortical and trabecular bones, their physical properties are markedly different to suit their local requirements. The thin cortical shell supported by trabecular bone at the ends of long bones is well suited to distribute the concentrated loads in the joints, whereas the tubular cortical midshaft is better suited to support the large torsional and bending loads applied to this area.

In the absence of a cartilage scaffolding, new bone is formed by mesenchymal osteoblasts through intramembranous ossification. Endochondral ossification occurs in the presence of a preexisting calcified cartilage matrix. Long bones and vertebrae increase in size through a combination of these two processes. For example, in a long bone, ossification of the shaft is an intramembranous process, while endochondral ossification increases long-bone length by cartilage proliferation at the growth plate. The rate of

remodeling changes with age, a 2-year-old boy remodels 50% of his skeleton annually, while an older adult does only 2–5% each year.

The initial step in cortical bone remodeling is the removal of portions of osteons through the action of osteoclasts. The osteoclasts form shallow depressions in the bone called *Howship lacunae* (*resorption bays*). After the bone is removed, osteoblasts secrete osteoid, which mineralizes with the osteoblasts inside and transforms them into osteocytes. The osteocytes regulate bone remodeling from external mechanical stimuli. New lamellae are then deposited in concentric layers until a new osteon is formed.

Cortical bone is remodeled on the periosteal, endosteal, and Haversian canal surfaces. Bone width growth occurs on the periosteal surface, the diameter of the medullary canal is determined through endosteal activity, and together they determine the overall thickness of the bone cortex. Haversian canal surfaces are responsible for the density of the cortex.

Cancellous bone is remodeled five times more than the cortical bone because it has a larger surface area. In actively forming or remodeling cancellous bone, the direction of deposition is determined by a row of osteoblasts on one border of the trabecula. The deposition of new bone by osteoblasts is counterbalanced by osteoclast removal of bone on the opposite surface of the trabecula. This coordinated formation and resorption can shift trabecula within a bone while maintaining mass homeostasis.

1.4 Effect of Mechanics on the Bone

Mechanical integrity depends on a precise balance between osteoblast and osteoclast activity, resulting in bone formation and resorption. In the normal adult skeleton, bone formation and resorption are balanced in a state called *coupling* where net bone formation equals net bone resorption. Any alteration to this complex balance results in clinical diseases such as osteoporosis, in which resorption action is predominant. Other examples include primary hyperparathyroidism in which osteoclastic activity is abnormal or osteopetrosis in which the bone formation is predominant.

Exercise improves and maintains bone mass, while inactivity leads to atrophy. In immobilized limbs, disuse osteoporosis sets in rapidly and the cortex thins. After resuming weight-bearing, the bone gradually rebuilds and cortical mass and thickness are restored. Disuse osteoporosis can be either localized or generalized. In generalized disuse osteoporosis, bone loss is extensive in the cancellous bone of the axial skeleton and weight-bearing long bones. Bone loss due to immobilization does not continue indefinitely and plateaus at approximately 50% of initial values.

Bone mass decreases with age and is one of the causes of increased fractures in older people. The magnitude of bone density varies between individuals as it depends on genetic, mechanical, nutritional, and hormonal factors. If the bone density and structure become so depleted that the skeleton can no longer withstand the mechanical stresses of everyday life, pathologic fractures also known as fragility fractures may result. When considering the postmenopausal period, lower bone density values are usually found in women. Beginning at 35 years of age, cortical bone loss begins in both sexes. A reduction of 0.3 and 0.5% of total bone density per year is reported. The compressive strength of bone is proportional to the square of its apparent density, meaning for a density decrease by a factor of 2, its compressive strength decreases by a factor of 4. In men, the loss of bone density with age is progressive, while in women, it decreases dramatically before menopause.

1.5 Bone Mass Homeostasis

The skeleton acts as an effective mineral bank for the body to store ionized calcium and phosphate in a metabolically stable and usable manner. In order to clinically evaluate mineral metabolism, laboratory tests must be conducted to determine the levels of calcium (total and unbound), phosphorus, and magnesium. There are four basic mechanisms of bone mass regulation: stimulation

and inhibition of bone formation, and stimulation and inhibition of bone resorption. Markers of bone formation include specific bone alkaline phosphatase, osteocalcin, and N-terminal propeptide of type I collagen. Bone resorption markers include free and total pyridinoline, free and total deoxypyridinoline, telopeptide N with cross-links with type I collagen, and telopeptides C with cross-links.

1.5.1 Calcium

The body is able to regulate serum calcium levels with exceptional accuracy despite variations in daily calcium intake. Primary regulation of serum calcium concentration is under the control of parathyroid hormone (PTH), calcitonin, 1,25-dihydroxy vitamin D (1,25-$(OH)_2$D), and osmotic gradient that provides passive calcium transport. Secondary factors such as hormones, proteins, phosphate, and blood pH also contribute to the regulation of serum calcium concentration. When calcium levels fall below physiological range, it is restored to normal through the PTH/vitamin D system, which increases calcium absorption in the gastrointestinal tract, reabsorption in the kidneys, and resorption of bone. Through the balance of bone formation and resorption, around 1000 mg/day of calcium is exchanged between bone and extracellular fluid. Most of the calcium in bones does not readily diffuse into the extracellular compartment, so when the body needs to utilize calcium from its mineral reserves, it must resorb bone. In order to counteract the resorption, bone formation increases and osteoblasts are stimulated; however, this response may not occur in the elderly. Calcium is introduced by the diet, and its absorption in the bowel is regulated by the body's needs, vitamin D status, age, and calcium consumption. Only some of the calcium secreted into the intestine is absorbed; much of it is excreted as unabsorbed calcium. The kidneys filter around 8000 mg of calcium daily and reabsorb over 95% under the effects of PTH. Due to this, calcium absorption is better determined using urinary calcium level than calcium intake.

1.5.2 Phosphate

Unlike calcium, phosphate is present in almost all foods and dietary deficiencies are uncommon. Absorption of phosphate is less selective than that of calcium and is partly dependent on vitamin D metabolites. Renal excretory mechanisms control phosphate ion levels. Additionally, renal absorption of phosphate is inversely related to PTH concentration. Phosphorus is present in crystalline form in the skeleton (85%), and the remaining 15% is in extracellular fluid and intracellular phosphorylated intermediates.

1.6 Hormones

Bone metabolism is largely regulated by parathyroid gland, which produces PTH; the thyroid gland, which secretes calcitonin; and the kidney, which provides 1,25-$(OH)_2$D from less active vitamin D metabolites. Other hormones such as thyroid hormones, estrogens, insulin, growth hormone also play a role. These hormones act collectively to regulate calcium and phosphate levels in serum and extracellular fluid by adjusting the ion absorption in the gut and kidneys and rate of bone resorption.

1.6.1 Vitamin D

About half of our vitamin D is obtained from diet, with the other half originating from a reaction in our skin with ultraviolet radiation. A diet high in oily fish can help prevent vitamin D deficiency. Vitamin D from the skin and diet is metabolized in the liver to 25-hydroxyvitamin D (25-(OH)D), which is used to determine a patient's vitamin D status. 25-(OH)D is metabolized in the kidneys by the enzyme 25-hydroxyvitamin D-1 α-hydroxylase to its active form, 1,25-$(OH)_2$ vitamin D. The renal production of 1,25-$(OH)_2$ vitamin D is regulated by plasma PTH levels and serum calcium and phosphorus levels. The vitamin D metabolite 1,25-dihydroxyvitamin D (1,25-$(OH)_2$D or calcitriol) helps regulate calcium and phosphate

serum levels by increasing the efficiency of their absorption in the intestine and renal tubules as well as osteoclastic bone resorption by stimulating osteoclast precursors.

1.6.2 Parathyroid Hormone (PTH)

PTH is secreted from the parathyroid glands to regulate Ca and P levels in the blood. When a dip in calcium is detected, the parathyroid glands increase PTH secretion, which then leads to an increase in calcium levels and suppression of PTH secretion creating a feedback loop. PTH raises calcium levels by promoting calcium entry from the bone, renal tubules, and intestine. This is achieved by increasing bone resorption, enhancing renal reabsorption of calcium, and increasing the synthesis of 1,25-$(OH)_2$D, which activates calcium absorption in the small intestine. Renal reabsorption of phosphate is inversely related to PTH concentration, and high PTH decreases phosphate reabsorption. Compared to other secretory systems, parathyroid cells contain minimal amounts of PTH and as such regulation of secretion occurs at the gene level and through cell proliferation. Apart from calcium and phosphate concentrations, PTH secretion is also controlled by the active form of 1,25-$(OH)_2$D and fibroblastic growth factor-23 (FGF23). Serum levels of calcium and phosphate posttranslationally affect PTH by increasing and decreasing PTH mRNA levels, respectively. FGF23 can also bind to Klotho proteins on parathyroid cells suppressing PTH gene expression and secretion. PTH is also essential for normal mineralization of bone in the later stages of fracture healing.

1.6.3 Calcitonin

Calcitonin is a polypeptide hormone produced by parafollicular cells of the thyroid gland. Calcitonin's principal function is to inhibit osteoclast resorption of bone in response to elevated serum calcium levels. The production of this hormone is stimulated by elevated serum calcium levels and inhibited by decreased calcium levels. Calcitonin may have analgesic properties on bone pain in patients with osteoporosis and in bone resorption processes (Paget's disease).

1.6.4 Growth Hormone (GH)

Secretion of growth hormone in the pituitary gland is regulated by neurosecretory neurons in the hypothalamus. GH stimulates bone formation and is necessary for the maintenance of normal bone mass. GH has an important function throughout the process of fracture healing by binding to cell membrane receptors in the tissues involved in the regenerative response. Individuals with a GH deficiency can show symptoms like short stature and dwarfism, and conversely gigantism and tall stature result from high or excess GH levels.

1.6.5 Insulin

Insulin stimulates the synthesis of the bone matrix and promotes the correct mineralization. Much like GH, insulin has an important function throughout the process of fracture healing by binding to cell membrane receptors in the tissues involved in the regenerative response.

1.6.6 Leptin

Leptin regulates bone mass by stimulating bone formation and resorption through sympathetic signaling pathways. One through AP-1 favors osteoblast proliferation, and another one, through the molecular clock, inhibits osteoblast proliferation.

1.6.7 Androgens

Androgen deficits produce a decrease in the secretion of calcitonin and calcitriol through its osteoblast-like receptors. Androgens counteract PTH (bone resorption) activity, may stimulate an increase in bone mass, and prevent Ca loss.

Medications that lower the levels of androgens increase the rates of bone loss. The exact role of androgens in maintaining skeletal mass is not well understood.

1.6.8 Estrogens

Estrogen receptors are found in both osteoblasts and osteoclasts. Estrogen decreases osteoclasts' sensitivity to PTH, causing estrogen deficiency to be the major cause of bone loss in early postmenopause. Development of osteoporosis in postmenopausal women is linked with estrogen deficiency. Estrogen plays an important role in regulating RANK ligand (RANKL), a molecule critical in osteoclast formation and activity. Decreasing estrogen decreases RANKL regulation and osteoclast activity increases.

1.6.9 Glucocorticoids

At physiological concentrations, glucocorticoids act as modulators of bone remodeling. At high doses, they inhibit osteoblasts and the synthesis of insulin-like growth factors, inducing bone loss.

1.7 Metabolic Diseases

Metabolic diseases affect bone as a regulatory organ of mineral metabolism. The etiology is very varied, but a common pathologic mechanism is present. Metabolic diseases usually produce an imbalance between bone formation and resorption (Table 1.1). These conditions could be quantitative alterations, altering the mineral content (osteoporosis, Südeck's dystrophy, osteosclerosis, renal osteodystrophy, osteopetrosis) or qualitative diseases, modifying the structure or the shape of the bone (osteomalacia, rickets, Paget's disease).

1.7.1 Osteoporosis

Osteoporosis is the decrease in bone mass per unit of bone volume, without affecting mineralization, to what is considered normal for a certain age, sex, and race, with the structure and content of bone mineral being normal. Osteoporosis is fully described in Chap. 2.

Table 1.1 Analytics in some metabolic diseases

Anemia ↑ VSG	Myeloma Pathologic fracture
↑ Ca	Hyperparathyroidism
Ca	Tumoral hypercalcemia Malabsorption Vitamin D deficit
P	Osteomalacia
↑ Alkaline phosphatase	Vitamin D deficit Liver diseases Paget's disease
↑ Transaminases	Liver diseases
Albumin	Malnutrition Liver diseases Renal diseases
25-(OH) vitamin D	Vitamin D deficit
FPTH	Hyperparathyroidism
TSH	Hyperthyroidism
Ca 24-h urine Hypercalciuria	Hyperparathyroidism Hyperthyroidism Tumoral hypercalcemia

1.7.2 Südeck's Dystrophy or Reflex Sympathetic Dystrophy

Südeck's dystrophy is painful osteoporosis, located in the skeletal area related to previous trauma, fracture, sprain, or inflammatory process, where venous stasis occurs in bone sinusoidal spaces. It occurs in 2–3% of patients with limb injuries, and pain develops shortly after fracture or joint injury has healed. The first clinical manifestations are redness, swelling, hyperhidrosis, and hyperesthesia of the limb at the injury site as well as proximally and distally. The precise pathophysiology of this disorder is not yet understood, but disinhibition of the sympathetic nervous system in the area of injury is one theory.

1.7.3 Renal Osteodystrophy

Kidney pathologies affect the skeleton, especially patients undergoing hemodialysis or treatment with drugs that prevent hydrolyzing

25-dihydroxyvitamin D to form the active form ($1,25\text{-}(OH)_2D$) or retain phosphates by not being filtered by the kidney.

1.7.4 Paget's Disease

In Paget's disease, the patient presents excessive osteoclastic activity, intense bone resorption, and substitution of the marrow by hypervascular fibrous tissue. The repair bone is a disorganized bone with poor mechanical conditions. This condition is fully described in Chap. 3.

1.7.5 Osteopetrosis or Albers-Schönberg Disease or Marble Bone Disease

Osteopetrosis is characterized by defective osteoclast function and impaired bone resorption, leading to bone fragility with an increased risk of fracture. It presents a diffuse increase in skeletal density and closure of the trabecular spaces (marrow spaces). Three distinct types of osteopetrosis are seen in humans, autosomal dominant (adult, benign, Albers-Schönberg disease), autosomal recessive (infantile, malignant), and X-linked inheritance patterns. Autosomal dominant is associated with limited disability and a normal life span; however, autosomal recessive is often fatal during childhood.

1.7.6 Osteomalacia and Rickets

Nutritional rickets is an infrequent metabolic bone disease defined by impaired mineralization of osteoid resulting from severe vitamin D deficiency (<10 ng/mL). Osteomalacia (adult rickets) has several additional causes to severe vitamin D deficiency.

Childhood rickets presents with impaired growth, bowed extremities, and height usually below the third quartile. Low weight is not typically seen unless there is a simultaneous nutritional deficiency. Affected children are typically lethargic and irritable. The head will present with abnormalities such as softness or skull deformity. A thoracic exam may show flaring and deformity of the ribs, funnel chest, pigeon breast, or nodules at costochondral junctions. Examination of the extremities will expose abnormalities such as symmetric enlargement of the ends of the long bones, bowleg, and, less frequently, knock-knee. Other manifestations include frequent fractures, respiratory infections, and chronic cough. The prevalence of childhood rickets varies highly between different regions of the world. Treatment for childhood rickets is achieved through vitamin D and calcium supplementation.

It is more difficult to establish a diagnosis for adult osteomalacia as the changes may be much smaller than in childhood rickets. In the early stage, patients may be asymptomatic with only biochemical changes. The most sensitive of these changes is an elevated serum total alkaline phosphate, which if observed should lead to obtaining a bone-specific alkaline phosphate (BSAP). An elevated BSAP should suggest osteomalacia; this can be confirmed with quantitative bone histomorphometry. Once a diagnose is established, biochemical testing can determine the etiology, and the abnormalities can be corrected.

Patients with adult and advanced osteomalacia present with generalized weakness, bone pain, easy fatigability, and a general feeling of discomfort or illness. Physical findings are limited; however, in more serious and long-standing cases, tenderness of bony prominences, muscle weakness strong enough to affect gait, bowleg, or kyphosis may be seen. Rickets presents with a very characteristic histological appearance of the epiphysial plate. Rachitic epiphysial plates show a greatly increased axial height due to an increased number of cells in the maturation zone, which have lost their columnar organization. There are also significant bone changes associated with osteomalacia; however, these changes are not unique to osteomalacia and are seen in other metabolic bone disorders. These changes include thin cortices and small and irregular trabeculae displaying osteoclastic reabsorption. Most characteristic is the presence of a wide zone of unmineralized bone surrounding the mineralized trabeculae. There are a number of categories of rachitic and osteomalacic conditions with different causes and manifestations.

The classic and most understood cause of nutritional rickets and osteomalacia is a severe vitamin D deficiency. Causes for this deficiency can be a result of diet, lack of sunlight, or malabsorption due to a variety of gastrointestinal conditions. Other conditions that may result in this deficiency include premature infants whose immature livers cannot adequately convert vitamin D to 25-(OH)D and chronic use of anticonvulsants, which may decrease 25-(OH)D by interfering with liver enzyme systems. Nutritional disorders that interfere with calcium absorption may also lead to a similar syndrome; these include excessive ingestion of oxalate, citrate, or phosphate. Conditions in which the gut wall is damaged or rapid transit of gastrointestinal contents occurs may also lead to rachitic or osteomalacic syndrome due to a deficiency in vitamin D, calcium, or both.

Due to vitamin D fortification in foods, genetic or acquired rachitic and osteomalacic conditions are seen more frequently than those associated with vitamin D deficiencies. These genetic and acquired conditions are often resistant to high therapeutic doses of vitamin D and are renal in origin. The renal conditions, vitamin D-resistant rickets and osteomalacia due to proximal tubular defects, vitamin D-resistant rickets and osteomalacia due to proximal and distal renal tubular defects, and vitamin D-resistant rickets due to renal tubular acidosis are described in Table 1.2.

Table 1.2 Renal rachitic conditions

Category of rachitic and osteomalacic conditions	Subtype	Causes and manifestations	Additional information
Vitamin D-resistant rickets and osteomalacia due to proximal tubular defects	Type I	Sex-linked dominant genetic disorder in which the renal tubule does not reabsorb phosphate	
	Type II	Same as Type I but the defect is broader and involves both phosphate and glucose	
	Type III	Same as Type II but the reabsorptive defect also includes glucose and various amino acids	
Vitamin D-resistant rickets and osteomalacia due to proximal and distal renal tubular defects	Type I	Result of the failure of tubular reabsorption of phosphate combined with the loss of fixed base, including calcium	Broader tubular defects and may interfere more severely with normal metabolism. The functions of the distal tubule are significantly altered. Results in a mild-to-moderate secondary hyperparathyroidism (which in turn worsens the bone lesions and intensifies the hypophosphatemia). Almost completely independent of vitamin D and treatment with high doses of the vitamin have little effect
	Type II	Almost identical to Type I but has an additional defect in the metabolism of cytosine	
	Type III	Presents with many of the manifestations of Type I disease but may also have a broad range of ocular and neurologic abnormalities	
	Type IV	Symptoms include profound motor weakness and very high urinary concentrations of glycine	
Vitamin D-resistant rickets due to renal tubular acidosis	Type I (distal or classic)	Proton exchange is defective	Common mechanism is the kidney's inability to substitute hydrogen ions for fixed base. Characterized by metabolic acidosis and an alkaline urine. The mechanisms are not yet completely understood. Chronic acidosis alone can deplete calcium and phosphate from the bones but it is considered to be a minor mechanism
	Type II (proximal)	Due to a failure of reabsorption of bicarbonate in the proximal tubule	
	Type IV (generalized distal)	Excretion of both hydrogen and potassium ions in the distal tubule is defective	

Another category of conditions is vitamin D-dependent (pseudo-deficiency) rickets and osteomalacia. This category of rickets is characterized by abnormalities of vitamin D metabolism. The defect is also almost always inherited and involves either a failure of conversion of 25-(OH)D to 1,25-$(OH)_2$D or an insensitivity to the patient's 1,25-$(OH)_2$D. In both of these circumstances, vitamin D supplementation in standard or therapeutic doses does not increase reabsorption of calcium by the renal tubule, resulting in hypocalcemia and rachitic manifestations. A specific disorder that fits into this category is oncogenic osteomalacia. This syndrome is a result of very small, subcutaneous, mostly benign mesenchymal tumors that develop in adults. These tumors secrete FGF23 and induce renal phosphate wasting and hypophosphatemia while inhibiting renal production of 1,25-$(OH)_2$D.

Some of the rachitic disorders are life threatening, such as vitamin D-resistant rickets and osteomalacia due to proximal and distal tubular defects; other disorders are mild and may not require prolonged treatment. In patients with classic rachitic or osteomalacic changes in the epiphyseal growth plates and bones, renal tubular acidosis should be suspected as the underlying cause of the disease. Treatment of rachitic and osteomalacic syndromes due to renal tubular acidosis should focus on the primary process rather than on the bone disease.

1.7.7 Vitamin C Deficiency (Scurvy)

Scurvy is a disorder caused by vitamin C deficiency and can lead to multiple complications, including lethargy, bone pain, impaired wound healing, myalgia, and impaired bone growth. Vitamin C deficiency manifests symptomatically after 8–12 weeks of inadequate intake. Although vitamin C deficiencies are prevalent, even in industrialized countries, scurvy is rare. Prevalence is variable depending on the region of the world.

The clinical manifestations of scurvy are caused by ascorbic acids' role in collagen synthesis. Vitamin C is necessary for the formation of mature collagen, which is the main component of blood vessel walls, skin, and basement membrane zone separating the epidermis from the dermis. Vitamin C facilitates hydroxylation and cross-linking of procollagen, and a lack of vitamin C decreases transcription of procollagen. Additionally, a lack of vitamin C leads to epigenetic DNA hypermethylation and inhibition of the transcription of various types of collagens found in skin, blood vessels, and tissue. Rheumatologic problems occur, such as hemarthrosis and subperiosteal hemorrhage resulting from vascular fragility from impaired collagen formation. Osseous pathology also occurs and presents with fractures in brittle bones from disrupted endochondral bone formation. Vitamin C is essential for matrix elaboration in both cartilage and bone. Vitamin C plays an important role in the differentiation of chondrocytes (cartilage cells). Vitamin C also plays a role in osteoblast fate determination and proliferation. Vitamin C's antioxidant properties minimize the concentration of reactive oxygen species, which increases osteoclast differentiation and bone resorption. Deficiency of vitamin C results in a failure of formation of the osteoid matrix and in disruption of endochondral bone formation. The treatment for scurvy is vitamin C supplementation.

Take-Home Message

- Metabolic diseases affect bone as a regulatory organ of mineral metabolism, mainly calcium and phosphorus.
- Bone mass is a main contributor to the maintenance of adequate levels of calcium in the blood.
- Mechanical integrity depends on a balance in the regulation of bone resorption and bone formation (*coupling*).
- The regulation of bone metabolism is fundamentally determined by three hormones: vitamin D, PTH, and calcitonin. Other hormones (thyroid hormones, estrogens, and hypothalamic control) have a secondary influence.
- The etiology of the metabolic diseases is very varied, but they produce a local or

general remodeling with a reduction (osteoporosis) or an increase (osteosclerosis) of bone density.
- Metabolic alteration can be quantitative, altering the mineral content, or qualitative, modifying the structure or shape of the bone.

Summary

Bones constitute a part of the musculoskeletal system that defines the shape of the body and protects vital organs. Bones are essential in movement, acting as a rigid lever for the action of muscles. In addition, they have a hematopoietic function and act as phosphorus and calcium (P/Ca) homeostasis regulators. Bone is a living structure that grows, develops, and is continually modified during life due to the coordinated functions of its cells—osteoblasts, osteocytes, and osteoclasts. In addition, the skeleton is essential for the immune system and is a storehouse of minerals, calcium and phosphates, among others, and essential substances for nerve conduction, muscle and heart contraction, clot formation, and many other metabolic patterns. Bone mass contributes to the maintenance of adequate levels of calcium in the blood. The bone tissue is made up of an organic component (30%) and inorganic component (70%). The formation or remodeling of the bone is regulated by the bone cells that repair and adapt to the conditions of each person at all times.

Mechanical integrity depends on a balance in bone resorption and formation regulation and osteoblasts and osteoclasts' coordinated activities (*coupling*).

Metabolic diseases affect bone as a regulating organ of mineral metabolism, mainly calcium and phosphorus. The etiology is very varied, but they produce a local or general remodeling with a reduction (osteoporosis) or an increase (osteosclerosis) of bone density.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Osteoporosis:
   (a) Is the decrease in bone mass per unit of bone volume, without affecting mineralization, to what is considered normal for a certain age, sex, and race, with the structure and content of bone mineral being normal
   (b) Is the increase in bone mass per unit of bone volume, without affecting mineralization, to what is considered normal for a certain age, sex, and race, with the structure and content of bone mineral being normal
   (c) Is the decrease in bone mass per unit of bone volume, affecting mineralization, with bone alteration
   (d) Is due to excessive consumption of vitamin D
2. The main cause of osteoporosis:
   (a) Is idiopathic
   (b) Is due to excessive consumption of vitamin D
   (c) Is due to hypoparathyroidism
   (d) Is due to excessive physical activity
3. PTH:
   (a) Is secreted from the parathyroid glands, regulates Ca and P levels in the blood, and modulates the activity's specific kidney and bone cells. It increases resorption, stimulates osteoclastic function, releases Ca directly from the bone, stimulates reabsorption of Ca in the renal glomerulus, and inhibits phosphate reabsorption.
   (b) Is secreted from the thyroid, regulates P levels in the blood, and modulates the activity's specific kidney and bone cells. It increases resorption, stimulates osteoclastic function, releases Ca directly from the bone, stimulates reabsorption of Ca in the renal glomerulus, and inhibits phosphate reabsorption.
   (c) Is secreted from the parathyroid glands, regulates Ca and P levels in the blood, and modulates the activity's specific kidney and bone cells. It decreases resorption, stimulates osteoblastic function, releases Ca directly from the bone, stim-

ulates reabsorption of Ca in the renal glomerulus, and inhibits phosphate reabsorption.
   (d) Is secreted from the parathyroid glands, regulates Ca and P levels in the blood, and modulates the activity's specific kidney and bone cells. It increases resorption, stimulates osteoclastic function, increases Ca bone uptake, stimulates extrusion of Ca in the renal glomerulus, and promotes phosphate reabsorption.
4. Osteoclasts:
   (a) Reabsorb bone, forming the so-called Howship lacunae, while osteoblasts fill them by secreting osteoid or non-mineralized bone tissue
   (b) Produce bone, forming the so-called Howship lacunae, while osteoclasts fill them by secreting osteoid or non-mineralized bone tissue
   (c) Produce bone by secreting osteoid or non-mineralized bone tissue
   (d) Are stimulated by vitamin D
5. Osteomalacia:
   (a) Occurs due to vitamin D deficiency, due to either vitamin D resistance, impaired vitamin D synthesis or metabolism, metabolic acidosis, hypophosphatemia, intestinal malabsorption, kidney disorders, heavy metal poisoning, surgery, anticonvulsant treatments (phenytoin), or low content of vitamin D in the diet
   (b) Occurs due to vitamin D excess, metabolic alkalosis, kidney disorders, heavy metal poisoning, surgery, anticonvulsant treatments (phenytoin), or low content of vitamin D in the diet
   (c) Occurs due to vitamin D deficiency, due to either vitamin D resistance, impaired vitamin D synthesis or metabolism, metabolic alkalosis, hyperphosphatemia, intestinal malabsorption, liver disorders, heavy metal poisoning, surgery, anticonvulsant treatments (phenytoin), or high content of vitamin D in the diet
   (d) Is due to metabolic alkalosis

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Osteoporosis

2

Francesca Giusti and Maria Luisa Brandi

Overview

Osteoporosis is a progressive systemic skeletal disease characterized by low bone mass and microarchitectural deterioration of bone tissue, with a consequent increase in bone fragility and susceptibility to fracture.

2.1 Definition

The World Health Organization (WHO) defines osteoporosis as a "progressive systemic skeletal disease characterized by low bone mass and microarchitectural deterioration of bone tissue, with a consequent increase in bone fragility and susceptibility to fracture."

This "silent," common, condition represents a growing major public health problem, severely impacting the quality and duration of life.

In Italy, about 18.5% of women and 10% of men are estimated to be osteoporotic; the number of osteoporotic patients is expected to increase by 25% in the next 20 years.

2.2 Classification

Osteoporosis is classified in primary or idiopathic osteoporosis and secondary osteoporosis.

Primary osteoporosis is the most common type, due to a direct alteration/defect in bone remodeling, and it includes the following

- *Inherited juvenile osteoporosis* in which the bone loss occurs from infancy to adolescence as a direct consequence of genetic mutations in genes involved in bone homeostasis, causing a reduced amount and impaired quality of the fibrous component of bone (e.g., leading to osteogenesis imperfecta).
- *Idiopathic osteoporosis of the elderly* caused by an aging-dependent increased bone loss. This form of osteoporosis is characterized by a specific skeletal disease pattern, including prevalent trabecular bone loss and perforation, compared to cortical bone loss, leading to site-specific fracture risks at vertebral bodies and at the distal radius. Women are mainly affected, since the postmenopausal estrogen depletion severely enhances the high bone turnover and the bone loss occurring with aging. In the postmenopausal period, estrogen deficiency leads to bone loss through both

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bone marrow expansion and endosteal resorption, whereas periosteal apposition occurs, mainly in response to mechanical stress, to counteract reduced bone strength. It is difficult to predict the individual clinical outcome because of the interpersonal variability in the rate of bone loss after menopause.

Conversely, secondary osteoporosis is due to a spectrum of other conditions, which indirectly alter the normal bone metabolism and homeostasis, and it affects about two-thirds of men, over 50% of premenopausal women, and 20% of postmenopausal women.

Secondary osteoporosis can derive from endocrine (hypogonadism, hypocortisolism, hyperparathyroidism, acromegaly, diabetes mellitus), hematological (thalassemia, multiple myeloma), gastrointestinal (malabsorption, celiac disease), rheumatic (rheumatoid arthritis, systemic lupus erythematosus, ankylosing spondylitis, scleroderma), kidney (renal failure, chronic tubular acidosis), and collagen disorders, from prolonged immobilization, or from medications, such as glucocorticoids, aromatase inhibitor anticoagulants, diuretics, and others.

2.3 Risk Factors

The pathogenesis of osteoporosis is multifactorial. Individual predisposition to osteoporosis and personal fracture risk are results of several independent endogenous and exogenous factors, singularly exerting different, and synergic, degrees of risk. The concomitant presence of multiple risk factors increases the probability to develop osteoporosis.

Osteoporosis risk factors can be classified into non-modifiable and modifiable agents. The first ones include age, gender, menopause, ethnicity, hormonal profile, and genetic factors (familiarity). Modifiable factors mainly depend on lifestyle and dietary habits and include sedentary lifestyle, smoking, excessive consumption of alcohol, insufficient intake of calcium and vitamin D, low consumption of fresh fruits and vegetables, excessive intake of protein, sodium and caffeine, excessive thinness, and petite constitution. Changes in these modifiable risk factors help in reducing the personal risk of osteoporosis development and fragility fracture occurrence.

Peak bone mass (PBM) is the highest level of bone mass, achieved by the age of 25–30 years as a result of normal growth, and it determines resistance or susceptibility to osteoporosis and fractures. About 40–50% of bone mass is achieved by the age of 18 years. At skeletal maturity, women have 10–15% lower bone mass than men. PBM is the result of the interaction of various factors (genetic variants, hormonal profile, ethnicity, nutrition habits, lifestyle, physical exercise), the same ones defined as risk factors for osteoporosis. Environmental factors modulate the expression of the genetic potential to achieve PBM, already establishing, during childhood, adolescence, and young adulthood, an important part of the future individual risk for osteoporosis and fragility fracture.

2.4 Prevention

Therapeutic lifestyle is an essential part for the primary prevention and/or the management of osteoporosis. This includes a balanced diet; exposure to sunlight; reduction of smoking and intake of alcohol, caffeine, and salt; and adequate, constant, physical activity. Appropriate resistance, weight-bearing aerobics, and core-stabilizing exercisers are needed to maintain muscle strength and bone health. Balance exercises are necessary to improve equilibrium and prevent falls.

Nutrition plays a key role in the prevention and management of osteoporosis.

Adequate daily intake of calcium, vitamin D, and protein is the gold standard, along with good sun exposure.

An increase in dietary calcium intake through the consumption of calcium-rich foods (milk, yogurt, cheese) represents the first step to correct a negative calcium balance. The recommended intake of calcium (RNI) is at least 1000 mg daily in men and women over 50 years. Calcium-fortified dairy products provide at least 40% of

the RNI of calcium (400 mg) per portion and are valuable options. When dietary sources of calcium are not sufficient to provide daily requirements, calcium supplements can be administered.

Vitamin D insufficiency and deficiency are common among older people and have detrimental effects on bone health and neuromuscular function. Serum levels of 25(OH) vitamin D of less than 20 ng/mL are associated with mineralization defects. A serum target of 30 ng/mL and a daily dietary intake of 800 IU of vitamin D are recommended in men and women over 50 years, mostly for individuals at high risk of fracture. Fortified dairy products provide 200 IU of vitamin D3 per portion. A medical supplementation is strongly recommended when the correct vitamin D requirement is not granted by the diet. The use of 25(OH) vitamin D3 supplements should be preferred in conditions characterized by impaired 25-hydroxylation, obesity, and malabsorption and when a rapid correction of vitamin D status is needed.

The use of combined calcium and vitamin D3 supplementation has been proven to significantly reduce total and hip fracture rates in both community-dwelling and institutionalized middle-aged to older adults.

Both caloric and protein intakes decrease with age. Protein intake has a positive impact on bone health at all ages. A combination of protein supplementation and resistance exercises results in greater gains in muscle mass and strength and helps in preventing falls. The recommended average daily intake of protein is at least 1.0–1.2 g/kg/BW, including at least 20–25 g of high-quality protein (such as protein supplied by dairy products) with each main meal (breakfast, lunch, dinner) during the day.

2.5 Diagnosis

Medical history should gather information regarding family history of osteoporosis and/or fragility fractures, personal history of previous fractures, nutritional habits, level of physical activity, lifestyle, use of medications affecting bone metabolism, and, for women, duration of ovarian estrogen production.

Dual-energy X-ray absorptiometry (DXA), performed at lumbar spine (L1–L4) and hip (total hip or femoral neck) every 18–24 months, is the gold standard for quantitative assessment of bone mineral status in adults. DXA is performed on distal forearm only if both lumbar and femoral assessments are impractical or inaccurate, in severely obese patients, and in patients with hyperparathyroidism. T-score defines standard deviation (SD) in the measured bone density value with respect to that of a 35-year-old young adult of the same gender, while for the Z-score, the comparison is made with someone of the same age, gender, ethnicity, height, and weight. The WHO has defined T-score criteria for the classification of osteoporosis (T-score at or below −2.5 SD), osteopenia (T-score between −1.0 and −2.5 SD), and normal bone (T-score over −1.0 SD).

Conventional X-ray of thoracic and lumbar spine is performed for the detection of prevalent vertebral fractures. Vertebral variations are evaluated by a semiquantitative method measuring anterior, middle, or posterior heights of the dorsal and lumbar vertebral bodies in lateral projection, via conventional radiography or with vertebral fracture assessment by DXA. Vertebral fractures, the most common osteoporotic fractures, are an indicator of a high risk for future fractures, even when the T-score does not meet the threshold for osteoporosis. They are often asymptomatic when they first occur, may remain undiagnosed for many years, and be incidentally discovered by a spine X-ray examination performed for other reasons.

Trabecular bone score (TBS) is a textural index that measures pixel gray-level variations in the lumbar spine DXA image, providing an indirect index of trabecular microarchitecture. Variability in the 2-dimensional projected DXA image is presumed to correlate with absorption parameters in 3-dimensional bone according to a mathematical relationship. High TBS values correlate with homogeneous (normal) bone texture, while low values are indicative of more variable (weaker) bone texture. TBS predicts fracture risk

independent of BMD and enhances fracture risk prediction capabilities of FRAX®.

Quantitative computed tomography (QCT) measures BMD, bone mineral content (BMC), and true bone density expressed in g/cm^3, obtaining a clean data from any osteoarthritic processes. Unlike other instrumental methods, it involves higher radiation dose, has reduced accuracy, and is relatively expensive. Peripheral QCT (pQCT) screens skeletal peripheral segments (forearm and tibia) and allows a 3-dimensional reconstruction of trabecular bone, providing information on bone microarchitecture. High spatial resolution pQCT (HR-pQCT) is a new technique able to display also trabecular bone microstructure.

Biochemical evaluation of bone metabolism: All women with postmenopausal osteoporosis, at high fracture risk, should undergo an appropriate medical evaluation to identify coexisting medical conditions that cause or contribute to bone loss. Because of the high prevalence of causes of secondary osteoporosis, even in apparently healthy postmenopausal women, biochemical screenings should be considered for all women with osteoporosis, since they provided useful information in 40–85% of women who did not have clinical evidence of secondary osteoporosis.

Recent guidelines suggest classifying laboratory tests in two groups

1. First-level exams: Blood cell count, erythrocyte sedimentation rate (ESR), serum calcium (corrected for albumin), serum phosphate, serum protein electrophoresis, serum creatinine, alkaline phosphatase, and 24-h urinary calcium.
2. Second-level exams: Serum 25-hydroxyvitamin D, serum parathyroid hormone (PTH), serum ionized calcium, serum thyroid-stimulating hormone (TSH), free kappa and lambda light chains, anti-tissue transglutaminase antibodies, urinary free cortisol, serum cortisol after 1 mg dexamethasone suppression, serum testosterone and SHBG (in men), serum tryptase (or urine N-methylhistamine), ferritinemia, bone marrow aspiration and biopsy, and undecalcified iliac crest bone biopsy.

Bone turnover markers (BTM) are used to quantify the degree of bone resorption and bone formation and to monitor response to anti-fracture therapies. Bone formation markers include bone alkaline phosphatase, osteocalcin, and propeptides of procollagen type I (P1NP) in the serum. Bone resorption markers include CTX and NTX in the serum, and pyridinoline, deoxypyridinoline, and CTX in the urine. Many studies confirmed that serum P1NP and serum CTX are the most reliable markers, respectively, for bone formation and bone resorption, both at baseline evaluation and in the follow-up.

These laboratory tests cannot be used to diagnose osteoporosis.

2.6 Therapy

Several drugs are approved for the treatment of postmenopausal osteoporosis and prevention of fracture occurrence. They are classified into anti-resorption agents (bisphosphonates, denosumab, and selective estrogen receptor modulators), anabolic molecules (abaloparatide and teriparatide), and a novel monoclonal antibody against sclerostin (romosozumab).

All these therapies have been shown to reduce the risk for vertebral fractures; some of them also reduce the risk for non-vertebral fractures, including hip fractures.

In the personal setting of osteoporosis therapy, anti-fracture drugs have to be chosen taking into account the personal conditions of each patient (sex, age, presence of comorbidity, severity of osteoporosis), and they have to be associated with the supplementation of calcium and vitamin D to obtain the recommended daily doses, rarely reached with diet alone.

2.6.1 Bisphosphonates

Bisphosphonates (BPs) are analogs of inorganic pyrophosphate, able to reduce bone turnover by blocking osteoclast activity. All BPs approved for treatment of postmenopausal osteoporosis

showed a broad spectrum of anti-fracture efficacy and can be administered orally or intravenously.

Orally administered BPs (the most commonly used are alendronate 70 mg weekly, risedronate 35 mg weekly, or ibandronate 150 mg monthly) are poorly absorbed (0.5–5%) in the gastrointestinal tract and have side effects, such as upper gastrointestinal symptoms and bowel disturbance. They must be taken after a prolonged fast (usually in the morning, soon after arising), swallowed with a full glass of water, without taking any other medication, food, or beverage (other than water) for at least 30 min. Recent data indicate that the use of proton pump inhibitors in combination with oral BPs may reduce anti-fracture effectiveness.

Zoledronate (zoledronic acid) is given as a single intravenous infusion (5 mg/year), over a minimum period of 15 min, to be repeated yearly. Side effects of zoledronate include an acute-phase reaction (usually in patients receiving their first dose) characterized by fever and muscle aches—a flu-like illness—lasting several days and gastrointestinal symptoms. An increase in atrial fibrillation can be rarely reported as a serious adverse event.

BPs are contraindicated in patients with hypocalcemia, gastrointestinal disease, and renal impairment (serum creatinine above 200 μmol/L or creatinine clearance below 30 mL/min) and in those who are pregnant or lactating. Adverse effects of long-term BP therapy are osteonecrosis of the jaw (ONJ) and atypical fractures. ONJ incidence is low for both oral and intravenous BP therapies for osteoporosis, on the order of 1/10,000–1/100,000 patients per year. Risk factors include dental pathologic conditions, invasive dental procedures, and poor dental hygiene. A preventive oral examination should be done in patients being considered for treatment with these agents. If significant dental issues are present, delaying the initiation of BP or denosumab therapy, until the dental issues have been corrected, should be considered. In patients already receiving BPs or denosumab who require invasive dental procedures, there is no evidence that discontinuing or interrupting treatment will change the outcome or reduce the risk of ONJ. Nonetheless, stopping should at least be considered for patients undergoing extensive invasive dental procedures such as extraction of several teeth. Atypical fragility fractures of the subtrochanteric region are another rare event that seems to be estimated in 1/1000 women with osteoporosis treated for up to 3 years on 100 osteoporotic fractures prevented. Patient under BP therapy who reports a persistent thigh or groin pain should interrupt the treatment. It has been hypothesized that these patients may have very low bone turnover, although this point has not been rigorously substantiated. A causal relationship has not been established. Guidelines recommended that the risk of such events should be reassessed after 5-year therapy for alendronate, risedronate, or ibandronate and after 3 years for zoledronate. In patients at a high risk of osteoporotic fracture, a continuation of treatment without the need for further assessment can generally be recommended.

2.6.2 Denosumab

Denosumab is a humanized monoclonal antibody against RANKL, able to inhibit osteoclast recruitment, maturation, and survival. It is approved for the treatment of osteoporosis in postmenopausal women at increased risk of fracture, in women with breast cancer treated with aromatase inhibitors, and in men under antiandrogen treatment for prostate cancer and is given as a subcutaneous injection of 60 mg once every 6 months. Its action is uniform on all skeletal structures, despite bone turnover degree, granting a greater pharmacological activity in the cortical bone than other therapies. The chronic therapy is associated with a continuous densitometric increase of mineral bone mass, in which a BMD plateaus after 3–4 years of administration, particularly at the cortical level. The anti-resorption effect ceases immediately upon the interruption of treatment.

Denosumab is contraindicated in women with hypocalcemia or hypersensitivity to any of the constituents of the formulation. It does not need dose adjustment in patients with renal impairment. Its use is not recommended during preg-

nancy or in young individuals (age ≤18 years). Side effects include skin infection, predominantly cellulitis, and hypocalcemia.

It is very important to evaluate an alternative drug if treatment is discontinued and/or the patient maintains a high risk of fracture.

2.6.3 Selective Estrogen Receptor Modulators

Selective estrogen receptor modulators (SERMs) are synthetic ligands of estrogen receptors, which exert agonistic effects in bone and liver and antagonistic effects in the breast and the genitourinary tract.

SERMs currently approved in Italy for the prevention and treatment of osteoporosis in postmenopausal women are raloxifene and bazedoxifene. They are contraindicated in women with childbearing potential, a history of venous thromboembolism or unexplained uterine bleeding, liver and kidney failure, or climacteric symptoms.

Raloxifene inhibits bone resorption, and it is administered to the dose of 60 mg daily.

Bazedoxifene is able to prevent the loss of bone mass at a dose of 20 mg/day in normal and osteopenic women.

Conversely, estrogen therapy is no longer indicated for osteoporosis therapy or prevention of osteoporosis.

2.6.4 Abaloparatide and Teriparatide

Abaloparatide (modified PTH-related peptide 1–34) and teriparatide (recombinant human PTH 1–34) are two anabolic agents approved by the FDA for initial treatment of women with postmenopausal osteoporosis who are at high risk of fracture or have failed or resulted intolerant to previous osteoporosis therapy. Teriparatide is also approved for the treatment of glucocorticoid-induced osteoporosis and treatment of osteoporosis in men. Both are injected subcutaneously: abaloparatide is administered at a dose of 80 μg daily and teriparatide is given at 20 μg daily; they both cannot be used for more than 24 months. Available data demonstrate that treatment with teriparatide or abaloparatide should be regularly followed by anti-resorption therapy (BPs or denosumab followed by an oral BP) to avoid a rapid decrease in BMD upon discontinuation of the two drugs in the following months.

Before treatment with either these molecules, it is recommended to measure serum calcium, PTH, 25(OH)D, and alkaline phosphatase (to rule out Paget's disease). Both these drugs demonstrated to increase BMD and reduce the risk of vertebral and non-vertebral fractures in women with postmenopausal osteoporosis. Side effects of both abaloparatide and teriparatide generally are mild and transient and include nausea, orthostatic hypotension, and leg cramps. Hypercalcemia can occur, but usually is mild, asymptomatic, and transient.

Both abaloparatide and teriparatide should not be used

- In patients at increased risk of developing osteosarcoma; development of this tumor has been reported, in some studies, in rats treated with very high doses of both drugs.
- In patients with primary or any form of secondary untreated or unresolved hyperparathyroidism.
- In patients with Paget's disease of bone, open epiphyses, a history of irradiation involving the skeleton, or an unexplained elevation of bone alkaline phosphatase.

2.6.5 Romosozumab

Romosozumab is a monoclonal antibody directed against sclerostin. Sclerostin binds with the Wnt receptor and inhibits the differentiation of precursor cells into mature bone-forming osteoblasts. Blocking sclerostin binding to osteoblasts allows osteoblast activity to increase. BTMs suggest an early anabolic effect, bone density increases are dramatic, and biopsies indicate an anabolic effect through both modeling (increase in cross-sectional area) and remodeling (bone repair).

Approval of romosozumab for postmenopausal women at high risk of fracture was based on significant reductions in radiographic vertebral fractures at 12 and 24 months.

Romosozumab is an option for patients previously treated with teriparatide or abaloparatide, and future retreatment with romosozumab may be possible. Romosozumab can be used in patients with prior radiation exposure. Romosozumab should not be used in patients at high risk for cardiovascular events or who have had recent myocardial infarction or stroke.

Romosozumab has also been studied in men but is not currently approved for male osteoporosis.

Take-Home Message

- Osteoporosis is the most common metabolic disease of the skeleton, characterized by a reduction of bone mass and qualitative changes of bone microarchitecture, which increase bone fragility and susceptibility to fractures.
- Peak bone mass is observed between 20 and 30 years of age and is largely determined by genetic factors.
- Symptoms usually occur only late, manifesting as musculoskeletal pain and the final endpoint of the disease, the low-trauma fracture.
- Patient is defined as normal with a T-score higher than −1 SD, osteopenic with a T-score between −1.0 and −2.5 SD, or osteoporotic if the T-score is less than −2.5 SD.
- Physical exercise improves bone health and increases muscle strength, coordination, and balance, reducing the risk of falling and fractures. Medical therapy consists of drugs acting on skeletal remodeling to restore the correct balance between bone resorption and new formation.

Summary

Osteoporosis is the most common metabolic disease of the skeleton, characterized by a reduction of bone mass and qualitative changes of bone microarchitecture, which increase bone fragility and susceptibility to fractures. Peak bone mass is observed between 20 and 30 years of age and is largely determined by genetic factors. During life, other risk factors, such as environmental factors, eating habits, lifestyle, comorbidities, and bone-altering medications, can accumulate being, together, decisive for the onset of the disease.

Osteoporosis is a silent disease; symptoms usually occur only late, manifesting as musculoskeletal pain and the final endpoint of the disease, the low-trauma fracture. The most common fragility fractures in osteoporosis involve lumbar spine, proximal femur (neck), and wrist.

Diagnosis of osteoporosis is made by bone densitometry measurement at lumbar spine and proximal femur, through T-score value. Patient is defined as normal with a T-score higher than −1 SD, osteopenic with a T-score between −1.0 and −2.5 SD, or osteoporotic if the T-score is less than −2.5 SD. In the presence of a densitometric diagnosis of osteoporosis, the X-ray screening of the spine is indicated for vertebral fracture assessment. Urine and serum dosages of bone turnover markers help in determining the degree of bone turnover.

Osteoporosis can benefit from corrections in eating habits, such as the correct daily intake of calcium and vitamin D. Physical exercise improves bone health and increases muscle strength, coordination, and balance, reducing the risk of falling and fractures. Medical therapy consists of drugs acting on skeletal remodeling to restore the correct balance between bone resorption and new formation. The great majority of approved anti-fracture drugs (bisphosphonates, denosumab, SERM) act by inhibiting osteoclast activity and, thus, reducing the osteoporosis-related increased bone resorption. Anabolic drugs (teriparatide or abaloparatide) act by stimulating osteoblast function and promoting new bone formation. Romosozumab is a novel monoclonal

antibody aimed to increase new bone formation and, to a lesser extent, reduce bone resorption. In patients with vertebral or femoral fractures, in addition to drug therapy for preventing further fracture events, it is necessary to plan both surgical and rehabilitative approaches for, respectively, the acute treatment of fracture complications and for the management of post-fracture neuromotor outcomes.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. A marked increase in urinary elimination of hydroxyproline may occur?
   (a) Osteoporosis
   (b) Paget's disease
   (c) Hypothyroidism
   (d) Achondroplasia
2. What are the recommended anatomic sites for performing bone densitometry in the diagnosis of osteoporosis?
   (a) Lumbar spine and femoral neck
   (b) Lumbar spine and wrist
   (c) Lumbar spine and hand
   (d) Wrist
3. Which of these statements is NOT correct?
   (a) A frequent osteoporosis fracture is that of the diaphysis femoral
   (b) The risk of osteoporosis fracture increases after menopause
   (c) The risk of fracture increases if the T-score of the bone mineralometry (DEXA) is less than −3
   (d) Vertebral fracture is very frequent in women aged 60–70 years
4. Osteoporosis is defined by a T-score:
   (a) At or below −2.5 SD
   (b) Between −1.0 and −2.5 SD
   (c) Over −1.0 SD
   (d) Over −0.5 SD
5. Bisphosphonates are usually adopted for:
   (a) Postmenopausal osteoporosis
   (b) Juvenile osteoporosis
   (c) Osteoporosis with a T-score of −2.5
   (d) Osteoporosis with a T-score of −1.0

Further Reading

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Paget's Disease of Bone

3

Francesca Marini and Maria Luisa Brandi

Overview
Paget's disease of bone (PDB) is an osteometabolic disease characterized by focal abnormal bone remodeling, which gradually leads to bone deformity and pathological fracture. This disease presents an incomplete penetrance even among mutation carriers, and a high inter- and intrafamilial genetic heterogeneity.

3.1 Disease Definition

Paget's disease of bone (PDB) is an osteometabolic disease characterized by focal abnormal bone remodeling, which gradually leads to bone deformity and pathological fracture. At the cell level, the disease begins with osteoclast-driven excessive bone resorption followed by increased compensatory osteoblast activity, resulting in a non-organized bone architecture (woven bone), replacing the normal tissue, which is weaker, less compact, more vascularized, accompanied with bone marrow fibrosis, and more susceptible to deformity and fracture.

PDB condition consists mainly of three consecutive phases

- The initial phase, in which there is a high prevalence of bone resorption and bone vascularization. Bone appears osteolytic to a radiograph control.
- The mixed phase, an osteosynthetic phase characterized by a predomination of new bone formation. Novel bone appears structurally abnormal with thickening of the cortex, indistinguishability of the cortico-medullary border, and accentuation of the trabecular pattern. The radiography scan shows a mixed aspect of the bone.
- The late phase, characterized by thickening of long bones, increased bone volume, and poor biomechanical efficiency of the tissue. Bone appears osteosclerotic to a radiograph control.

3.2 Epidemiology

PDB is the second most common metabolic bone disease in elderly people, after osteoporosis.

It shows a specific geographical/ethnical distribution, with a higher prevalence in the United Kingdom (about 1–2% of British people are estimated to be affected by the age of 55) and other European countries such as Italy, France, Greece,

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and Spain, being, conversely, rare in Scandinavia and Asia. A high PDB prevalence is reported also in people of British descent who have migrated to Australia, New Zealand, South Africa, Canada, and the United States.

PDB affects both sexes, but a higher prevalence in males was reported in most series. This condition is rare before the age of 50 (0.3 cases per 10,000 persons/year are reported among women aged 55–59 years and 0.5 cases among men of similar age; after the age of 85 years, these rates rise to 5.4 in females and 7.6 in males).

3.3 Clinical Presentation

PDB preferentially affects the axial skeleton. The most commonly affected bone sites are the pelvis (70%), femur (55%), lumbar spine (53%), skull (42%), and tibia (32%), but the disease can potentially occur at any skeletal site.

PDB can manifest in two main forms

- Monostotic PDB (only one bone segment is affected), affecting mainly the long bones. This form is less common and rarely symptomatic.
- Polyostotic PDB (more than one bone site is affected). This form is more frequent, symptomatic, and often associated with pain and complications.

Bone and joint pains are most common symptoms. Bone pain is usually deep within the affected bone, dull or aching, and constant and worse at night or during rest. Joint pain is usually due to the articular cartilage damaged by the abnormal bone growth, often in association with swelling and stiffness (secondary osteoarthritis), and it manifests prevalently at the awakening, with a possible bit improvement when the person starts to move.

The direct affection of bones can result in bone deformity (scoliosis, kyphosis, curved legs), fragility fractures, hearing loss, osteomyelitis, and, if the skull is involved, mandibular prognathism, migration of the teeth, tooth missing, headache, vertigo, and noise in the ears.

Abnormal bone growth at the spine can compress nearby nerves, resulting in a variety of clinical symptoms like sciatica, peripheral neuropathy, numbness and/or tingling of arms and legs, reduction of limb movements, loss of balance, and loss of bower and/or bladder control.

Cardiovascular complications have been reported, mainly due to a higher propensity of PDB patients to endocardial, aortic valve, and arterial calcification.

3.4 Clinical Diagnosis

PDB patients are usually incidentally diagnosed by X-ray, bone scan, and/or blood tests performed for other medical problems.

3.4.1 Radiological Diagnosis

Instrumental diagnosis is done by radiographs and bone scintigraphy.

Classical radiology allows to visualize enlarged bones and to identify the osteolytic, mixed, or osteosclerotic tissue and, thus, classify the phase of PDB development.

The total-body bone scintigraphy permits to assess the correct location, extension, and activity of skeletal lesions.

Bone biopsy may be required only when X-ray images result not to be able to confirm or exclude the presence of PDB.

Given the elderly age of PDB patients, a DXA analysis should be performed to assess a possible coexistence of osteopenia or osteoporosis.

3.4.2 Biochemical Screenings

The result of a blood test for bone metabolism biomarkers, often performed for other reasons, can be a first alert to doctors about the possibility that a person has PDB, which needs to be confirmed by instrumental scan.

Serum total (ALP) and/or bone-specific alkaline phosphatase (BALP) are generally increased in PDB patients, as markers of increased osteoblast activity. A mild increase of ALP is not specific of PDB, and it can be caused by some other conditions, like liver disease or an active fracture healing process. An ALP value twice higher than the normal upper level is strongly indicative of a PDB, mostly in patients with concomitant normal levels of calcium and phosphate and no kidney disease.

ALP and BALP levels are also used to monitor progression of the disease and response to treatments.

Also, levels of the amino-terminal and the carboxy-terminal propeptides of type 1 procollagen and osteocalcin may be increased in PDB patients.

3.5 Genetics of PDB and Genetic Diagnosis

The exact cause of PDB is still unknown. A combination of environmental and genetic factors is suspected to contribute to the development of the disease. Given the ethnical prevalence of the disease and the fact that it is maintained despite people's migrations to other countries, genetic factors surely have an important role in PDB etiology. Indeed, up to 40% of individuals with PDB have at least one first-degree affected relative; transmission appears to follow an autosomal dominant pattern with incomplete penetrance.

Genome-wide linkage analyses on PDB families identified seven possible candidate loci.

Currently, the PCR-based sequencing screening of *SQSTM1* gene is the only genetic test for late-onset PDB. Up to 50% of familial cases and 10% of sporadic cases have been estimated to be the carrier of an *SQSTM1* mutation. The identification of a mutation enables the genetic screening for the family members of an index case and allows the recognition of mutation carriers at asymptomatic level, providing the opportunity to perform a clinical surveillance to precociously detect the onset of the first PDB-related sign/symptom and, thus, decrease the morbidity associated with the disease. However, the incomplete penetrance and the genetic heterogeneity of the disease may limit the advantages of the genetic test.

3.6 Therapy

Medical therapy for PDB is usually limited to symptomatic patients, with the strong recommendation to immediately start the treatment at the first occurrence of any symptom (usually bone pain). Asymptomatic patients can be administered therapy in case of significant biochemical abnormalities or bone imaging indicating the risk of ongoing complications from untreated disease.

PDB medical treatment consists of two main categories of drugs: analgesics and nonsteroidal anti-inflammatory drug to treat bone and articular pain, and bisphosphonates (BPs) to normalize bone remodeling.

Various BPs have been licensed for the treatment of PDB, as resumed in Table 3.1.

The intravenous administration allows a better compliance and avoids gastrointestinal side effects, but it requires to be performed in a medical center. Oral BPs can be taken by the patient

Table 3.1 Bisphosphonates used in the medical therapy of PDB

Drug	Posology	Effect on serum alkaline phosphatase levels (response outcome)
Alendronate	Oral, 40 mg/day for 6 months	Reduction of 73–79% in 6 months
Pamidronate	Intravenous, 60 mg/day for 3 days or a single infusion of 60 mg/day every 3 months	Reduction of 53% in 6 months, after two injections of 60 mg/day every 3 months
Risedronate	Oral, 30 mg/day for 2 months	Reduction of 69% in 6 months
Zoledronic acid	Intravenous, 5 mg in a single dose	Reduction of about 80% in 6 months

himself at home, but they can cause dyspepsia and occasionally esophageal ulceration, granting a lesser compliance.

The follow-up of BP treatment includes the periodical dosage of serum ALP (every 3–6 months) until the normalization of its value. After remission, ALP can be measured once a year.

Some patients can benefit from supportive therapies, such as physiotherapy involving exercises and techniques that can help in reducing pain and improving movements.

Surgical therapy is applied for the resolution/correction of skeletal and articular severe damages, such as bone deformation of the legs, fractures, and degenerative arthritis, and in the rare occurrence of a sarcomatous degeneration of the tissue.

Take-Home Message

- Paget's disease of bone (PDB) is the second most common metabolic bone disease in elderly people, after osteoporosis.
- PDB is characterized by focal abnormal bone remodeling, preferentially affecting the axial skeleton, which gradually leads to bone deformity and pathological fracture.
- Genetic causes being responsible for up to 40% of individuals with PDB have at least one first-degree affected relative.

Summary

Paget's disease of bone (PDB) is an osteometabolic disease characterized by focal abnormal bone remodeling, which gradually leads to bone deformity and pathological fracture. PDB is the second most common metabolic bone disease in elderly people, after osteoporosis. At the cell level, the disease begins with osteoclast-driven excessive bone resorption followed by increased compensatory osteoblast activity, resulting in a non-organized bone architecture (woven bone), replacing the normal tissue, which is weaker, less compact, more vascularized, accompanied with bone marrow fibrosis, and more susceptible to deformity and fracture.

PDB condition consists mainly of three consecutive phases

- The initial phase, in which there is a high prevalence of bone resorption and bone vascularization.
- The mixed phase, an osteosynthetic phase characterized by a predomination of new bone formation.
- The late phase, characterized by thickening of long bones, increased bone volume, and poor biomechanical efficiency of the tissue.

PDB preferentially affects the axial skeleton. The most commonly affected bone sites are the pelvis (70%), femur (55%), lumbar spine (53%), skull (42%), and tibia (32%), but the disease can potentially occur at any skeletal site. PDB can manifest in two main forms: monostotic and polyostotic.

PDB patients are usually incidentally diagnosed by X-ray, bone scan, and/or blood tests performed for other medical problems. A combination of environmental and genetic factors is suspected to contribute to the development of the disease. Medical therapy for PDB is usually limited to symptomatic patients, with the strong recommendation to immediately start the treatment at the first occurrence of any symptom (usually bone pain). Asymptomatic patients can be administered therapy in case of significant biochemical abnormalities or bone imaging, indicating the risk of ongoing complications from untreated disease. PDB medical treatment consists of two main categories of drugs: analgesics and nonsteroidal anti-inflammatory drug to treat bone and articular pain, and bisphosphonates (BPs) to normalize bone remodeling.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. A marked increase in urinary elimination of hydroxyproline may occur?
   (a) Osteoporosis
   (b) Paget's disease
   (c) Hypothyroidism
   (d) Achondroplasia
2. Researchers believe that Paget's disease may be caused by:
   (a) Virus
   (b) Parasite
   (c) An abnormal gene
   (d) A + C
3. Paget's disease most often strikes in which age group?
   (a) Children under age 12
   (b) Teens
   (c) Adults under 30
   (d) Adults over 40
4. Which bones are most commonly affected by Paget's disease?
   (a) Spine
   (b) Pelvis
   (c) Skull
   (d) A + B + C
5. A healthcare provider can best diagnose Paget's disease with which test?
   (a) Bone scan
   (b) X-ray
   (c) Alkaline phosphatase blood test
   (d) A combination of all of the above

Further Reading

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Part II

Systemic Diseases of the Musculoskeletal System

4 Endocrine Abnormalities Affecting the Musculoskeletal System

Vincenzo Denaro, Alessandro Mazzola, and Umile Giuseppe Longo

Overview

Endocrine glands are specialized organs that secrete hormones into the endocrine signalling system. Endocrinopathies are a heterogeneous group of pathologies that result in hormonal disturbances and can affect nearly every body system, including the musculoskeletal system. Radiological evaluation of these conditions can demonstrate typical appearances of the bones and soft tissues. This chapter focuses on the typical musculoskeletal findings of acromegaly, gigantism, pituitary dwarfism, hyperthyroidism, hypothyroidism, hyperparathyroidism, hypoparathyroidism, hypercortisolism and hypocorticosurrenalism. The clinical manifestations of each of these endocrine disorders, along with a brief discussion of the pathophysiology and radiological findings, will be discussed.

4.1 Hypersomatotropism (Acromegaly and Gigantism)

4.1.1 Definition

Hypersomatotropism is a condition due to uncontrolled secretion of GH. If the hormonal hypersecretion is present prior to physeal fusion, then gigantism results. Alternatively, if the increased levels of GH are present after skeletal maturity, then acromegaly is the resultant condition.

4.1.2 Epidemiology

Hypersomatotropism is the second most frequent cause of hyperpituitarism (overactive pituitary gland) after prolactinomas. Gigantism occurs during puberty, whereas acromegaly affects adults.

4.1.3 Aetiology/Pathogenesis

More than 90% of cases of hypersomatotropism are caused by benign pituitary adenomas producing

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GH. Very rarely, it could be due to pituitary carcinomas or malignant ectopic hormonal secretion.

4.1.4 Diagnosis

4.1.4.1 Clinical Diagnosis

Prior to physeal fusion, excess GH results in amplification of the normal process of endochondral bone formation that occurs in the physeal growth plates during puberty. It leads to an exaggerated longitudinal bone growth that is clinically identified as gigantism. Conversely, after the growth plates have closed, an excess GH can only stimulate endochondral ossification and bone formation at the chondro-osseous junctions resulting in a clinical syndrome called acromegaly. Laboratory tests are needed to confirm these clinical diagnoses.

4.1.4.2 Musculoskeletal Involvement in Gigantism and Acromegaly

Gigantism is always not proportioned: patients present very long limbs, a small cranium and height significantly above the average. Skeletal deformities such as flatfoot, knee valgus and kyphosis are possible.

Acromegaly determines alterations in acral bone and soft tissues (face, hands and feet), but also in internal organs (thyroid, heart, liver and adrenal glands). In this case, new bone formation is not longitudinal but circumferential due to hormonal stimulation of the intramembranous periosteal ossification. The head presents pronounced brow protrusion, often with ocular distension (frontal bossing), prognathism, macroglossia and spaced teeth. Hands, feet, ears and nose result to be enlarged, and carpal tunnel syndrome is frequent. Erdheim's syndrome consists of subperiosteal ossification in the front surface of the vertebral body, mainly in the dorsal rachis. It leads to dorsal kyphosis and lumbar hyperlordosis resulting in chronic back pain. Patients may also develop the acromegalic arthropathy of the limbs, which consists of proliferation and hypertrophy of the articular cartilage. Radiographic findings show hypertrophic epiphyses and abundant osteophytosis. Joints most commonly affected are the knee, shoulder, hip, elbow, metacarpophalangeal of the thumb and metatarsophalangeal of the hallux. Acromegalic osteochondropathy is the hypertrophy of cartilage that connects ribs to the sternum, determining para-sternal deformities.

4.1.5 Treatment

Treatment depends on the aetiology of GH excess. Various treatments involving surgery and drugs have been used to treat gigantism and acromegaly.

4.2 Hyposomatotropism (Pituitary Dwarfism or GH Deficiency)

4.2.1 Definition

Pituitary dwarfism is a condition in which a delay of growth results in short stature. It is due to GH deficiency.

4.2.2 Epidemiology

GH deficiency is a rare disorder, with estimated prevalence being approximately 1 in 4000.

4.2.3 Aetiology/Pathogenesis

The aetiology of GH deficiency can be divided into congenital and acquired. Acquired causes include midline tumours, cranial irradiation, traumatic brain injury, central nervous system infections or inflammation. Congenital causes include genetic mutations and structural brain malformations.

4.2.4 Diagnosis

Pituitary dwarfism is a form of harmonic short stature (abnormal stature with normal body proportions). The lack of GH production results in reduced ossification of the growth plates. GH deficiency should be considered when height is more than 3 standard deviations (SD) below the mean and height velocity is <6 cm/year before

the age of 4, <5 cm/year from 4 to 8 years of age or <4 cm/year before puberty onset.

Differential diagnosis with hypothyroid dwarfism is based on the absence of mental retardation and myxoedema in pituitary dwarfism. Radiographic findings show thinned cortical bone in long-bone diaphysis. Laboratory tests are needed to confirm diagnosis.

4.2.5 Treatment

GH deficiency is treated by replacing it with recombinant human growth hormone (rHGH), manufactured by recombinant DNA technology.

4.3 Hyperthyroidism

4.3.1 Definition

Hyperthyroidism occurs when the thyroid gland overproduces thyroid hormones.

4.3.2 Epidemiology

The prevalence of overt hyperthyroidism ranges from 0.2 to 1.3% in iodine-sufficient parts of the world.

4.3.3 Aetiology/Pathogenesis

The most common causes of hyperthyroidism are the Graves-Basedow disease (an autoimmune disorder in which thyroid-stimulating antibodies overstimulate the thyroid), the toxic adenoma and the toxic multinodular goitre (single or multiple secreting thyroid nodules).

4.3.4 Diagnosis

Hyperthyroidism determines irritability, nervousness, increased heart rate, tremors, weight loss, diarrhoea and heat intolerance. The locomotor system is also affected, with patients experiencing fatigue and muscle weakness. Thyroid hormones stimulate osteoclasts to increase bone resorption leading to osteoporosis and pathologic fractures.

4.3.5 Treatment

Treatment depends on the aetiology of hyperthyroidism. Antithyroid drugs and surgery are the most common forms of treatment in order to relieve symptoms.

4.4 Hypothyroidism

4.4.1 Definition

Hypothyroidism is a disorder of the thyroid gland that does not produce enough thyroid hormones [triiodothyronine (T3) and thyroxine (T4)].

4.4.2 Epidemiology

The prevalence of overt hypothyroidism in the general population varies between 0.3 and 3.7% in the USA. Hypothyroidism occurs more frequently in women, in older people (>65 years) and in patients with autoimmune diseases.

4.4.3 Aetiology/Pathogenesis

Worldwide, iodine deficiency is the most common cause of thyroid enlargement and goitre. Indeed, iodine is an essential component of thyroid hormones. In iodine-sufficient areas, the most common cause of hypothyroidism is chronic autoimmune thyroiditis (also known as Hashimoto's disease). Iatrogenic hypothyroidism can be permanent, when induced by surgical treatment leading to permanent thyroid damage, or reversible, when caused by drugs interfering with thyroid function.

4.4.4 Diagnosis

Hypothyroidism can be congenital or autoimmune. These two forms affect patients in differ-

ent periods of life and have separate clinical patterns. Clinical evaluation, epidemiology and laboratory tests are needed in order to make a correct diagnosis.

4.4.4.1 Congenital Hypothyroidism

Congenital hypothyroidism is a thyroid hormone deficiency present at birth. It is generally due to maternal iodine deficiency during pregnancy or congenital defects in thyroid development. Clinical manifestations of this condition at birth are reduced interest in nursing, hypotonia, macroglossia (large tongue), prolonged jaundice, umbilical hernia and persistence of open fontanels. As the infant grows, the locomotor system develops cretinoid epiphyseal dysgenesis. It refers to the absence of or delayed appearance and development of epiphyseal growth centres. Furthermore, the formation of epiphyses arises from multiple loci of ossification. Radiographic findings show diffuse and scattered calcifications into the epiphyseal cartilage. This condition is generally bilateral and is more common in capital epiphyses. It may improve with adequate therapy but is frequently confused with the chondrodystrophies described by Legg, Perthes and others. Congenital hypothyroidism results in disharmonic dwarfism (short stature with disproportion of different parts of the body) called cretinism. The head is dramatically bigger than normal and presents deformities (saddle nose and hypertelorism). Other deformities include short neck, stubby limbs and delayed teething. It differs from pituitary dwarfism for some focal aspects (Table 4.1).

4.4.4.2 Autoimmune Hypothyroidism

Autoimmune hypothyroidism is more common in adult women and is determined by congenital and environmental trigger factors. Typical manifestations of hypothyroidism include goitre, reduced cold tolerance, tiredness, depression, constipation, slow heart rate and weight gain. Hashimoto's thyroiditis is the most common form of autoimmune hypothyroidism. It is characterized by antithyroid autoantibodies that target and gradually destroy the gland. Patients with hypothyroidism frequently develop carpal tunnel syndrome and other nerve entrapment syndromes because of soft tissue fluid build-up that puts pressure on the nerves. They may also present reduced osteotendon reflexes and muscle hypotrophy.

Table 4.1 Differential diagnosis between cretinism and pituitary dwarfism

Cretinism	Pituitary dwarfism
Disproportionate dwarfism	Proportionate dwarfism
Mental retardation	No mental retardation
Abnormal development of the nervous system	Normal development of the nervous system
Reproductive system is affected	Reproductive system may be unaffected

4.4.5 Treatment

Treatment targets include normalization of TSH, T3 and T4 concentrations. Levothyroxine monotherapy is the treatment of choice. However, specific treatment depends on the aetiology and differs for primary and central hypothyroidism but also for congenital and autoimmune hypothyroidism.

4.5 Hyperparathyroidism

4.5.1 Definition

Hyperparathyroidism is a condition that occurs when the parathyroid glands overproduce PTH. Primary hyperparathyroidism derives from parathyroid defects; secondary hyperparathyroidism is a response of the parathyroids to chronic hypocalcaemia that results in hyperplasia of these glands.

4.5.2 Epidemiology

Overall, the prevalence of primary hyperparathyroidism in the USA was estimated at 0.86%. This disease most often presents as an asymptomatic disorder. The most common causes of primary hyperparathyroidism are a sporadic, single parathyroid adenoma (85% of cases), parathyroid hyperplasia (15% of cases) and very rarely parathyroid carcinoma (<1% of cases). Secondary hyperparathyroidism is typical in patients with chronic kidney failure or severe vitamin D deficiency.

4.5.3 Aetiology/Pathogenesis

In advanced stages of the disease, bones become lightweight and soft. It determines typical bone deformities such as long bones curving, antero-posterior flattening of the chest and vertebral collapses. The bone marrow is replaced by a highly vascularized fibrous tissue, and the formation of bone cysts filled with serohematic fluid is frequent. Other classic manifestations of hyperparathyroidism are brown tumours: they are cyst-like lesions in bone composed of fibrous tissue, woven bone and supporting vasculature. They are secondary to osteoclast hyperactivity followed by osteoblast reparative bone deposition. The mass, that is not neoplastic, breaks through under the periosteum and causes severe pain. The characteristic brown coloration results from hemosiderin deposition into the osteolytic cysts. Histologically, brown tumours show aggregation of osteoclasts, reactive giant cells and hemosiderin.

4.5.4 Diagnosis

Asymptomatic primary hyperparathyroidism is the most common presentation. When the disease becomes symptomatic, renal and skeletal manifestations occur. Osteitis fibrosa cystica is the term given to the pathognomonic skeletal features of primary hyperparathyroidism that are evident by routine skeletal radiograph. It is present in 5% of patients with primary hyperparathyroidism and includes some typical features due to excess of PTH (Table 4.2).

Radiological findings of osteitis fibrosa cystica include salt-and-pepper degranulation of the skull, thinning of the lateral half of the clavicle, subperiosteal resorption of the distal phalanges, bone cysts and brown tumours.

Renal disorders are present in 20% of cases and include nephrolithiasis, nephrocalcinosis and hypercalciuria secondary to persistent hypercalcemia. In secondary hyperparathyroidism, chronic kidney failure symptoms are generally prevalent and skeletal manifestations are less evident.

Blood tests show high levels of PTH, hypercalcaemia and hypophosphataemia in primary hyperparathyroidism. In secondary hyperparathyroidism, there are high levels of PTH but low/normal serum calcium levels.

Because of the specific involvement of the cortical bone, computerized bone mineralometry (MOC–DEXA) shows low bone density in the distal radius (rich of cortical bone) with normal values in the lumbar spine (rich of trabecular bone).

Table 4.2 Clinical features of osteitis fibrosa cystica

Subperiosteal bone resorption of distal phalanges
Demineralization of cranial bones with osteoblastic lesions
Bone cysts
Thoracic and pelvic deformities
Brown tumours in long bones
Chondrocalcinosis and pseudogout in joints
Pathologic fractures due to osteoporosis and osteitis fibrosa cystica

4.5.5 Treatment

Treatment of choice in primary hyperparathyroidism is generally the parathyroidectomy (removal of the parathyroid adenoma). In secondary hyperparathyroidism, treatment must be directed at the underlying cause of hypocalcaemia.

4.6 Hypoparathyroidism

4.6.1 Definition

Hypoparathyroidism is a disorder characterized by low serum calcium levels due to insufficient secretion of PTH. Pseudohypoparathyroidism is a less common disorder due to target organ resistance to PTH.

4.6.2 Epidemiology

The prevalence of hypoparathyroidism was estimated at 37 per 100,000 person-years in the USA. Primary hypoparathyroidism is generally

due to genetic parathyroid defects, whereas secondary hypoparathyroidism occurs when the glands are destroyed, impaired or surgically removed.

4.6.3 Aetiology/Pathogenesis

Several conditions may lead to hypoparathyroidism. It generally occurs in case of surgical removal of the glands, congenital agenesis of parathyroids or familial predisposition.

4.6.4 Diagnosis

Hypoparathyroidism is characterized by reduced bone resorption, hypocalcaemia, hyperphosphataemia and low PTH levels. Analysis of the urine shows hypocalciuria and hypophosphaturia. Radiological findings may show osteosclerosis and increased radiopacity. Other clinical manifestations are due to chronic hypocalcaemia: the typical presentation of low calcium levels is increased neuromuscular irritability, symptoms of which include perioral numbness, muscle spasms, acral tingling and prolongation of the QTc intervals on ECG. This condition is also known as hypocalcaemic tetany, and Chvostek's and Trousseau's signs can be elicited during examination when hypocalcaemia is suspected. The Chvostek's sign is positive when muscles of the face twitch in response to tapping of the ipsilateral facial nerve. The Trousseau's sign is evoked by inflating a blood pressure cuff on the upper arm above systolic blood pressure for 3 min, showing carpal spasm, with wrist and thumb flexion.

4.6.5 Treatment

Treatment target is the normalization of serum calcium levels. Chronic hypocalcaemia is generally managed by the administration of calcium carbonate and vitamin D. The management of acute hypocalcaemia requires intravenous administration of calcium gluconate 10%, followed by slow calcium infusion over 24 h.

4.7 Hypercortisolism

4.7.1 Definition

The adrenal cortex produces three different types of steroid hormones. Therefore, three different clinical manifestations are possible in case of hypercortisolism: Cushing's syndrome (excess of cortisol), hyperaldosteronism (excess of aldosterone) and congenital adrenal hyperplasia (excess of androgens). These syndromes have multiple common features because steroid hormones have similar structure and, consequently, similar effects. Cushing's syndrome is the form of hypercortisolism that mainly affects the musculoskeletal system.

4.7.2 Epidemiology

Annual incidence of Cushing's disease is estimated to be 2.4 cases per million, with a prevalence of 39.1 cases per million. The disease is more common in women, and the female:male ratio is 3:1.

4.7.3 Aetiology/Pathogenesis

The aetiology of high levels of glucocorticoids can be exogenous, caused by overuse of cortisol medication, or endogenous. Endogenous hypercortisolism is due to pituitary overproduction of ACTH in 70% of cases (it is called "Cushing's disease" and generally refers to a pituitary adenoma). "Cushing's syndrome", instead, occurs in 30% of cases and is referred to as primary adrenal dysfunctions (secreting adrenal hyperplasia, adenoma or carcinoma). Very rarely, ectopic production of ACTH may determine hypercortisolism.

4.7.4 Diagnosis

Hypertension, overweight (central obesity of the trunk), hirsutism, impaired glucose tolerance, thinning of the skin, red striae on the trunk, frequent infections, menstrual disorders and depres-

sion are typical signs and symptoms of hypercortisolism. As regards the locomotor system, the excess of cortisol causes atrophy of type II muscle fibres, leading to loss of muscle mass and fatigue. Furthermore, cortisol increases protein catabolism: it alters the deposition of new bone matrix, generally rich in proteoglycans and glycoproteins. Osteoblast becomes unable to contrast bone resorption, and severe osteoporosis occurs. Radiographic findings include diffuse reduced radiopacity and wedge fractures of the vertebrae.

4.7.5 Treatment

The treatment of hypercortisolism depends on the aetiology. Surgical resection of the causative tumour is the treatment of choice for endogenous hypercortisolism. Exogenous hypercortisolism should be treated by gradually interrupting cortisol administration.

4.8 Hypocorticosurrenalism

4.8.1 Definition

Addison's disease, also known as primary hypocorticosurrenalism, is an endocrine disorder characterized by inadequate production of both cortisol and aldosterone, resulting in primary adrenal insufficiency.

4.8.2 Epidemiology

The estimated incidence of Addison's disease in Europe ranges from 4.4 to 6.2 new cases/million/year.

4.8.3 Aetiology/Pathogenesis

At present, autoimmunity is the predominant cause of Addison's disease accounting for 75–96% of all the cases. In the past, tuberculosis of the adrenal glands was a common cause of hypocorticosurrenalism. Primary hypocorticosurrenalism is associated with elevated ACTH levels, whereas secondary hypocorticosurrenalism is generally due to pituitary dysfunctions and presents low ACTH levels.

4.8.4 Diagnosis

The acute form of hypocorticosurrenalism is a life-threatening condition resulting in hypovolemic shock. The chronic form is characterized by fatigue, nausea, abdominal pain, weight loss and orthostatic hypotension. Musculoskeletal manifestations are possible, including myalgia, backache and delayed skeletal maturation.

4.8.5 Treatment

Treatment depends on the aetiology of hypocorticosurrenalism. Administration of the missing glucocorticoids and mineralocorticoids is necessary to relieve symptoms.

Take-Home Message

- Several endocrine disorders may affect the musculoskeletal system altering bone metabolism and growth.
- Endocrinopathies may also occur in adult patients, affecting the locomotor system and leading to muscle atrophy, osteoporosis and increased risk of pathologic fractures.
- Acromegaly and gigantism are two very similar conditions, but they affect patients in different periods of life.
- Pituitary dwarfism and cretinism are two common causes of short stature.
- In advanced stages, hyperparathyroidism determines severe damages to the locomotor system and kidneys.
- Cushing's syndrome leads to osteoporosis, muscle atrophy and central obesity.

Summary

- Hypersomatotropism is a condition due to uncontrolled secretion of GH. If the hormonal hypersecretion is present prior to physeal fusion, then gigantism results. Alternatively, if the increased levels of GH are present after skeletal maturity, then acromegaly is the resultant condition.
- Pituitary dwarfism is a condition in which a delay of growth results in short stature. It is due to GH deficiency.
- In patients with hyperthyroidism, thyroid hormones stimulate osteoclasts to increase bone resorption leading to osteoporosis and pathologic fractures.
- Congenital hypothyroidism results in disharmonic dwarfism (short stature with disproportion of different parts of the body) called cretinism.
- Differential diagnosis between pituitary dwarfism and cretinism is based on the proportions of different parts of the body and on the development of the nervous system and reproductive system.
- Hyperparathyroidism determines typical bone deformities such as long bones curving, anteroposterior flattening of the chest and vertebral collapses.
- Classic manifestations of hyperparathyroidism are brown tumours.
- Chvostek's and Trousseau's signs are classic manifestations of hypocalcaemia in patients with hypoparathyroidism.
- Cushing's syndrome leads to muscle mass loss, muscle weakness and osteoporosis.
- Severe fatigue is a hallmark of Addison's disease.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Patients with Cushing's syndrome present:
   (a) Hypoglycaemia
   (b) Osteoporosis
   (c) Hypotension
   (d) Muscle hypertrophy
   (e) Muscle atrophy
2. Which syndrome is frequent in patients with acromegaly?
   (a) Carpal tunnel syndrome
   (b) De Quervain's syndrome
   (c) Marfan's syndrome
   (d) Erdheim's syndrome
   (e) Sjogren's syndrome
3. What is a brown tumour of the bone?
   (a) Focal bony lesions due to bone remodelling
   (b) A malignant neoplasm
   (c) A form of bony callus
   (d) A bone metastasis
   (e) Cyst-like lesions composed of fibrous tissue and supporting vasculature
4. Clinical findings of pituitary dwarfism are:
   (a) Disproportion of parts of the body
   (b) Mental retardation
   (c) Altered development of the nervous system
   (d) Dysfunction of the reproductive system
   (e) Harmonic short stature
5. Cretinism occurs in case of:
   (a) Hashimoto's thyroiditis
   (b) Total thyroidectomy
   (c) Graves-Basedow disease
   (d) Thyroid adenoma
   (e) Maternal iodine deficiency during pregnancy
6. Patients with Addison's disease present:
   (a) Myalgia
   (b) Hypovolemic shock
   (c) Fatigue
   (d) Renal insufficiency
   (e) Internal bleeding

Further Reading

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The Musculoskeletal System in Blood Diseases

5

Vincenzo Denaro, Maria Cristina Sangiovanni, Sergio De Salvatore, and Umile Giuseppe Longo

Overview

Haematological diseases may involve the musculoskeletal system. Diagnosis is clinical and radiographic. Treatment requires a multidisciplinary approach tailored to the patient.

5.1 Multiple Myeloma

5.1.1 Definition

Multiple myeloma (MM) is a neoplastic proliferation of monoclonal, malignant plasma cells in the bone marrow. It belongs to neoplasms known as paraproteinaemia/plasma cell dyscrasias/monoclonal gammopathies. The main characteristic of these malignancies is the abnormal production of monoclonal immunoglobulin IgG (60%) or IgA (25%), named the "M component".

5.1.2 Epidemiology

MM accounts for 1% of all cancers and is the second most common haematologic malignancy after lymphoma. The mean age at diagnosis is 65 years, and <3% of patients are younger than 40 years.

5.1.3 Pathogenesis

Risk factors include radiation, chromosomal mutation of the immunoglobulin heavy-chain switch region (chromosome 14), MYC and RAS over-expression, and dysregulation of p53 and Rb1 oncosuppressor. Moreover, occupational exposure was identified as a possible risk factor.

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5.1.4 Diagnosis

Pathologic fractures are the most common clinical manifestation of MM. Plasma cell proliferation and osteoclast activation-induced bone resorption decrease the strength of the bone segment, resulting in pathologic fractures. In vertebral fractures, the onset could be acute with compression of myeloradicular structures and neurological signs or progressive producing skeletal deformities.

Pain is common, and it is the first symptom of myeloma. The spine (especially the lumbar spine) and the ribs are the most frequent sites involved. Patients with MM refer that pain worsens with movement and regresses with rest, affecting the quality of life.

The diagnosis of myeloma is based on the presence of a classic triad consisting of bone marrow plasmacytosis >10%, M component in serum (>3 g protein), and presence of at least one of the CRAB symptoms (C = high levels of calcium, R = renal failure, A = anaemia, B = bone lesions).

Osteolytic lesions could be observed through conventional radiographs and confirmed by the absence of uptake on scintigraphy examination. The geode presence is characteristic in MM: it is a radiolucent area with sharp margins and without sclerotic edge. The skull ("salt-and-pepper" skull), pelvis, ribs, humerus, and proximal femur are the typical locations. However, other types of radiographic patterns may be present:

- An extensive bone resorption process could be found at the level of transverse apophyses or vertebral pedicles ("winking owl" sign).
- The diffuse lytic pattern of the spine is associated with vertebral collapses.
- Voluminous lesions (ribs, long bones, pelvis, and skull) are due to intraosseous proliferation of plasma cells. Cortical erosion and subperiosteal bone neo-apposition are usually present.

Magnetic resonance imaging (MRI) often reveals bony lesions not detectable by X-ray and allows to assess a possible involvement of myeloradicular structures.

5.1.5 Treatment

Orthopaedic treatment should be part of a multidisciplinary approach, which includes radio-chemotherapy.

5.1.5.1 Conservative Treatment

In appendicular skeletal injuries, the conservative treatment includes the use of orthoses and assisted walking aids. Moreover, stable vertebral fractures without myeloradicular involvement require orthoses. Bracing aims to support the spine and protect the segment at risk of fracture. It is also indicated as a bridge to surgery in the pre- and postoperative period.

5.1.5.2 Surgical Treatment

Osteosynthesis is indicated in high-risk fracture and pathological fractures of the appendicular skeleton. Vertebral decompression and stabilisation are indicated in unstable lesions and in case of neurological involvement (Fig. 5.1). A biopsy is always performed during the surgical procedure.

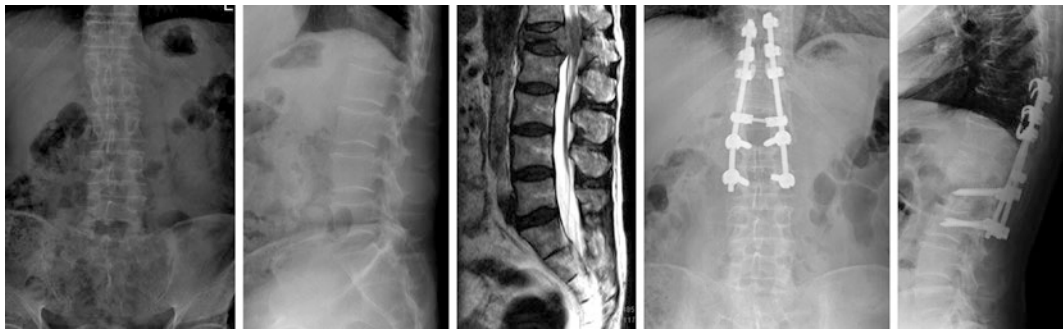

Fig. 5.1 Vertebral decompression and stabilisation of a pathological vertebral fracture caused by myeloma

5.2 Haemoglobinopathies

5.2.1 Sickle Cell Disease or Drepanocytosis

5.2.1.1 Definition

Sickle cell disease is an autosomal recessive hereditary form of chronic haemolysis. It is characterised by sickle cells in blood and recurrent painful crises.

5.2.1.2 Epidemiology

The highest prevalence of sickle cell disease worldwide is reported in sub-Saharan Africa, followed by the Middle East and Indian subcontinents.

5.2.1.3 Pathogenesis

Drepanocytosis is caused by a point mutation in the β-globin gene (at position 6), with glutamate-valine substitution. The mutated haemoglobin is known as HbS and polymerises in a deoxygenated state within the red blood cell. Precipitates of haemoglobins stiffen the cell membrane, and hence, erythrocytes lose elasticity causing haemolytic anaemia and vaso-occlusive phenomena.

5.2.1.4 Diagnosis

The most common clinical manifestations include the following

- Painful crises caused by intermittent ischaemia episodes with acute pain, fever, tachycardia, and anxious state. Hypoxia, dehydration, acidosis, infection, and fever could induce crises.
- Splenic infarction typically occurs in the first 18–36 months, resulting in infection from Pneumococcus or Haemophilus.
- Chronic arthropathy, aseptic necrosis of the femoral head, and osteomyelitis are also common.

Diffused bone lesions in sickle cell disease are caused by bone marrow hyperplasia. Patients experience thickening of the diploë, especially at frontal and parietal level, with a "brush skull" pattern. The long bones are also involved and appear expanded, with thinned cortical and rarefied bony trabeculae. Diffuse thrombosis causes ischaemic necrosis resulting in focal bone lesions. On X-ray, these lesions are characterised by a radiolucent area surrounded by a radiopaque edge. Progressive joint necrosis could result in degenerative arthrosis. Characteristic haemolytic anaemia, erythrocyte morphology, and intermittent episodes of ischaemic pain are suspicious for sickle cell disease. The diagnosis is confirmed by haemoglobin electrophoresis, sickle cell test, and peripheral blood smear.

5.2.1.5 Treatment

An acute sickle cell pain crisis is treated with fluids, analgesics, oxygen, red blood cell transfusions, or exchange transfusions. Folic acid replacement and hydroxyurea are used to decrease the vaso-occlusive pain crisis frequency. Bone marrow transplantation can be curative in severe case.

5.2.2 Thalassaemia Syndromes

5.2.2.1 Definition

Thalassaemia is a hypochromic, microcytic anaemia due to congenital underproduction of α- or β-globin chains of the haemoglobin molecule.

5.2.2.2 Epidemiology

The α-thalassaemia reported the highest frequency across the tropical belt area. The β-thalassaemia is less common in sub-Saharan Africa and spreads across the rest of the tropical belt at varying frequencies.

5.2.2.3 Pathogenesis

Four genes regulate α-globin synthesis. Four different types of α-thalassaemia can be distinguished due to the type of the deletion of the α-globin gene: α+ trait thalassaemia (one gene deleted), α0 thalassaemic trait (two genes deleted), HbH disease (three genes deleted), and foetal hydrops with Bart's haemoglobin (deletion of all four α-globin genes, incompatible with life).

β-Globin synthesis is regulated by two genes (one pair of alleles). Two different β-thalassaemia can be distinguished: thalassaemic trait (β0/β heterozygosity) and thalassaemia major or Cooley's disease (homozygous mutations of both genes resulting in absent production of beta chains, β0/β0).

5.2.2.4 Diagnosis

The onset of Cooley's disease symptoms is at 6 months when the switch from foetal to adult haemoglobin occurs. Extramedullary haematopoiesis and expansion of haematopoietic marrow within flat bones cause the clinical manifestations. Growth failure, hepatosplenomegaly, jaundice, and bony deformities are frequent. Hypertrophy of the maxillaries, consequent protrusion of the cheekbones, widening and flattening of the root of the nose, and teeth alterations constitute a typical pattern named "facies from Tamia". Involvement of the long bones is characterised by enlargement of the medullary canal due to the cortical thinning. Moreover, an expansion of bone segments may occur, especially in hand (metacarpals and phalanges with a barrel-like appearance), distal femur, and anterior portion of the ribs. Rarefied trabeculae are observed into the cancellous bone. However, in late stages, new bone production occurs, and trabeculae assume an irregular distribution. In severe cases, growth retardation may be present.

5.2.2.5 Treatment

Patients with β-thalassaemia major require multiple blood transfusions. Otherwise, the iron overload needs to be treated with deferasirox. Splenectomy and bone marrow transplantation are useful treatment.

5.3 Lymphomas and Leukaemia

5.3.1 Hodgkin's Lymphoma

5.3.1.1 Definition

Hodgkin's lymphoma is a malignant neoplastic transformation of lymphocytes. It is histologically characterised by Reed-Sternberg cells that spread to contiguous areas of lymph nodes.

5.3.1.2 Epidemiology

Hodgkin's disease has a bimodal age distribution, affecting patients <20 years old and >60 years old.

5.3.1.3 Diagnosis

The lymph nodes could appear enlarged, painless, rubbery, non-erythematous, and non-tender. Patients may also develop "B symptoms", represented by night sweats, 10% weight loss, and fevers. Pruritus is not included in the "B" symptoms. Cervical, supraclavicular, and axillary lymphadenopathy are the most affected sites in the early stages. Spleen, skin, gastric, lung, central nervous system (CNS), and other organs may be involved.

Bone localisation occurs in 10–15% of cases due to a direct invasion from adjacent lymph nodes or by the haematogenous or lymphatic spread. Localised pain, deformities (hump) with myeloradicular syndromes, and pathological fractures may occur. The spine is the most frequent site involved. The vertebral soma may assume a purely osteolytic appearance or present diffuse sclerosis, assuming the "ivory vertebra" characteristic pattern.

Excisional lymph node biopsy is essential for the diagnosis. X-ray, computed tomography scan (CT), and MRI are used to assess the extension of the disease. In vertebral localisation, MRI also identifies the boundaries of the lesion and its relationship to myeloradicular structures. The pattern of lesions on radiographic and CT examination is variable (usually lytic); however, after successful radio-chemotherapeutic treatment, bone localisations become sclerotic. Lymphangiography and laparotomy are no longer routinely used for staging. Scintigraphy may help evaluate multiple lesions.

5.3.1.4 Treatment

Treatment is based on chemotherapy and radiation therapy. The orthopaedic treatment principles of patients with lymphoma are similar to myeloma, previously described.

5.3.2 Non-Hodgkin's Lymphoma

5.3.2.1 Definition

Non-Hodgkin's lymphoma (NHL) is a neoplastic transformation of both B and T cells. Neoplastic cells accumulate in lymph nodes and extralymphatic organs. In cases of musculoskeletal involvement, the most frequent sites are femur, tibia, humerus, pelvis, and spine.

5.3.2.2 Epidemiology

NHL is the fifth most frequently diagnosed cancer in the United Kingdom. The worldwide incidence of NHL is 19.6 per 100,000 people per year.

5.3.2.3 Pathogenesis

Human immunodeficiency virus (HIV), hepatitis C, Epstein-Barr, human T-lymphotropic virus type I (HTLV-I), *Helicobacter pylori*, and other infections predispose to NHL development.

5.3.2.4 Diagnosis

NHL has similar findings with Hodgkin's lymphoma. The main difference between Hodgkin's and NHL is the localisation. The cervical and supraclavicular node localisation occurs in only 10–20% of NHL patients, whereas it occurs in 80–90% of the Hodgkin's cases. Extra-lymphatic site, CNS, and blood involvement is usually more common in NHL. The prognosis is worse in case of metastases. The radiological pattern is characterised by blurred edge osteolysis with minimal periosteal reaction. The cortical bone may be disrupted, and, in the advanced stages of the disease, neoplastic tissue may invade adjacent soft tissues. The excessive weakening of the bone can lead to pathological fractures. Bone scintigraphy and PET scans are highly sensitive to detect skeletal localisation.

5.3.2.5 Treatment

The NHL is highly radiosensitive; therefore, surgical treatment is usually not necessary. In case of vertebral localisations, neurological involvement, or fractures, surgical treatment is required.

5.4 Leukaemia

5.4.1 Acute Leukaemia

5.4.1.1 Definition

Leukaemia is the common name for several malignant disorders present with increased numbers of leucocytes in the blood and/or the bone marrow. The derangement of the pluripotent stem cells in bone marrow leads to acute leukaemia.

5.4.1.2 Epidemiology

Leukaemia may affect all ages, with different distribution for each type of the disease. Acute lymphoblastic leukaemia is most common in early childhood.

5.4.1.3 Diagnosis

The clinical presentation is characterised by pain at palpation, diffuse redness, and joint swelling. The pain becomes constant in the late stages and is typically localised in the metaphyseal region. Acute osteomyelitis and joint rheumatism are alternative diagnoses to exclude. Spontaneous fractures are rare, and the vertebrae are the most commons site involved. The bone lesions present a characteristic radiological pattern with radiolucent bands in the metaphyseal region. This aspect is due to the bone resorption and the compressive action of the leukemic tissue that avoid bone formation. The cortical of the long bones is usually not affected. The radiographic examination may also reveal periosteal reaction phenomena and pathological fractures (e.g. vertebral collapses). The periosteal reaction in long bones, hands, and feet can provide an "onion bulb" morphology.

5.4.1.4 Treatment

Haematologic treatment is based on chemotherapy. The orthopaedic treatment of fractures requires stabilisation and fixation.

5.4.2 Chronic Leukaemia

5.4.2.1 Definition

Chronic leukaemia is a chronic lymphoproliferative disorder, composed by monomorphic lymphocytes. It involves peripheral blood, bone marrow, and lymphoid organs.

5.4.2.2 Epidemiology

The incidence of chronic leukaemia increases by age. In Europe, the median age at diagnosis is 55–60 years.

5.4.2.3 Diagnosis

Bone lesions in chronic leukaemia are less frequent than in acute leukaemia. Localised bone lesions radiographically appear as radiolucent areas, with undefined margins and shapes. They are usually located in ribs, vertebrae, sternum, and skull. Pain, pathological fractures (especially vertebrae), and swelling are common signs. Radiographic examination shows increased bone

transparency due to thinning of the trabeculae and the cortical bone. In rare cases, some osteosclerotic lesions could appear. This condition is asymptomatic and is detected by radiographic examination due to the marked radiopacity of bony segments.

5.4.2.4 Treatment

Haematologic treatment is based on chemotherapy. The orthopaedic treatment of fractures requires stabilisation and fixation.

5.5 Haemophilia

5.5.1 Definition

Haemophilia is an X-linked recessive Mendelian disorder, characterised by the deficiency of factor VIII (haemophilia A) or factor IX (haemophilia B), resulting in an increased risk of bleeding.

5.5.2 Epidemiology

Haemophilia affects approximately 1/10,000 people. Haemophilia A (1/6000 males) is more common than haemophilia B (1/30,000 males).

5.5.3 Diagnosis

Recurrent joint haemarthrosis is the most frequent manifestation and occurs between 4 and 10 years of age. Generally, bleeding has a synovial origin. In the early stages, haemarthrosis resolves spontaneously, without residual joint injury. The haemarthrosis onset is acute and associated with pain and swelling. After a few hours, the synovial cavity fills with blood resulting in joint locking. A palpatory sensation of crackling may be appreciated. After a first episode, the full healing requires about 3 weeks; otherwise, recurrences are frequent. Further episodes lead to progressive joint injury and chronic arthropathy, characterised by synovial hyperplasia, cartilage resorption, and haemorrhagic areas in the cancellous epiphyseal bone. Ankylosis and capsule retraction occur in late stages.

The clinical pattern differs depending on the affected joint. Involvement of the hip and knee is extremely painful and results in joint locking. Elbow and finger involvements result in early ankylosis. Conversely, the shoulder does not experience ankylosis. In the early stages of the disease, radiographic examination in haemarthrosis reveals a distended joint cavity with diffuse radiopacity. In chronic arthropathy, the first radiologic signs are narrowing of the joint line associated with osteoporosis and subchondral osteosclerosis. Subchondral and subperiosteal haemorrhages could change the epiphysis appearance, with single or multiple geodesic lesions surrounded by a sclerotic rim. In long-standing forms, the epiphyses progressively collapse to mechanical load, and the joint line disappears.

5.5.4 Treatment

Haemophilia patients should be treated by a multidisciplinary team. Arthrocentesis is contraindicated for haemarthrosis due to the high risk of bleeding and recurrence. Therefore, joint immobilisation with orthoses is the correct treatment. Moreover, blood or plasma transfusions are required to restore coagulation factors. In the chronic arthropathy stage, pain therapy and education of the patients are performed. In chronic synovial proliferation and recurrent haemarthrosis, synovectomy (open or arthroscopic) is indicated to reduce bleeding episodes. Total joint replacement is performed in cases of severe joint injury.

Take-Home Message

- Haematological disorders are frequently involved in the musculoskeletal system.
- The haematologic patient is difficult to treat and requires a multidisciplinary approach.
- Nowadays, haemophiliacs, those with the joint disease, could be treated with excellent results, comparable to those suffering from degenerative arthritis.

Summary

- Myeloma is a haematologic neoplasm due to the proliferation of plasma cells derived from a single clone. Bone lesions are usually lytic, without a sclerotic rim. Vertebral collapses may cause neurological symptoms due to compression on myeloradicular structures. The diagnosis of myeloma is based on the presence of a classic triad. Conventional radiographs allow assessment of isolated or diffuse osteolytic lesions. Radiological lesions are determined by bone resorption induced by plasma cell proliferation. The most frequent radiographic pattern is the geodesic image. Orthopaedic treatment must be placed in the context of a multidisciplinary approach, including the use of radio- and chemotherapy.
- Sickle cell anaemia is an inherited haemoglobinopathy characterised by a point mutation globin gene. Generalised bone lesions are caused by bone marrow hyperplasia. The affected subject presents thickening of the diploë, especially at the frontal and parietal level, with a "brush skull" pattern.
- Thalassaemia syndromes are genetic alterations in the synthesis of α- or β-globin chains. Ineffective erythropoiesis results in increased erythropoietin production, ectopic haematopoiesis, erythroblastic hyperplasia, bone alterations, and osteopenia.
- Hodgkin's lymphoma is a malignant lymphoma that could involve the musculoskeletal system. The most frequent radiologic appearance is the osteolytic lesion with blurred margins.
- Primitive and secondary lesions of non-Hodgkin's lymphomas could affect the musculoskeletal system. The prognosis is worse if the involvement is secondary. The classic radiologic appearance is osteolysis with blurred margins, and few or absent periosteal reaction.
- Bone lesions in acute leukaemia occur primarily in children. The typical bone lesions are radiolucent due to bone resorption. The most distinctive aspect of resorption is the presence of radiotransparent bands at the metaphyseal site.
- Bone lesions during chronic leukaemia are less frequent than acute leukaemia. The lesions are due to bone resorption and may be localised or diffused.
- Haemophilia is an X-linked recessive Mendelian disorder, characterised by a severe insufficiency of the coagulation cascade due to total or partial lack of factor VIII (haemophilia A) or IX (haemophilia B). Haemarthrosis is the most frequent manifestation. The rate of recurrence is high, and this condition could lead to progressive joint deformation.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Myeloma:
   (a) Is the most frequent paraproteinaemia
   (b) Produces lytic-type bone lesions
   (c) Is characterised by the presence of a monoclonal component in the serum or urine
   (d) Can lead to pathologic fractures
   (e) Is typical of adolescence
2. Sickle cell anaemia:
   (a) Can give haemolytic anaemia
   (b) Can give vaso-occlusive phenomena at the bone level
   (c) Usually has indolent vaso-occlusive syndromes
   (d) Is diagnosed with the useful Hb electrophoresis
3. β-Thalassaemia syndromes:
   (a) They are always associated with severe anaemia
   (b) At the skeletal level, they may involve both growth and bone development
   (c) A typical deformity is the brush skull
4. α-Thalassaemia syndromes:
   (a) Deletion of all four genes is compatible with life
   (b) Four different forms can be distinguished
   (c) They typically give lytic lesions at the level of the spine
5. Lymphomas:
   (a) Hodgkin's lymphoma can give secondary manifestations at the skeletal level

(b) The first symptom with which Hodgkin's lymphoma tends to manifest is localised pain at the affected site
(c) Hodgkin's lymphomas can affect both primary and secondary bone tissues
(d) Non-Hodgkin's lymphomas can affect bone tissue both primitively and secondarily
(e) The sites most frequently affected by non-Hodgkin's lymphoma are the femur, tibia, humerus, pelvis, and spine

Further Reading

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The Musculoskeletal System in Neurological Pathologies

6

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Overview
Several neurological diseases could affect the musculoskeletal system. This chapter aims to highlight the osteoarticular involvements in neurological pathologies.

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6.1 Neurofibromatosis

6.1.1 Definition

The term "neurofibromatosis" (NF) includes two different types of the disease: type 1 (NF1) and type 2 (NF2). Von Recklinghausen described NF1 in 1882 for the first time. The main characteristic of NF1 is the presence of multiple neurofibromas (a benign tumour that originates from peripheral nerves). Other multiple tumour and nontumor manifestations could be present. A wide range of phenotypic variability could be manifested, also within the same family. NF1 usually tends to progress over time. The NF2 is rarer than NF1 and involves the brain, ocular nerves, and auditory nerves.

6.1.2 Epidemiology

NF1 affects approximately 1/3000 people worldwide; instead, NF2 usually affects only 1/300 individuals, with an incidence of 1/2500 live births. Due to the higher incidence and the greater musculoskeletal involvement, only NF1 will be treated in this chapter.

6.1.3 Pathogenesis

The neurofibromatosis is a genetic disease that involves neuro-ectodermal and mesodermal tis-

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sues. Both types of NF are genetically based, and the pattern is autosomal dominant. The NF1 gene (chromosome 17) is an oncosuppressor involved in the production of the neurofibromin. The mutation of the NF1 gene could cause dysregulation of the RAS/MAPK pathway. Approximately 50% of patients with NF1 present new mutations and have no parents affected. In rare cases, the parents of affected children could present germline mosaicism. The risk of NF1 transmission to any offspring is 50%.

6.1.4 Diagnosis (Clinical and Imaging)

The musculoskeletal system is often involved in NF1. Skeletal manifestations in NF1 include dystrophic and non-dystrophic alterations in the spine and dysplastic alterations in the tibia (congenital tibial pseudoarthrosis) and other bone segments. The spine is the most frequently involved bony segment (about 50% of cases) being the location of multiple dystrophic deformities, often localized in the cervical tract (30%). The diagnosis of NF1 is based on family history and careful clinical examination. Identifying at least two diagnostic criteria recognized by the National Institute of Health (NIH) is sufficient to make the diagnosis of NF1. The criteria include the most frequent clinical manifestations and familiarity (Table 6.1). The imaging with X-ray and MRI is also required to diagnose NF1 properly.

Table 6.1 Diagnostic criteria for neurofibromatosis type 1 (National Institute of Health, NIH)

Two or more of the following criteria
1. ≥6 Café-au-lait macules >5 mm in maximum diameter in pre-pubertal subjects and >15 mm in maximum diameter in post-pubertal subjects
2. ≥2 cutaneous or subcutaneous neurofibromas or ≥1 plexiform neurofibroma
3. Lentiginosis (or "freckling") in the axillary or inguinal regions
4. Optic nerve glioma
5. ≥2 Lisch nodules
6. Typical bony lesion (e.g. sphenoid dysplasia or cortical thickening of a long bone, with or without pseudarthrosis)
7. At least one first-degree relative with neurofibromatosis I diagnosed by the above criteria

6.1.4.1 Spinal Disorders

The dystrophic alterations of the spine include characteristic radiographic abnormalities: pencilling of the transverse processes and the ribs, short segment curves with tight apex, vertebral scalloping, severe apical rotation, and kyphoscoliosis. Scoliosis in NF1 could have two patters: non-dystrophic (similar to idiopathic scoliosis) and dystrophic.

The non-dystrophic pattern is the most common in NF1 patients. The prognosis is good, but 60% of cases progress into the dystrophic pattern. Risk factors for the pattern degeneration are age <7 years old and Cobb angle greater than 30°. The treatment is comparable to idiopathic scoliosis. Observation is required for Cobb angle <20°, bracing for Cobb between 20° and 45°, and surgical treatment for cases with Cobb angle >45°.

The dystrophic pattern is the less common form, and the prognosis is poor. It is characterized by rapid progression, development of disabling deformities, and neurological complications, often despite treatment. Dystrophic scoliosis includes two distinct subtypes, based on the type of curvature: type I is coronal plane scoliosis associated with a sagittal plane kyphosis <50°. In contrast, type II is characterized by severe sagittal plane deformities with kyphosis >50°. In both cases, observation is recommended for curves <20°, while surgery is indicated for curves >30°, as the use of bracing has proven to be ineffective. A posterior arthrodesis is performed for type I; instead, an anterior or anterior-posterior combined approach is required for type II. Due to the high risk of scoliosis, all patients with NF1 required screening and observation every 6 months. A magnetic resonance imaging (MRI) is required before surgery to exclude paravertebral tumours (present in 40% of cases).

6.1.4.2 Congenital Tibial Pseudoarthrosis

Congenital pseudoarthrosis is rare in the general population (1/190,000 live births) but occurs in 75% of patients with NF1. It is characterized by a progressive anterolateral bowing of the tibia, with cortical thickening and bone marrow narrowing. It occurs within the first year of life or,

more rarely, at birth. Without treatment, it is associated with spontaneous tibial fractures (25% of cases within the first year of life and 100% of cases before the eighth year) and consequent pseudoarthrosis. Histological studies have shown an increase in spindle cell elements with myofibroblastic differentiation and reduced osteocyte elements, an expression of a reduced regenerative capacity of the bone. The treatment is based on orthoses; however, for severe cases, surgical treatment is required. The orthoses are useful to prevent the onset of fractures in dysplastic bone, improve walking, and delay surgery in fracture cases. However, orthoses alone are not sufficient to achieve fracture consolidation. In the case of multiple fracture recurrences, amputation may be necessary.

6.1.4.3 Extraskeletal Manifestations

The main extraskeletal manifestations of NF1 include cutaneous or subcutaneous neurofibromas, plexiform neurofibromas (benign tumours that infiltrate the peripheral nerve and adjacent tissues), optic nerve gliomas, pigmented iris nodules (Lisch nodules), flat and hyperpigmented skin lesions (café-au-lait macules), and lentigines (or "freckling"). In NF1 patients, a higher incidence of psychiatric disorders (about 30%) and motor deficits is described. Moreover, NF1 is associated with increased cardiovascular risk, higher incidence of melanomas, pheochromocytomas, endocrine tumours of the pancreas, and soft-tissue sarcomas.

6.1.5 Treatment

Management of NF1 comprises observation, surgical excision of progressive lesions, and genetic counselling. Dermal neurofibromas can be resected by plastic surgery, use of CO_2 laser, or electrodesiccation. Optic gliomas do not usually require treatment, but symptomatic, progressive lesions are generally treated with chemotherapy. Radiation therapy is avoided in young children due to vascular complications and tumour malignancy caused by radiation. The treatment of NF1 scoliosis and congenital tibial pseudoarthrosis has been previously described.

6.2 Neuropathic Arthropathy

6.2.1 Definition

Mitchell first described neuropathic arthropathy in 1831. In 1868, Charcot was the first to report the main characteristic of this disorder in detail; therefore, it is also known as "Charcot arthropathy". Nowadays, the first cause of Charcot arthropathy is diabetes mellitus. However, in the past, the most common cause of neuropathic arthropathy was the advanced syphilis infection. After the development of the first antibiotics, tertiary syphilis with joint involvement has become infrequent. Therefore, tabetic arthropathy is often not recognized.

Neuropathic arthropathy is a chronic degenerative disorder (caused by a neurological disease) characterized by progressive destruction of one or more joints, with the loss of joint congruence and alteration of the whole bone architecture. Nociceptive sensitivity deficit is the common feature of these pathologies. It exposes the joint to repeated microtrauma, resulting in microfractures, subluxations, and dislocations. Furthermore, although many subjects experience nociceptive sensitivity deficit, only a small percentage of them develop neuropathic arthropathy. The incidence of this condition depends on the leading neurological cause. The diagnosis is made on clinical and neurological examinations.

6.2.2 Pathogenesis

Several factors may cause neuropathic arthropathy:

- *Tabes dorsalis*. Syphilis represents one of the most frequent causes. Lower limbs are frequently involved.
- *Diabetic neuropathy*. It is the most frequent cause. All the peripheral joints are involved, in particular the foot.
- *Syringomyelia*. It is associated with neuropathic arthropathy, especially of the large joints.
- Other causes may be neurological disorders, like spina bifida, myelomeningocele, spinal injury or compression, peripheral nerve injury, and neurological infection from

Mycobacterium leprae (leprosy). Lastly, chronic alcoholism should be included.

6.2.3 Tabetic Arthropathy

6.2.3.1 Definition

Tabes dorsalis is a neurological disease characterized by the progressive demyelination of the neural tracts primarily in the dorsal root ganglia of the spinal cord. Antibiotics have markedly reduced the occurrence of tertiary syphilis as the leading cause; however, tabes dorsalis still represents one of the most common causes of neuropathic joint disease. Approximately 5–10% of patients with tabes experience tabetic arthropathy. The nociceptive deficit causes the insurgence of this condition.

6.2.3.2 Epidemiology

The incidence of tabes dorsalis is around 250/100,000 patients with syphilis and 5000/100,000 patients with neurosyphilis.

6.2.3.3 Diagnosis (Clinical and Imaging)

The clinical characteristic of tabetic arthropathy is the lack of spontaneous pain at mobilization or walking. The patient, not feeling pain, continues to walk, being only limited by mechanical obstacle, further aggravating the lesions. The clinical and radiological signs frequently do not correspond.

The first symptom is a sudden haemarthrosis with or without joint instability. However, in other cases, the onset of swelling is slow and progressive.

The most frequent localizations are knee, foot, and hip. Shoulder, elbow, and hand are rarely affected.

In the tabetic foot, diffuse osteoarticular lesions are present. In detail, four deformities are present: shortening, thickening of the inner rim, a complete collapse of the plantar vault, and metatarsal deviation.

Vertebral involvement is common but painless; therefore, it is frequently not diagnosed. Rarely, increased lumbar lordosis and/or hump may be present. In some cases, arthropathic changes to the spine may result in compression of myeloradicular structures with neurologic syndromes.

Swelling, hypermobility, and progressive joint oedema are the most frequent signs on clinical examination. In the late stages of the disease, considerable joint destruction is observed, with numerous intra-articular free bodies and crepitus on joint mobilization.

A radiologic pattern of marked joint disruption is present at the onset of the symptoms. The subchondral bone is always exposed. The radiological features are similar to those of osteoarthritis and include irregularity of the joint surface, sclerosis areas alternating with radiotransparent areas, and free intra-articular bodies. Other findings include intra-articular free bodies, osteophytes, synovial proliferation, capsular distension and thickening, ligamentous distension and rupture (resulting in joint instability), tissue calcification, and fistula formation. This condition could lead to a joint subluxation or dislocation.

6.2.3.4 Treatment

Due to the late diagnosis, in many cases, tabetic arthropathy does not respond to therapy. Conservative therapies include the use of orthoses to stabilize the joint. Surgical options include prosthetic replacement, arthrodesis, and amputation for untreatable cases. The high rate of postsurgical complications, e.g. loosening/poor integration of prosthetic components, pseudoarthrosis, and infections, makes the risk of surgery in these patients relatively high.

6.2.4 Syringomyelic Arthropathy

6.2.4.1 Definition

The formation of a cyst or cavity within the spinal cord is named syringomyelia. This cyst, named “syrinx”, could expand and elongate over time, destroying the spinal cord. Syringomyelia is usually localized to the cervical spine; therefore, all joints can be involved, although the upper limb is more frequently affected.

6.2.4.2 Epidemiology

Syringomyelic arthropathy occurs less frequently than tabetic arthropathy. The percentage of syringomyelic subjects with arthropathy is approximately 25–30%.

6.2.4.3 Diagnosis (Clinical and Imaging)

Syringomyelic arthropathy usually occurs in individuals with symptomatic syringomyelia. In rare cases, it may be the first symptom of the disease. The main sign is joint destruction without pain. The symptomatology is similar to that of tabetic arthropathy with haemarthrosis, joint instability, and mechanical functional limitation. Physical examination reveals swelling with signs of local inflammation and limitation to mobilization. The radiographic findings are similar to tabetic arthropathy. The most frequent location is the shoulder, where osteolytic lesions may induce the disappearance of the humeral head. Voluminous periarticular calcifications are often visible. The elbow is often involved with a hypertrophic pattern, and osteophytes and periarticular calcifications may reduce the range of motion.

6.2.5 Diabetic Neuropathy

6.2.5.1 Definition

Diabetic neuropathy is a frequent complication of diabetes, affecting approximately one-third of diabetic patients. Risk factors include age, duration of disease, and blood glucose control. Diabetic neuropathies are a set of signs and symptoms due to peripheral nerve dysfunction, occurring in diabetic patients in which all other possible causes of neuropathy have been excluded. The joint involvement (diabetic neuropathy) is frequent in the late stages of the disease.

Diabetic arthropathies are usually located in the feet. Rarely, the ankle and knee may be involved.

This condition must be distinguished from bacterial arthropathies of the foot, which represent a further complication of diabetes. Neuropathy may be asymptomatic or cause pain and paraesthesia. Lesions are mainly osteolytic with considerable bone destruction and joint osteophytes. In the late stages, lesions could result in a foot deformity: shortening of the hindfoot or forefoot, flattening of the longitudinal vault, and widening of the foot.

6.2.5.2 Epidemiology

The incidence ranged from 0.08 to 13% in the diabetic population.

6.2.5.3 Pathogenesis

Diabetic neuropathy has a multifactorial aetiology. Hyperglycaemia and metabolic alterations are the primary cause of damage and are responsible for endoneurial ischaemia and hypoxia. All nerve components are involved (Schwann cell and myelin, axon, and endoneurial capillaries). Myelinated and unmyelinated fibres are decreased in proportion to the severity of the clinical pattern. Besides, endoneurial microangiopathy correlates with the severity of the disease.

6.2.5.4 Classification

Diabetic neuropathy can be distinguished into symmetric forms and asymmetric forms

- Symmetric forms include chronic sensorimotor polyneuropathy, acute painful neuropathy, and autonomic neuropathy.
- Asymmetric neuropathies include mononeuropathy, radiculopathy, and amyotrophy.

The most common form is chronic sensorimotor polyneuropathy.

6.2.5.5 Diagnosis (Clinical and Imaging)

Symptoms differ depending on the type of neuropathy. Diabetic neuropathies are diagnosed by evaluating symptoms, neurological examination, and electrodiagnostic tests (electromyography and motor and somato-sensory evoked potentials).

6.2.6 Chronic Sensorimotor Polyneuropathy

It is the most frequent form (80% of cases) and is characterized by asymmetrical and distal local-

ization, predominant in the lower limbs. In the early stages, only the sensory component is involved; instead, the motor component is injured later. Sensory symptoms in patients with polyneuropathy include paraesthesia and hyperaesthesia, or numbness and hypoaesthesia. Moreover, it is possible to observe a "glove" or "stocking" hypoaesthesia. Unstable gait and absence of tendon reflexes (e.g. Achilles) could be present. Asthenia and hypotrophy of the intrinsic muscles of hand and foot appear in late stages.

6.2.7 Acute Painful Neuropathy

Symmetric polyneuropathy is characterized by acute onset and pain, with mild impairment of the sensory and motor components.

6.2.8 Mononeuropathies

They occur in 15% of diabetic neuropathic patients. The cranial nerves are usually involved, and motor deficit, areflexia, and pain are the most common signs.

6.2.9 Radiculopathies

They are classified into monoradiculopathies (acute, good prognosis) and polyradiculopathies (gradual onset, high risk of recurrence). The thorax and abdomen are mainly affected, and pain, dysaesthesia, and paraesthesia are common symptoms. The symptomatology of radiculopathies can simulate visceral pathologies.

6.2.10 Amyotrophy

It is a rare condition with an insidious onset. It is characterized by pain, asthenia, and muscular hypotrophy in the lower limbs. Moreover, a deficit of the patellar and Achilles reflexes is present.

6.2.10.1 Treatment

Nowadays, no valid therapies are reported in the literature. Metabolic control is fundamental to prevent the onset and evolution of neuropathy. In painful forms, symptomatic treatment is partially effective. In these cases, the treatment includes drugs (tricyclic antidepressants, gabapentin, tramadol) and physical therapy (TENS and physiotherapy).

6.3 Neurogenic Para-Articular Ossifications

Neurogenic para-articular ossifications occur primarily in patients with severe spinal cord interruption syndrome associated with paraplegia. The lesions are localized to the large joints (hip, knee, and shoulder), and the most frequent clinical manifestation is the joint locking. In symptomatic cases, surgical excision followed by early active mobilization is the treatment of choice; however, recurrences are frequent.

6.4 Charcot–Marie–Tooth Disease

6.4.1 Definition

Charcot–Marie–Tooth disease (described by Jean-Martin Charcot, Pierre Marie, and Howard Henry Tooth) is a hereditary neurologic syndrome involving the peripheral nervous system.

6.4.2 Epidemiology

The incidence rate varies on a geographic basis and ranges from 9.7/100,000 to 82.3/100,000 population.

6.4.3 Pathogenesis

There are several forms of the disease. Type 1A (CMT1A), caused by duplication on chromo-

some 17 of a gene that produces a protein (PMP22) in peripheral nerve myelin, is the most common (50% of cases). Demyelination and formation of "onion bulbs", consisting of concentric coils of Schwann cells around the axon, intermingled with collagen, are common lesions in all forms of the disease.

6.4.4 Diagnosis (Clinical and Imaging)

The main symptoms are weakness, progressive and symmetrical atrophy of the muscles (feet, legs, and hands), skeletal deformities (feet and ankle), and gait alteration. Clinical manifestations generally occur within the first or second decade of life. A characteristic sign of Charcot–Marie–Tooth disease is the cavus foot with an elevation of the medial longitudinal arch. Claw deformity of the toes and hindfoot varus is often present. The involvement of the upper limbs (progressive weakness, cramps, claw deformity of the hands) appears when the symptoms of the lower limbs have become apparent.

A family history of diabetes and diabetic neuropathy is frequent. Electromyography shows a uniform and diffuse slowing of nerve conduction. Radiographic examinations of the feet in orthostatism generally show cavus foot, forefoot adduction, hindfoot varus, and first metatarsal plantar flexion.

6.4.5 Treatment

Flexible foot deformities can be managed conservatively with the use of orthoses. Conservative treatment is not effective in the long term due to the progressivity of the pathology. The most common surgical treatment for the cavus foot is plantar fasciotomy and casting, but tendon transposition surgeries could be performed in severe cases. In rigid deformities, corrective osteotomies at the forefoot, midfoot, or calcaneus are necessary. In untreatable cases, triple arthrodesis may be required.

Take-Home Message

- Neurological pathologies could affect the musculoskeletal system leading to progressive joint destruction, and the common characteristic is a deficit in nociceptive sensitivity.
- The most common neurological pathologies related to musculoskeletal disorders are neurofibromatosis, neuropathic arthropathy, and Charcot–Marie–Tooth disease.
- The skeletal manifestations of NF1 include spinal abnormalities and dysplastic changes in the long bones, especially the tibia.
- Neuropathic arthropathy is most frequently associated with syphilis, diabetes, and syringomyelia.
- The Charcot–Marie–Tooth disease consists of progressive, symmetrical weakness and atrophy of the muscles of the feet, legs, and hands.

Summary

- Neurofibromatosis type 1 (or Von Recklinghausen syndrome) is an autosomal dominant genetic syndrome caused by the loss of the oncosuppressor gene NF1, responsible for cutaneous, visceral, and musculoskeletal manifestations. The skeletal manifestations of neurofibromatosis include spinal abnormalities (scoliosis, kyphosis, erosion of the vertebral margins, and thinning of the ribs) and dysplastic changes in the long bones, especially the tibia (with or without pseudoarthrosis).
- Neuropathic arthropathy is a chronic degenerative disorder characterized by progressive destruction of one or more joints, with loss of joint congruence and alteration of the entire bony architecture. It is most frequently associated with syphilis (syphilitic tabes dorsalis), diabetes (diabetic neuropathy), and syringomyelia. The common feature is a deficit in

nociceptive sensitivity. This deficit exposes the joint to repeated microtrauma, resulting in microfractures, subluxations, and dislocations.

- Diabetic neuropathy is a set of signs and symptoms due to peripheral nerve dysfunction. These conditions are divided into symmetric (chronic sensorimotor polyneuropathy, acute painful neuropathy, and autonomic neuropathy) and asymmetric (mononeuropathies, radiculopathies, and amyotrophy). All nerve components (Schwann cell and myelin, axon, and endoneurial capillaries) are involved.
- The Charcot–Marie–Tooth disease consists of demyelination of peripheral nerves. Symptoms generally occur within the first or second decade of life. Progressive, symmetrical weakness and atrophy of the muscles of the feet, legs, and hands are the most common signs. These conditions could be associated with skeletal deformities of the feet and ankle and gait alterations.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Tabetic arthropathy:
   (a) Is the second most frequent form of neuropathic arthropathy
   (b) Affects joints in patients with sensory deficit
   (c) Is more common than diabetic neuropathy
   (d) Rarely results in joint instability
   (e) Causes progressive joint destruction
2. Syringomyelic arthropathy:
   (a) Occurs in more than 60% of patients with syringomyelia
   (b) Has a lower frequency than tabetic arthropathy
   (c) Is one of the initial symptoms of syringomyelia
   (d) Has a radiographic pattern similar to tabetic arthropathy
   (e) Is typically painful
3. Diabetic neuropathy:
   (a) Is a rare complication of diabetes
   (b) Has a higher frequency in long-standing diabetics
   (c) Affects only the axon
   (d) Is associated with a condition of endoneurial ischaemia
   (e) Has a multifactorial pathogenesis
4. Chronic sensorimotor polyneuropathy:
   (a) Is the second most frequent form of diabetic neuropathy
   (b) Is characterized by a symmetrical and distal localization
   (c) Affects mainly the upper limbs
   (d) May result in glove or stocking hypoaesthesia
   (e) May result in the absence of tendon reflexes
5. Mononeuropathies:
   (a) These are the leading form of diabetic neuropathy in frequency
   (b) These most frequently involve the spinal nerves
   (c) These cause pain, motor deficit, and are flexia
   (d) The oculomotor is the most affected cranial nerve
   (e) The trigeminal nerve is the most affected cranial nerve

Further Reading

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The Musculoskeletal Involvement in Rheumatic Disorders

7

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Overview

Rheumatic diseases represent a heterogeneous group of pathologies strictly associated with disability, low quality of life and increased mortality. They include a wide spectrum of disorders that can involve many tissues, with a predominant involvement of the musculoskeletal system. In clinical practice, many patients suffer from joint pain and other symptoms that are generally attributed to arthritis. Rheumatic diseases are divided into different ways, but a simple form of classification is based on their pathogenesis: inflammatory and degenerative. Inflammatory rheumatisms can have an autoimmune, infective or metabolic trigger mechanism. In the autoimmune group, rheumatoid arthritis (RA) and seronegative spondyloarthropathies (SpA) are both common causes of functional impairment in developed countries.

7.1 Rheumatoid Arthritis

7.1.1 Definition

RA is a chronic, symmetric and erosive polyarthritis. Its remittent joint involvement worsens over time causing irreversible deformity and ankylosis that results in disability.

7.1.2 Epidemiology

RA has a prevalence of 0.5–1% in adults. The disease is more frequent in women (F:M = 3–4:1) and elderly people.

7.1.3 Aetiology/Pathogenesis

RA is considered an autoimmune disease. Probably, the coexistence of a genetic predisposition (HLA-DR4 allele) and environmental factors may trigger an aberrant T lymphocyte activation in

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the synovial tissue. The rheumatoid factor (RF) and anti-citrullinated protein antibodies (ACPAs) are present in 70% of patients with RA. The resulting inflammation leads to proliferation and neoangiogenesis in the synovial membrane, which thickens and forms the synovial pannus. The increasing pannus, together with inflammatory cells and enzymes, promotes subchondral bone erosion and cartilage degradation.

7.1.4 Diagnosis

7.1.4.1 Clinical Diagnosis

RA typically affects diarthrosis (joints with synovial membrane), involving firstly distal joints and secondly the proximal ones. Joints bilaterally and symmetrically become swollen, tender and warm and morning stiffness limits their movement. Moreover, patients often report constitutional symptoms (myalgia, fever, weight loss, fatigue and depression).

7.1.4.2 Musculoskeletal Involvement in RA

Hand

In the early stages, RA affects the metacarpophalangeal (MCP) and the proximal interphalangeal (PIP) joints in a bilaterally symmetrical distribution. As a rule, the distal interphalangeal (DIP) joints are spared. Over time, RA determines progressive and irreversible joint deformity, range of motion becomes limited and interosseous muscles atrophy. Typical deformities of the hand are the following:

- The "ulnar drift": Fingers show a deviation toward the ulnar side due to a volar subluxation of the PIP. It determines a lateral dislocation of the extensor digitorum tendons.
- The "swan neck" deformity: It is the hyperextension of the PIP joints with fixed flexion of the DIP joints.
- The "boutonnière" deformity: It is a persistent flexion of the PIP joint with hyperextension of the DIP joint.
- The "hitchhiker's thumb": It is a flexion of the MCP joint with hyperextension of the IP joint.
- The volar subluxation of the base of the metacarpal bones.
- The "camel's back" deformity: It is the volar subluxation of the base of the metacarpal bones and the volar subluxation of the PIP on the metacarpal heads.

Wrist

- The "piano key" sign: When the subluxated ulnar styloid moves up and down on applying pressure.
- The "sign of benediction": The fourth and fifth fingers show a clawed-like appearance with hyperextension of the MCP joints and flexion of the PIP and DIP joints. It is due to the dorsal subluxation of the distal ulna that may produce the rupture of the common extensor tendon and of the extensor digiti minimi tendon.

Elbow

The elbow joint may become swollen and painful. Over time, the joint may develop ankylosis in flexion.

Shoulder

Passive and active mobilization may be painful in patients with RA. In advanced stages, lesions to the rotator cuff and to the tendon of the caput longum are possible.

Hip

Patients rarely report pain in the groin, but it can also be in the buttock or be a knee pain. Hip joint involvement may be difficult to distinguish from coxarthrosis and avascular necrosis of the hip.

Knee

The knee is the large joint most frequently affected. It presents joint effusion and swelling. Sometimes, a popliteal cyst (also known as Baker's cyst) may be observed due to synovial fluid collection behind the knee.

Ankle and Hindfoot

The ankle may present perimalleolar swelling that limits movements of flexion/extension. Frequently, a retrocalcaneal bursitis and rheumatoid nodules in the Achilles tendon can be observed.

Forefoot

In RA patients, it is frequent to observe a plantar subluxation of metatarsal heads. Other typical forefoot deformities are the following:

- The "triangular forefoot" deformity: It consists of the flattening of the plantar surface and plantar subluxation of the metatarsal heads with bunion.
- The "claw toe" deformity is hyperextension of the metatarsophalangeal (MTP) joint with flexion of the IP.

Cervical Spine

The cervical spine involvement is not frequent, but is possible and affects the atlas-axis joint. It is a diarthrosis, and its synovial inflammation may result in focal erosion of the odontoid process of the axis, leading to atlantoaxial subluxation and, consequently, a potentially fatal cervical spinal cord compression. The resulting cervical myelopathy may determine sensory and motor deficits, requiring an immediate surgical treatment.

7.1.4.3 Diagnostic Imaging

In the early stages of the disease, imaging may be negative for joint alterations. As RA progresses, X-rays of the affected joints may gradually show a reduction of the normal joint space, bone erosions and juxta-articular osteoporosis. For an early diagnosis, magnetic resonance imaging (MRI) and ultrasound (US) have a higher sensitivity than X-rays in the detection of initial lesions. In particular, MRI can identify the bone marrow oedema (a predictor of rapid radiological progression of RA), whereas US allows to study synovial hypertrophy and joint inflammation.

7.1.5 Treatment

Surgery becomes mandatory in case of advanced stages of RA and medical treatment failure. It may be important to prevent pain and to ameliorate patients' quality of life. Surgical procedures on the affected structures include synovectomy, capsulotomy, tenorrhaphy and osteotomy. Severely impaired joint functions may require joint replacement surgery.

7.1.5.1 Hand Deformities

Hand deformities may require different surgical approaches

- Procedures on soft tissues (synovectomy, tenosynovectomy, capsulotomy, tenotomy, tenodesis, tendon transfer and ligament reconstruction)
- Joint resection arthroplasty
- Joint replacement surgery
- Arthrodesis

7.1.5.2 Forefoot Deformities

Surgery is required when conservative treatment with orthosis fails. Surgical forefoot procedures include:

- Joint resection arthroplasty (resection of the metatarsal heads and the base of proximal phalanges)
- Osteotomy (in order to realign toes)
- Procedures on soft tissues
- MTP total joint replacement
- Arthrodesis

7.1.5.3 Hindfoot Deformities

In case of conservative treatment failure, surgery is necessary to ease pain coming from the subastragalic or the astragalus-scaphoid joint. In the absence of valgus hindfoot, the best choice is the astragalus-scaphoid arthrodesis. The double arthrodesis (arthrodesis of subastragalic and astragalus-scaphoid joint) is preferred in the presence of painful valgus hindfoot. When the ankle is severely affected, an ankle arthrodesis or a total ankle replacement may be performed.

7.1.5.4 Large Joints (Shoulder, Knee, Hip, Elbow)

Synovectomy is a helpful procedure in patients with RA. It consists of the surgical removal of the hypertrophic synovial membrane in order to prevent cartilage degradation. Over time, the synovial pannus recurs and the procedure may be repeated. It is performed in the knee when pain and swelling do not respond to medications for at least 6 months.

Patients with RA may require joint replacement in advanced stages. It may be necessary to alleviate symptoms and to restore normal range

of motion. In comparison to arthrosis, joint replacement in RA presents some disadvantages: low-quality bone (due to inflammation and long-term steroid administration) and younger patients. It implies that these patients are very likely to undergo revision surgery during their lifetime.

Arthrodesis is the artificial induction of joint ossification between two bones by surgery. It is performed to relieve intractable pain. However, it induces important functional limitation. It is a procedure better tolerated in small joints, whereas it remains the last option in large joint treatment.

7.2 Ankylosing Spondylitis

7.2.1 Definition

AS is the prototypical disease of the seronegative SpA. It is characterized by inflammatory back pain with progressive spinal stiffness.

7.2.2 Epidemiology

AS is more frequent in men (F:M = 1:2), and it has the greatest association with the HLA-B27 allele (90–95% of patients with AS).

7.2.3 Aetiology/Pathogenesis

Similarly to other SpA, it remains incompletely understood. The tumour necrosis factor (TNF) is the most important inflammatory mediator in AS; in fact, it is a specific target of modern therapies.

7.2.4 Classification

AS is an axial SpA. It means that arthritis primarily affects the sacroiliac joints and the spine.

7.2.5 Diagnosis

7.2.5.1 Clinical Diagnosis

AS is generally diagnosed many years after the onset of symptoms. In fact, it is characterized by periods of flare and dormancy that make it difficult to distinguish inflammatory back pain from mechanical back pain. Moreover, patients with AS frequently experience extra-articular manifestations, such as acute anterior uveitis and aortic valve insufficiency. Sacroiliac joint should be observed, as in the early stages of the disease, the degenerative change of SpA appears in this joint. Furthermore, lumbar and cervical spines are involved.

7.2.5.2 Musculoskeletal Involvement in AS

Typical sites of injury in AS are

- Sacroiliac joints
- Interapophyseal joints
- Costovertebral joints
- Sternoclavicular joint
- Sternocostal joints
- Intervertebral discs and ligaments

Spine

Patients with AS report chronic pain and stiffness in the back due to sacroiliitis that awakens them in the early morning hours. Gradually, inflammation moves from the sacroiliac joints to the rachis in an ascending manner. AS may affect any part of the spine, often with pain referred to one or the other buttock or the back of the thigh. Due to long-term inflammation, the spinal entheses and the sacroiliac joints ossify leading to ankylosis and irreversible rigidity. Ossification of the outer fibres of the fibrous ring of the intervertebral discs results in the formation of bony growths called syndesmophytes. When this process affects the whole spine, it determines the classic “bamboo spine” X-ray pattern. Due to vertebral fusion, in advanced stages, patients may develop a severe cervical kyphosis and loss of lumbar lordosis that gives to the spine a forward curve. It leads to a compensatory flexion contracture of the hips and knees in order to maintain forward vision.

Chest

Sternoclavicular, sternocostal and costovertebral joint involvement may determine chest pain. In advanced stages, the ankylosis of these joints and spine deformity may cause restrictive lung disease.

Extra-Axial Joint Involvement

Other common manifestations include peripheral joint involvement (up to 50% of patients; mainly shoulders, hips and knees), enthesitis (up to 70% of patients) and dactylitis (up to 8% of patients).

7.2.5.3 Diagnostic Imaging

Sacroiliac Joints

Erosion is seen soonest on the iliac side, and then the joint may appear abnormally wide (pseudowidening) because of progression of the erosions. It is followed by bony sclerosis on both sides and, eventually, fusion or ankylosis of the sacroiliac joints.

Spine

Initial lesions are seen where the annulus fibrosus (outer layer of the vertebral disc) inserts into the vertebral body and "shiny corners" develop. Subsequently, the squaring of the vertebrae appears. Over time, these outer layers become ossified and syndesmophytes form. These bony growths determine vertebral fusion and the "bamboo spine" occurs.

However, the use of modern advanced imaging technology as MRI with STIR (short tau inversion recovery) sequences shows inflammatory changes years before X-rays. It allows an early diagnosis and to start treatment quickly, in order to prevent deformity.

7.2.6 Treatment

When daily activities become severely impaired, surgery may be an option, including a joint replacement. Laminectomy is the most commonly performed procedure. It consists of the removal of the vertebral lamina in order to relieve pressure on nerve roots. Rarely, osteotomy may be necessary to treat severe cervical spine deformity.

7.3 Psoriatic Arthritis

7.3.1 Definition

PsA is a chronic inflammatory arthritis occurring in patients with psoriasis. In 10–15% of patients, arthritis can precede skin disease ("PsA sine psoriasis").

7.3.2 Epidemiology

In contrast to AS, there is no gender predilection. HLA-B27 is seen in 10–25% of patients with PsA. Patients with PsA or psoriasis in 40% of cases have a first-degree relative with the disease.

7.3.3 Aetiology/Pathogenesis

The pathogenesis of psoriasis and PsA remains unknown. The axial form of PsA is frequently associated with the HLA-B27 allele, whereas the peripheral form is associated with the HLA-DR4.

7.3.4 Classification

In most cases, PsA affects peripheral joints. Moll and co-workers described five clinical patterns in PsA (Table 7.1).

Table 7.1 Classification of PsA according to Moll and co-workers

Patterns of PsA	Affected joints	Frequency
Asymmetric oligoarthritis	≤4 peripheral joints (often dactylitis)	70% of cases
Symmetric polyarthritis	≥5 peripheral joints (resembling RA)	5–20% of cases (poor prognosis)
Distal interphalangeal predominant	DIP (often nails)	5–10% of cases
Arthritis mutilans	PIP and DIP (advanced destructive changes, digit "telescopic")	5% of cases
Axial predominant	Joints of the spine and sacroiliac joints	4% of cases in isolation; 50% of cases associated with peripheral disease

7.3.5 Diagnosis

7.3.5.1 Clinical Diagnosis

Skin changes consistent with psoriasis generally occur before the onset of PsA, but the arthritis can precede the rush in 15% of cases. Unlike RA, DIP joint involvement is a distinctive feature of PsA. Dactylitis and enthesitis occur in 30–40% of patients and are considered classic manifestations of PsA.

7.3.5.2 Musculoskeletal Involvement in PsA

In 70% of cases, patients present an asymmetric oligoarticular disease that results in joint pain and morning stiffness. This form is generally mild and is often associated with dactylitis. The axial form of PsA may be associated with peripheral arthritis. Unlike AS, the sacroiliac joints frequently show a monolateral involvement, and syndesmophytes do not affect the whole spine, justifying a better prognosis.

Dactylitis is considered a hallmark clinical feature of PsA and most commonly involves the third and fourth toes. It is a uniform swelling of an entire digit, due to inflammation of digital joints (MCP, PIP, DIP) and soft tissues. Similarly, enthesitis is more common in PsA than other forms of SpA. It is the inflammation of the entheses (the insertion sites of tendons and ligaments to the bone surface) and frequently affects the Achilles tendon insertion or the plantar fascia.

7.3.5.3 Diagnostic Imaging

A typical feature of PsA is the involvement of fingers and toes. Peripheral radiologic findings are similar to RA, with reduction of the joint space and bone erosions. Osteolysis is generally milder than RA, except for the arthritis mutilans. In this form, resorption of bone, with destruction of the joint, is observed as the "pencil-in-cup" radiographic finding and leads to redundant, overlying skin with a telescoping motion of the digit. The axial involvement resembles AS but is less severe. Dactylitis and enthesitis are investigated with MRI and US.

7.3.6 Treatment

Psoriatic arthritis acts from a pathogenetic point of view as an arthrosis, with structural alterations of the diarthrosis. Therefore, the treatment is similar to that of arthrosis and can result in prosthetic replacement, which is recommended if PsA has determined severe damage to the joints of the hands and feet that limits everyday activities.

7.4 Reactive Arthritis

7.4.1 Definition

ReA, also known as Reiter's syndrome, is an aseptic inflammatory arthritis that occurs within 4 weeks following an extra-articular bacterial infection. The classic clinical triad of conjunctivitis, nongonococcal urethritis and arthritis following gastrointestinal or urogenital infection is seen only in one-third of patients.

7.4.2 Epidemiology

ReA is associated with the HLA-B27 in 60% of cases. The most common triggers are intestinal (with Salmonella, Shigella, Campylobacter) and genitourinary infections (Chlamydia, Mycoplasma).

7.4.3 Aetiology/Pathogenesis

All of SpA are thought to be triggered by a preceding infection, but it has been demonstrated only in ReA. Synovial fluid cultures are negative, suggesting that ReA is caused by an autoimmune response. The exact mechanism of interaction between the microorganism and the immune system remains unclear.

7.4.4 Diagnosis

7.4.4.1 Clinical Diagnosis

ReA should be suspected in young adults presenting with arthritis within 4 weeks following a urogenital or enteric infection.

7.4.4.2 Musculoskeletal Involvement in ReA

ReA generally presents an asymmetric oligoarticular arthritis predominantly involving the large joints of lower limbs. Dactylitis, enthesitis, bursitis and tenosynovitis are possible. Axial involvement is less common; it tends to be asymmetric and associated with HLA-B27. In 50% of cases, ReA is self-limiting, but recurrent or chronic arthritis may occur.

7.4.4.3 Diagnostic Imaging

In the early stages, X-rays may reveal the presence of juxta-articular osteoporosis. Chronic arthritis may result in bone erosions and deformity. Sacroiliac joints and spine may show a monolateral involvement.

7.4.5 Treatment

Infection must be treated if present. Similarly to other SpA, surgical treatment is considered in advanced stages.

7.5 Gout

7.5.1 Definition

Gout is a dysmetabolic disorder due to persistently elevated levels of uric acid in the blood. It determines recurrent attacks of a painful, red, hot and swollen joint.

7.5.2 Epidemiology

Gout is more frequent in men. In women, it generally occurs in the post-menopausal period.

7.5.3 Aetiology/Pathogenesis

Gout attack is an acute arthritis caused by MSU crystals. Microcrystalline aggregation takes place when uric acid hematic levels overcome 7 mg/dL. In this condition, uric acid crystallizes in the form of MSU and precipitates in joints. Cellular inflammatory response against these deposits (called tophi) occurs, and the resulting inflammation causes arthritis.

7.5.4 Diagnosis

7.5.4.1 Clinical Diagnosis

When the disease starts, recurrent acute arthritis episodes are frequently interspersed with asymptomatic phases. The gout attack presents a sudden and rapid onset (peak reached in 24–48 h), usually at night-time. Joint pain is described as burning, violent and intolerable. The surrounding skin appears swollen, red and hot, and desquamation may be observed. Gout attack is self-limiting in 5–10 days. The diagnosis is clinical and may require the analysis of the synovial fluid in order to identify MSU microcrystals.

Articular chondrocalcinosis (also known as "pseudogout") is a microcrystalline arthropathy very similar to gout. It is determined by aggregation of calcium PPD crystals and causes recurrent attacks of acute arthritis. The most frequently affected joint is the knee, followed by the shoulder and the ankle. Pseudogout should be suspected in patients with acute synovitis in a large joint and normal levels of uric acid.

7.5.4.2 Musculoskeletal Involvement in Gout

Typically, the acute arthritis episode affects the first MTP joint. The inflammatory process involving the hallux is also called "podagra". Less frequently, the knee, the wrist and the fingers may be involved. Over time, gout becomes a chronic condition. Tophi are specific manifestations of this phase and occur after 10 years from the first acute attack. They typically form in the helix, retro-olecranon bursa, hands and feet. MSU crystal deposits in the joints are responsible for carti-

lage ulcers, marginal osteophytes and geodes. Moreover, the synovial tissue develops a chronic inflammation in response to MSU deposits. Symptoms are similar to arthrosis, with painful rigidity alleviated by rest.

7.5.4.3 Diagnostic Imaging

X-rays do not show any joint alteration in acute attacks. In chronic gouty arthropathy, tophi are typically encountered as eccentric, juxta-articular soft-tissue nodules. Bony erosions with sclerotic margins and overhanging edges are commonly seen in proximity. Pseudogout typically shows linear or granular calcifications in the hyaline cartilage and fibrocartilage, mainly in the knee.

7.5.5 Treatment

In gout, medications to lower uric acid are usually effective. Colchicine, nonsteroidal anti-inflammatory drugs and corticosteroids could be used in adult patients with acute episodes. Chronic kidney disease, two or more acute episodes per year, urolithiasis, tophus, chronic gouty arthritis and joint degenerations are proper indications for long-term urate-lowering therapy. Allopurinol and febuxostat are used to prevent flare-ups, although febuxostat is associated with an increase in all-cause and cardiovascular mortality and is therefore not routinely recommended. Indications for surgery are sepsis control in case of ulcerated tophi, pain control and functional impairment. The most common procedures are arthroscopic shaver and open tophectomy. For infiltrative tophi affecting joints or tendons, sharp debridement and curettage are preferred. Complete joint resection may be performed when tophi have destroyed the cartilaginous surfaces of the joint causing pain and impaired function.

7.6 Hydroxyapatite Crystal Deposition Disease

7.6.1 Definition

HADD is a disorder characterized by periarticular and intra-articular deposition of HA crystals.

7.6.2 Epidemiology

The disease is more frequent in women and in middle-aged individuals.

7.6.3 Aetiology/Pathogenesis

It is a disease of uncertain aetiology. It is believed that HA crystals begin to accumulate in injured tendons (secondary to trauma) via fibrocartilaginous metaplasia. Crystals may deposit in any tendon of the body, but most commonly in the tendons of the rotator cuff.

7.6.4 Diagnosis

7.6.4.1 Clinical Diagnosis

Patients typically report shoulder pain, weakness and decreased range of motion. The acute form can be preceded by traumatic events and is usually self-limiting in a few weeks. The chronic form may cause adhesive capsulitis (also known as “frozen shoulder”) with severe shoulder stiffness.

7.6.4.2 Musculoskeletal Involvement in HADD

Calcifications are generally located in the supraspinatus tendon (80% of cases). In some cases, arthrosis can be co-present with HADD leading to a worse prognosis. An example is the “Milwaukee shoulder” (apatite-associated destructive arthritis) that is the coexistence of gleno-humeral arthrosis, deposits of HA in the synovial membrane and rotator cuff tears. In advanced stages, marked erosions of the humeral head or degeneration of cartilage, capsule and bursae may occur.

7.6.4.3 Diagnostic Imaging

The radiographic appearance of HADD can be divided into three stages of calcification: the formative, resting and resorptive phases. The formative and resting phases show round-to-ovoid soft-tissue calcification with well-defined margins. The resorptive phase shows ill-defined calcification with a comet tail-like shape. It may mimic a periosteal reaction.

7.6.5 Treatment

Surgical removal of calcifications may be considered for cases refractory to conservative treatment. An arthroscopic approach is generally preferred. It allows calcification removal with or without acromioplasty of the shoulder. Frequently, patients also require a rotator cuff repair because of tendon injures caused by the surgical removal of calcification or the disease itself.

Take-Home Message

- Rheumatoid arthritis is a chronic, symmetric and erosive polyarthritis with an additive and progressive character that may evolve into articular deformities and ankylosis.
- The wrist and the joints of the hand are the most frequently involved, followed by metatarsophalangeal and proximal interphalangeal of the foot.
- The radiographic signs of rheumatoid arthritis consist of erosions (one of the most representative characteristics), progressive narrowing of the joint line due to the destruction of cartilage, juxta-articular osteopenia and, in advanced stages, complete loss of the normal joint relationships.

Summary

- Rheumatoid arthritis is a chronic, symmetric and erosive polyarthritis with an additive and progressive character that may evolve into articular deformities and ankylosis.
- The most involved joints in case of RA are diarthrosis.
- The wrist and the joints of the hand are the most frequently involved (proximal interphalangeal and metacarpophalangeal joints), followed by metatarsophalangeal and proximal interphalangeal of the foot.
- The radiographic signs of rheumatoid arthritis consist of erosions (one of the most representative characteristics), progressive narrowing of the joint line due to the destruction of cartilage, juxta-articular osteopenia and, in advanced stages, complete loss of the normal joint relationships.
- The seronegative spondyloarthropathies represent a group of pathologies characterized by axial and peripheral joint involvement. Seronegativity (absence of rheumatoid factor) is a typical feature. They include ankylosing spondylitis, psoriatic arthritis, reactive arthritis and arthritis associated with chronic enteropathies.
- Ankylosing spondylitis mainly involves the axial skeleton (sacroiliac and spine joints), leading to a progressive fibrosis and ossification (ankylosis) of the affected structures.
- Psoriatic arthritis is an inflammatory disease which occurs in people suffering from psoriasis or predisposed to psoriasis.
- Following Moll and co-workers' classification, joint involvement in psoriatic arthritis can be distinguished into five different subtypes.
- Reactive arthritis is an autoimmune aseptic arthritis triggered by an extra-articular infection.
- Microcrystalline arthropathies are caused by the accumulation of microcrystals in joints. Deposits of monosodium urate (gout), calcium pyrophosphate dihydrate (chondrocalcinosis) and hydroxyapatite are the causative agents of these diseases.
- Gout is due to persistently elevated uric acid levels in the blood that form crystals of monosodium urate in the joints (tophi). It determines recurrent attacks of acute arthritis.
- Chondrocalcinosis or pseudogout is similar to gout but is caused by the deposition of calcium pyrophosphate dihydrate (PPD) crystals.
- Hydroxyapatite deposition disease is due to deposition of hydroxyapatite crystals in the joint and periarticular structures. There are three stages of the disease: the formative, resting and resorptive ones. It can result in a destructive arthropathy of the shoulder.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. In rheumatoid arthritis:
   (a) The articular involvement is generally asymmetrical
   (b) Most patients have an involvement of hand joints
   (c) Knee involvement is rare and asymptomatic
   (d) The "ulnar drift" hand deformity is due to DIP joint involvement
   (e) Cervical spine lesions are belatedly radiographically visible
2. Rheumatic foot deformities:
   (a) Are usually well tolerated by patients and scarcely disabling
   (b) Are treated with plaster treatments
   (c) May be treated with prosthetic implants
   (d) May be initially treated conservatively
   (e) May be treated with joint resection arthroplasty
3. Ankylosing spondylitis:
   (a) Symptoms generally occur after the age of 45
   (b) Hands are frequently involved
   (c) Chronic back pain is often the first manifestation
   (d) Syndesmophytes frequently affect the hip joint
   (e) MRI allows an early diagnosis of sacroiliitis
4. Psoriatic arthritis:
   (a) It generally precedes skin lesion occurrence
   (b) It is usually an asymmetric oligoarticular disease
   (c) Dactylitis is extremely rare
   (d) DIP joint involvement cannot affect nails
   (e) Arthritis mutilans is rare
5. Which one of the following is a reactive arthritis?
   (a) Arthrosis
   (b) Rheumatoid arthritis
   (c) Psoriatic arthritis
   (d) Ankylosing spondylitis
   (e) Reiter's syndrome
6. Gout:
   (a) It is caused by deposition of calcium pyrophosphate dehydrate crystals
   (b) It typically affects the knee
   (c) It is more frequent in men
   (d) The first episode usually involves the first MTP joint
   (e) The acute form is characterized by the presence of tophi

Further Reading

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Part III

Infections

8 Osteomyelitis and Infectious Diseases

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Overview
This chapter provides introductory information about three main bone and joint infectious disorders: osteomyelitis, septic arthritis, and periprosthetic joint infections. Divided into three sections, this chapter defines each of these diagnoses and systematically breaks them down into their epidemiology, etiology/pathogenesis, classification, diagnosis, and treatment.

8.1 Osteomyelitis

8.1.1 Highlights (Table 8.1)

Definition
– Infection of the **bone** or **bone marrow**

Epidemiology
– **Most common pathogen**: *S. aureus*
– **Risk factors**: immunosuppression, micro- or macroangiopathy, systemic infections

Etiology/pathogenesis
– **Biofilm** formation through **hematogenous**, **direct**, or **contiguous** spread

Classification
– **Timing** (acute, subacute, chronic)
– Stage and host status (**Cierny-Mader** classification)

Diagnosis
– **Clinical presentation**: inflammation, fever, malaise, chills, weight loss
– **Testing**: lab studies, blood culture and biopsy, histologic evaluation, imaging

Treatment
– **Surgical** (debridement and irrigation, resection, plastic surgery, amputation)
– **Medical** (antibiotic therapy and hyperbaric oxygen)

8.1.2 Definition

Osteomyelitis is defined as the inflammation (***-itis***) of the bone (***-osteo***, from the Greek word *osteon*) or bone marrow (***-myel***, from the Greek *myelos*).

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Table 8.1 Summary table

	Osteomyelitis	Septic arthritis	PJI
Definition	Infection of the **bone** or **bone marrow**	Infection of the **joint**	Infection of the **prosthesis**
Epidemiology	**Pathogens**: *S. aureus* and other bacterial species **Risk factors**: Immunosuppression, angiopathy, systemic infections	**Pathogens**: Bacteria, viruses, fungi **Risk factors**: Age >80, comorbidities, history of crystal arthropathy, endocarditis or recent bacteremia, IV drug user, recent joint surgery	**Pathogens**: *S. aureus*, *S. epidermidis*, *coagulase-negative Staphylococcus* spp., *Streptococcus* **Risk factors**: Male sex, prior surgeries, and comorbidities
Etiology	**Mechanism**: Biofilm formation **Pathways**: Hematogenous, direct, contiguous	**Mechanism**: Proteolytic enzyme release **Pathways**: Hematogenous, direct, contiguous	**Mechanism**: Biofilm formation **Pathways**: Hematogenous, direct, contiguous, recurrent infections
Classification	**Timing**, **stage**, **host status**	By **organism**	**Acute** vs. **chronic**
DX	Positive blood cultures, elevated serum levels, and abnormal imaging findings in periosteum, cortical bone, or medullary cavity **Symptoms**: Inflammation, pain, limited ROM, inability to bear weight, fever, malaise, chills, and weight loss **Testing**: Lab tests, culture and biopsy, histologic evaluation, imaging	Cloudy and purulent synovial fluid with WBC >50,000, elevated serum levels, and abnormal imaging findings in joint space **Symptoms**: Swollen joint, fever, erythema, effusion, localized pain, inability to bear weight, or joint passive ROM **Testing**: Joint aspiration, cultures, lab tests, imaging	At least two positive cultures with the same microorganisms. Blood markers elevated (CRP, ESR) **Symptoms**: Pain, edema, wound drainage, surgical site erythema, effusion **Testing**: Lab tests, joint aspiration, histopathology, frozen sections, imaging
TX	Antibiotics, debridement and irrigation, resection, amputation	Debridement, irrigation, antibiotics	Antibiotics, debridement and irrigation, one-stage and two-stage revision
Differential DX	Cellulitis, abscess, myositis, gout, toxic synovitis, septic arthritis, bursitis, Charcot arthropathy, fracture, malignancy, and osteonecrosis	Transient synovitis, crystallopathy, gout, pseudogout, acute inflammatory arthropathy	Aseptic failure

8.1.3 Epidemiology

Osteomyelitis is a serious condition caused by microorganisms that reach the bone and cause infection. The most common pathogen is the skin commensal ***Staphylococcus aureus*** and other prevalent species including coagulase-negative *Staphylococci*, aerobic *Gram-negative bacilli*, *Enterococcus*, and *Streptococcal* species.

Osteomyelitis has a heterogeneous presentation across the skeleton in patients of all ages. Patients with immunosuppression, micro- and macroangiopathy (e.g., diabetes, cardiovascular disorders, and smoking), or systemic infections are considered at higher risk of infection.

8.1.4 Etiology/Pathogenesis

Osteomyelitis occurs when planktonic bacteria or "free-bacteria" attach to an inert substrate and enter a no-growth or "sessile" phase to create a matrix for **biofilm**. These bacterial complexes are harder to penetrate for antibiotics, thus reducing their efficacy.

Its pathogenesis relies on hematogenous spread (i.e., bacteremia) from another site of the body to the bone. However, penetrating injuries, open fractures, and surgical contamination can be causes of osteomyelitis and bone abscess (Fig. 8.1).

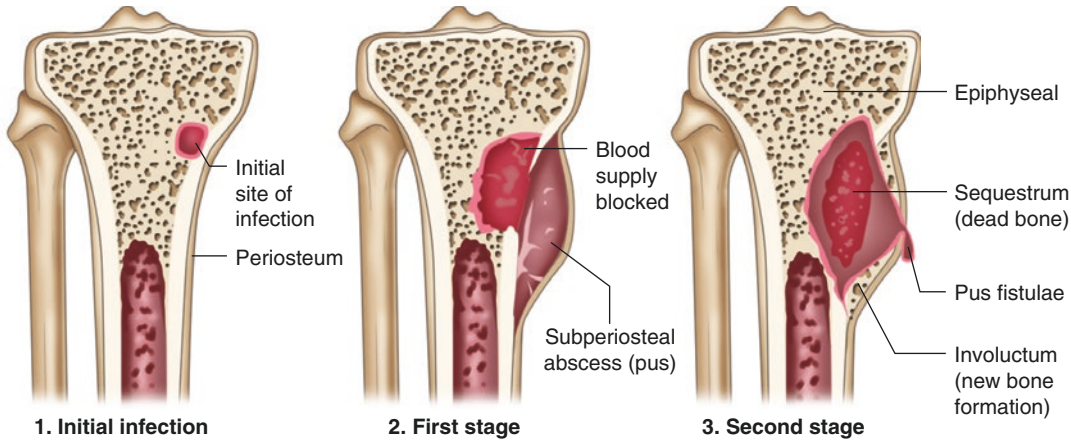

Fig. 8.1 Development of osteomyelitis

8.1.5 Classification

Osteomyelitis can be classified by timing, staging, mechanism, and host status.

Based on **timing classification**, the disorder can be divided into *acute* (when the infection is found within 2 weeks of onset), *subacute* (when found in between 1 and several months), and *chronic* (when the infection is prolonged for months/years). In the acute and subacute phases, the body reacts with a cascade of pro-inflammatory cytokines and leukocyte recruitment. In the chronic stage, there may be more severe consequences such as sequestrum and involucrum formation.

Cierny-Mader classification defines the *stage* of the disease (*stage I:* medullary; *stage II*: superficial; *stage III*: localized; *stage IV:* diffuse) and guides treatment based on location, bone involvement, and *host status* (*A host*: normal host; *B host*: systemic, local, or systemic/local compromise; *C host*: tx worse than disease).

8.1.6 Diagnosis

Osteomyelitis clinically presents with inflammation, fever, malaise, chills, and weight loss, nonspecific **symptoms** that can however show systemic involvement. Other common **signs** to look for in a physical examination are inflammation (e.g., erythema, hyperthermia, swelling), drainage with the presence of sinus tract, local soft-tissue breakdown, and ulceration. These are also nonspecific and may not be present in chronic cases.

There are several testing and imaging techniques that can be used to reach a diagnosis. Finding a positive **culture** via sterile biopsy technique is considered the gold standard for diagnosis. Elevated **serum counts** (ESR, CRP, and WBC) are nonspecific findings but can confirm systemic involvement. **Histologic evaluation** can be helpful in finding acute and/or chronic inflammation, bone necrosis, and new bone formation. **Radiographs** can detect bone abnormalities such as bone lucency, sclerotic rim, enlarged cortical bone and periosteum, elevation of periosteum, osteopenia, and loss of normal architecture (**CT scans** can assist in the diagnosis and surgical planning). These are usually helpful to detect necrotic bones. **MRIs** are great to identify early osteomyelitis and localizing infection. Specifically, this imaging technique can detect signs of penumbra. Finally, **nuclear medicine** is sometimes utilized when radiographs are normal and MRI is not an option.

8.1.7 Treatment

The treatment goal is to eradicate local infection, limiting local and systemic morbidity and recurrence of disease. Medical therapy alone does not cure osteomyelitis, and thus surgical intervention is often required.

Debridement and irrigation are usually suggested when an acute osteomyelitis is diagnosed. **Resection** may be needed in certain chronic cases where large bone segments are affected and there is the need of advanced reconstructive techniques (e.g., Ilizarov and Masquelet). **Plastic surgery** is sometimes needed. **Amputation** is usually suggested when the affected bone segment has a poor reconstructive potential.

Medical therapy includes **antibiotic** treatment for 4–6 weeks.

8.2 Septic Arthritis

8.2.1 Highlights (Table 8.1)

Definition
- Infection of the **joint**

Epidemiology
- **Pathogens**: bacteria, viruses, fungi
- **Risk factors**: age >80 years, comorbidities, history of crystal arthropathy, endocarditis or recent bacteremia, IV drug user, and recent joint surgery

Etiology/pathogenesis
- Release of **proteolytic enzymes** causing irreversible **cartilage destruction** within 8 h through **hematogenous**, **direct**, or **contiguous** spread

Classification
- By **pathogen** (bacterial, viral, fungal)

Diagnosis
- **Clinical presentation**: red hot swollen joint, fever, erythema about the joint, joint effusion, localized joint pain, inability to bear weight, or joint passive ROM
- **Testing**: joint aspiration, blood cultures, lab studies, imaging

Treatment
- **Surgical** (debridement, irrigation, resection, plastic surgery, amputation)
- **Medical** (antibiotic therapy and hyperbaric oxygen)

8.2.2 Definition

Septic arthritis is an infection within a native joint space that is a surgical emergency.

8.2.3 Epidemiology

Septic arthritis can be bacterial, viral, or fungal. **Bacterial infections** are caused by several species. *Staphylococcus* species include *S. aureus* (>50% of cases), *methicillin-resistant S. aureus*, and *S. epidermis*. In IV drug users, another prevalent bacterium could be *Pseudomonas*. *Neisseria gonorrhoeae* (~20% of cases), instead, is most commonly observed in sexually active young adults with a history of unprotected sex. *Gram-negative bacilli* (~10–20% of cases) are common in infants, IV drug users, elderly, and immunocompromised patients with diabetes.

Viral infections account for 1% of all septic arthritis and are often caused by *Parvovirus B19, Hep B, HCV, HIV, Alphaviruses,* and *HTLV-1*. **Fungal infections** are induced by species of *Candida, Coccidioides immitis, Blastomyces dermatitidis, Histoplasma capsulatum, Sporothrix schenckii, Cryptococcus neoformans,* and *Aspergillus Fumigatus*.

Common **risk factors** include age >80 years, comorbidities, history of crystal arthropathy, endocarditis or recent bacteremia, IV drug user, and recent joint surgery.

8.2.4 Etiology/Pathogenesis

In septic arthritis, proteolytic enzymes (matrix metalloproteinases) are released from polymorphonuclear leukocytes and cause irreversible cartilage destruction within 8 h.

Pathogenesis relies on hematogenous **spread**, **direct inoculation**, and intra-articular **contiguous** spread.

8.2.5 Classification

Septic arthritis is a medical and surgical **emergency** that can be caused by a great variety of

pathogens. As a result, its classification is mostly done by **organism type** (bacterial, viral, and fungal).

8.2.6 Diagnosis

Septic arthritis presents with common **symptoms** such as a red hot swollen joint, localized joint pain, fever (common but not necessary for diagnosis), inability to bear weight or lift, erythema about the joint, joint effusion, or inability to tolerate joint passive ROM.

Joint fluid aspiration is considered the "gold standard" for diagnosis and treatment. Key findings include a cloudy and purulent joint fluid appearance and elevated WBC counts (septic arthritis is evident when >50,000, but infections can occur at lower counts). **Blood cultures** are important to identify the pathogen and direct treatment. **Serum studies** are also helpful for diagnosis (WBC >10,000 with left shift; ESR and CRP are commonly elevated, though ESR can be normal in very acute presentations). **Radiographs** are often normal, but these can show effusion/widened joint space, periosteal elevation or cortical thickening, and loss of joint space. **MRIs** are only used when there is concern for bone involvement. These can show effusion and osteomyelitis.

8.2.7 Treatment

Septic arthritis is considered an orthopedic emergency that requires immediate surgical intervention through **debridement** and **irrigation**. **IV antibiotic therapy** is initiated before receiving definitive cultures based on patient age and risk factors. The therapy is then adjusted based on culture results.

8.3 Periprosthetic Infections

8.3.1 Highlights (Table 8.1)

Definition
- Infection of the **prosthesis**

Epidemiology
- **Pathogens:** *S. aureus* and *S. epidermidis* (**USA**), coagulase-negative *Staphylococcus* spp., *S. aureus*, *Streptococcus*, and *Enterococcus* organisms (**Europe**)
- **Risk factors**: male sex, prior surgeries, operating environment, comorbidities

Etiology/pathogenesis
- **Biofilm** formation through **hematogenous**, **direct**, or **contiguous** spread

Classification
- **Early** vs. **delayed** infection

Diagnosis
- **Clinical presentation:** pain, induration or edema, wound drainage, surgical site erythema, and effusion
- **Testing:** lab studies, joint aspiration, histopathology, frozen sections, imaging

Treatment
- **Surgical** (debridement and irrigation, one- and two-stage implant replacement)
- **Medical** (antibiotic therapy)

8.3.2 Definition

A periprosthetic joint infection (PJI) is an infection involving the joint prosthesis and adjacent tissue.

8.3.3 Epidemiology

Arthroplasty is a prevalent and successful elective surgery for hip and knee osteoarthritis. Despite the improved surgical techniques and infection prevention, PJIs' incidence rate remains between 0.6 and 2.4%. Notoriously difficult to treat and the main cause for revision surgeries, PJIs remain a devastating complication.

S. aureus and *S. epidermidis* are the most common pathogens in the United States, while Europe has a high prevalence of coagulase-negative *Staphylococcus* species, followed by *S.*

aureus, *Streptococcus*, and *Enterococcus* organisms.

Risk factors include male sex, prior surgeries, operating environment, comorbidities (morbid obesity, diabetes, malnutrition, smoking, alcohol consumption, immunocompromising diseases, drug use, nasal carriage of *S. aureus*) and other factors (joint infections, septicemia, active cutaneous or deep tissue infections, and blood transfusions).

Several **preventive measures** are now taken preoperatively (e.g., glycemic control, skin decontamination, decolonization), intraoperatively (e.g., antibiotic prophylaxis, cutaneous preparation, ultraclean operative environment, and blood conservation), and postoperatively (e.g., refined anticoagulation and improved wound dressing) to minimize the risk of infection.

8.3.4 Etiology/Pathogenesis

Microbes undergo transformation in the presence of a prosthesis and form biofilms. The main pathways of infection are **direct seeding, hematogenous spread**, and **recurrent infection**.

8.3.5 Classification

PJIs can be classified into early or delayed-onset infections. **Early post-interventional infections** are caught within 3 months postoperatively. *Symptoms* include pain, induration or edema, wound drainage, surgical site erythema, and effusion. Most common *pathogens* are *Propionibacterium acnes*, *enterococci*, and coagulase-negative *staphylococci*. This type of infection occurs during implantation or in the setting of wound dehiscence and should be managed without delay.

Delayed-onset infections occur after more than 12 months postoperatively. It presents with an acute set of *symptoms* in a previously asymptomatic joint. Persistent joint pain and fever occur in <50% cases. This might present similarly to aseptic failures, but persistent pain is associated with infection and weight-bearing pain, and motion is indicative of aseptic failure. The most common *pathogens* are *S. aureus*, *Gram-negative bacilli*, and *β-hemolytic streptococci*. This type of infection is most commonly caused by hematogenous and contiguous spread and requires removal of the prosthesis.

8.3.6 Diagnosis

Diagnosis is usually confirmed with at least two positive cultures with the same microorganisms. Minor diagnostic criteria include elevated ESR and CRP, high WBC count change on leukocyte esterase test strip, high PMN%, positive histological analysis, and a single positive culture.

Isolating organisms residing in biofilm structures can be challenging, thus posing an issue in the diagnosis and treatment of PJIs. There is no single test or diagnostic with ideal sensitivity and specificity for the diagnosis of PJI, and multiple tests are often utilized before reaching a diagnosis. **Joint aspiration** and culture analysis remain the best diagnostic methods, but culture yields negative result in about 7–50% of PJI cases and sensitivity of cultures remains at 39–70%. **Microbiological studies** are good for diagnosis but require the collection of at least 3–5 specimen cultures to confirm the disorder. **Histopathological studies** are sensitive to detect culture-positive infections, but these provide only a moderately accurate diagnosis. **Radiographs** are the most common first-step imaging, and findings include a wide band of radiolucency at the metal-bone interface with bone destruction. **Bone marrow scintigraphy** combined with **indium-labeled WBC** has shown superior accuracy to diagnose infection. **PCR** is good to identify isolated and difficult-to-culture bacteria and can be used to identify genes responsible for biofilm formation and antibiotic resistance. However, multiple studies have shown that PCR has a similar or lower sensitivity than traditional cultures. Preliminary studies have shown **next-generation sequencing (NGS)** as a potentially more accurate and faster technique for the detection of microorganisms, but further studies are needed to assess the validity of the results.

8.3.7 Treatment

There are several approaches to treat PJIs. **Debridement with retention** is mostly performed in acute cases and only when the wound can be closed. Higher success rates are shown with early treatment (within 30-day onset), low-virulence organisms, and healthy patients. **One-stage implant replacement** is very common in Europe. This is a single operation which includes the removal of the old prosthesis and reimplantation of a new one. This is a reasonable option when effective antibiotic treatment is available. **Two-stage implant replacement** is the most common operation in the United States. This procedure consists of the removal of the prosthesis and the subsequently delayed reimplantation of a second prosthesis. A *short-interval* approach (2–4 weeks) is good for patients with easily treatable pathogens, while a *long-interval* approach (>8 weeks) is good for patients with antibiotic-resistant or unknown organisms, with a presence of a sinus tract, or with nonviable soft-tissue coverage.

Long-term suppressive oral antibiotics can be prescribed when prosthesis removal is not an appropriate option (e.g., poor general health, when removal would result in poor functional outcomes, or patient preferences). The goal of this medical therapy is to achieve asymptomatic functioning prosthesis and not necessarily infection eradication.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. How can you differentiate between osteomyelitis and septic arthritis?
   (a) Osteomyelitis is confirmed with abnormal imaging findings in the periosteum, cortical bone, or medullary cavity. Septic arthritis is confirmed with purulent synovial fluid and abnormal imaging findings in the joint space.
   (b) Septic arthritis is confirmed with abnormal imaging findings in the periosteum, cortical bone, or medullary cavity. Osteomyelitis is confirmed with purulent synovial fluid and abnormal imaging findings in the joint space.
   (c) Septic arthritis is confirmed with normal imaging findings in the periosteum, cortical bone, or medullary cavity. Osteomyelitis is confirmed with purulent synovial fluid and abnormal imaging findings in the joint space.
   (d) Osteomyelitis is confirmed with normal imaging findings in the periosteum, cortical bone, or medullary cavity. Septic arthritis is confirmed with purulent synovial fluid and abnormal imaging findings in the joint space.
2. Why is septic arthritis considered a medical emergency?
   (a) The sudden release of proteolytic enzymes in the joint space causes irreversible cartilage damage within 8 h of infection.
   (b) The sudden release of proteolytic enzymes in the joint space causes irreversible cartilage damage within 2 h of infection.
   (c) The sudden release of proteolytic enzymes in the joint space causes irreversible cartilage damage within 12 h of infection.
   (d) The sudden release of proteolytic enzymes in the joint space causes irreversible cartilage damage within 24 h of infection.
3. What are the challenges in diagnosing PJI?
   (a) Biofilm formation makes it harder to detect and treat the infection.
   (b) High cost of laboratory
   (c) Operator skill level
   (d) Timing
4. What are the incidence and mortality rate of osteomyelitis?
   (a) Incidence: 2/10,000 people. Mortality: when untreated, it can lead to death, but long-term mortality remains unclear.
   (b) Incidence: 20,000 cases each year in the United States. Mortality: *S. aureus* infections up to 50%.

(c) Incidence: in between 0.6 and 2.4%. Mortality: 2–7% of cases.
(d) Incidence: in between 6 and 24%. Mortality: 15–35% of cases.

5. Are physicians more likely to underdiagnose or overdiagnose these infectious disorders?
   (a) Osteomyelitis, septic arthritis, and PJIs are underdiagnosed due to inaccurate testing, low incidence, and similar clinical presentations to more common diagnoses.
   (b) Due to the high frequency, osteomyelitis, septic arthritis, and PJIs always receive a timing diagnosis.
   (c) Osteomyelitis, septic arthritis, and PJIs are overdiagnosed as they exhibit distinct clinical features that make them stand out.
   (d) Physicians rarely encounter challenges in diagnosing osteomyelitis, septic arthritis, and PJIs due to clear and straightforward testing methods.

Take-Home Message

- Three diagnoses with similar **pathogens**, **risk** factors, **etiology**, and **clinical** presentation:
  - Main pathogens: *S. aureus* and skin bacteria
  - Risk factors: immunosuppression and comorbidities
  - Pathogenesis: osteomyelitis and PJI similar cellular mechanism (**biofilm** formation). Septic arthritis caused by sudden release of **proteolytic enzymes**
  - Common symptoms: inflammation, erythema, localized pain, fever, malaise
- **Diagnosis**:
  - Diagnostic steps: recognize inflammation (history, PE, labs), identify pathogens (culture, biopsy, histological examination, joint aspiration), determine location (imaging).
  - Lab findings are nonspecific, and further testing or imaging is usually required to identify the location of the infection.
- Common **treatment** options: surgery + antibiotic therapy.

Further Reading

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Part IV

Pediatric Orthopedics

9 Hereditary Diseases of Orthopedic Interest

Federico Canavese

Overview

This chapter describes the characteristic musculoskeletal manifestation and treatment principles for common hereditary disorders of interest to the orthopedist. In particular, the following disorders are discussed: achondroplasia, hypochondroplasia, arthrogryposis, trisomy 21, Ehlers-Danlos syndrome, neurofibromatosis type 1, osteogenesis imperfecta, osteopetrosis, melorheostosis, mucopolysaccharidosis, Gaucher disease, Klippel-Feil syndrome, and Sprengel's deformity.

9.1 Achondroplasia

Achondroplasia is a rhizomelic short-limbed (disproportionate) dwarfism syndrome and is the most common skeletal dysplasia (incidence: 1/25,000 live births worldwide). Achondroplasia (Fig. 9.1) has to be differentiated from rachitic patients with proportionate dwarfism.

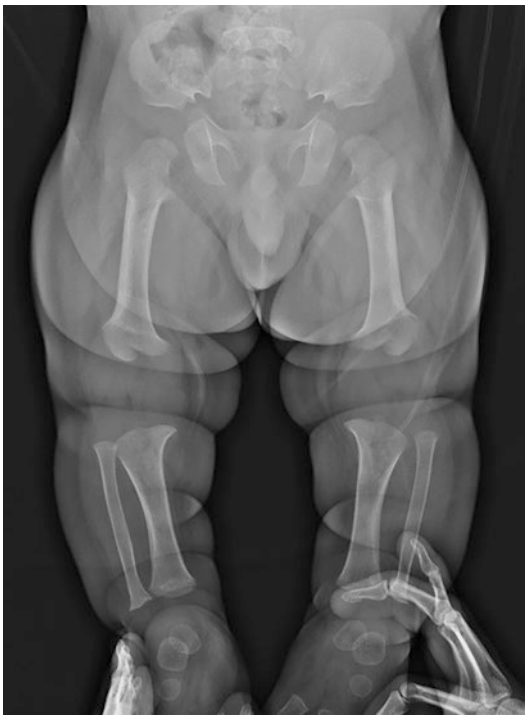

Fig. 9.1 Achondroplasia, lower extremities

Achondroplasia is secondary to defective endochondral ossification due to a mutation in the fibroblast growth factor receptor 3 (FGFR3) gene. Inheritance is autosomal dominant, although in about 80% of cases, it is due to a de novo mutation (parents of average stature). If one parent has achondroplasia, there is a 50% chance

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with each pregnancy of passing it on to offspring. Homozygous achondroplasia (both parents with achondroplasia transmit the mutation; 25% risk) is incompatible with life.

Life expectancy in individuals with achondroplasia is slightly decreased compared to the general population, potentially due to cardiovascular disease. Management of orthopedic and non-orthopedic problems is multidisciplinary, and anticipatory care is warranted.

9.1.1 Clinical Features

Patients with achondroplasia typically show macrocephaly with frontal bossing, midfacial hypoplasia with a depressed nasal bridge, short stature, and disproportionate body habitus (short limbs and long and narrow trunk); adult males and females reach an average height of 131 cm and 124 cm, respectively. Hands are typically short (brachimetacarpia) and trident shaped.

Hydrocephalus, dental crowding, and chronic otitis media leading to conductive hearing loss are also relatively common at this stage; in older children, midfacial hypoplasia with tonsil and adenoid hypertrophy can cause obstructive sleep apnea.

Overweight and obesity are common issues; weight gain must be monitored in childhood in order to avoid later complications.

9.1.2 Orthopedic Manifestations

Orthopedic manifestations of achondroplasia are typical and develop at different stages of the patient's life. The neuromotor development is frequently delayed due to the coexistence of hypotonia, large head, short limbs, and short neck. In addition, during infancy, thoracolumbar kyphosis is a common finding although it resolves over time in about 90% of cases.

Genu varum often occurs in childhood as well; progressive and symptomatic forms can be treated by surgery (guided growth or osteotomy, depending on the severity of the deformity and the age of the patient). Limb-lengthening procedures (humerus, femur, and tibia) are controversial.

During infancy, the presence of a smaller and/or abnormally shaped foramen magnum can cause spinal cord compression or vertebral artery compression leading to central apnea; neurosurgical advice is warranted if imaging of the head and neck and polysomnography are abnormal. Activities at risk of injury to the craniocervical junction should be avoided.

During adulthood, patients with achondroplasia frequently develop lower lumbar spinal stenosis with the associated neurological deficit; lumbar laminectomy is usually performed to treat symptomatic spinal stenosis. Pregnant women with achondroplasia must deliver by caesarian section due to small pelvis size.

9.2 Hypochondroplasia

Hypochondroplasia is a skeletal dysplasia characterized by disproportionate body habitus, short stature, short arms and legs, broad short hands and feet, mild joint laxity, mild lumbar lordosis, limited extension of the elbow joints, and macrocephaly. The skeletal features are very similar to those seen in achondroplasia but tend to be milder.

Hypochondroplasia is caused by the same mutation of achondroplasia (FGFR3 gene), and transmission is autosomal dominant. However, the majority of patients with hypochondroplasia have parents of average height and manifest the disease as the result of a de novo mutation. An individual with hypochondroplasia who has a partner of average stature is at a 50% risk of having a child with hypochondroplasia; final adult height varies between 132 and 147 cm, and life expectancy is normal.

9.3 Arthrogryposis Multiplex Congenita

The term "arthrogryposis" describes a diffuse group of syndromes, including, but not limited to, amyoplasia (the most common form of arthro-

gryposis involving all four limbs), Larsen syndrome, distal arthrogryposis, and pterygia syndromes.

Arthrogrypotic syndromes are rare, nonprogressive congenital disorders characterized by multiple joint contractures associated with *akinesia* (reduced or absent joint movements), muscle weakness, and connective tissue fibrosis that can be either generalized or limited to upper and/or lower extremity. The limbs are typically tubular, and skin creases are frequently reduced or absent. Transmission of arthrogrypotic syndromes can be autosomal dominant, autosomal recessive, or X-linked recessive.

9.3.1 Larsen Syndrome

Larsen syndrome is a rare arthrogrypotic syndrome with an estimated birth prevalence of 1/100,000. Inheritance is autosomal dominant; if one parent has Larsen syndrome, there is a 50% chance with each pregnancy of passing it on to offspring. Life expectancy in individuals with Larsen syndrome is comparable to the general population.

The thumb of patients with Larsen syndrome is typically short due to the presence of a broad, short, and spatula-shaped distal phalanx. All infants with Larsen syndrome must have their cervical spine evaluated to rule out cervical spine instability (life-threatening; surgical stabilization required), or cervical kyphosis (myelopathy); scoliosis is relatively frequent Clubfoot and numerous joint dislocation at birth are frequent.

Other typical clinical signs are flattened facial appearance (midface, in particular), depressed nasal bridge, and a prominent forehead. Ocular hypertelorism, cataract, and cleft palate (midline) are other relatively frequent clinical signs.

9.3.2 Distal Arthrogryposis

The term distal arthrogryposis characterizes a group of 11 autosomal dominant disorders that mainly involve the distal parts of the limbs. Features shared among all distal arthrogryposes include a consistent pattern of hand (camptodactyly or pseudocamptodactyly, hypoplastic or absent flexion creases, overriding fingers, ulnar deviation of the wrist) and foot involvement and limited involvement of proximal joints.

9.3.3 Pterygia Syndromes

The term pterygium syndrome characterizes a group of several disorders (autosomal dominant or recessive) with *akinesia* and joint contractures due to webbing of the skin (pterygium). The webbing typically affects the skin of the neck, fingers, forearms (elbow), inner thighs, and popliteal fossa.

9.4 Down Syndrome (Trisomy 21)

Down syndrome is caused by the presence of a third copy (partial or total) of chromosome 21; the average incidence is about 1/700 live births although it increases with the maternal age, with the highest risk in pregnant women aged 40 years and older (1/100).

Patients with Down syndrome have a typical facial dysmorphism, muscular hypotonia, joint laxity, as well as various cardiac (atrioventricular canal), gastrointestinal (duodenal atresia, Hirschsprung disease, celiac disease), neurosensorial (congenital cataract, conductive hearing loss, early-onset Alzheimer disease), endocrine (hypothyroidism, diabetes type 1), and tumoral diseases (leukemia); a variable degree of intellectual disability completes the clinical picture.

Life expectancy is above the age of 60 years in developed countries.

9.4.1 Clinical Features

Patients with Down syndrome typically have short stature, up-slanting palpebral fissures, epicanthus, flat neck, round face, small nose, and bilateral single palmar crease; these clinical signs

can be however relatively mild and are not pathognomonic of the condition. Neuromotor development is delayed.

9.4.2 Orthopedic Manifestations

The spine is often affected in patients with Down syndrome. Upper cervical spine instability is frequent (contact sports to be avoided), scoliosis is present in about 50% of cases, while spondylolisthesis is detected in 5–10% of patients. Treatment varies according to the severity of the deformity (brace/surgery). The pelvis is frequently abnormal with flattened acetabulum and iliac wings; progressive hip dysplasia may begin during later childhood leading to loss of containment and dislocation in some cases; slipped capital femoral epiphysis is relatively frequent and is secondary to hormonal imbalance (hypothyroidism).

Hyperlaxity is responsible for plano-valgus foot deformity and recurrent patella dislocation (lateral); foot deformity, if symptomatic, is generally treated by fusion while patella dislocation rarely requires treatment (frequently asymptomatic; recurrence rate above 90%).

9.5 Ehlers-Danlos Syndrome

Ehlers-Danlos syndrome (EDS) is the most prevalent heritable connective tissue disorder with a prevalence of 1/20,000 live births. EDS is characterized by defective elastin and collagen formation leading to fragile hyperelastic skin, generalized joint hypermobility, wound healing difficulty, and vascular fragility; scars are typically widened and atrophic.

EDS has been classified into 13 types, with variable clinical manifestations. The inheritance of the classical form of EDS is autosomal dominant, and the risk of transmission to offspring from an affected parent is 50%; other forms can be recessive.

Life expectancy in individuals with EDS is normal although it can be reduced in some cases due to vessel rupture (spontaneous dissection or rupture of medium-sized arteries). Quality of life depends on the severity of the disease.

9.5.1 Clinical Features

The diagnosis of EDS is based on the simultaneous presence of skin hyperextensibility, atrophic scarring, and either generalized joint hypermobility or three or more of the following minor criteria: skin fragility, easy bruising, soft/doughy skin, molluscoid pseudo-tumors, subcutaneous spheroids, hernia, epicanthus, complications of joint hypermobility, and positive family history (first-degree relative); some patients may have blue sclera. Regular cardiac assessment (echocardiography) is recommended to diagnose aortic root dilation and mitral valve prolapse.

9.5.2 Orthopedic Manifestations

Orthopedic manifestations of EDS are of variable severity. Individuals with EDS have a frequent subluxation and occasional dislocation of large and small joints. Individuals with EDS frequently complain of on/off pain involving joints, limbs, spine, and neck; some patients can develop abnormal gait. Patients with upper cervical spine instability and basilar impression (lower brain pushing down into the spine) require surgical treatment. The presence of scoliosis or kyphoscoliosis is also frequent, and treatment varies according to the severity of the deformity (brace and/or surgery).

Pain relief is the primary goal for patients with EDS. Painful joint instability usually responds to conservative treatment. If this is unsuccessful, surgical intervention may help; however, surgery is not the first option and must be carefully evaluated as the incidence of bleeding complications, wider and poorly esthetic scars, and less predictable healing is high.

9.6 Marfan's Syndrome

Marfan's syndrome is an autosomal dominant connective tissue disorder with a prevalence of 1/5000 (no difference between sexes); if one parent has Marfan's syndrome, there is a 50% chance with each pregnancy of passing it on to offspring.

Patients have a life expectancy close to that of the general population if they are properly followed up and adequately treated.

9.6.1 Clinical Features

Patients with Marfan's syndrome are typically tall due to excessive length of the extremities (*dolichostenomelia*, arm span larger than height), joint hypermobility, and arachnodactyly of hands (Fig. 9.2) and feet (excessive length of the fingers/toes); patients are usually thin due to little subcutaneous fat.

9.6.2 Orthopedic Manifestations

Orthopedic problems are often the first sign of the disease and include scoliosis (early onset)

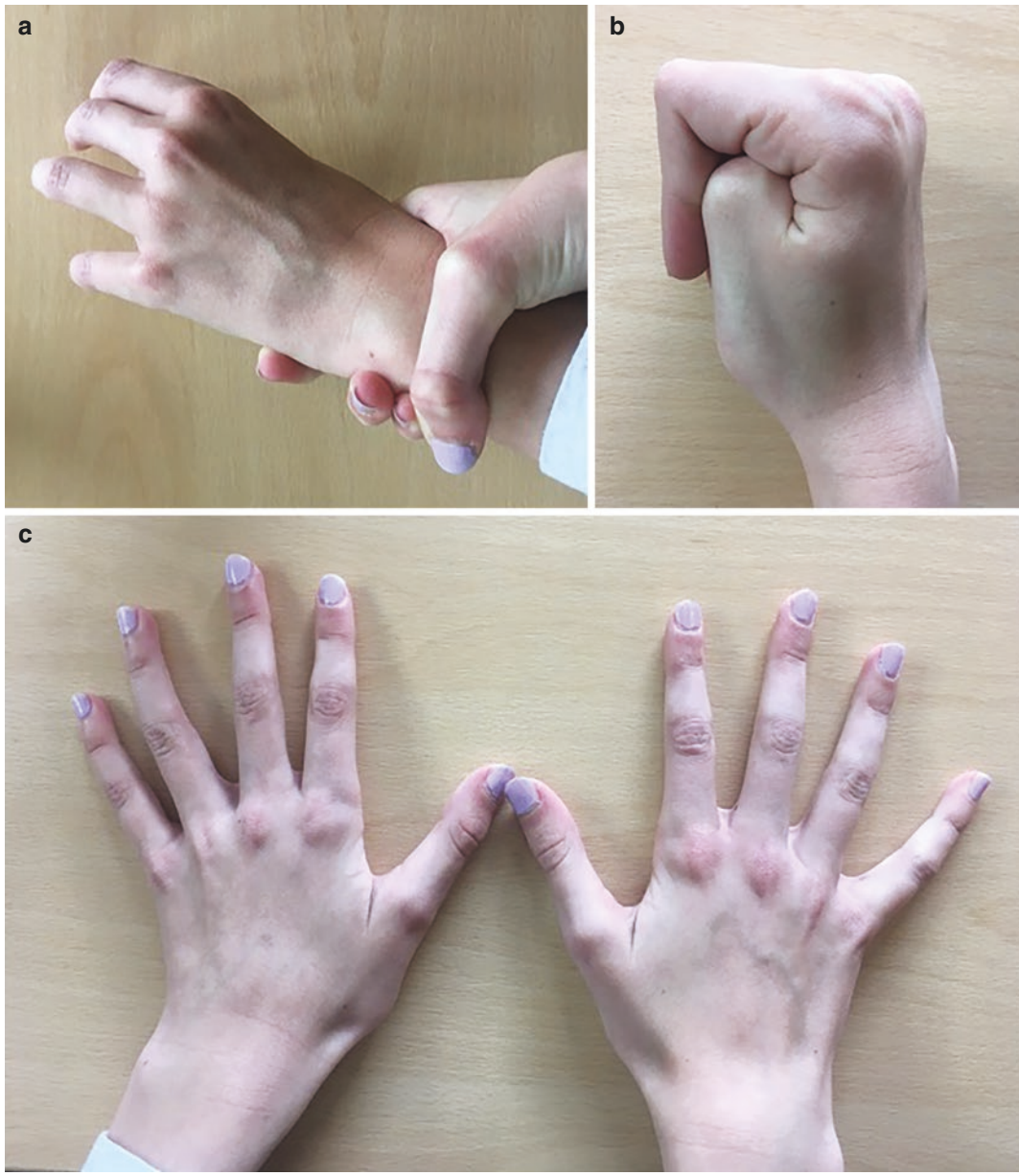

Fig. 9.2 Marfan's syndrome, joint laxity (**a**, **b**), and arachnodactyly of hands (**c**)

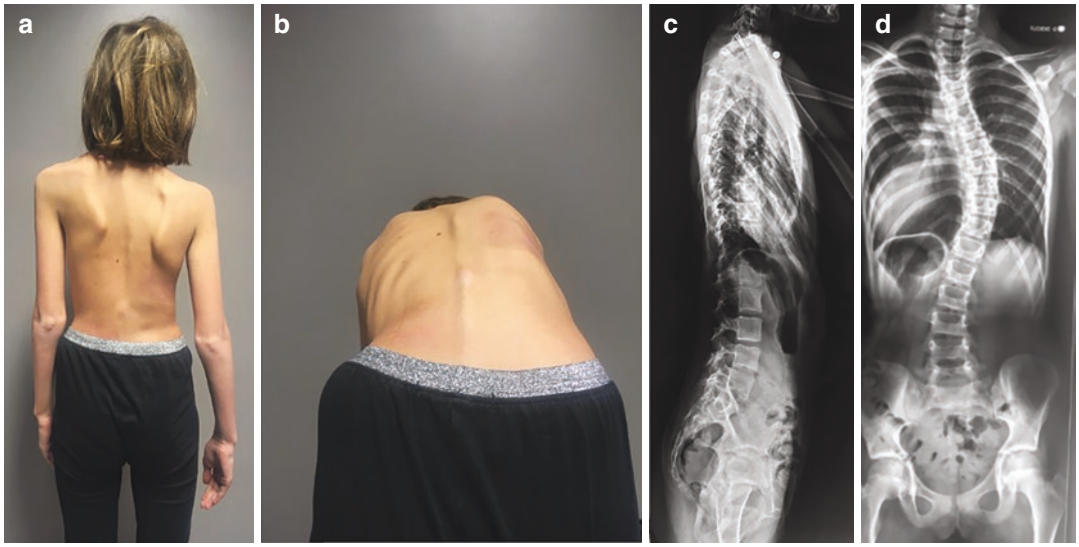

Fig. 9.3 Marfan's syndrome, early-onset scoliosis, clinical (**a**, **b**) and radiographic (**c**, **d**) appearance

(Fig. 9.3), pectus carinatum or excavatum, protrusio acetabui, dislocation, or subluxation of the patella; other typical skeletal signs are flattened facial appearance and micrognathism or malar hypoplasia.

Surgery can be offered for skeletal anomalies (vertebral column stabilization in the case of scoliosis or reparation of thoracic deformities).

9.7 Neurofibromatosis Type 1

Neurofibromatosis (Fig. 9.4) is discussed in Chap. 7.

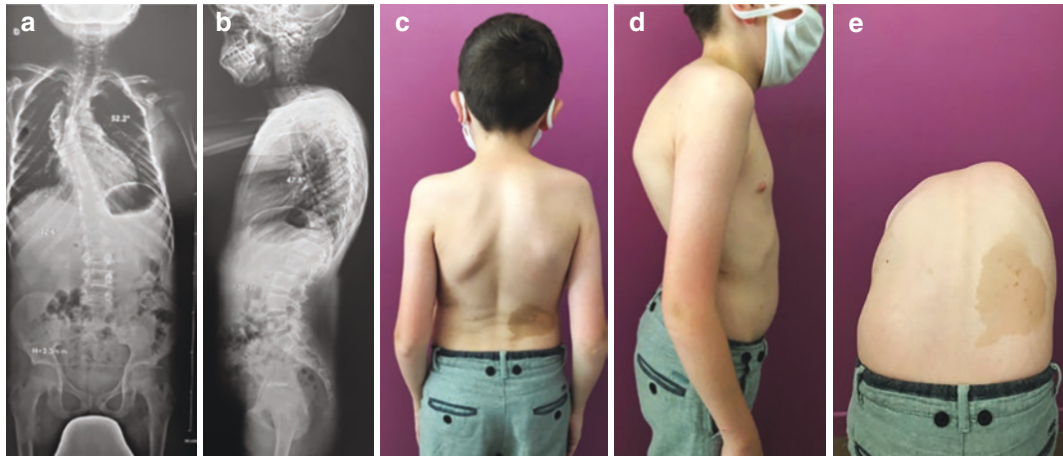

Fig. 9.4 Neurofibromatosis type 1, dystrophic scoliosis; anteroposterior (**a**) and lateral (**b**) view; clinical picture (**c**, **d**); typical café-au-lait spot (**e**)

9.8 Osteogenesis Imperfecta

Osteogenesis imperfecta (OI) is a group of disorders characterized by increased bone fragility and multiple spontaneous fractures; the disease is also known as "brittle bone disease," "glass bone disease," or Lobstein disease.

OI is caused by mutations in the COL1-A1 and COL1-A2 genes encoding the α1 and α2 chains of type 1 collagen; the inheritance is autosomal dominant; autosomal recessive forms are much more infrequent (<5%), are caused by different genetic mutations, are always severe, and are associated with marked hypotonia. The prevalence of the disease is 1/10,000 live births.

Five clinical subtypes of OI with different degrees of severity have been identified: types I–V. Life expectancy is related to the severity of the disease and the respiratory status associated with spinal deformities.

9.8.1 Clinical Features

The characteristic clinical signs of OI are spontaneous fractures, short stature, blue sclera, and dentinogenesis imperfecta; however, not all forms of the disease have all clinical signs present simultaneously.

In particular, type I patients have normal or mild short stature, blue sclera without skeletal deformity, and dentinogenesis imperfecta; patients with type II present multiple fractures at birth (long bones, ribs), severe skeletal deformity, and dark blue sclera; this form of the disease is lethal. Patients with type III and type IV have similar clinical presentation with short stature, triangular face, scoliosis, grayish sclera, and dentinogenesis imperfecta. Type V patients have mild short stature, dislocation of the radial head, mineralized interosseous membranes, and hyperplastic callus; patients have white sclera and no dentinogenesis imperfecta; this type of OI is extremely rare with a reported prevalence of less than 1/one million.

9.8.2 Orthopedic Manifestations

Orthopedic manifestations of OI are multiple fractures (olecranon fracture in children is rare and its occurrence should alter the clinician), bone deformity (Fig. 9.5), scoliosis and kyphosis, and dislocation of the radial head. Surgical management is essential for the correction of bone and spinal deformities. Long-bone fractures can be prevented by endo-medullary osteosynthesis in association with multilevel osteotomy to

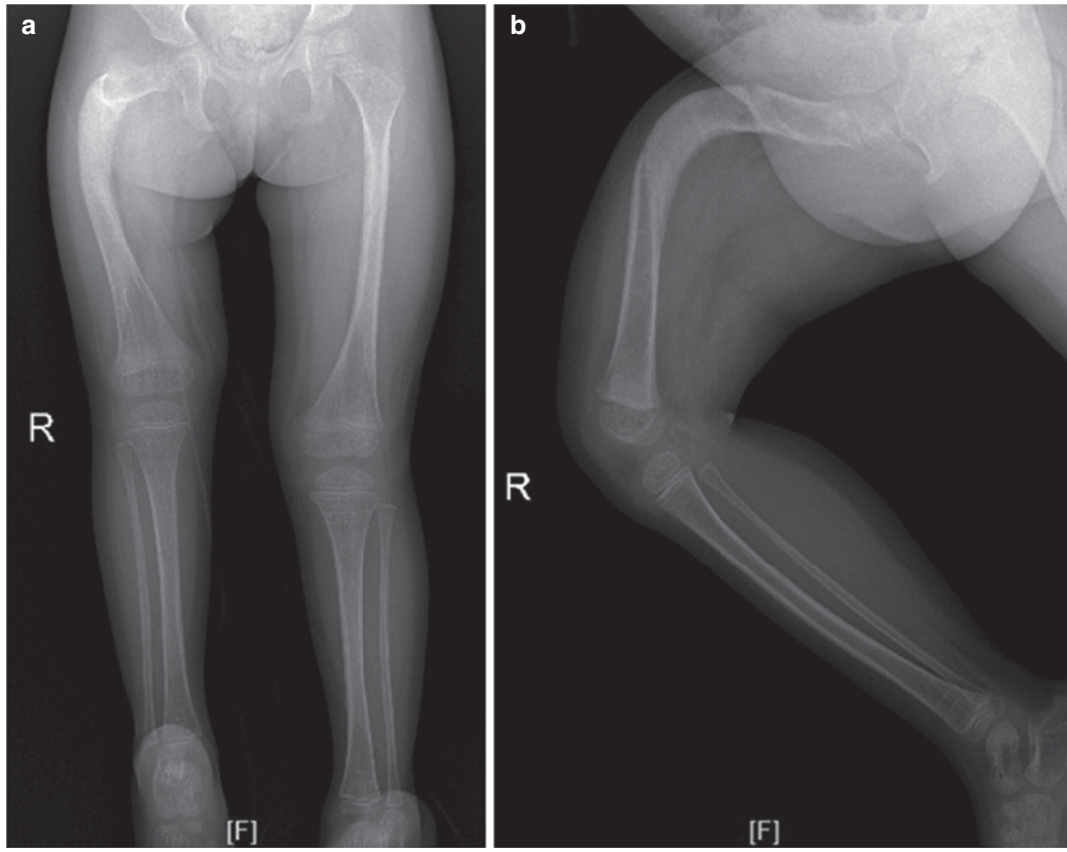

Fig. 9.5 Osteogenesis imperfecta, deformity of right femur (**a**, **b**)

realign the bone and to prevent fractures. Spine surgery is challenging due to the severity of the deformity and to the poor bone quality and stock; complications are frequent.

9.9 Osteopetrosis

The term osteopetrosis ("marble bone" or Albers-Schönberg disease) describes a group of rare, heritable conditions characterized by increased bone density and narrow medullary canal due to failure of osteoclastic resorption. The autosomal dominant form of the disease has a prevalence of about 1/20,000 births, while the autosomal recessive (malignant osteopetrosis; severe) has an incidence of 1/250,000 births; X-linked forms of the condition have also been described.

The onset of skeletal manifestations such as fractures and osteomyelitis in late childhood or adolescence is typical of the autosomal dominant form, while short stature, compressive neuropathies (optic and facial), hypocalcemia and tetanic seizures, and life-threatening pancytopenia are typical of the autosomal recessive variant.

The severe forms of osteopetrosis are associated with reduced life expectancy (death from repeated hemorrhage or overwhelming infection); on the other hand, milder and late-onset forms have normal life expectancy.

9.9.1 Orthopedic Manifestations

Multiple fractures are frequent (generalized osteosclerosis; fragility) and are characterized by

slow healing and increased rate of nonunion due to diminished osteoclastic resorption and absence of vascular bone marrow (failure of remodeling).

Long bones have widened cortices and narrow medullary canal; the skull is thickened and sclerotic, and the spine has characteristic stripped appearance due to sclerotic end plates (rugger jersey spine).

Patients with osteopetrosis are prone to osteomyelitis (in particular after teeth extraction); deafness is another frequent finding.

Treatment of osteopetrotic conditions is symptomatic. In case of fracture, orthopedic surgeons must consider careful intramedullary fixation as medullary canal is small or absent (drill rather than ream).

9.10 Melorheostosis

Melorheostosis is a rare connective tissue disorder (prevalence: 1/1,000,000; sporadic disease) characterized by a unilateral sclerosing bone dysplasia (monostotic, polyostotic, or monomelic abnormal proliferation of compact haversian bone distorting the contour of bone periosteal and endosteal surfaces). Melorheostosis is not life-threatening but can greatly impact the quality of life of affected individuals due to chronic pain.

9.10.1 Orthopedic Manifestations

Patients with melorheostosis typically develop joint contractures and stiffness, bone and joint deformities, and chronic pain, usually aggravated by activities.

On plain radiographs, melorheostosis typically appears as linear lesions of increased bone density along the major axis of the tubular bones leading to increased thickness of the cortices ("dripping wax along a candle" appearance). In polyostotic forms, the lesions are typically continuous along the same side of the affected limb and "jump" across the joints (Fig. 9.6).

There is no curative treatment for melorheostosis. Management aims to relieve pain, correct deformity, and restore range of motion. Surgical options include limb and tendon lengthening, fasciotomies, capsulotomies, osteotomies, excision of fibrous tissue and/or hyperostosis, contralateral epiphysiodesis, arthrodesis, callotasis, and amputation. Surgical treatments are best performed after skeletal maturity although deformities can recur.

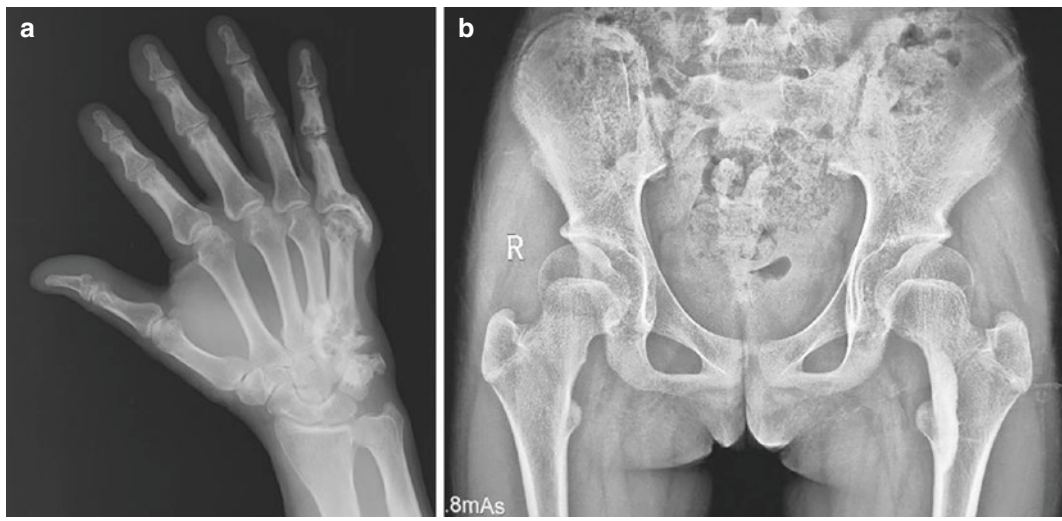

Fig. 9.6 Melorheostosis, polyostotic form with ossification crossing the joint (**a**); monostotic form (left proximal femur, increased thickens of the cortex; **b**)

Extra-skeletal manifestations include edema, hyperpigmented skin patches, circumscribed and linear scleroderma, vascular tumors, and malformations.

9.11 Mucopolysaccharidosis

Mucopolysaccharidosis (MPS) is a group of lysosomal storage diseases, with variable clinical presentation and different mode of transmission. Skeletal manifestations are common, and multiple orthopedic surgeries are frequently needed. Children with MPS are at risk of surgical and anesthetic complications due to cervical spine instability (Fig. 9.7), cervical stenosis, airway compromise, and underlying cardiac disease; in addition, general anesthesia may be problematic due to intubation difficulties. Neurophysiological monitoring is strongly recommended, even for non-orthopedic procedures.

9.11.1 Mucopolysaccharidosis Type 1

MPS type 1 includes three distinct syndromes, with different degrees of severity.

9.11.1.1 Hurler Syndrome

Hurler syndrome is the most severe form of MPS type 1. Prevalence is about 1/100,000, and transmission is autosomal recessive.

Patients with Hurler syndrome have cognitive impairment, multiple skeletal abnormalities, cardiomyopathy and valvular abnormalities, respiratory problems (enlarged tonsils and adenoids, nasal secretion), enlarged liver and spleen, and reduced life expectancy. The facies is characteristic with large head with bulging frontal bones, depressed nasal bridge with broad nasal tip and anteverted nostrils, full cheeks, and enlarged lips.

9.11.1.2 Orthopedic Manifestations

Patients with Hurler syndrome have short stature, thickened skull, shortened long bones, odontoid hypoplasia (70% of cases), vertebral abnormalities, severe thoracolumbar kyphosis (gibbus deformity; 80% of cases), scoliosis, acetabular dysplasia (90% of cases; associated with hip dislocation in about 40% of patients), genu valgum (70% of cases), and short/broad digits (the term *dysostosis multiplex* includes all bone abnormalities) (Fig. 9.8). Sixty percent of patients can develop carpal tunnel syndrome; if carpal tunnel

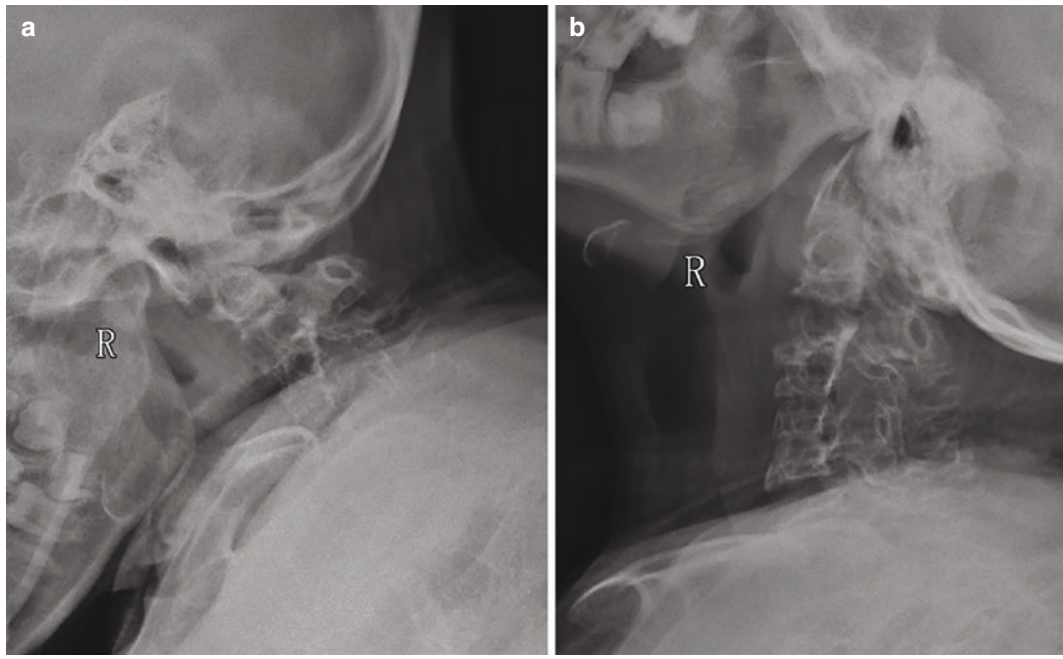

Fig. 9.7 Mucopolysaccharidosis type 1, upper cervical spine instability; flexion (**a**) and extension (**b**) radiographs

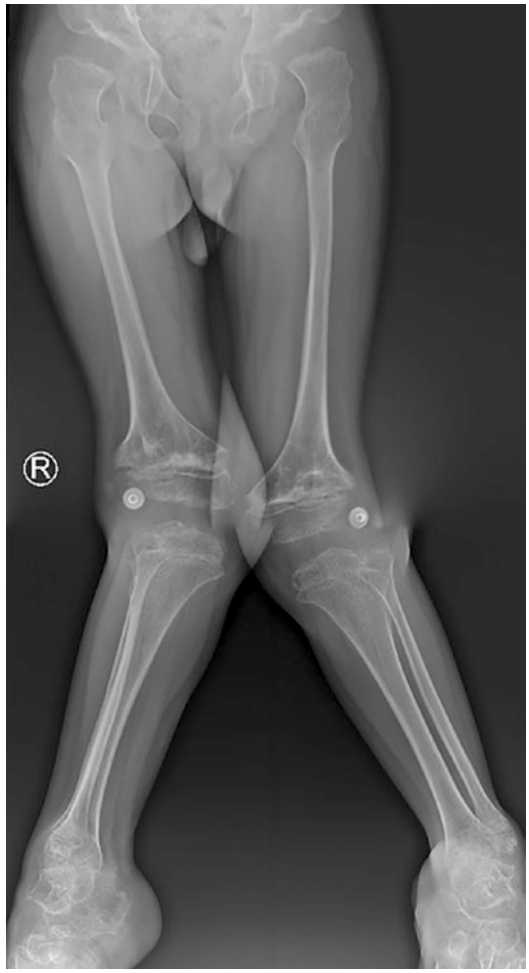

Fig. 9.8 Mucopolysaccharidosis type 1, multiple lower extremity deformity

syndrome is detected in a child (extremely rare), MPS should be ruled out.

Early surgical intervention (usually between 4 and 8 years of age) to address the multiple orthopedic problems is indicated despite the high rate of complications.

9.11.1.3 Hurler-Scheie Syndrome

Hurler-Scheie syndrome is the intermediate form of MPS type 1 characterized by musculoskeletal alterations (see Hurler syndrome) and a delay in motor development. Transmission is autosomal recessive.

9.11.1.4 Scheie Syndrome

Scheie syndrome is the mildest form of MPS type 1; symptoms usually appear after the age of 5 years. Patients have almost normal height and no intellectual deficiency. Compression of the cervical spinal cord, caused by glycosaminoglycan infiltration of the dura, requires neurosurgical intervention. Transmission is autosomal recessive.

9.11.2 Mucopolysaccharidosis Type 2

MPS type 2 (Hunter syndrome) is an X-linked recessive disorder with a prevalence of 1/150,000. There are two forms of MPS type 2: A (severe) and B (attenuated).

Patients with the severe form are expected to survive until the second decade of life while those with the attenuated form may have a longer life span.

9.11.3 Orthopedic Manifestations

Orthopedic involvement is common in patients with Hunter syndrome and leads to severe restrictions in motion and impaired quality of life. Musculoskeletal manifestations include arthropathy, contractures and joint stiffness (prevalence of joint contractures is variable; hand/wrist: 65%, elbow: 60%, shoulder: 55%, knee: 45%, ankle: 35%), claw hands (55% of cases), carpal tunnel syndrome, trigger finger, kyphosis and scoliosis (35% of cases), short limb, and acetabular and proximal femur dysplasia; spine instability and spinal cord compression may also occur (15% of cases). It is important that concurrent disease manifestations (multisystem involvement) are identified and taken into account prior to any orthopedic procedure.

9.11.4 Mucopolysaccharidosis Type 3

Orthopedic problems are mild or absent. Life expectancy is reduced.

9.11.5 Mucopolysaccharidosis Type 4

MPS type 4 (Maroteaux–Lamy syndrome) is characterized by distinctive and severe spondylo-epi-metaphyseal dysplasia, dysplastic odontoid process (atlantoaxial instability and risk of spinal cord compression), joint hyperlaxity, and short stature.

9.11.6 Mucopolysaccharidosis Type 6

MPS type 6 (Maroteaux–Lamy syndrome) is characterized by short stature, *dysostosis multiplex*, restricted joint movement, flexion contractures, and degenerative joint disease.

Skeletal manifestations of patients with MPS type 6 are kyphosis, cranio-cervical stenosis, multilevel cervical stenosis, atlantoaxial instability (with resultant myelopathy and spastic quadriparesis), hip dysplasia, and short bones. Due to the development of femoral head osteonecrosis and collapse, hip surgery is usually not recommended. Patients with MPS type 6 can also develop genu valgum severe enough to warrant surgery.

9.12 Gaucher Disease

Gaucher disease is an inherited lysosomal storage disorder characterized by hepatosplenomegaly, anemia, bleeding disorders, thrombocytopenia, musculoskeletal problems, growth retardation, and, in most severe cases, pulmonary compromise.

The disease is particularly frequent among the Ashkenazi Jewish (incidence: 1/1000), while the prevalence in the general population is 1/100,000. Transmission is autosomal recessive.

9.12.1 Orthopedic Manifestations

Patients with Gaucher disease are at risk for pathologic fracture (osteopenia and osteoporosis due to progressive storage of glucocerebroside in the bone marrow), abnormal bone remodeling and delayed healing, increased intraoperative bleeding, and infection.

Avascular bone necrosis (especially hips), cortical thinning, lytic bone lesions, osteosclerosis, and acute osteomyelitis (relatively rare) are the other orthopedic manifestations of the disease. Avascular necrosis and osteomyelitis have similar clinical presentation and warrant careful and accurate diagnosis to ensure proper treatment. In particular, the so-called *Gaucher crisis* must be recognized to avoid overtreatment; *Gaucher crisis* (pseudo-osteomyelitis) is characterized by acute intense pain (distal femur, proximal tibia, and proximal femur, most frequently), mild localized tenderness and swelling, increased WBC, ESR, and CRP. The crisis is caused by subperiosteal or intramedullary canal bleeding with blood under pressure causing pain. Bone infarcts and avascular necrosis can lead to progressive functional impairment or even destruction of joints.

9.13 Sprengel's Deformity

Sprengel's deformity is a rare malformation characterized by an underdeveloped and abnormally high scapula incompletely descended to its normal position during the embryonic development.

9.13.1 Clinical Signs

The abnormally high scapula is mostly unilateral; clinically and radiographically, the scapula is high (asymmetry of scapular height) and has abnormal shape, reduced size, and limited motion; in particular, patients with Sprengel's deformity have limited shoulder abduction (rotation of the scapula leaves the glenoid facing down).

Sprengel's deformity can be associated with other abnormalities, such as deformities of vertebral bodies (association with Klippel-Feil syndrome in 25% of cases), fused or absent ribs, or genitourinary anomalies.

9.13.2 Orthopedic Manifestations

An omovertebral bone between the cervical spine and the elevated scapula can be present in 25% of cases; scoliosis is detected in about 40% of patients with Sprengel's deformity.

Surgical treatment of Sprengel's deformity aims to draw the scapula down to a lower position and to excise the omovertebral bone, if present (Woodward procedure).

9.14 Klippel-Feil Syndrome

Klippel-Feil syndrome (KFS) is characterized by a short and hypomobile neck (fixed torticollis) due to the presence of congenitally fused cervical vertebrae (failure of segmentation during embryogenesis); mode of inheritance is unknown.

The prevalence of KFS is 1/10,000, and it can be associated with Sprengel's deformity in about 25% of cases.

9.14.1 Clinical Features

The classic clinical signs of KFS are low posterior hairline, short neck, and limited neck range of motion.

9.14.2 Orthopedic Manifestations

Patients with KFS can develop neck pain, radiculopathy (nerve root impingement), and/or myelopathy (congenital cervical stenosis). Surgical intervention may be required in cases with cervical or cranio-vertebral instability and is associated with an increased risk of spinal cord injury.

Take-Home Message

- Although rare, hereditary disorders represent a complex chapter of pediatric orthopedics as musculoskeletal problems are frequent and can significantly impact the quality of life of affected individuals.
- Specific orthopedic problems are typical for each type of disease; important and common manifestations of orthopedic interest are spine deformity and instability, chest deformity, upper and lower extremity deformity, and joint contractures. In particular, cervical spine involvement must be promptly recognized as to avoid potential life-threatening complications; lower limb deformity is generally complex, and recurrence rate is high.
- Specialized treatment is required due to the complexity of surgical indications, the high frequency of concomitant extra-skeletal manifestations, and the high rate of complications.

Summary

This chapter describes the characteristic musculoskeletal manifestation and treatment principles for common hereditary disorders of interest to the orthopedist. In particular, the following disorders are discussed: achondroplasia; hypochondroplasia, arthrogryposis, trisomy 21, Ehlers-Danlos syndrome, neurofibromatosis type 1, osteogenesis imperfecta, osteopetrosis, melorheostosis, mucopolysaccharidosis, Gaucher disease, Klippel-Feil syndrome, and Sprengel's deformity.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. What is the most common skeletal dysplasia?
   (a) Achondroplasia (incidence: 1/25,000)
   (b) Arthrogryposis
   (c) Osteogenesis imperfecta
   (d) Kniest dysplasia
2. In patients with achondroplasia, what is the expected outcome of thoracolumbar kyphosis?
   (a) Favorable as it tends to resolve over time in more than 90% of cases
   (b) Not favorable as it tends to increase over time in more than 90% of cases
   (c) It remains stable throughout life in most cases
   (d) It is typically seen only in a minority of cases
3. What are the typical signs of arthrogrypotic syndromes?
   (a) Joint contractures, akinesia, muscle weakness, tubular limbs, skin creases reduced or absent
   (b) Short and hypomobile neck (fixed torticollis) due to the presence of congenitally fused cervical vertebrae
   (c) Hyperextension of all major joints, decreased muscle tone, and elongated limbs
   (d) Progressive muscle hypertrophy, elongated fingers, and excessive skin folds
4. Severe thoracolumbar kyphosis (gibbus deformity) is frequently seen in what hereditary condition?
   (a) Mucopolysaccharidosis type 1 (Hurler syndrome), in about 80% of cases
   (b) Mucopolysaccharidosis type 6, in about 80% of cases
   (c) Marfan syndrome, in about 80% of cases
   (d) Osteogenesis Imperfecta, in about 80% of cases
5. If carpal tunnel syndrome is detected in a child, what disorder has to be suspected?
   (a) Mucopolysaccharidosis; carpal tunnel syndrome is an extremely rare finding in normal children; it is present in about 60% of children with mucopolysaccharidosis
   (b) Sprengel's deformity; carpal tunnel syndrome is an extremely rare finding in normal children; it is present in about 50% of children with Sprengel's deformity
   (c) Legg-Calvé-Perthes disease
   (d) Osteogenesis imperfecta

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10 Congenital Deformities of the Appendicular and Axial Skeleton

Ole Rahbek, Peter Heide Pedersen, and Søren Kold

Overview

- Congenital deformities of the appendicular and axial skeleton are rare diseases. Some diseases have impact on both the skeleton and internal organs.
- Congenital deformities can result from generalized skeletal dysplasia as in achondroplasia, leading to shortening of all limbs.
- Congenital disease can also be a result of non-generalized limb abnormalities such as fibular hemimelia, leading to one-sided limb deformity.

10.1 Introduction

Congenital deformities of the appendicular and axial skeleton are rare diseases. Some diseases mainly affect the skeleton (such as achondroplasia), while others (such as Marfan's syndrome) have impact on both the skeleton and internal organs. Congenital deformities can result from generalized skeletal dysplasia as in achondroplasia, leading to shortening of all limbs. Congenital disease can also be a result of non-generalized limb abnormalities such as fibular hemimelia, leading to one-sided limb deformity. An overview of congenital limb anomalies is given in Table 10.1.

The etiology is often multifactorial as in clubfoot but can also be inherited as in vitamin D-resistant rickets. Different classification systems are available for most congenital diseases and should ideally be capable of guiding treatment. The diagnosis is made from a thorough clinical examination. Some diagnoses are supplemented by ultrasound (as in developmental dysplasia of the hip in the newborn) or supplemented by standard X-rays of affected skeletal sites. Some deformities can be diagnosed from prenatal ultrasound, and inherited diseases are backed by detailed family history and laboratory testing for genetic disease. In syndromes, MRI or ultrasound may be warranted to detect pathologies in internal organs or in the central nerve system. CT scans are commonly used for 3D visualization of bony deformities for preoperative planning.

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Table 10.1 The table gives an overview of congenital limb anomalies

Dysmelia—congenital limb anomalies			
Types	Subtypes	Definition	Examples
Amelia		Deficiency of one or more limbs	Arm or leg missing
Meromelia		A part of a limb is missing	
	Ectrodactyly	Deficiency or absence of one or more central digits of the hand or foot	Cleft hand or cleft foot
	Phocomelia	Malformation of a limb or limbs	Short limbs. Due to genetics or drug exposure in fetal life, e.g., thalidomide
	Hemimelia	Approximately half of a limb is missing	Fibular and tibial hemimelia, radial or ulnar clubhand
Polymelia		Extra limb	Having five or more limbs
Polydactyly		Extra finger or toe	Most common site is on the ulnar side of the hand. Often underdeveloped
Syndactyly		Two or more digits are fused	In the simple form, digits are fused with soft tissue and in complex forms the bones are fused. Fusion can be partial or complete
Oligodactyly		Less than five fingers or toes on a hand or foot	Can be seen isolated or as part of hemimelia or ectrodactyly
Brachydactyly		Short digit	Is most often an isolated condition, but can also be seen with many congenital syndromes
Amniotic band syndrome		Strands of the amniotic sac separate and entangle digits or limbs in fetal life	A variety of anomalies that include amputations and deformities dur the constricting band
Generalized dysmelia		All limbs involved	Short stature diseases, Marfan's syndrome
	Proportionate	Both limbs and torso involved	Osteogenesis imperfecta, spondyloepiphyseal dysplasia congenita, mucopolysaccharidoses
	Disproportionate	Only limbs are involved	Achondroplasia is most common

10.2 Infantile Torticollis

The disease usually manifests itself in the first year of life with a skewed position of the head (0.5% of infants). The cause may be an abnormality in the cervical spine (X-ray examination of the cervical spine) but is most often due to pathological changes in the sternocleidomastoid muscle possibly due to a compromised blood supply. In the early stages, the disease manifests itself as a limited thickening with a tilted head position as a result. The disorder is treated in milder cases with manipulation and if needed supplementary injection of botulinum toxin. More severe cases are treated with surgical elongation of the sternocleidomastoid muscle. This topic is fully described in Chap. 13.

10.3 Klippel-Feil Syndrome/ Brevicollis

Klippel-Feil syndrome (KFS) is characterized by the abnormal fusion of two or more cervical vertebrae. It is a rare disorder with an incidence of 1 in 40,000 births. The cause of most cases of KFS is unknown, but it can be inherited in both an autosomal recessive and an autosomal dominant pattern. The disorder can be associated with other skeletal deformities such as Sprengel's deformity, scoliosis, and hyper-kyphosis. Clinically, the fused vertebrae result in a short neck with a limited range of motion. Patients often appear with a low hairline at the back of the neck. There is no cure for KFS, and treatment is focused on managing symptoms. Some require restriction of activities, while others may require surgery.

10.4 Congenital Spinal Stenosis

Congenital spinal stenosis (CSS) is a narrowness of the spinal canal caused by dysplasia of bony structures. Short pedicle length and smaller distance between pedicles result in a smaller cross-sectional spinal canal area. The prevalence is 2–4%. The etiology of CSS is unknown. The symptoms are mostly observed in the fourth and fifth decades of life with neurogenic complaints, which are accompanied by degenerative changes of the spine. Symptomatic cases can be treated with surgical decompression with or without fusion as in acquired spinal stenosis.

10.5 Sacralization (Lumbosacral Transitional Vertebrae)

Sacralization is a common deformity that includes both lumbarization of the most superior sacral segment and sacralization of the lowest lumbar segment. Most often, the L5 fuses either fully or partially on either side of the sacrum, or on both sides. The prevalence in the general population has been reported at 4–36%.

The cause of sacralization is not known; there may be a genetic predisposition. Figure 10.1 shows a full, bilateral, sacralization of the L5 to the sacrum in conjunction with other congenital deformities of the spine.

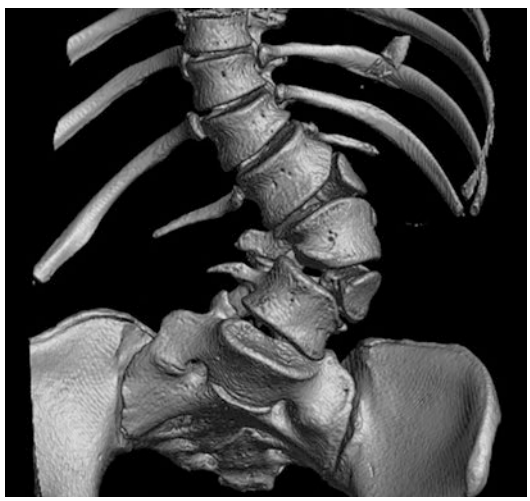

Fig. 10.1 A three-dimensional CT reconstruction of the lumbar spine of a 14-year-old patient with congenital scoliosis. The sinistro-convex scoliotic curve has a Cobb angle close to 80°. Notice the fully segmented hemivertebrae at the L1 and L3 levels on either side of the L2 curve apex. The T12, L2, and L4 vertebrae show partial deformation. Furthermore, a full bilateral sacralization of the L5 to the sacrum is present

An anterior-posterior X-ray of the lumbar spine is needed for diagnosis. The disorder is often asymptomatic and may be recognized when treating other spinal disorders.

10.6 Congenital Spondylolisthesis/Dysplastic Spondylolisthesis

It is a rare disorder characterized by insufficiency of the lumbosacral articulation and disc complex, resulting in ventral displacement of the L5 vertebra. Posterior elements are intact (no spondylolysis). Severe dysplastic spondylolisthesis is more common among girls. Genetic predisposition is a significant causative factor. Dysplastic spondylolisthesis, unlike isthmic spondylolisthesis (with spondylolysis), presents with a high occurrence of nerve root pressure owing to the intact lamina and patients are at risk of neurologic impairment in the presence of only minor degrees of slip. Symptoms usually present during adolescence with low back pain and abnormal gait secondary to hamstring shortness. Radiating pain below the knee or cauda equina symptoms are suggestive of high-grade spondylolisthesis. Diagnosis is based on clinical assessment and lateral X-rays of the lumbosacral junction. Low-grade spondylolisthesis is usually treated with nonsurgical management. High-grade spondylolisthesis and those with neurological impairment are generally treated with fusion with or without postural reduction and decompression.

10.7 Spina Bifida

Spina bifida is a general term describing defects that occur when there is incomplete closure of the spine and the membranes around the spinal cord. Spina bifida may be found in all areas of the spine but is most common in the lumbosacral area. There are three main types: spina bifida occulta (prevalence 12%), meningocele, and myelomeningocele, the latter two with a combined incidence of 0.1–5 per 1000 births. Insufficient intake

of folic acid during pregnancy is a key risk factor. Occulta is the least severe type, usually only involving defects in the posterior bony structures, and presents with no or mild symptoms. A hairy patch or hyperpigmented skin area might be present over the defect.

Myelomeningocele, also known as open spina bifida, is the most severe type of spina bifida. Clinical findings often include paraplegia, poor ability to walk, impaired bladder and bowel control, and hydrocephalus. Diagnosis may occur either before or after a child is born. Before birth, it is done by blood tests, amniocentesis, or ultrasound. Standard surgery for meningocele and open spina bifida is surgical closure after delivery to prevent further damage of the nervous tissue and infection.

10.8 Congenital Scoliosis

Congenital scoliosis is the presence of an abnormal curvature of the spine at birth. Hemivertebra, Fig. 10.1, is the most common cause of congenital scoliosis. The incidence is 3 in 1000 births. The etiology of congenital scoliosis is likely multifactorial and poorly understood.

There are two main causes of congenital scoliosis

1. Failure of formation of portions of one or more vertebra
2. Lack of segmentation of parts of the vertebral column

Both causes can occur at multiple levels and sides of the vertebral column, separately or in combination. The deformities often lead to asymmetrical growth responsible for the preliminary intrauterine development of scoliotic curvature. Congenital scoliotic curvature in the frontal plane is often seen with concurrent hyper-kyphosis or hyper-lordosis in the lateral plane.

Congenital scoliosis is highly associated with other congenital abnormalities (60%). Genitourinary anomalies are observed in 20% of congenital scoliosis and heart defects in 10% of cases. Intraspinal abnormalities are found in 18% of congenital scoliosis.

Diagnosis is based on clinical history taking and medical imaging. Full-spine digital or biplanar X-rays are standard imaging options. CT scan might be needed for preoperative assessment of 3D curvature and vertebral morphology. Full-spine MRI is mandatory before surgical treatment to rule out intraspinal abnormalities.

Treatment consists mainly of observation and surgical intervention including removal of deformed vertebrae, most commonly one vertebra, in combination with posterior instrumented fusion. Surgery is often performed in early childhood to avoid secondary curve formation as most curves progress.

10.9 Sprengel's Deformity

This disorder is characterized by a high and rotated scapula, due to a failed fetal migration of the scapula to the posterior surface of the thorax. The disease is predominantly hereditary. It is more common in girls. The child has usually no complaints. The thoracoscapular motility is reduced, while the humeroscapular motility is usually normal. The indication for orthopedic surgical treatment is mainly cosmetically based and is reserved for patients with pronounced deformity. It should be done before the child is 5 years old and consists of a release of the abnormal thoracic-scapular musculature from the scapula and repositioning. Klippel-Feil syndrome must be ruled out as it is associated with Sprengel's deformity.

10.10 Congenital Dislocation of the Radial Head

This is the most common congenital elbow disorder, and in more than 50% of cases, it is accompanied by other malformations. The condition is due to lack of embryonic development of radial head. It is bilateral in 30% of cases.

Clinically, the congenital dislocation is quite asymptomatic. The condition usually manifests itself in toddlers because of limitations in range of motion. Secondarily, cubitus valgus may develop. Radiographic examination gives the

diagnosis. In the child, treatment is rarely indicated in the congenital condition, but if the dysplastic caput in the adult gives rise to nuisances, it can be excised.

10.11 Congenital Proximal Radioulnar Synostosis

Congenital elbow joint synostoses are rare. Most common is synostosis between the proximal part of the radius and the ulna. Elbow flexion and extension are free, but the rotational movements are severely restricted, and usually the hand is pronated or hyper-pronated. Radiographic examination gives the diagnosis.

Treatment is usually not indicated, except in the bilateral cases if the position of the hand causes disability. Corrective rotational osteotomy can optimize the fixed position of hand in cases with excessive pronation.

10.12 Polydactyly and Syndactyly

An extra finger (polydactyly) (2 per 1000 infants) is the most common congenital deformity of the hand. Certain types are with autosomal heredity.

Often, the extra finger is attached to the hand with soft tissue that contains only neurovascular structures. Best cosmetic results are obtained by surgical removal rather than strangulation. More rarely, a partially fully developed finger with joints and tendons is seen, which may require more advanced reconstruction.

In syndactyly, the fingers have not separated in fetal life and are still fused. If one finger impedes full movement of the other, the separation should take place before the age of 1 year. Otherwise, surgery is performed at the age of 1–2 years. Syndactyly in toes is rarely treated.

10.13 Developmental Dysplasia of the Hip (DDH)

DDH (0.5–1% of infants) is a disorder of abnormal development of the hip joint, the socket is poorly developed, and the head of the femur can be abnormally angled or rotated (Fig. 10.2). These abnormal anatomical conditions change the mechanical forces of the hip joint, which leads to instability and ultimately joint dislocation. Risk factors for DDH are breech position, family history, and female gender.

The DDH is fully described in Chap. 14.

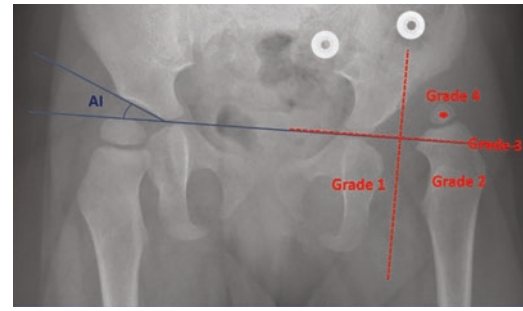

Fig. 10.2 Patient with a dislocated left hip. When acetabular dysplasia and grade of dislocation are analyzed, the first step is to draw the line of Hilgenreiner (blue line) intersecting both triradiate cartilages for reference. Perkin's line is drawn perpendicular to Hilgenreiner's line intersecting the superior acetabular corner (left hip). Acetabular index (AI) is the angle between the Hilgenreiner's line and a line intersecting the superior acetabular corner and the triradiate (right hip). Dislocation is graded according to Tönnis and is assessed by the location of the center of the femoral head (red dot) in relation to Hilgenreiner's and Perkin's lines. In this case, dislocation is 4 (the most severe)

10.14 Congenital Femoral Deficiency

Congenital femoral deficiency (1 in 50,000 births) affects both the femoral bone (shortening, deformity, nonunion) and the soft tissues (ligaments, joints, and muscles) of the femur. Correction of deformities and limb lengthening might be an option, whereas in more severe cases, amputation or conservative treatment might be recommended.

10.15 Fibular Hemimelia

Fibular hemimelia is the most common deficiency involving the long bones. However, it is still a rare disease with an incidence of 1 in 40,000 births.

The etiology is unknown, but the disease is not inherited. In fibular hemimelia, the fibula is underdeveloped or absent and is often associated with lateral disease of the foot, knee, and femur. Depending on the severity of the disease, fibular hemimelia might result in limb length discrepancy, equinovalgus deformity of the ankle and hindfoot, valgus deformity of knee, and absence of lateral toe rays. The main goal of treatment is to restore a plantigrade foot with normal limb length and normal mechanical axes of the lower Boyd. In severe cases, a Boyd amputation of the foot followed by orthoses is recommended.

10.16 Posteromedial Bow of the Tibia

This is a rare, non-hereditary deformity of the bones of the lower leg with a posteromedial curvature. Dysplasia also involves a slight to moderate shortening of the crus. The bone structure itself in the deformed area is normal.

The disorder is usually diagnosed at birth, with the deformity of the crus being conspicuous. In most cases, treatment is not indicated, as the curvature corrects spontaneously during the first years of life, so that the conditions are normalized at the age of 4–5 years. In case of persistent deformity, there may be an indication for a corrective osteotomy.

10.17 Congenital Pseudarthrosis of the Tibia

This is a very rare disease (1 per 140,000 births) caused by a localized pathological periosteum at the tibia and fibula. It will usually be part of neurofibromatosis or fibrous dysplasia. The bones are deformed, often anti-curved (anterolateral curvature). A pathological fracture is present at birth or may occur later, and pseudoarthrosis develops due to poor healing potential.

The treatment is difficult and involves a radical excision of the entire pathologically altered periosteum and reconstruction of the bone defect. Alternatively, amputation is performed.

10.18 Congenital Talipes Equinovarus (Clubfoot)

This is an intrauterine developmental anomaly with a rigid equinovarus deformity of the foot. The congenital clubfoot is one of the more common congenital deformities (1 per 1000 newborns). Genetic factors as well as environmental factors play a role. The clubfoot is bilateral in one-third of cases. Clubfoot is fully described in Chap. 15.

10.19 Metatarsus Adductus/Varus

Metatarsus adductus is a congenital deformity (1 per 1000 newborns) with isolated malposition in the tarsometatarsal joints, so that all five metatarsals deviate medially. The cause of the disorder is probably most often intrauterine placement, but there may be an inherited predisposition.

The diagnosis is made at birth. Apart from the cosmetic, the deformity is not symptomatic. There are two types, the postural type (metatarsus adductus), which is due to intrauterine placement, and an actual deformity (metatarsus varus). The postural type is redressed without any resistance to normal position, while the metatarsus varus is more rigid with high medial arch as well as a varus of the heel.

The postural type does not require any treatment and normalizes spontaneously within the first years of life. Metatarsus varus is treated primarily with plaster, splints, or a special corrective boot for 6–12 weeks. Surgery is therefore very rarely indicated and is reserved for metatarsus varus.

10.20 Congenital Vertical Talus

Congenital vertical talus is constituted by an irreducible dorsal dislocation of the navicular on the talus. It is also known as rocker-bottom or Persian foot. Three main characteristics are present: fixed equinovalgus hindfoot deformity and convex sole, medial prominence of the talar head, and abducted and dorsiflexed forefoot. Clinicians

may observe a peg-leg gait and other associated conditions (myelomeningocele, arthrogryposis, spinal muscular atrophy, neurofibromatosis, etc.).

A mild form of this disease is named "oblique talus." In this condition, the navicular reduced with plantar flexion. The proper treatment is observation and occasionally talonavicular pinning and Achilles lengthening.

In the congenital vertical talus, the navicular does not reduce on forced plantar-flexion X-ray lateral view. The proper treatment of congenital vertical talus includes manipulation and casting followed by limited surgery (percutaneous Achilles tendon lengthening). In case of casting failure, surgery needs to be performed at 6–12 months of age.

10.21 Achondroplasia

Achondroplasia (1 in 50,000 births) is transmitted as a dominant gene; however, the majority of cases (80%) are spontaneous mutations of the growth factor gene leading to abnormal endochondral ossification. Achondroplasia is fully described in Chap. 10.

10.22 Hereditary Multiple Exostoses (Hereditary Multiple Osteochondromas)

Multiple hereditary exostoses (1 in 50,000 births) are transmitted as a dominant gene resulting in benign bone tumors. The multiple osteochondromas primarily grow outward from the growth plates of the long bones but also arise from the scapula, spine, or pelvis. Synostoses between the fibula and tibia, as well as between the radius and ulna, result in deformities at the ankle and wrist. Bony deformities, restricted joint motion, and entrapment of nerves (particularly the peroneal nerve) indicate surgery. The risk of malignant transformation is about 5% and is only seen in adult patients. MRIs might be indicated for larger lesions with rapid progressive growth or axial lesions, making clinical examination difficult.

Take-Home Message

- Congenital deformities of the appendicular and axial skeleton are rare diseases.
- The etiology is often multifactorial as in clubfoot but can also be inherited.

Summary

- Congenital deformities of the appendicular and axial skeleton are rare diseases. Some diseases mainly affect the skeleton (such as achondroplasia), while others (such as Marfan's syndrome) have impact on both the skeleton and internal organs. Congenital deformities can result from generalized skeletal dysplasia as in achondroplasia, leading to shortening of all limbs.
- Infantile torticollis usually manifests itself in the first year of life with a skewed position of the head. The cause may be an abnormality in the cervical spine but is most often due to pathological changes in the sternocleidomastoid muscle possibly due to a compromised blood supply.
- Klippel-Feil syndrome is characterized by the abnormal fusion of two or more cervical vertebrae.
- Congenital spinal stenosis is a narrowness of the spinal canal caused by dysplasia of bony structures.
- Sacralization is a common deformity that includes both lumbarization of the most superior sacral segment and sacralization of the lowest lumbar segment.
- Congenital spondylolisthesis/dysplastic spondylolisthesis is a rare disorder characterized by insufficiency of the lumbosacral articulation and disc complex, resulting in ventral displacement of the L5 vertebra.
- Spina bifida is a general term describing defects that occur when there is incomplete closure of the spine and the membranes around the spinal cord. Spina bifida may be found in all areas of the spine but is most common in the lumbosacral area.

- Congenital scoliosis is the presence of an abnormal curvature of the spine at birth. Hemivertebra is the most common cause of congenital scoliosis.
- Sprengel's deformity is characterized by a high and rotated scapula, due to a failed fetal migration of the scapula to the posterior surface of the thorax.
- Congenital dislocation of the radial head is the most common congenital elbow disorder, and in more than 50% of cases, it is accompanied by other malformations.
- Fibular hemimelia is the most common deficiency involving the long bones. The fibula is underdeveloped or absent and is often associated with lateral disease of the foot, knee, and femur.
- Congenital pseudarthrosis of the tibia is a very rare disease caused by a localized pathological periosteum at the tibia and fibula.
- Congenital talipes equinovarus (clubfoot) is an intrauterine developmental anomaly with a rigid equinovarus deformity of the foot.
- Metatarsus adductus is a congenital deformity with isolated malposition in the tarsometatarsal joints, so that all five metatarsals deviate medially.
- Achondroplasia is transmitted as a dominant gene; however, the majority of cases (80%) are spontaneous mutations of the growth factor gene, leading to abnormal endochondral ossification.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Mention an inherited congenital deformity
   (a) Multiple hereditary exostoses
   (b) Adolescent idiopathic scoliosis
   (c) Blount disease
   (d) Perthes
2. Mention a congenital deformity that is most often monolateral:
   (a) Fibular hemimelia
   (b) Polydactyly
   (c) Syndactyly
   (d) Clubfoot
3. What are the risk factors for DDH?
   (a) Breech position, family history, and female gender
   (b) Breech position, family history, and male gender
   (c) Male gender, second born
   (d) Alcohol abuse during pregnancy
4. What is the treatment of clubfoot?
   (a) Ponseti's method with manipulation of the feet and plaster treatment. Hereafter brace treatment until the age of 4
   (b) Ponseti's method with manipulation of the feet and plaster treatment. Hereafter brace treatment until the age of 1
   (c) Surgical treatment
   (d) Conservative treatment with manipulation and observation
5. What is the main reason for functional impairment in achondroplasia?
   (a) Short arms resulting in problems with personal hygiene
   (b) Cosmesis
   (c) Psychological problems
   (d) Short arms resulting in problems with social relations

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Neuro-Orthopedic Pathologies: Myelomeningocele

11

Dmitry Popkov and Sergey Ryabykh

Overview

Myelomeningocele is the most common open neural tube defect characterized by failure of the neural tube to close in the thoracic or lumbosacral region during the fourth week of embryonic development and leading to the herniation of the meninges and spinal cord through a vertebral defect.

11.1 Definition and Epidemiology

Myelomeningocele (MMC) or open spina bifida (SB) is a severe congenital malformation of the central nervous system. MMC is the most common open neural tube defect characterized by failure of the neural tube to close in the thoracic or lumbosacral region during the fourth week of embryonic development and leading to the herniation of the meninges and spinal cord through a vertebral defect. Myelomeningocele is the most common presentation of spinal dysraphism and constitutes about 80% of the cases. MMC has an estimated birth prevalence of 3.4–4.6 cases per 10,000 live births.

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11.2 Etiology

MMC has a multifactorial etiology. Up to 16% of them are associated with a single gene or chromosomal anomalies (trisomy 18, Meckel-Gruber syndrome, Roberts, VATER syndrome, and X-linked neural tube defects among others). Maternal alcoholism and smoking and elevated glycemic index increase the risk of MMC. Low maternal folate and methionine intake and low levels of zinc, vitamin C, and vitamin B12 also increase its risk. Various folic acid antagonists like valproic acid methotrexate have been linked to MMC.

11.3 Pathogenesis of Orthopedic Disorders

The neurological level of lesion is an essential factor influencing ambulation, functional motor level, secondary orthopedic complications, treatment, outcome, and ultimate prognosis. Hip, ankle, and foot deformity in MMC patients results from muscle imbalance and paralysis around the joints and may present as contracture, subluxation, or dislocation. Hip dislocation can lead to pelvic obliquity and compensatory spinal abnormality. Deformity at the knee joint results from static forces of positioning, fibrosis of surrounding muscles, and muscle imbalance. Both ambulatory and nonambulatory patients with

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MMC frequently develop torsional deformities of the lower extremities involving the femur and/or the tibia. The femoral torsion present at birth in all newborns does not decrease normally with growth in a child SB due to abnormal gait and muscle disbalance. External tibial torsion is secondary and can be caused by muscle imbalance.

11.4 Classification

Thoracic level: Patients do not have active movement of the lower extremity muscles and have some abduction, external rotation, and flexion contracture of the hips.

Upper lumbar level (L1, L2): The active flexion of the hip is preserved, but the functional walking abilities are often lost with adult body mass. A contracture of the unopposed hip flexors and hip adductors typically develops in these patients that predisposes to hip dislocation.

Mid-lumbar level (L3, L4): These patients typically have normal strength in the hip flexors, knee extensors, and adductors but no function in hip extensors or abductors. Therefore, a flexion contracture of the hip and limitation of abduction frequently develop in these patients. This pattern of muscle imbalance also predisposes to the progressive subluxation of the hip. Prognosis for walking is positive, but the patients with L4 level of involvement have the most potential benefit for walking ability from orthopedic management of musculoskeletal disorders.

Low-lumbar level (L5): These patients retain quadriceps and medial hamstring function but lack function of the gluteus medius and maximus. Mostly, they community ambulation as adults. The typical orthopedic issues are represented by foot deformities.

Sacral level (S1–S3): Patients demonstrate both quadriceps and gluteus medius function. This group is subdivided into high-sacral and low-sacral level of involvement, distinguished by the presence of gastrocnemius-soleus strength in low-sacral patients. Patients with high-sacral level walk using ankle orthoses. Low-sacral level patients have no need for braces for independent walking.

The **diagnosis** of orthopedic disorders in MMC is clinical and based on physical examination (including active and passive range of motion, muscle weakness, markers of spinal dysraphism), observation of functional and walking abilities, neurological examination (identification of the level of paralysis), and visual and instrumented gait analysis. The Functional Mobility Scale (FMS) is a useful tool to describe functional mobility. It allows practical scoring of mobility over three distinct distances representing mobility in the home (5 m), at school (50 m), and in the community (500 m). Radiology (radiographs, scanner) contributes to establishing the diagnosis and needs for orthopedic corrections for joint dislocation, torsion femur or tibia deformity, or foot deformities and to monitoring spinal balance (kyphoscoliosis or lordoscoliosis). MMC is often associated with other defects such as Arnold-Chiari malformation, hydrocephalus, and primary or secondary syringomyelia. Evaluation by pediatric neurologist, urologist, and/or multidisciplinary team will be beneficial to diagnosis and complex treatment program.

11.5 Treatment

Orthopedic care of the child with spina bifida is made challenging by the presence of multiple medical comorbidities: central nervous system involvement, insensate skin, and bowel and bladder incontinence. Orthopedic care should be provided as a part of complex treatment by a multidisciplinary team (neurosurgery, urology, and physiatry). Complications in this population may relate to latex allergy or increased risk of postoperative infection.

Patients with MMC require the use of orthoses for ambulation. Orthoses are indicated for various purposes: maintenance of lower limb alignment and upright standing, prevention of deformity, progressive correction of mild contractures, facilitation of independent walking and self-care, and protection for the insensate limb.

Hip surgery implies proximal femoral osteotomy (mostly, femoral varus derotational osteotomy and pelvic osteotomy) along with open reduction and capsular plication in order to achieve and maintain reduction of paralytic hip dislocations. Another approach is based on main-

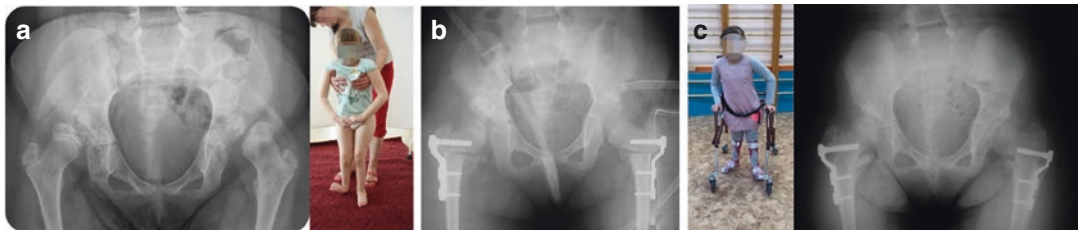

Fig. 11.1 A 9-year-old girl, L2–L3 level lesion: (**a**) before surgery, bilateral hip dislocation, secondary acetabular dysplasia, calcaneovalgus foot deformity, impaired standing; (**b**) staged bilateral proximal femoral osteotomy, capsular plication and acetabuloplasty; (**c**) in 12 months after surgery, improved self-standing abilities and centered hips

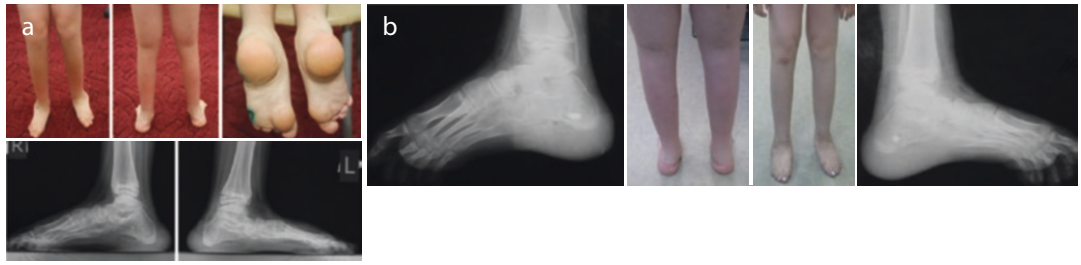

Fig. 11.2 Severe thoracolumbar kyphoscoliosis in an 8-year-old child, thoracic level lesion: (**a**) before surgery, Cobb angle over 110°; (**b**) after surgery, balance restored, posterior instrumentation

taining hip range of motion with contracture release only aimed to improve hip range of motion and the ability to ambulate.

Flexion knee contractures that interfere with ambulation or with transfers or sitting balance should undergo the knee flexor release including the hamstrings, gastrocnemius, and posterior capsule or even supracondylar extension osteotomy.

Torsional deformities for functional (for functional and sitting abilities) and cosmetic reasons (both in ambulatory and nonambulatory patients should be considered for surgery.)

Surgeries for clubfoot, equinus, vertical talus, cavus/varus/cavovarus, calcaneus/calcaneovalgus foot deformities, ankle valgus, and hindfoot valgus are multiple and depend on age and functional prognosis. They may include all spectrum of techniques (Fig. 11.1): tendon transfers, joint release, musculotendon lengthening, corrective osteotomies, and arthrodesis.

Spine procedures (Fig. 11.2) involve most frequently vertical expandable prosthetic titanium rib device application followed by posterior or anterior fusion with instrumentation.

Pressure ulcers (Fig. 11.3) and/or osteomyelitis of the underlying bone represent a specific issue in MMC orthopedic treatment. It requires staged surgeries involving debridement/fistulectomy and foot deformity correction with external fixators and plastic intervention (myocutaneous flap/local plastic) in the majority of cases. Another problem in SB patients is fractures following orthopedic surgery. They are related to disuse secondary osteoporosis.

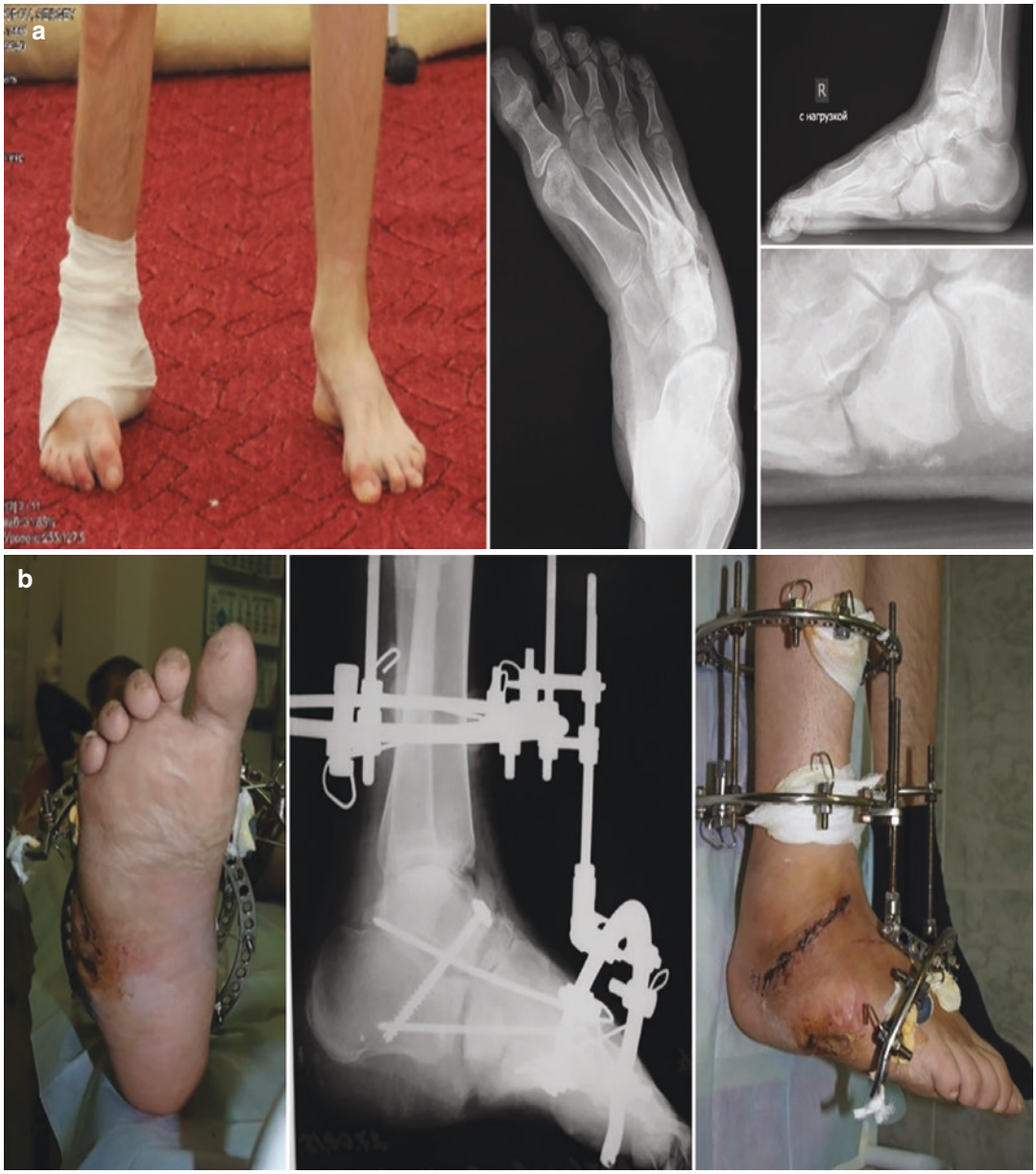

Fig. 11.3 A 18-year-old patient, L5-level lesion: (**a**) before surgery, varus-supination foot deformity, pressure ulcer over lateral mid-foot and osteomyelitis of the cuboid bone; (**b**) Ilizarov frame applied for gradual foot deformity correction followed by debridement and triple-foot arthrodesis, ulcer healed

Take-Home Message

Orthopedic complications in MMC patients are primarily related to neurological deficiency and muscle imbalance. The primary and major aim of surgery is favorable functional outcome. Orthopedic surgery in MMC patients is challenging and has high complication rate in postoperative period due to neurological lesion and multiple medical comorbidities.

Summary

Orthopedic complications in MMC patients are primarily related to neurological deficiency and muscle imbalance. The neurological level of lesion is an essential factor influencing prognosis for ambulation, functional motor level, secondary orthopedic complications, and outcome. Surgery in MMC should be aimed at functional result with regard to standing, gait improvement, prevention of skin pressure/ulcers, improvement of sitting ability, self-care, and facilitation of orthotic management. Indications for surgery should be approved by the multidisciplinary team, and there should be consensus on the postoperative rehabilitation plan.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Myelomeningocele is a congenital malformation of the central nervous system.
   (a) a congenital malformation of the central nervous system
   (b) an acquired malformation of the central nervous system
   (c) a congenital malformation of the peripheral nervous system
   (d) an acquired malformation of the peripheral nervous system
2. Orthopedic complications in SB children are
   (a) Related to muscle spasticity
   (b) Related to muscle imbalances
   (c) Linked to nerve dysfunction
   (d) Caused only by joint hypermobility
3. Which level of lesion is more severe?
   (a) Upper lumbar levels
   (b) Sacral levels
   (c) Thoracic levels
   (d) Lumbar levels
4. Specific issues of MMC are
   (a) Pressure ulcers and/or osteomyelitis of the underlying bone
   (b) Progressive walking impairment
   (c) Visual disturbances
   (d) Hearing loss
5. The failure of what muscle(s) in upper lumbar lesions predisposes to hip dislocation?
   (a) m.semimembranosus and m.semitendinosus
   (b) m.rectus femoris and m.gracilis
   (c) m.gluteus medius
   (d) m.adductor magnus

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Neuro-Orthopedic Pathologies: Cerebral Palsy

12

Dmitry Popkov and Sergey Ryabykh

Overview
Cerebral palsy refers to the spectrum of permanent movement and posture disorders resulting from perinatal nonprogressive damage of the central nervous system in early childhood. It is the most prevalent physical disability originating in childhood.

12.1 Definition and Epidemiology

Cerebral palsy (CP) is the most prevalent physical disability originating in childhood. This condition refers to the spectrum of permanent movement and posture disorders resulting from perinatal nonprogressive damage of the central nervous system in early childhood. The incidence of CP is of 2–3.6 cases per 1000 live births. The prevalence of CP is strongly associated with prematurity and low birth weight.

12.2 Classification

Depending on the topography of initial brain injury, there are four principal phenotypes of cerebral palsy, namely spastic, dyskinetic, ataxic, and mixed cerebral palsy. The spastic type is the most common form of the disease and accounts for about 80% of all diagnosed cases of CP. Anatomic classification of spastic CP involves spastic diplegia (the lower extremities are primarily involved with mild upper extremity involvement), spastic hemiplegia (which involves one half of the body), and spastic quadriplegia (all four limbs are significantly involved).

Four functional classification systems are used to assess the degree of disabilities (general or specific) in CP children with CP. The Gross Motor Function Classification System (GMFCS) describes gross motor function, especially the ability to walk. The Manual Ability Classification System (MACS) is used to describe the typical use of both hands and upper extremities. Communication Function Classification System (CFCS) is used to describe the ability for daily routine communication. The Eating and Drinking Ability Classification System (EDACS) is used to describe the eating and drinking function.

12.3 Pathogenesis of Orthopedic Disorders

The initial neuropathologic lesion is nonprogressive and over time interferes with normal development affecting the motor impairment. The motor difficulties may represent muscle spasticity, motor planning difficulties, and dystonia. The muscle

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spasticity causes muscle shortening, tendon contractures, bone deformities, joint subluxation, and dislocation resulting in permanent pathological gait pattern and/or abnormal posture. These secondary progressive musculoskeletal conditions require orthopedic surgical interventions.

The **diagnosis** of CP is clinical, including neurological and orthopedic examinations (goniometry, assessment of the spasticity) and visual and instrumented gait analysis. Radiology (radiographs, scanner) contributes to establishing the diagnosis and needs for orthopedic corrections (for example, Reimers index over 40% measured on pelvic radiograph justifies hip reconstruction surgery).

12.4 Treatment

The indications to perform goal-oriented orthopedic surgery are to improve function; prevent deformity; decrease pain from joint dislocation or subluxation; improve gait pattern; prevent skin pressure areas; improve sitting position, cosmesis, and hygiene; and facilitate orthotic management. In all cases, the surgeon should work closely with a team which might include physical and occupational therapists, neurologists, and physiatrists. Only conditions caused by permanent muscle retraction (shortening) should be considered for orthopedic surgery. Orthopedic surgery is contraindicated in early age (less than 5 years old) for motor disabilities due to only spasticity, especially in cases classified as I–III GMFCS levels.

A common practice consideration is to perform most procedures as a single event multilevel orthopedic surgery (SEMLS). SEMLS refers to the correction of all musculoskeletal deformities producing gait deviations in one session. SEMLS is a complex orthopedic surgical intervention using a combination of muscle tendon lengthening, tendon transfers, osteotomies, and/or joint stabilization procedures. It is usually based on detailed clinical assessment and instrumented gait analysis in ambulatory patients. SEMLS must be combined with an intensive postoperative rehabilitation program including appropriate bracing, strengthening, and gait training. SEMLS is not a uniform procedure and should be tailored to the CP patient's needs.

1. Musculotendinous lengthening involves a lengthening of the musculotendinous unit at the musculotendinous junction or through the fascia as a recession. Other procedures like Z-shape tendon lengthening or percutaneous fibromyotomies may be deleterious to the muscle and result in weakness.
2. Tendon transfers are used to take a muscle which is spastic and contributing to a deformity and to reposition it to perform joint balancing or serve as a functional transfer.
3. Osteotomies are indicated to manage deformities of the bones (for example, increased femoral anteversion and internal or external tibial torsion) resulting in alteration of the joint mechanics and gait abnormalities. Reconstructive surgery for hip dislocation (Fig. 12.1) necessitates either femoral varus

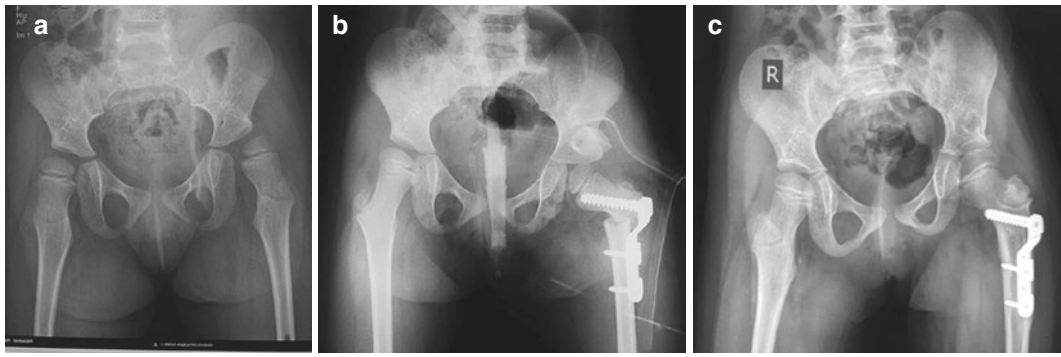

Fig. 12.1 Hip dislocation in a 6-year-old CP girl, GMFCS IV: (**a**) before surgery, Reimers index 54% in left hip; (**b**) femoral varus derotational osteotomy was associated with acetabuloplasty and adductor lengthening and iliopsoas release; (**c**) in 3 years after surgery, operated hip develops normally

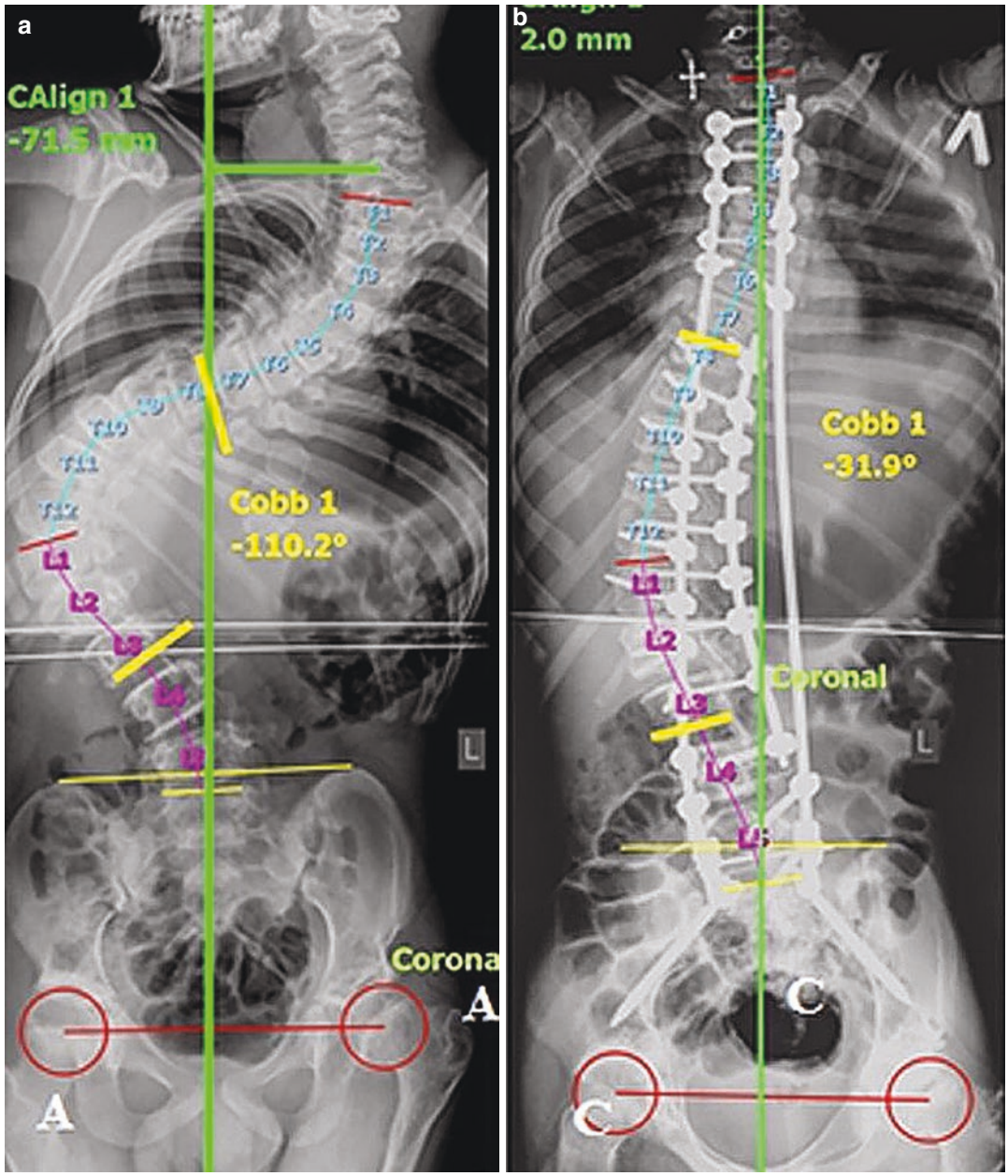

Fig. 12.2 Severe thoracolumbar scoliosis in a 15-year-old child, GMFCS V: (**a**) before surgery, Cobb angle 110°, severe frontal disbalance; (**b**) after surgery, balance restored, posterior instrumentation with pelvic fixation

derotational osteotomy or pelvic osteotomy, or both. Multiple deformities at the foot and ankle, which may interfere with ambulation, should be treated with osteotomies of the tibia and fibula, hindfoot, or forefoot to align the foot.

4. In some cases, arthrodesis is necessary to place a segment (i.e., foot) in an optimum position. The best outcomes in fusion in CP include arthrodesis of the first metatarsal phalangeal joint of the hallux. The Grice procedure (extra-articular fusions of the subtalar joint) is still used to help stabilize the hindfoot. The most common arthrodesis is a spine fusion (Fig. 12.2) to treat the scoliosis in older children with spastic quadriplegia.

Take-Home Message
Orthopedic disorders in CP are secondary and progressive. Surgery does not represent the primary approach in the treatment of cerebral palsy. Indications for surgery should be approved by the multidisciplinary team.

Summary
Secondary progressive orthopedic complications in CP children result over time in severe motor disabilities, including impaired gait, self-care, etc. Orthopedic surgery has an important role in the management of children and adults with cerebral palsy. It should be performed by orthopedic surgeons with experience and expertise in neuro-orthopedics. The principal aim of the surgery is to restore initial functional abilities injured and lost due to secondary orthopedic complications. Indications for surgery should be approved by the multidisciplinary team, and there should be consensus on the postoperative rehabilitation plan.

Questions
Multiple correct answers are possible. Answers available in the book back matter.

1. Cerebral palsy
   (a) is a stable encephalopathy
   (b) is not a stable encephalopathy
   (c) is due to genetic abnomalies
   (d) is only due to ischemic hypoxia
2. Orthopedic complications in CP children are
   (a) Primary and present since birth
   (b) Progressive and secondary
   (c) a+b
   (d) None of the previous
3. What appears first?
   (a) Muscle spasticity
   (b) Muscle shortenings
   (c) Fatigue
   (d) Muscle hypotrophy
4. Are percutaneous fibromyotomies recommended in young children?
   (a) Yes
   (b) No
   (c) Not at any age
   (d) Only in older patients
5. What value of Reimers index is the threshold for hip reconstructive surgery in CP children?
   (a) 35%
   (b) 50%
   (c) 75%
   (d) 90%

Further Reading

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Congenital Muscular Torticollis

13

Magdalena Maria Oledzka

Overview

Congenital muscular torticollis is one of the most common congenital musculoskeletal conditions. It is observed at birth or in infancy and characterized by a persistent head tilt toward the involved side with the chin rotated to the contralateral shoulder. Conservative physical therapy management can achieve excellent outcomes. In rare instances, surgical release of the SCM muscle may be warranted.

13.1 Introduction

Congenital muscular torticollis (CMT) is one of the most common congenital musculoskeletal conditions. The term "torticollis" comes from the Latin words *tortus* = twisted and *collum* = neck and refers to the clinical sign of an ipsilateral head tilt with a contralateral rotation. Other terms used in literature are "wry neck" and "twisted neck" (Fig. 13.1). It is observed at birth or in infancy and characterized by a persistent head tilt toward the involved side with the chin rotated to the contralateral shoulder due to the shortening or fibrosis of the involved sternocleidomastoid muscle (SCM). For example, right torticollis describes a child with the head tilted to the right and rotated to the left in a resting posture. Atypical presentations are possible, but rare, with a head tilt and rotation to the same side. The incidence of CMT has been on the rise and is reported in between 3.9% and 16% of infants, with a slightly higher prevalence in males. It is often accompanied by cranial deformation, and less frequently by developmental dysplasia of the hip (DDH), club foot, and metatarsus adductus.

The Academy of Pediatric Physical Therapy of the American Physical Therapy Association published an updated evidence-based clinical practice guideline (CPG) on Physical Therapy Management of Congenital Muscular Torticollis in 2018. The guideline provides instructions on screening, referral, monitoring, examination and evaluation, prognosis, first-choice treatment interventions, and discharge criteria. It is intended as a reference for healthcare professionals and families of infants with CMT to maximize outcomes for this specialized population. The CPG also introduced an eight-level severity grading system for CMT (CMT-SCS) that will be described later in the chapter.

Conservative physical therapy management of CMT, especially when cases are identified early and treatment is initiated immediately, can

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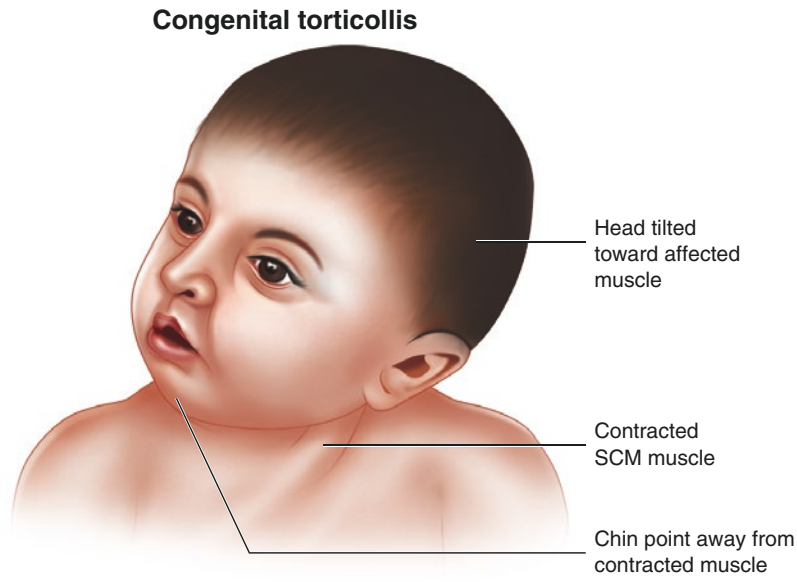

Fig. 13.1 Congenital muscular torticollis

achieve excellent outcomes in 90–99% of cases. In rare instances, surgical release of the SCM muscle may be warranted.

13.2 Etiology

There are several theories on the etiology of CMT and little agreement. The most accepted theories include an ischemic event during delivery, birth trauma, and intrauterine malpositioning. Higher incidence of CMT has been documented with breech presentation, oligohydramnios, difficult delivery, use of forceps during delivery, longer babies, and multiple pregnancy. An increased incidence of developmental dysplasia of the hip and metatarsus adductus in infants with CMT further supports the theory of intrauterine crowding as a likely cause. More recent immunohistochemical studies support an intrauterine maldevelopment of the fetal SCM theory.

Milder forms of CMT are often noted later in the first few months of life and have been linked to the introduction of the *Back to Sleep Campaign* in the early 1990s by the American Academy of Pediatrics and similar programs initiated in Europe. These programs were designed to decrease the incidence of sudden infant death syndrome (SIDS). However, while the success of these programs helped decrease the incidence of SIDS, it also correlated with an unintended increase in the incidence of cranial deformation and CMT.

13.3 Imaging and Pathophysiology

Ultrasonography is the preferred type of diagnostic imaging for infants with CMT as it is noninvasive, inexpensive, and fast. An ultrasonogram is done routinely primarily in the presence of SCM mass, in cases where clinical findings do not correspond with CMT, or when the infant does not respond to physical therapy treatment. Ultrasonographic imaging of the affected SCM may show focal or diffuse enlargement, mild to severe fibrosis, and various levels of atrophy. Based on echogenicity findings, four severity types of fibrosis were defined with type 1 being the mildest. Type 1 is defined as a fibrotic mass within otherwise healthy muscles. Type 2 is characterized by diffuse fibrosis mixing with healthy muscle, while type 3 is typified by diffuse fibrosis with almost no healthy muscle. Infants with

severe fibrosis in the entire SCM muscle (type 4) typically require longer treatment duration and are much more likely to require a surgical intervention than infants with mild fibrosis.

13.4 Cranial Deformation

A distortion of the skull shape often accompanies CMT, reported in up to 95% of CMT cases. This distortion results from prenatal or postnatal external compression forces on the infant's easily moldable skull. As mentioned earlier, since the *Back to Sleep* campaign was initiated, there has been a marked increase in the incidence of postnatal cranial deformation, documented in as high as 18% of infants.

Cranial deformation is classified into three types: plagiocephaly, brachycephaly, and dolichocephaly. **Deformational plagiocephaly** presents as a parallelogram shape of the skull with unilateral occipital flattening often accompanied by ipsilateral shift of the earlobe forward and forehead bossing. **Deformational brachycephaly** is characterized by central occipital flattening without earlobe or facial asymmetries. **Deformational scaphocephaly** is characterized by narrow and long skull shape.

Deformational plagiocephaly coexisting with CMT increases the risk of facial, ear, and mandibular asymmetry. Treatment for milder cases involves repositioning and physical therapy. For more severe cases, cranial orthosis may be prescribed for cranial remodeling (Fig. 13.2a). Cranial orthoses direct cranial growth to fill out in the areas that are flattened and to promote a more symmetrical head shape. Cranial orthosis is most effective when initiated between 4 and 6 months of age due to the rapid brain and skull growth during this period, but it can be used up to

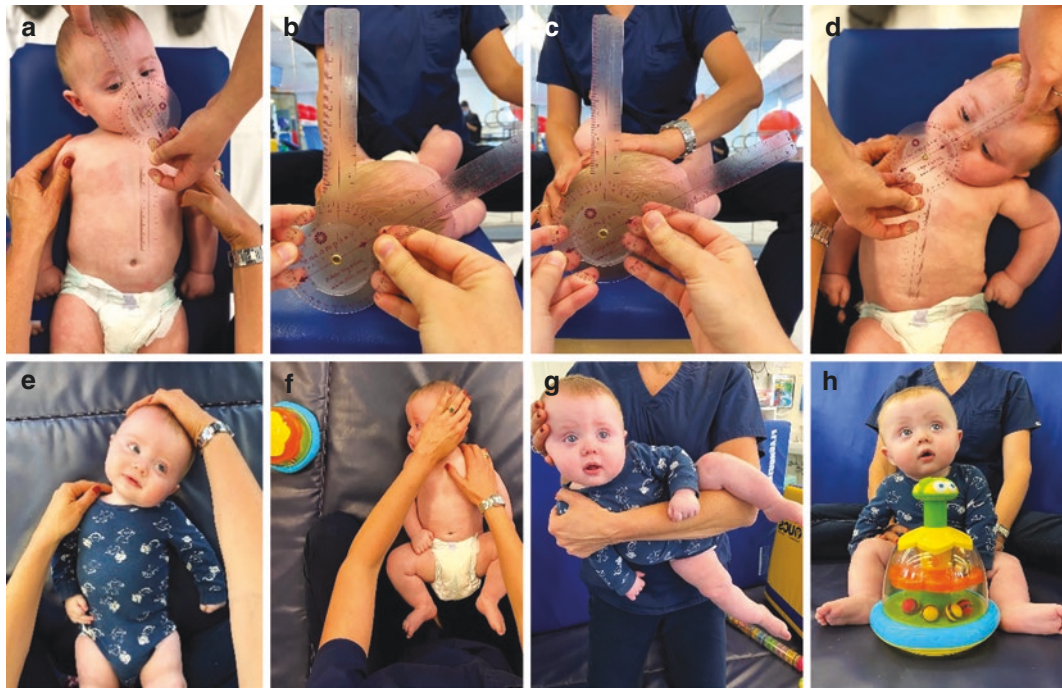

Fig. 13.2 An infant with right CMT. Top row: Cervical measurements in supine using a universal goniometer: (**a**) resting head tilt; (**b**) active rotation to the right; (**c**) passive rotation to the right; (**d**) passive lateral flexion to the left. Bottom row: (**e**) passive cervical lateral flexion stretch of the right SCM; (**f**) passive cervical rotation stretch to the right side; (**g**) side carry stretch of the tight right SCM; (**h**) sitting balance activity with focus on symmetrical weight-bearing through both buttocks; note the right head tilt

15–18 months of age with a longer treatment duration and lesser efficacy. It is vital to distinguish deformational plagiocephaly from craniosynostosis, which is characterized by an abnormal head shape due to the premature closure of one or more of the cranial sutures and requires surgical intervention.

13.5 Differential Diagnosis

The tilted head posture typical of congenital muscular torticollis can be caused by other disorders; therefore, it is critical to rule out other causative factors before arriving at the diagnosis of CMT. Up to 18% of children presenting with a sign of torticollis will have a diagnosis other than CMT. Assembling a comprehensive pregnancy and birth history, combined with a multisystem screening, is paramount. The differential diagnosis process requires taking a detailed history of the onset of the condition (congenital vs. acquired), assessing pain (painful vs. nonpainful), and examining the child's neurological, musculoskeletal, cardiopulmonary, gastrointestinal, and vision systems.

Neurologic causes of torticollis include central nervous system (CNS) tumors, brachial plexus injuries, brain stem and cerebellar gliomas, and agenesis of CNS structures. Red flags at the exam include abnormal posturing, abnormal muscle tone, brachial plexus injury, and pain during movement—these indicate that neurologic consultation is needed. Musculoskeletal conditions that present in infants with associated torticollis include Klippel-Feil syndrome (failure of segmentation of vertebra in cervical spine), clavicle fracture, infantile scoliosis, and atlantoaxial joint rotatory subluxation. Ocular conditions may cause asymmetrical posturing especially noticeable when an infant starts to sit independently. An infant with congenital ocular torticollis tilts and rotates their head to see better due to an imbalance of the extraocular muscles (most caused by paralysis of the superior oblique muscle). Other conditions include ocular apraxia, strabismus, and nystagmus. Sandifer syndrome, a gastrointestinal condition resulting from the hiatal hernia and gastroesophageal reflux (GER) disease, typically causes an infant to tilt their head to the right and arch their back due to discomfort after eating. Reflux and constipation may also cause the abnormal posturing.

Table 13.1 Differential diagnosis: congenital, acute, and developmental disorders with a clinical sign of torticollis

Differential diagnosis	
Skeletal origin	Atlantoaxial rotatory displacement (AARD) due to trauma
	Clavicle fracture
	Congenital scoliosis
	Failure of segmentation/formation
	Hemivertebrae
	Klippel-Feil syndrome
Neurogenic origin	Arnold-Chiari malformation
	Central nervous system tumors
	Ocular torticollis
	Paroxysmal torticollis
	Syringomyelia
Gastrointestinal origin	Sandifer syndrome
Inflammatory origin	Cervical adenitis
	Grisel syndrome (AARD due to inflammation)
	Upper respiratory infection
	Vertebral osteomyelitis and diskitis
Neoplastic origin	Cervical spinal cord tumor
	Posterior fossa tumor

Acquired torticollis occurs in older infants and children. It may be caused by trauma, ocular lesions, benign paroxysmal torticollis, Arnold-Chiari malformation, infections, dystonic syndromes, syringomyelia, and posterior fossa tumors. Sudden onset of torticollis and pain at rest or during gentle cervical range of motion movement are red flags and require immediate attention. See Table 13.1 for a summary.

13.6 Classification of CMT

Congenital muscular torticollis is subdivided into three groups based on clinical presentation: infants with a sternomastoid tumor, muscular torticollis, and postural torticollis. Infants in the sternomastoid tumor (SMT) group present with a palpable fibrotic mass or fibrous band within the

substance of the affected SCM, and restrictions in cervical range of motion (ROM). Fibrotic mass, also known as pseudotumor of infancy, appears in 25–55% of infants with CMT and varies in size from a small pea-size nodule to a large fibrotic nodule involving the whole muscle. Infants in the muscular torticollis (MT) group present with cervical active and passive ROM limitations due to tightness in the SCM but no palpable mass. Lastly, infants in the postural torticollis (POST) group, the mildest form, have a characteristic head tilt due to muscle strength imbalance, but no limitations in the cervical passive ROM. Congenital muscular torticollis can be graded using an eight-grade severity classification system, which is based on the infant's age at examination, the difference in cervical range of motion in passive rotation between sides, and the presence or absence of SCM mass. The CMT-SCS is first dichotomized as "early" (0–6 months) or "later" (older than 6 months). Each age category is separated further into severity grades classified as mild, moderate, and severe.

13.7 Changes in Body Structure and Their Effect on Development

Although the primary impairment in the diagnosis of CMT is the shortening or fibrosis of the involved SCM, there is a ripple effect on the surrounding tissues and the infant's development of early motor skills. Ipsilateral cervical and shoulder muscles (platysma, scalenes, trapezius, cervical extensors, and pectoral muscles), spinal accessory nerve (XI), cervicothoracic fascia, and facial features may be altered as well. An uncorrected head tilt and biomechanical restrictions caused by the imbalanced muscular forces on the developing infant's skeleton may cause secondary sequelae of worsening scoliosis and cranial asymmetries, an altered perception of the midline, and asymmetrical fine and gross motor development. In a developing infant with CMT, each movement is a result of the dynamic interaction among many subsystems such as postural control, muscle strength, perceptual processes (visual, vestibular, proprioceptive), and cognition in response to the body constraints caused by a tight SCM and limited cervical range of motion. Some infants display not only asymmetry of movement of a face, spine, rib cage, and extremities, but also a delay in motor skill acquisition. In most cases, the delay resolves by 10–12 months of age or onset of walking with physical therapy intervention; however, in some infants, it may persist into early childhood and require additional referrals to occupational and speech therapies.

13.8 Rehabilitation

The diagnosis of CMT is typically made by the pediatrician within an infant's first 6 months of life. Early identification and prompt referral to a pediatric physical therapist are critical for the best outcome—minimizing secondary sequelae and resulting in a shorter treatment duration. All infants even with mild symptoms of positional preference, facial asymmetries, cervical range-of-motion limitations, SCM mass, and craniofacial asymmetries should be referred to a pediatric physical therapist as soon as these symptoms are noted. Research has repeatedly shown that earlier intervention shortens treatment duration to achieve full cervical passive and active ranges of motion, reduces secondary impairments, and decreases the need for a surgical intervention. Cervical stretches which are part of a daily home program are easier to administer by parents when infants are younger; older infants are stronger and often less amenable. If left untreated, this condition will persist and most likely progress resulting in more established asymmetries in posture, movement patterns, and musculoskeletal structures.

13.8.1 Examination

A pediatric physical therapist gathers and documents an infant's birth and delivery history; performs a comprehensive examination including screening for nonmuscular causes and examining

body function and structures and developmental status; and determines a prognosis. Cervical measurements of the resting head tilt, and the active and passive cervical range of motion, are taken at the initial visit and at regular intervals to assess progress (see Fig. 13.2a–d). Cervical muscle strength is measured using the Muscle Function Scale (MFS). The test involves holding the infant in vertical suspension and tipping them horizontally to assess the amount of lateral head righting from the horizontal line that the infant can maintain for 5 s in front of a mirror. The MFS scores range from 0 to 4: a score of 0 describes an infant holding their head below the horizontal line, while a score of 4 describes an infant holding their head at least 75 degree above the horizontal line for 5 s or more. Healthy infants typically display symmetrical cervical strength, while infants with CMT show a discrepancy between sides. For example, an infant with right CMT may hold their head higher on the right side as compared to the left side due to overactive right SCM.

13.8.2 Treatment

Conservative, evidence-based physical therapy treatment consists of five interventions: passive cervical stretches, neck and trunk active ranges of motion, facilitation of symmetrical motor skill development, environmental adaptations, and caregivers' education. The frequency of therapy depends on the severity of findings from the initial examination, age of infant at the time of exam, and parents' ability to perform the home program diligently and correctly. Generally, the more severe cases will require a higher dosage of therapy and longer treatment duration.

During each physical therapy session, passive cervical stretches are performed by the therapist after gentle warm-up exercises for infants with cervical range-of-motion limitations (muscular and SMT presentations). The therapist and caregiver can engage the infant via eye contact, gentle speaking, and toys with visual and/or auditory components, all great ways to reassure the infant, keep their attention, and assist with head movement during stretches and active range-of-motion exercises. Engagement with the infant will help make the stretches more enjoyable and less stressful for the baby and caregivers. The three basic stretches include cervical rotation stretch in supine, lateral flexion stretch in supine, and side carry stretch (Fig. 13.2e–g). In all three stretches, the goal is to maintain a maximally elongated position of the involved SCM muscle into rotation or side flexion while stabilizing the infant's shoulders for a prolonged time (on average about 30 s). With the caregiver's increasing skill level, stretches can be advanced and performed in a variety of positions (supine, prone, supported sitting, or when carried).

Following passive stretches, active rotation of the head and trunk in a new lengthened position will strengthen the weak contralateral SCM muscle and surrounding neck and trunk muscles. Strengthening is achieved by intentional play, handling, positioning, and carrying the infant during daily care to encourage cervical rotation to the involved side and head and trunk righting on the opposite side. In addition, therapists facilitate symmetrical fine and gross motor development, minimize unbalanced sensory development due to side preference, and monitor for delays in acquisition of motor skills (Fig. 13.2h). Promoting prone positioning during awake time for infants (tummy time) is one of the staples of pediatric physical therapy treatment; it is especially important in infants with CMT and cranial deformities to minimize cranial deformation and strengthen the infant's posterior neck and trunk muscles.

Caregivers' education in handling the infant, repositioning, and environmental adaptations is not to be underestimated as it is the caregivers who are primarily responsible for daily exercises. Therapists teach caregivers methods of gentle cervical stretches, active strengthening exercises, handling and carrying techniques, and environmental adaptations. Parents demonstrate the exercises to the provider during physical therapy sessions to support their learning and mastery of these skills.

For infants who are slower to respond or with more severe presentations, there are supplemental physical therapy interventions available with less robust evidence. **Kinesiologic taping** to the

neck has been used by therapists to facilitate and/or inhibit muscles in the neck. The **tubular collar for torticollis** (TOT collar) can be used as an adjunct for therapy in recalcitrant cases of CMT (Fig. 13.3b). It is designed to decrease head tilt posture, assist in the maintenance of midline head control, and promote movement away from the side of tilt. The TOT collar should only be used with infants who have sufficient cervical range of motion and head control. It is worn only during active play or feeding time to prevent habitual tilting and increase times spend in midline. The use of **microcurrent**, a low-intensity, single-channel alternating current, applied superficially to the affected SCM has been promising. In recent years, two well-designed randomized controlled trials demonstrated a significant decrease of treatment duration in a group of infants who received microcurrent therapy in addition to traditional physical therapy intervention.

13.9 Nonconservative Interventions

In cases where the physical therapy intervention fails to achieve full resolution or in older children who began intervention after the age of 1 year, more invasive interventions may need to be considered. Botulinum toxin (BOTOX), a neurotoxin that acts as a neuromuscular block to reduce an overreactive muscle activity, thereby causing muscle atrophy, has been used as an adjunct to physical therapy. Botox may be injected into the affected SCM as well as the surrounding musculature. The injection creates a window of opportunity to strengthen cervical musculature on the opposite side with the goal of rebalancing musculature strength and achieving lasting midline position.

The surgical management of CMT typically involves older children with recalcitrant CMT due to severe fibrosis of the involved SCM. Indications for surgery include failure of traditional treatment, age 1 year or older, ROM limitations in the cervical range of motion of more than 15^0, a persistent visible head tilt, and persistent SCM mass [1]. Children with type 4 fibrosis are 13.4 times more likely to require surgery than those with type 1 fibrosis.

There is no established agreement among pediatric orthopedic surgeons on the optimal timing and technique for SCM release. Reported results in children younger than 3 are excellent; however, children treated surgically in the first year of life often require a second surgery because of recurrence related to hematoma or failure to comply with the daily home exercise program and bracing. Most surgeons agree that the optimal time is between 1 and 8 years of age. Surgical approaches fall into three categories: tendon lengthening, unipolar release of the clavicular attachment of SCM (distal), or bipolar release of both SCM muscle attachments (proximal and distal) (Fig. 13.3c).

From a developmental point of view, children with recalcitrant CMT have a prolonged experi-

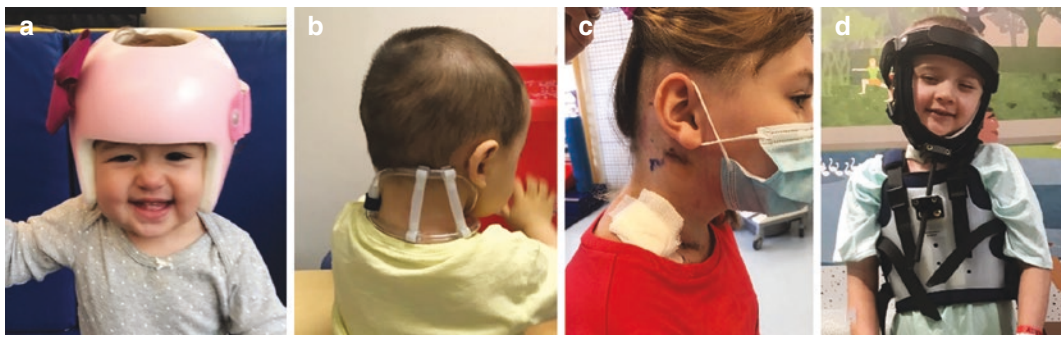

Fig. 13.3 (**a**) Infant wearing a cranial orthosis to correct plagiocephaly; (**b**) child wearing a TOT collar during play; (**c**) s/p few days after right SCM bipolar release; (**d**) child in a pinless halo s/p right SCM bipolar release

ence in asymmetrical posture without established midline, which may result in asymmetrical development of the visual, vestibular, and proprioceptive systems. After surgical release of the tight SCM, a physical therapy program with daily home exercise program and use of a brace to stabilize the neck and head in a new lengthened position are required for a successful outcome and to minimize recurrence of the tilt. There are several brace options used after surgery. Some surgeons use a soft cervical collar right after surgery, and some transition children to a pinless halo, to stabilize the head and neck in a slightly overcorrected position with the head laterally flexed slightly toward the unaffected side and rotated toward the involved side (opposite of the habitual position) for several weeks after surgery as the soft tissues heal (Fig. 13.3d). Rehabilitation guidelines are divided into three phases. Phase 1 is short in an acute postsurgical, hospital setting. It is followed by phase 2 in an outpatient rehabilitation clinic focused on gaining symmetrical cervical passive ROM, improving cervical strength, gaining symmetrical strength throughout the body, and establishing the midline. Phase 3 focuses on functional maintenance of the midline as the cervical bracing is discontinued. Postsurgical treatment duration ranges in literature from 4 to 6 weeks up to 11 months.

13.10 Outcomes

Infants with CMT benefit from early diagnosis and expeditious referral to pediatric physical therapy. It is well researched and documented that infants who start physical therapy early have better outcomes and shorter treatment durations, which minimizes stress on the caregivers and financial burden on the family and insurance payers. Infants are discharged from physical therapy when they achieve full passive and active cervical ranges of motion, when they can maintain head in midline 95% of the time, and when they have age-appropriate and symmetrical fine and gross motor skills. Parents are encouraged to continue to monitor their child as they grow, since occasional transient recurrence of tilt is possible. At times, short bouts of therapy may be needed over the next few years as the child grows to help rebalance the neck and trunk musculature.

Take-Home Message

- The broad term "torticollis" refers to the clinical sign of ipsilateral head tilt with contralateral rotation.
- Congenital muscular torticollis is subdivided into three groups based on clinical presentation: infants with a sternomastoid tumor, muscular torticollis, and postural torticollis.
- Once a diagnosis of CMT is made, an expeditious referral to a pediatric physical therapist specializing in the treatment of infants is warranted.
- Conservative physical therapy management of CMT achieves excellent outcomes in more than 90–99% of cases.
- If not addressed, this condition will most likely persist and even progress, resulting in more established asymmetries in posture, movement patterns, and musculoskeletal structures.

Summary

The broad term "torticollis" refers to the clinical sign of ipsilateral head tilt with contralateral rotation. Carefully analyzing possible differential diagnoses by attaining a thorough medical history including pregnancy and delivery and a clinical exam are imperative to make the correct diagnosis. Once a diagnosis of CMT is made, an expeditious referral to a pediatric physical therapist specializing in the treatment of infants is warranted. Conservative physical therapy management of CMT achieves excellent outcomes in more than 90–99% of cases. Prognosis varies based on the infant's age at initiation of treatment, severity of cervical range-of-motion restrictions, and presence of SCM mass. Earlier diagnosis, mild restriction in cervical range of motion, and absence of SCM mass correspond with faster and more complete resolution of

symptoms. If not addressed, this condition will most likely persist and even progress, resulting in more established asymmetries in posture, movement patterns, and musculoskeletal structures. Surgery is indicated for more severe cases with diffuse fibrotic involvement of the entire SCM muscle. Postsurgical physical therapy, use of a brace to stabilize neck in a new lengthened position, and family dedication to a daily home program ensure successful outcome of the surgery.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. The incidence of CMT ranges from:
   (a) 4–16% of infants
   (b) 16–20% of infants
   (c) 1–4% of infants
   (d) 4–12% of adolescents
2. Surgery for CMT is indicated:
   (a) For more severe cases with diffuse fibrotic involvement of the entire SCM muscle
   (b) For mild cases with localized involvement of the SCM muscle
   (c) In each case of CMT
   (d) As first-choice treatment for CMT
3. Conservative treatment:
   (a) Consists of five interventions: passive cervical stretches, neck and trunk active ranges of motion, facilitation of symmetrical motor skill development, environmental adaptations, and caregivers' education
   (b) Consists of three interventions: neck and trunk active ranges of motion, facilitation of symmetrical motor skill development, and environmental adaptations
   (c) Is not indicated in mild cases
   (d) Consists of two interventions: neck and trunk active ranges of motion and caregiver's education
4. The preferred diagnostic imaging for infants with CMT is:
   (a) Ultrasonography
   (b) MRI
   (c) X-ray
   (d) CT scan followed by MRI
5. CMT could be divided into:
   (a) Three groups: infants with a sternomastoid tumor, muscular torticollis, and postural torticollis
   (b) Three groups: adolescent with a sternomastoid tumor, muscular torticollis, and postural torticollis
   (c) Two groups: infants with a sternomastoid tumor and postural torticollis
   (d) Two groups: Muscular torticollis and postural torticollis

Further Reading

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14 Developmental Dysplasia (DDH) and Congenital Dislocation of the Hip (CDH)

Renata Pospischill and Sebastian Farr

Overview
Developmental dysplasia of the hip encompasses a wide spectrum of pathology ranging from a complete fixed displacement of the femoral head from its central position in the acetabulum at birth to asymptomatic acetabular dysplasia with an impaired ossification of the lateral epiphysis of the acetabulum.

14.1 Definition

Developmental dysplasia of the hip (DDH) encompasses a wide spectrum of pathology ranging from a complete fixed displacement of the femoral head from its central position in the acetabulum at birth (*congenital dislocation of the hip—CDH*) to asymptomatic acetabular dysplasia with an impaired ossification of the lateral epiphysis of the acetabulum (*DDH*).

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14.2 Epidemiology

The epidemiologic literature regarding DDH is vast and confusing due to different definitions of hip dysplasia, different methods of diagnosis (e.g., physical exam, plain radiographs, sonography), and different ages of the population studied.

On the basis of numerous studies, the incidence of DDH in Central Europe is published to be from 2% to 4% and from 0.5% to 1% for congenital dislocation of the hip. In other areas of the world, the condition of hip dislocation is virtually nonexistent. The reported incidence in Chinese and African newborns is almost 0%, whereas it is 1% for hip dysplasia. Due to the fact that in primitive Asian and African tribes newborns are traditionally carried by their caregivers on the side or the back with spread legs, hip dysplasia might be almost absent in these populations. This carrying tradition keeps the hips in an optimum position for stability and the developing acetabulum.

It is known that there is a prevalence of unilateral disease and the most affected side is the left side. The ratio of female:male is 4:1.

14.3 Etiology and Pathogenesis

The etiology of DDH is multifactorial, involving genetic and hormonal (***endogenous***) and/or intrauterine environmental (***exogenous***) factors (Table 14.1).

UG. Longo, V. Denaro (eds.), *Textbook of Musculoskeletal Disorders*,
https://doi.org/10.1007/978-3-031-20987-1_14

Table 14.1 Published risk factors for DDH and dislocation of the hip

Risk factors for DDH or dislocation of the hip	
Endogenous factors	Positive family history Female gender
Exogenous factors	Breech position Lower limb deformity (e.g., clubfoot, metatarsus adductus, flat feet) Muscular torticollis Facial asymmetry Oligohydramnios Other significant musculoskeletal abnormalities

A positive family history for DDH may be found in 12–33% of affected patients. General joint hypermobility is an evident condition associated with hip instability at birth. Among these, female patients are predominantly affected (female:male ratio of 12:1). This is thought to be because of the greater susceptibility of girls to maternal relaxin hormone, which increases ligamentous laxity.

Any condition that leads to a tighter intrauterine space and, consequently, less room for fetal movements may be associated with DDH. This is increasingly observed in cases of breech position or oligohydramnios (female:male ratio of 2:1). Other intrauterine molding abnormalities, such as muscular torticollis and foot deformities (e.g., metatarsus adductus), lend support to the theory that mechanical factors play a role in the pathogenesis.

14.4 Normal Hip Development

At birth, the acetabulum consists of a bony and cartilaginous portion. The cartilaginous portion is composed of the hyaline cartilage of the acetabular roof and the fibrocartilaginous acetabular labrum.

The hyaline cartilage of the acetabulum is made up of epiphyseal cartilage and functions as a major growth plate, not only in the shape of a hemisphere within the roof but also in the shape of a "Y" within the triradiate cartilage, which divides the three parts of the pelvis (ilium, ischium, and pubis).

The fibrocartilaginous labrum is the most peripheral part of the acetabulum and bridges the acetabular incisura as the transverse ligament.

In neonates, the femoral head, greater trochanter, and proximal portion of the femoral neck are of hyaline cartilage. These structures are separated from the bony shaft by the epiphyseal plate (chondro-osseous junction), and development occurs through a combination of appositional and epiphyseal growth. Within the center of the femoral head, the proximal femoral ossification center appears at approximately 5–7 weeks after birth (seen by sonography) and continues to enlarge until adult life.

The normal development of the hip depends on a tight growth fit between the femoral head and the acetabulum. Any damage to the cartilaginous epiphyseal area may lead to growth disturbance.

14.5 Hip Development in DDH

Developmental dysplasia of the hip (DDH) is a gradually progressive disorder of the hip that occurs in different forms at different ages.

In case of a hip dysplasia present at birth due to either endogenous or exogenous factors, a delayed ossification of the lateral acetabular epiphysis results in an inadequate contouring of the acetabular roof. The most common pathological change in severe dysplastic hips is a hypertrophied ridge of acetabular cartilage in the posterior, inferior, and superior aspects of the acetabulum. The labrum is everted.

If dysplasia is not treated, the femoral head starts to dislocate secondarily from its central position as a result of increasing extension of the hip during the first months after birth. In hips that remain dislocated, the acetabular roof becomes more oblique and the concavity flattens. The acetabulum is filled with fatty tissue known as pulvinar, the transverse ligament thickens, the teres ligament elongates, and the inferior capsule of the hip develops an hourglass shape. These changes are secondary barriers and may impede successful reduction.

The displacement progress is usually in a cranio-dorsal direction. As a consequence, the iliopsoas tendon and the gluteal muscles are shortened and the affected hip develops a flexion

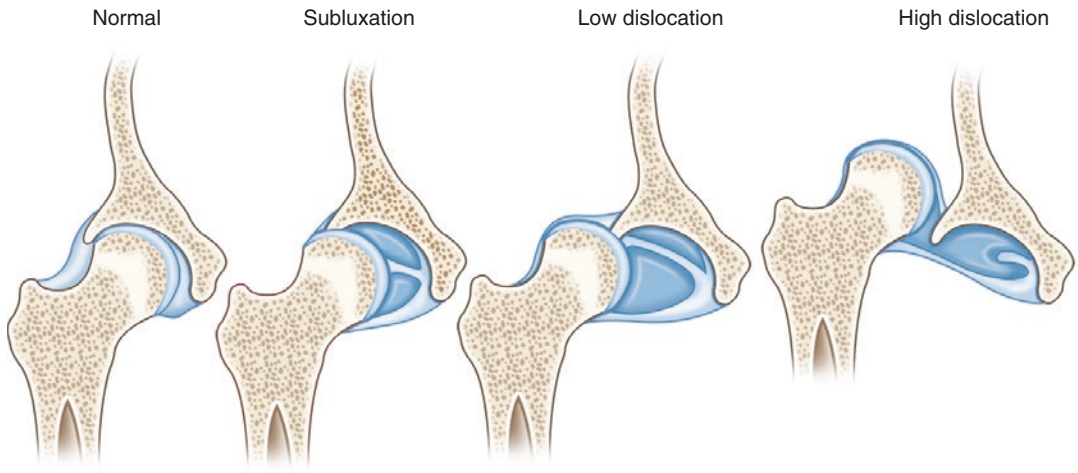

Fig. 14.1 Different stages of hip dysplasia

contracture. This leads to an abnormal pelvic tilt combined with a hyperlordosis of the lumbar spine in a walking child (Fig. 14.1).

Most abnormalities in DDH are on the *acetabular side*. Changes on the *femoral side* are secondary to abnormal pressure on the femoral head from the acetabulum or ilium during dislocation and may lead to avascular necrosis of the femoral head.

The introduction of general ultrasound screenings of neonates uncovered a high proportion of immature hips at birth (approx. 30%) that do not require treatment and usually resolve spontaneously. Those hips need to be monitored until they are fully matured and the complete ossification is caught up.

If acetabular dysplasia heals up during subsequent growth and treatment in severe cases, there remains the risk that residual acetabular dysplasia arises again during the pubertal growth spurt.

14.6 Diagnosis

14.6.1 Clinical Diagnosis

Physical examination of the neonate must be carried out with the infant placed supine in a comfortable and warm setting.

In case of unilateral dislocation, when **inspecting** the neonate, shortening of the thigh (*Galeazzi's sign*) is best appreciated by placing both hips in 90° of flexion and comparing the height of the knees. Increasingly visible thigh folds on the affected side could be present, but an asymmetry of *skin folds* is a common normal variant and may not necessarily indicate hip dislocation.

When **palpating** the hip, dislocation in the neonate is diagnosed by eliciting the Barlow or the Ortolani sign (Fig. 14.2).

Both tests begin with the hips and knees flexed to 90°.

The *Barlow test* assesses the potential for dislocation of a hip. Holding the patient's thigh in adduction, the examiner applies a posteriorly directed force. The test is positive, if the hip dislocates during the maneuver.

The *Ortolani test* is an attempt to reduce a dislocated hip (a reverse of the Barlow test). The examiner holds the patient's thighs and gently abducts the hip while lifting the greater trochanter with two fingers. The test is positive, if the dislocated femoral head slips into the acetabulum with a palpable "clunk."

An additional sign of DDH in the neonate or infant is *limited abduction* as a clinical manifestation of adductor muscle shortening. In the walking child, a *limb-length discrepancy*, a *limp*, a *waddling gait*, a *Trendelenburg sign*, and a *hyperlordosis of the lumbar spine* are noticed.

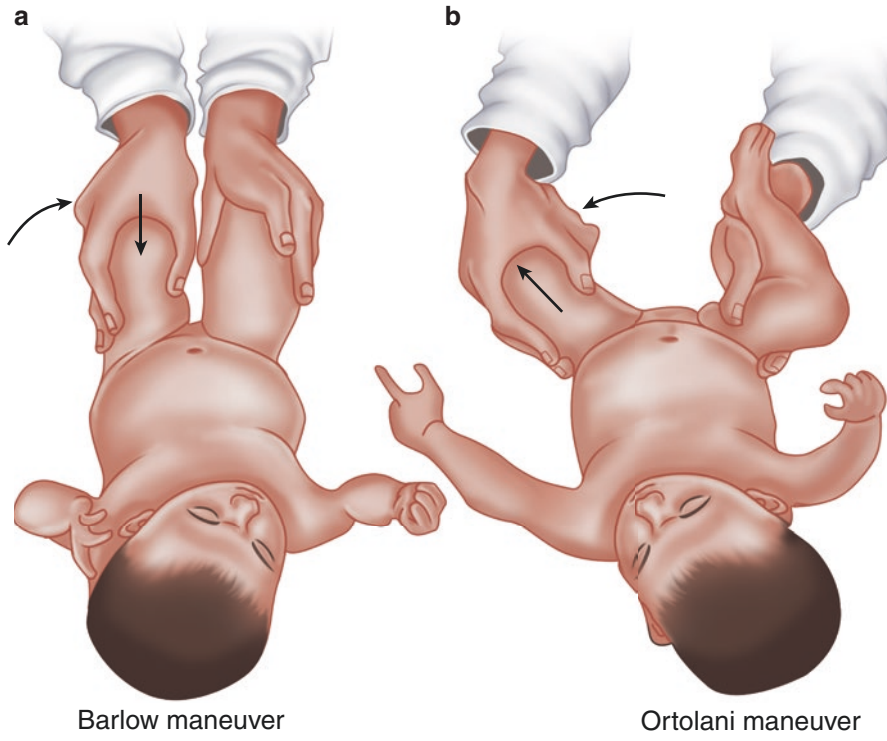

Fig. 14.2 Testing for the Barlow (**a**) and the Ortolani sign (**b**)

14.6.2 Hip Sonography in the Neonate

In the 1980s, **Graf** developed a **sonographic screening technique** for the infant hip younger than 6–8 months, when the acetabulum and the proximal femur are predominantly cartilaginous and thus not visible on plain radiographs. In this age group, these cartilaginous structures are best visualized with ultrasound and therefore DDH could be diagnosed earlier and more precisely.

The morphological assessment, as described by Graf, focuses primarily on the evaluation of the acetabular development and the anatomical characteristics of the hip joint. This is accomplished by measuring two angles on the ultrasound image achieved from a lateral approach:

The α angle, which measures the slope of the superior aspect of the bony acetabulum

The β angle, which evaluates the cartilaginous part of the acetabulum

With the α and β angles, the neonate hip is classified into *four types* (differentiated according to the α angle) and *several subtypes* (differentiated according to the age and β angle) taking into consideration the steepness of the bony acetabulum, the maturation of the cartilaginous acetabulum, the centering of the femoral head, and the age of the patient (Fig. 14.3):

Type I hips (Ia and Ib) are normal. Ia and Ib are differentiated according to the β angle.

Type II hips are either immature (IIa, IIa+, and IIa−) or dysplastic and stable (IIB, IIc stable) or dysplastic and unstable (IIc unstable, D). IIa, IIa+, IIa−, and IIb are differentiated according to age and IIc and D according to the β angle.

Type III and type IV hips are dislocated and differentiated according to the density (IIIa and IIIb) or the position (IV) of the cartilaginous part of the acetabulum.

Hip sonography in the neonate is used to monitor acetabular development during abduction treatment. The number of radiographs can be

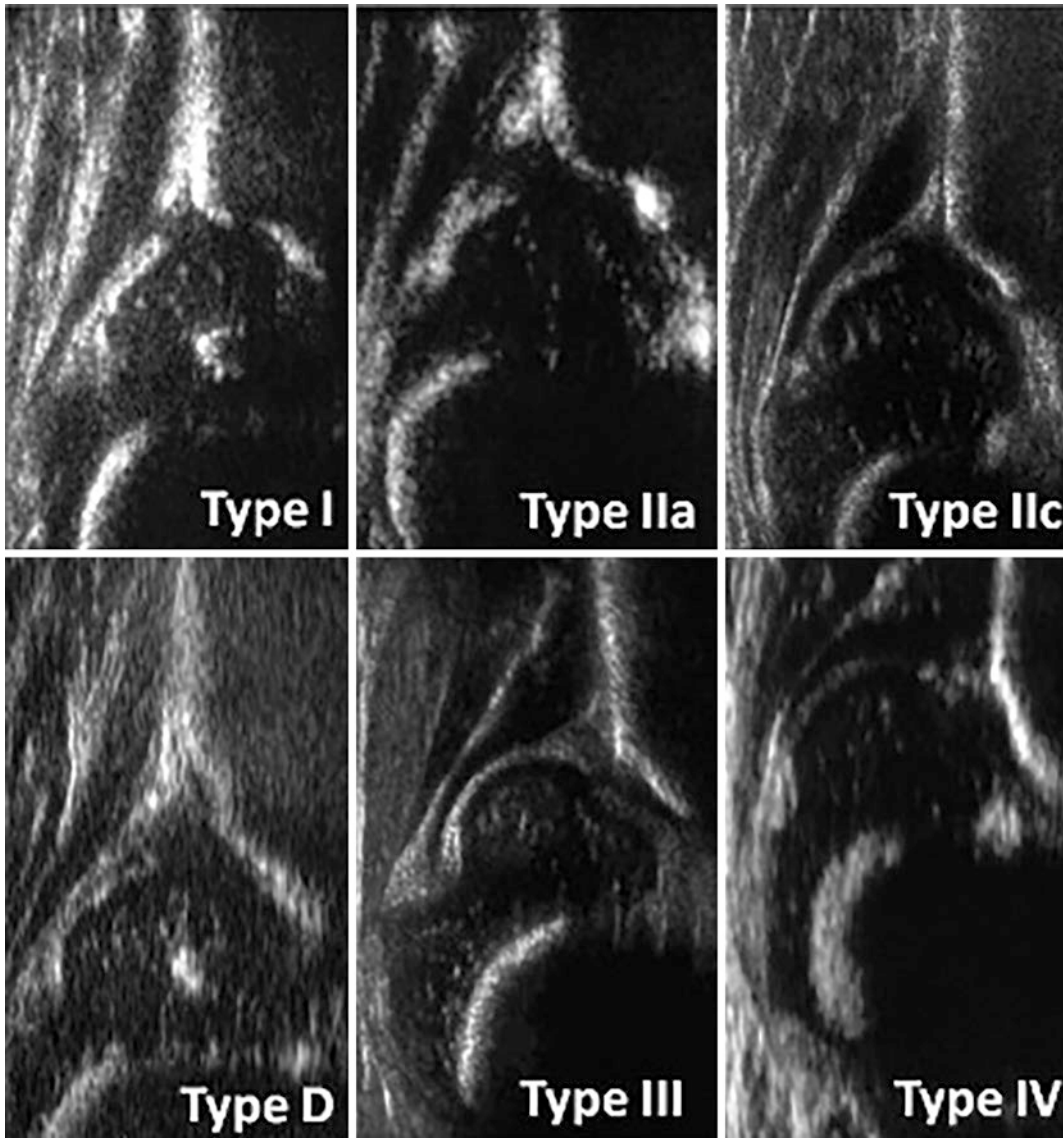

Fig. 14.3 Ultrasound examples of the infant hip types according to Graf

minimized, and the clinician is able to detect failure of treatment earlier.

A problematic aspect of this technique is that this is a static examination with a focus on morphological assessment and that reproducibility of the β angle measurements was not high. However, the classification system shows high agreement, and thus, Graf's method has a substantial value as morphological examination of the neonate hip.

In Central Europe, **general ultrasound screening** for all neonates is established for early detection of DDH. Several studies describe the cost-effectiveness of early treatment and, as a consequence, show low rates of surgeries in infants (open reduction of hip dislocation) in countries with a general ultrasound screening program. On the other hand, early screening uncovers a high percentage (approx. 30%) of immature hips at birth (type IIa), which usually recover spontaneously but need to be monitored until full maturation (type Ia or Ib).

If general screening is not available, the hip sonography should at least be performed in neonates with presenting risk factors (Table 14.1) or clinical suspicion of DDH.

14.6.3 Radiographic Diagnosis

Radiographs are recommended for an infant once the proximal femoral head ossifies, usually after 6 months. At this age, the AP view is normally recorded.

An AP view of the pelvis with both hips can be interpreted by using several classic lines:

The *Hilgenreiner's line* is a horizontal line drawn through the top of the triradiate cartilage.

The *Ombrédanne's line* is a vertical line drawn through the most lateral ossified margin of the acetabulum and is perpendicular to the Hilgenreiner's line. Four quadrants are formed, and in a normal hip, the femoral head is in the lower inner quadrant. In a high hip dislocation, the center of the femoral head moves to the upper outer quadrant.

The *Shenton's line* is a curved line drawn from the medial aspect of the femoral neck to the superior border of the obturator foramen. In normal hips, the Shenton's line is a continuous contour. In a dislocated hip, the line is "broken" because the femoral head is displaced upwards in the X-ray. Then the line consists of two separate arcs.

The *acetabular index (AC angle)* is the angle formed between Hilgenreiner's line and line drawn joining the triradiate cartilage and the most lateral ossified margin of the roof of the acetabulum. At birth, the AC angle is up to 30°, at 1 year no more than 25°, and at 3 years under 20°.

14.6.4 Arthrography of the Hip

Hip arthrography remains valuable for visualizing the result of a closed reduction and the position of the femoral head below the cartilaginous part of the acetabulum after a hip dislocation. Soft tissue obstacles to reduction in front of the femoral head are evaluated. This can also be useful prior to an attempted open reduction to assess any reduction obstacles.

14.7 Treatment

The main goal in the management of DDH is to obtain and maintain a concentric reduction of the femoral head within the acetabulum to provide a best fit of these two components within the joint for normal hip development. Observations on healthy infants have shown that the bony acetabulum grows in a specific way. The potential for maturation is greatest during the first 6 weeks of life and begins to level out after the end of the third month. The later the diagnosis, the more difficult it is to achieve reduction and normal development, as conservative treatment is not successful in a walking child and the potential for acetabular and proximal femoral remodeling decreases with age. Residual acetabular dysplasia remains. A therapeutic strategy for congenital dislocation of the hip according to age is established (Table 14.2).

Table 14.2 Management of congenital dislocation of the hip according to age

Treatment strategy of congenital hip dislocation	
<1 year	Closed reduction (Pavlik harness, traction) Immobilization (spica cast, Pavlik harness) for 6 weeks Maturation (Pavlik harness, abduction splint) until normal acetabular development
Age 2	Preliminary overhead traction Successful closed reduction Spica cast for 6 weeks Maturation (abduction splint) until normal acetabular development
	Failed closed reduction Open reduction and femoral shortening osteotomy Concomitant acetabular procedure in a child older than 18 months Spica cast for 6 weeks Maturation (abduction splint) until normal acetabular development
Ages 3–8	Open reduction with femoral shortening (and derotation) osteotomy and concomitant acetabular procedure (Pemberton or Salter) Hip cast for 6 weeks
>8 years	No reduction attempt

14.7.1 Conservative Treatment

After determining the patho-anatomical situation of the bony and cartilaginous parts in the infant's hip joint by ultrasound, an adequate therapy correlated to these findings is followed and three stages of treatment are defined:

The **reduction** is needed in all decentered hip joints (type D, III, and IV according to Graf). This is possible manually, through a reduction device (e.g., Pavlik harness) or through traction.

The *Pavlik harness* is made of shoulder and lower leg straps and places the legs in the so-called human position and safe zone (hips in flexion of 110° and abduction of 45°). The dislocated hips reduce themselves spontaneously as a result of the baby's movements. A precondition for a successful reduction with the Pavlik harness is an infant with normal motor skills. In case of a dislocated hip with additional adductor contractions, prior to reduction, an adductor tenotomy or traction is applied.

By *overhead traction*, reduction (closed reduction) is achieved spontaneously. A weight of around 15% of the baby's body weight is attached to the infant's legs for traction. The hips are positioned in a flexion of more than 90°, and abduction is increased gradually up to 60° after 10 days.

During the reduction phase, excessive abduction of more than 60° should be avoided to decrease the risk of developing an avascular necrosis of the femoral head.

The **retention** treatment (immobilization phase) must be applied after successful reduction. During this phase, the still unstable hip joint has the goal to maintain reduction. The deformed cartilage of the acetabulum ossifies and becomes congruent with the femoral head, the stretched joint capsule shrinks, and normal hip development starts. Any retention device that provides the hips to be placed in at least 100° of flexion and a maximum abduction of 45° for at least 4 weeks is suitable. Various plastic splints (Tuebinger splint, Rosen splint, Frejka pillow) as well as the Pavlik harness and at least a hip spica cast fulfil these requirements. During the retention/immobilization phase, the baby is not allowed to struggle or to extend the legs in order to prevent redislocation.

The **maturation** phase starts in all joints that have completed the retention phase as well as in stable but dysplastic hips with incomplete ossification of the acetabulum (type IIc stable, IIb, IIa−). The maturation device (e.g., Pavlik harness) maintains the retention position while allowing an increasing amount of movement in the hip joint. The squatting position is needed until they are completely healed and the hyaline cartilage is ossified to type I.

14.7.2 Surgical Treatment

Treatment of DDH in children after walking age is more challenging and usually requires surgery:

The **open reduction** is needed regularly in teratologic hip dislocation or only if closed reduction is unsuccessful in the first year. After the second year, the longer the hip is dislocated, the more likely it is that secondary changes in the hip joint impede a deep centering of the femoral head.

A concomitant **femoral shortening osteotomy** is needed in a high dislocation to reduce abnormally high pressure after open reduction.

A **femoral intertrochanteric varus and derotation osteotomy** is usually performed due to increased anteversion of the femoral neck to improve joint stability after open reduction.

A concomitant **pelvic osteotomy** is recommended with an open reduction, because the potential for acetabular development is markedly diminished. In children older than 2–3 years, the most common procedure accompanying open reduction is innominate osteotomy as described by *Pemberton (acetabuloplasty)* or by *Salter* (Fig. 14.4).

14.7.3 Sequelae and Complications After Treatment

Following reduction of a dislocated hip, the acetabulum remodels in response to the position

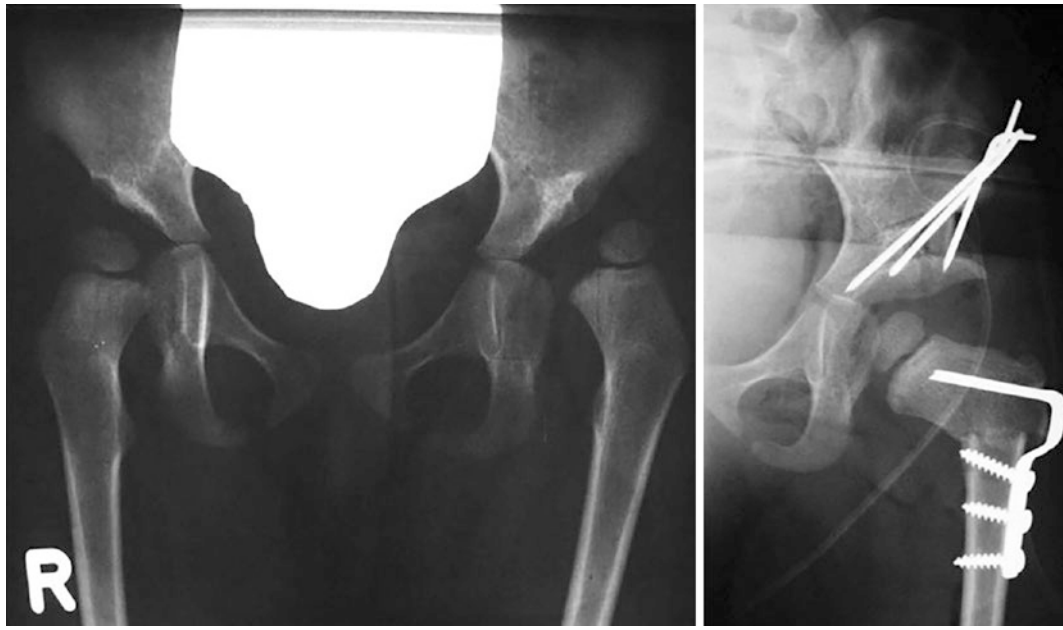

Fig. 14.4 AP X-ray in a 22-month-old child with unilateral hip dislocation of the left hip. An intertrochanteric varus derotation osteotomy and a Pemberton acetabuloplasty were performed

and pressure of the centered femoral head. There is good evidence that if the acetabular dysplasia is still present at 5 years after treatment, further spontaneous remodeling will be insufficient. **Residual acetabular** as a sequela remains, and a pelvic osteotomy should be performed to ensure adequate development of the hip.

Avascular necrosis (AVN) occurs after the treatment of DDH when excessive abnormal pressure is applied for an extended period to the femoral head. The diagnosis is made when the femoral head fails to ossify or to grow within the first year after reduction. Some of the anatomical changes of AVN are a major cause of long-term disability and lead to early degenerative joint disease.

Take-Home Message

- Developmental dysplasia of the hip (DDH) is a spectrum of gradually progressive disorders of the hip that occur in different forms.
- Most abnormalities in DDH are on the acetabular side.
- The white race and the female sex are more frequently associated with the condition, and the risk factors include gender, positive family history, oligohydramnios, and breech position.
- Ultrasound examination in infants younger than 6 months is used to monitor the acetabular development during treatment.
- The main goal in the management of DDH is to obtain and maintain a concentric reduction of the femoral head.
- In infants, conservative treatment with reduction braces, splints, plaster casts, or traction methods (closed reduction) is indicated.
- In cases of late diagnosis or if a secondary deterioration after successful treatment occurs, particularly after walking age, reduction of the femoral head is achieved by surgical management.

Summary

Developmental dysplasia of the hip (DDH) encompasses a wide spectrum of pathology ranging from a complete fixed displacement of the femoral head from its central position in the acetabulum at birth to asymptomatic acetabular dysplasia with an impaired ossification of the lateral epiphysis of the acetabulum. On the basis of numerous studies, the incidence of DDH in Central Europe is published to be from 2% to 4% and from 0.5% to 1% for congenital dislocation of the hip. The etiology of DDH is multifactorial, involving genetic and hormonal (endogenous) and/or intrauterine environmental (exogenous) factors. A positive family history for DDH may be found in 12–33% of affected patients. In case of a hip dysplasia present at birth due to either endogenous or exogenous factors, a delayed ossification of the lateral acetabular epiphysis results in an inadequate contouring of the acetabular roof. If dysplasia is not treated, the femoral head starts to dislocate secondarily from its central position as a result of increasing extension of the hip during the first months after birth. In hips that remain dislocated, the acetabular roof becomes more oblique and the concavity flattens. Most abnormalities in DDH are on the acetabular side. Physical examination of the neonate must be carried out with the infant placed supine in a comfortable and warm setting. In the 1980s, Graf developed a sonographic screening technique for the infant hip younger than 6–8 months, when the acetabulum and the proximal femur are predominantly cartilaginous and thus not visible on plain radiographs. The main goal in the management of DDH is to obtain and maintain a concentric reduction of the femoral head within the acetabulum to provide a best fit of these two components within the joint for normal hip development. The potential for maturation is greatest during the first 6 weeks of life and begins to level out after the end of the third month. The later the diagnosis, the more difficult it is to achieve reduction and normal development, as conservative treatment is not successful in a walking child and the potential for acetabular and proximal femoral remodeling decreases with age. Residual acetabular dysplasia remains.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. The incidence of DDH in Europe ranges from:
   (a) 2–4% of infants
   (b) 1–2% of infants
   (c) 10–15% of infants
   (d) 5–10% of adolescents
2. The etiology of DDH:
   (a) Is multifactorial, involving genetic, hormonal, and intrauterine environmental factors
   (b) Is hereditary
   (c) Is due to intrauterine environmental problems
   (d) Is due to hormonal deficiency
3. In case of a hip dysplasia present at birth:
   (a) A delayed ossification of the lateral acetabular epiphysis results in an inadequate contouring of the acetabular roof
   (b) A delayed ossification of the medial acetabular epiphysis results in an inadequate contouring of the acetabular roof
   (c) A delayed ossification of the lateral acetabular epiphysis results in a pathologic alteration of the femoral head
   (d) A delayed ossification of the medial acetabular epiphysis results in a pathologic alteration of the femoral head
4. Ortolani test:
   (a) Is an attempt to reduce a dislocated hip. The examiner holds the patient's thighs and gently abducts the hip while lifting the greater trochanter with two fingers. The test is positive, if the dislocated femoral head slips into the acetabulum with a palpable "clunk"
   (b) Assesses the potential for dislocation of a hip. Holding the patient's thigh in adduction, the examiner applies a posteriorly directed force. The test is positive if the hip dislocates during the maneuver
   (c) Is a shortening of the thigh evaluable by inspection
   (d) Is possible to perform only after the first 3 months

5. Type I of Graf classification:
   (a) Ia and Ib are normal and differentiated according to the β angle
   (b) Are either immature (Ia) or dysplastic and stable (Ib) or dysplastic and unstable (Ic)
   (c) Are dislocated and differentiated according to the density (Ia) or the position (Ib) of the cartilaginous part of the acetabulum
   (d) Represent a pathologic condition

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Congenital Clubfoot

15

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Matthew B. Dobbs, and Pier Francesco Costici

Overview
Clubfoot (also named congenital talipes equinovarus) is the most common inherited musculoskeletal birth defect. While the treatment method requires precision and patience, if done properly, the results are excellent in long term in terms of foot function and mobility.

15.1 Definition

Clubfoot (also named congenital talipes equinovarus) is one of the most common congenital foot deformities. Four primary alterations characterize clubfoot: midfoot cavus, midfoot adductus, hindfoot varus, and hindfoot equinus. If properly managed and diagnosed early, clubfoot reports a successful correction rate.

15.2 Epidemiology

The incidence of congenital talipes equinovarus ranges between 0.5 and 2 cases per 1000 births. A geographical variation influences this data; Hawaiians and Maori reported >6.8 per 1000 births. Males have a double risk compared to females. A bilateral presentation occurs in 50% of cases, while in the case of unilateral disease, the right foot is the most affected.

15.3 Pathogenesis

While the precise etiology of isolated clubfoot is multifactorial, genetic and environmental factors play a role. Maternal smoking and maternal diabetes reported a high correlation with clubfoot. Seasonal variation has also been reported due to a change in maternal temperature during embryonic growth. Genetic factors are responsible for

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many isolated clubfoot deformities, with several genes and genetic pathways identified to date. In the literature, 24.4% of patients have an associated family history, but the inheritance mode has not been recognized. In monozygotic twins, an incidence of 33% has been described, compared to 3% in dizygotic twins. In 80% of patients, clubfoot is an isolated condition; however, in the remaining part of cases, it is associated with other conditions (arthrogryposis, congenital myotonic dystrophy, myelomeningocele, amniotic band sequence, and other genetic syndromes). In 7% of cases, the patients are affected by other congenital anomalies, and in 7.6%, neurological disease is present. The tarsal bone is in a position of maximal flexion and adduction.

The grade of severity ranges from mild postural forms to severe rigid deformities. The talus is in plantar flexion and medially deviated; however, the calcaneus is in varus and is rotated medially around the talus. The navicular and the cuboid are displaced medially. Dense collagen fibers could be observed in structures medially and posteriorly of the talus with a histological exam. The Achilles tendon, tibialis posterior, tibionavicular, and calcaneonavicular ligaments are usually affected.

15.4 History and Physical

The intrauterine ultrasound of the fetus could diagnose the clubfoot. However, in low developed countries, ultrasound screening is not frequently available; therefore, the diagnosis occurs at birth. Midfoot cavus, midfoot adductus, hindfoot varus, and equinus are present with clubfoot. The severity of these deformities could differ depending on the grade of clubfoot. Several classification systems have been proposed in the literature, but the Pirani system is the most commonly adopted due to its reproducibility and validity. This classification system assesses six-foot items (three for the hindfoot and three for the midfoot). These items could be scored from 0 or 0.5 to 1, depending on severity. Therefore, the Pirani system could range from an overall score of 0–6.

The Pirani system has been used in the previous years as a prognostic tool. In fact, higher Pirani scores at the beginning of the treatment are related to a higher number of casts required and increased risk of Achilles tenotomy. However, it has been proved that the Pirani scoring system is not valid as a prognostic tool, but it is purely descriptive. Treatment should be guided by physical exam and not classification systems. The score developed by Dimeglio is more accurate, but the prognostic validity of this classification has not been demonstrated. The most important advantage of Dimeglio classification is the inclusion of a score for muscle function.

In a unilateral presentation, the affected foot is smaller than the other, and a reduction of the calf could be present. A complete evaluation of the patient needs to be performed to exclude other associated diseases. In particular, the back examination is required to exclude the presence of spina bifida or myelomeningocele. Hip assessment should be performed to evaluate the presence of developmental dysplasia (DDH), as patients with clubfoot reported a risk 25 times greater to have DDH. However, recent studies demonstrated that clubfoot is not positional and not related with increased risk of DDH.

15.5 Clinical and Radiological Evaluation

As previously mentioned, the evaluation of clubfoot should begin during the prenatal period with ultrasound and prosecute with clinical evaluation at birth (Fig. 15.1). Foot X-rays are useless in the first postnatal period, as many of the tarsal bones are cartilaginous and should be assessed by radiographs. The only indication for plane X-rays for clubfoot is in previously operated feet or older kids, in whom the need for bony reconstruction is assessed.

Metatarsus varus could be confused with congenital clubfoot, but this is a benign form that does not usually require treatments.

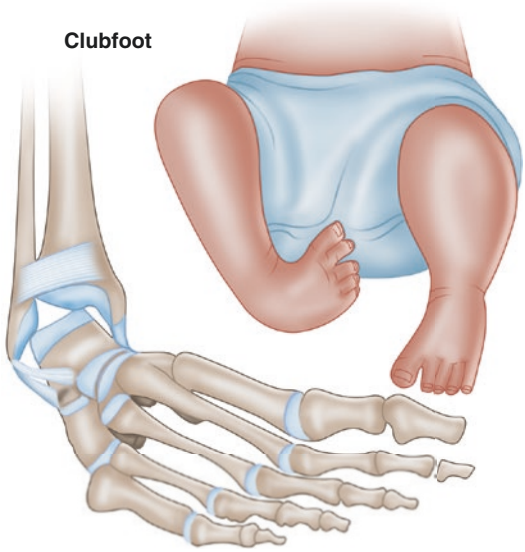

Fig. 15.1 Aspect of congenital clubfoot

15.6 Conservative and Surgical Treatment

The gold standard for clubfoot is the Ponseti technique, developed in the 1940s. This technique is divisible into two phases. In the early stage, the progressive casting of the foot aims to correct the deformity, while in the second phase, the focus is on the maintenance of the correction. The corrective treatment should start in the first 10 days of life, but it could be adopted for 2 years. It is necessary to change the casts every 4–5 days, and usually, 6–8 casts are required to correct the foot (depending on the stiffness of the deformity). The prolonged time of treatment is essential also for soft tissue adaptation. Fiber glass and plaster of Paris are typically used for casting. However, the latter is cheaper and easily moldable. Usually, the knee needs to be flexed at 90°. However, in situation of complex and atypical feet, the knee could be flexed at 110°.

The correction of the deformity needs to follow a precise order. The first casts aim to correct the cavus. Recent studies reported that the Ponseti method has also been used successfully on adolescents and adults.

The alignment between the forefoot and hindfoot should be adjusted by elevating the first metatarsal. The second stage focuses on the abduction of the foot around the talus. With sequential casting, a progressive abduction of the forefoot more than 60° relative to the tibia should be reached. However, this correction is not achievable in all feet and an attempt to perform this correction in rigid feet can result in a midfoot breach. With this procedure, it is possible to obtain also a hindfoot varus correction. Clinicians must be aware that the direct pressure of the hindfoot could not correct the varus. Moreover, pronation should also be avoided to recreate a cavus deformity, preventing the calcaneus' abduction. The third phase aims to dorsiflex the foot after the abduction is fully reached. A percutaneous Achilles tenotomy followed by casting (60° abduction and 20° dorsiflexion × 3 weeks) is indicated in cases of dorsiflexion <10° (80% of cases).

Once the phase of correction is achieved, it is mandatory to start the phase of maintenance. Abduction bracing should be set at the degree of 45 abduction achieved on the last cast with boot and bar (Dennis Brown brace) should start immediately after the last cast. The brace should be worn for 24 h/day/3 months, then 18 h/day/3 months, and then 14 h/day for 3 months, an than 12 h/day unit five years of age. Brace intolerance is frequent both in the child and the parents. However, in the case of poor compliance, 80% of recurrences are reported (compared to 6% of compliant patients). If an early recurrence appears, further castings are required.

Surgical treatment is required only for recurrent relapses or residual deformities and includes tibialis anterior transfer (child >30 months with dynamic supination). Other procedures as Z lengthening of the Achilles tendon may also be required.

15.7 Prognosis

The Ponseti method could reach a recurrence rate of 30%, but only 20% of patients will not require Achilles tenotomy and 80% anterior tibialis ten-

don transfer. An overall success rate is reported in 80% of patients treated by the Ponseti technique. However, children with unilateral deformity could result in a foot 1 or 2 sizes smaller than the healthy side.

15.8 Complication

The most feared and frequent complication is recurrence. It could be caused by improper treatment with casting or low compliance during the bracing period. Infection and other complication related to soft tissue may occur (mechanical weakness, overcorrection) after surgical intervention.

15.9 Conclusions

Clubfoot constitutes a challenging disease, but if properly managed, it could result in excellent outcomes. A multidisciplinary approach is required to face this disease properly. Neonatologist, radiologist, orthopedics, plasters, and parents need to work together, and the compliance of the family is mandatory.

Take-Home Message

- Four primary alterations characterize clubfoot: midfoot cavus, midfoot adductus, hindfoot varus, and hindfoot equinus.
- The incidence of clubfoot ranges between 0.5 and 2 cases per 1000 births.
- While the precise etiology of isolated clubfoot is multifactorial, genetic and environmental factors play a role.
- The gold standard for clubfoot is the Ponseti technique.
- The Ponseti method could reach a recurrence rate of 30%, but only 20% of patients will not require Achilles tenotomy and 80% anterior tibialis tendon transfer.
- The most feared and frequent complication is recurrence.

Summary

Clubfoot (also named congenital talipes equinovarus) is one of the most common congenital foot deformities. Four primary alterations characterize clubfoot: midfoot cavus, midfoot adductus, heel/hindfoot varus, and hindfoot equinus. The incidence of clubfoot ranges between 0.5 and 2 cases per 1000 births. A geographical variation influences this data. The precise etiology of clubfoot is still discussed, but genetic and environmental factors are probably involved in developing this condition. Maternal smoking and maternal diabetes reported a high correlation with clubfoot. Moreover, improper in utero positioning could be another risk factor. In the literature, 24.4% of patients have an associated family history.

Several classification systems have been proposed in the literature. The Pirani system could be used as a descriptive tool. Foot X-rays are useless in the first postnatal period, as many of the tarsal bones are cartilaginous and should be assessed by radiographs. The gold standard for clubfoot is the Ponseti technique, developed in the 1940s. This technique is divisible into two phases. In the early stage, the progressive casting of the foot aims to correct the deformity, while in the second phase, the focus is on the maintenance of the correction. The corrective treatment should start in the first 10 days of life, but it could be adopted for 2 years. The Ponseti method has a recurrence rate of 2%, but only 20% of patients will not require Achilles tenotomy and 80% anterior tibialis tendon transfer. The most feared and frequent complication is recurrence. It could be caused by improper treatment with casting or low compliance during the bracing period. A multidisciplinary approach is required to face this disease properly.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. In which of the following situations can Achilles tendon lengthening be indicated?

(a) Congenital equine clubfoot supinated adducted varus
(b) Congenital ana dysplasia
(c) Flat foot
(d) Talus valgus foot

2. In congenital clubfoot, which alteration(s) is/are present?
   (a) Medial and plantar deviation of the anterior portion of the talus
   (b) Medial subluxation of the scaphoid on the talus
   (c) Contracture of the Achilles tendon
   (d) All previous alterations
3. In the arthrogryposis are frequent
   (a) Congenital dislocation of the hip
   (b) Congenital clubfoot
   (c) Congenital twisted hand
   (d) All of these deformities
4. The initial treatment of congenital clubfoot consists of:
   (a) Ponseti method
   (b) Surgical lengthening of the Achilles tendon
   (c) Application of night braces
   (d) Application of plaster appliance
5. Which type of congenital deformity could be confused with clubfoot?
   (a) Equinus varus adductus barbed
   (b) Metatarsus varus
   (c) Talus valgus
   (d) Reflexive valgus

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Flatfoot and Pes Cavus

16

Nusret Köse and Zeynel Mert Asfuroğlu

Overview

Flatfoot and pes cavus are common foot deformities in children. These two deformities can be considered as opposites of each other. There is a loss of height of the longitudinal arches due to the laxity of soft tissues in flatfoot. In contrary, in pes cavus, the height of the longitudinal arches is increased and the soft tissues of the foot are rigid.

16.1 Introduction

Flatfoot and pes cavus are common foot deformities in children. These two deformities can be considered as opposites of each other (Fig. 16.1). There is a loss of height of the longitudinal arches due to the laxity of soft tissues in flatfoot. In contrary, in pes cavus, the height of the longitudinal arches is increased and the soft tissues of the foot are rigid. The most frequently seen type of flatfoot is flexible type, which is a physiological condition with low accompanying comorbidities. Pes cavus is usually a pathological condition accompanied by comorbidities, especially neurological diseases. In flatfoot, except for palliation of symptoms, treatment is generally unnecessary, whereas in pes cavus, surgical treatment may be needed.

The common point is that both of these two conditions cause parents to worry and seek solutions. In both deformities, the goal is obtainment of painless plantigrade feet.

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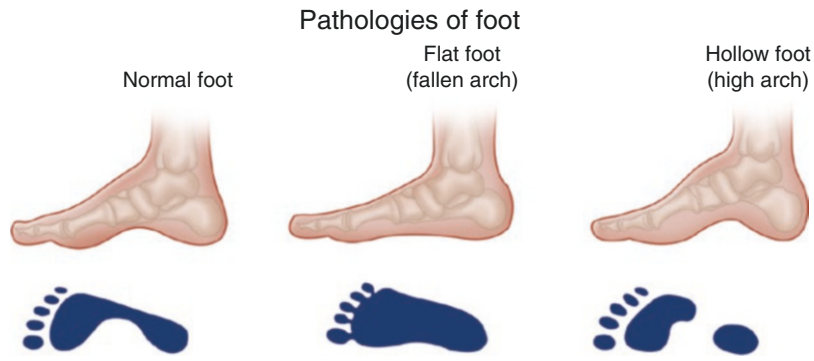

Fig. 16.1 Different type of footprint

16.2 Definition and Clinical Symptoms

The foot structure has three arches including two longitudinal (medial and lateral) and one transverse (anterior). The longitudinal arches create foot architecture and provide stability to the bones and other soft tissues (Fig. 16.2). Flatfoot is defined as an abnormally low or absent longitudinal arch, and pes cavus is described as an abnormal high medial longitudinal arch. Valgus foot, pronated foot, and pes planus are synonyms for the flatfoot. Flexible flatfoot is more common and is considered a normal foot shape in infants and in up to 20% of adult population. Flexible flatfoot is usually not associated with any other underlying disease in a child and is always bilateral. It has a positive correlation with increased body mass index. The child has a visible arch when he/she is not standing. He/she also has a normal mobility of the hindfoot (subtalar inversion and eversion). Generalized ligamentous laxity may be seen in patients [1, 2]. Subtalar joint is usually involved and hypermobile. Secondary to hypermobility, increased valgus of the hindfoot and hyperpronation of the forefoot are seen. Shortened Achilles tendon occurs as a compensatory mechanism.

Flatfoot can be briefly divided into three subgroups: flexible flatfoot, flexible flatfoot with short Achilles tendon, and rigid flatfoot. It is important to understand that "flexible flatfoot" and "physiological flatfoot" are not the same concepts. In the physiological flatfoot, the foot position changes unconsciously to compensate the increased anteversion of the femoral neck, resulting in arc flattening and increased valgus of the heel.

Children who have neuromuscular diseases or increased ligamentous laxity such as Marfan syndrome, Ehlers-Danlos syndrome, and Down syndrome may have a bilateral flatfoot. These cases are initially flexible but with time may progress to a rigid deformity. Rigid flatfoot is uncommon and may be unilateral or bilateral. These children have persistent flattening of the longitudinal arch in non-weight-bearing, and subtalar motion may be restricted. In rigid flatfoot, all the pathoanatomic features are accompanied by underlying bony abnormalities. Rigid flatfoot may be associated with tarsal coalition, vertical talus, pes calcaneovalgus, compensatory metatarsus adductus, skewfoot, trauma, infection, arthritis, poliomyelitis, cerebral palsy, myelodysplasia, and neoplasms.

Pes cavus is not as common as flatfoot. Pes cavus occurs due to the disruption of the balance between intrinsic and extrinsic muscle groups. Unlike flatfoot, generalized ligamentous rigidity is seen in most part of the foot. Plantar flexion of the first metatarsal, pronation of the forefoot, and contraction of the plantar fascia are essential pathoanatomical features. Secondary to rigidity, varus of the hindfoot and calcaneal deformity may occur. Although there are some idiopathic and traumatic cases in etiology, the majority are due to a neurological disease. The prevalence of pes cavus is approximately 8–10% in general population. It may be associated with hereditary sensory motor neuropathy (e.g., Charcot-Marie-

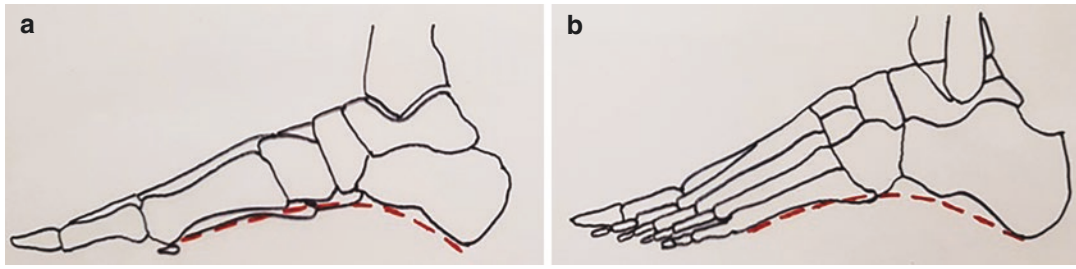

Fig. 16.2 (**a**) Medial longitudinal arch of the foot. (**b**) Lateral longitudinal arch of the foot

Tooth disease), spinal cord defect (e.g., spina bifida), acquired neuropathy, arthrogryposis, myelodysplasia, Friedreich ataxia, cerebral palsy, and sequel of trauma and compartment syndrome [3, 4].

16.3 Physical Examination

Both flatfoot and pes cavus are mainly diagnosed by clinical examination. Plain radiographs should be used to support the clinical diagnosis, especially for the evaluation of a rigid deformity. Clinical examination of the flatfoot reveals that the hindfoot (the heel) is in significant valgus alignment and midfoot in abduction positions. It gives the medial malleolus a prominent appearance, and the foot is rotated outward in relation to the leg. Thus, when viewed from the back of the child while standing, "too many toes on the lateral side of the foot" sign could be seen. To differentiate between flexible and rigid flatfoot, Jack's test (Hubscher maneuver) can be done. The test is performed with the child weight-bearing, with the foot flat on the ground, while the examiner dorsiflexes the hallux and watches for an increasing concavity of the arches of the foot. Arch formation (a positive test) results from the flatfoot being flexible. A negative result (lack of arch formation) results from the flatfoot being rigid. Contracture of the Achilles tendon with limited ankle dorsiflexion can often accompany a flexible flatfoot in an adolescent. The recommended radiographic views include weight-bearing anteroposterior foot X-rays for evaluation of the talar head coverage and talocalcaneal angle, weight-bearing lateral foot X-rays for evaluation of lateral talocalcaneal, Meary's and Hibbs' angles, and weight-bearing oblique foot to rule out any other bony anomalies (e.g., tarsal coalition) (Figs. 16.3 and 16.4).

Clinical examination of the pes cavus should begin with palpation of the plantar fascia as detection of contraction in this structure is important for diagnosis. Initial deformity is in the forefoot followed by subsequent changes in the hindfoot. The most important clinical test for pes cavus is "Coleman's lateral block" test. Coleman's block test evaluates hindfoot flexibility and pronation of forefoot. The test is performed by placing the patient's foot on wood block, 3–4 cm thick, with the heel and lateral border of foot on the block and bearing full weight, while the first, second, and third metatarsals are allowed to hang freely into plantar flexion and pronation. If deformity is flexible, calcaneus (hindfoot) will improve into neutral or valgus positions. If deformity is rigid, it will not improve. Various neurological disorders may be accompanied by pes cavus. Thus, detailed neurological examination should be performed for every patient. In the case of suspicion, patient can be consulted to the pediatric neurology department. The recommended radiographic views include weight-bearing anteroposterior foot to evaluate the hindfoot position and weight-bearing lateral foot to evaluate the Meary's and Hibbs' angle (Figs. 16.3 and 16.4). In atypical conditions such as unilateral deformity, spinal magnetic resonance imaging is required to rule out neurological disorders. The most common underlying conditions for flatfoot and pes cavus are summarized in Table 16.1.

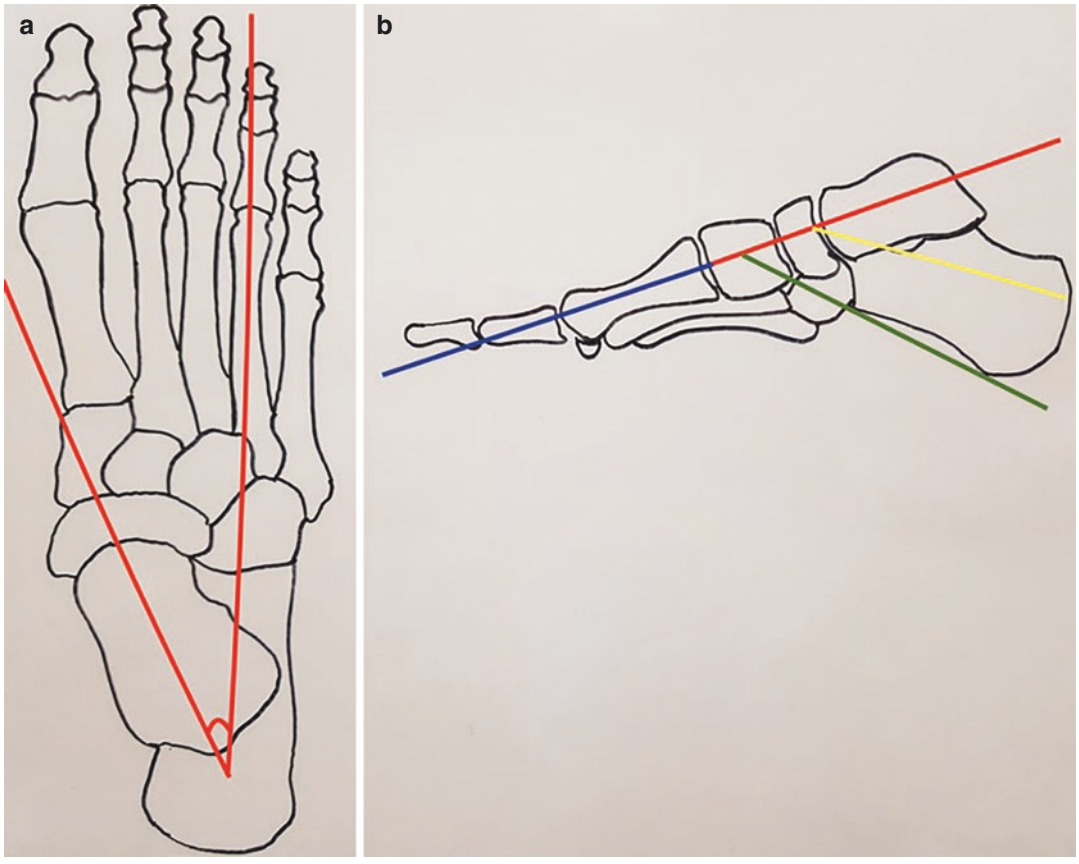

Fig. 16.3 (**a**) Talocalcaneal angle on the anteroposterior foot (N = 15–35°). (**b**) The Meary's angle is the angle between blue and red lines (N = 0–5°), the Hibbs' angle is the angle between red and yellow lines ($N < 45°$), the talocalcaneal angle is the angle between red and green lines (N = 25–45°)

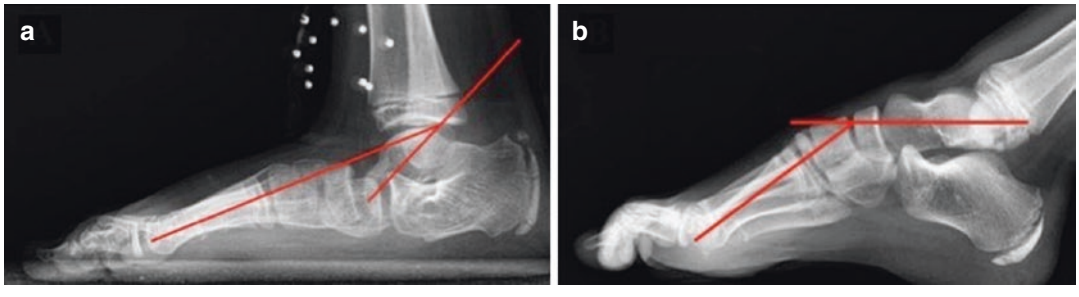

Fig. 16.4 (**a**) The lateral X-ray view of flatfoot. The parallelism between talus and first metatarsal is impaired, and arc height is decreased. (**b**) The lateral X-ray view of pes cavus. The parallelism between talus and first metatarsal is impaired, and arc height is increased

Table 16.1 The most common underlying conditions for flatfoot and pes cavus

Flatfoot	Pes cavus
Iatrogenic	Charcot-Marie-Tooth
Vertical talus	Poliomyelitis
Ligamentous hyperlaxity syndromes (Down, Ehlers-Danlos, Marfan)	Cerebral palsy
	Spina bifida
	Tethered cord
Tarsal coalition	Split cord (diastematomyelia)
Accessory navicular bone	
Cerebral palsy	Myopathy
Myelodysplasia	Friedreich ataxia
Poliomyelitis	Arthrogryposis
Peripheral nerve injuries	Residual deformity of pes equinovarus
Spinal cord injuries	
Guillain-Barré syndrome	
Peroneal spastic flatfoot	
Rupture of tibialis tendon	

16.4 Treatment

Flexible flatfoot is a normal variation of the shape of the foot that rarely causes disability or requires treatment. In these children, leg aches and uneven shoe wear may occur. Treatment is not necessary in asymptomatic patients, but pain, disability, and need for treatment are more common with a contracted Achilles tendon in flexible flatfoot. If there is Achilles contraction, stretching exercises, casting, or botulinum toxin A injection can be tried.

Although young children typically are asymptomatic, parents may be concerned about the appearance of the feet. Parents should be advised that flattening of the longitudinal arch will improve most often and custom-made shoes and ortheses are not therapeutic and not effective in the reconstruction of the arches. These can be used in palliation of the pain in children with painful feet. Surgical treatment is performed only in cases of extreme pain that do not respond to conservative treatment. It is not performed before the ages of 8–10 years. Subtalar arthrodesis, arthroereisis, medial sliding calcaneal osteotomy, and lateral colon lengthening can be performed for correction of the hindfoot valgus. Subtalar arthrodesis (complete fusion) and arthroereisis (staple fixation only without fusion) are obsolete techniques and are almost never performed today. Opening-wedge cuboid osteotomy can be performed for correction of the midfoot abduction. Opening-wedge cuneiform flexion osteotomy can be performed to provide plantar flexion of the first ray. Triple arthrodesis (subtalar, talonavicular, and calcaneocuboid joints) can be considered as a last resort in the symptomatic flatfoot in adults.

The main goal of pes cavus treatment is to achieve plantigrade, mobile, and painless foot. Conservative treatment is rarely performed. Stretching exercises and custom-made shoes can be used for symptomatic treatment in mild cases. Soft tissue surgery, osteotomy, and arthrodesis can be performed as surgical treatment. Release of the plantar fascia and intrinsic muscles, lengthening of the peroneus longus tendon, peroneus longus-to-peroneus brevis tendon transfer, and finger extensor tendon rerouting transfer for hammer toes are the most preferred soft tissue surgeries. Soft tissue surgeries are generally indicated in flexible cavus feet according to the Coleman's block test. If the foot is rigid according to the Coleman's block test, in addition to soft tissue surgery, bony procedure should be done. Dorsal closing-wedge extension osteotomy of the first and/or other metatarsal bones, tarsometatarsal wedge osteotomies, and posterior or lateral sliding osteotomy of the calcaneus are the most preferred bony procedures. In older cases or adults, arthrodesis (especially triple arthrodesis) is done as a salvage procedure.

Take-Home Message

- Physiological flatfoot does not require treatment; the parents should be informed and be calmed down about the condition.
- Conditions that may accompany both flatfoot and pes cavus should be kept in mind.
- Neurological origin must be thought and be evaluated in pes cavus.
- Diagnosis and planning of the treatment in both conditions are based on clinical examination. Thus, clinical examination should be performed carefully.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. How does physiological flatfoot occur?
   (a) The foot position changes unconsciously to compensate the increased anteversion of the femoral neck, resulting in longitudinal arc flattening and increased valgus of the heel
   (b) The foot position changes unconsciously to compensate the increased retroversion of the femoral neck, resulting in longitudinal arc flattening and increased valgus of the heel
   (c) The foot position changes unconsciously to compensate the increased anteversion of the femoral neck, resulting in longitudinal arc flattening and increased varus of the heel
   (d) The foot position changes unconsciously to compensate the increased retroversion of the femoral neck, resulting in longitudinal arc flattening and increased varus of the heel
2. In the diagnosis of flatfoot, what angle measurements are assessed on the full-weight-bearing lateral foot X-ray?
   (a) Lateral talocalcaneal, Meary's, and Hibbs' angles
   (b) Calcaneal inclination angle and tibiotalar angle
   (c) Tibiotalar angle and kite angle
   (d) Medial talocalcaneal and kite angle
3. How to distinguish between flexible and rigid flatfoot by clinical examination?
   (a) In flexible flatfoot, the height of the longitudinal foot arches increases with passive dorsiflexion of the first metatarsophalangeal joint. This event is not seen in rigid type. This test is named as Jack's test
   (b) In rigid flatfoot, the height of the longitudinal foot arches increases with passive dorsiflexion of the first metatarsophalangeal joint. This event is not seen in flexible type. This test is named as Jack's test
   (c) In flexible flatfoot, the height of the longitudinal foot arches increases with active dorsiflexion of the first metatarsophalangeal joint. This event is not seen in rigid type. This test is named as Jack's test
   (d) In rigid flatfoot, the height of the longitudinal foot arches increases with active dorsiflexion of the first metatarsophalangeal joint. This event is not seen in flexible type. This test is named as Jack's test
4. What is the most common neurological disease accompanying pes cavus?
   (a) Charcot-Marie-Tooth disease (hereditary sensory and motor neuropathy)
   (b) Diabetes mellitus with neurological involvement
   (c) Tabes dorsalis
   (d) Lateral amyotrophic sclerosis
5. Which clinical test should be performed before planning surgical treatment in pes cavus?
   (a) Coleman's block test. The test is performed by placing "1-inch block" under the lateral side of the foot. First, second and third toes should remain out of the block medially. If deformity is flexible, calcaneus (hindfoot) will correct to neutral or valgus position. If deformity is rigid, it will not correct. Soft tissue procedures are preferred in flexible type, while rigid types require bony procedures
   (b) Coleman's block test. The test is performed by placing "1-inch block" under the medial side of the foot. First, second, and third toes should remain out of the block medially. If deformity is flexible, calcaneus (hindfoot) will correct to neutral or valgus position. If deformity is rigid, it will not correct. Soft tissue procedures are preferred in flexible type, while rigid types require bony procedures
   (c) Jack's test
   (d) Adam's test

Further Reading

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Scoliosis

17

R. Compagnon and F. Accadbled

Overview
Scoliosis is the most common back deformity in pediatric population. Etiology of AIS still remains unknown, but its highly multifactorial nature is widely accepted.

17.1 Introduction

Scoliosis is a structural three-dimensional deformation of the spine with coronal Cobb angle greater than 10°. The coronal deviation is associated with a loss—or even an inversion—of the physiological sagittal curvatures and a vertebral rotation. It is a frequent condition affecting between 1% and 3% of the population, which most often begins during growth and which can progress very quickly during periods of rapid spinal growth (Fig. 17.1). It is essential to have an annual screening for all children until the end of growth.

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UG. Longo, V. Denaro (eds.), *Textbook of Musculoskeletal Disorders*,
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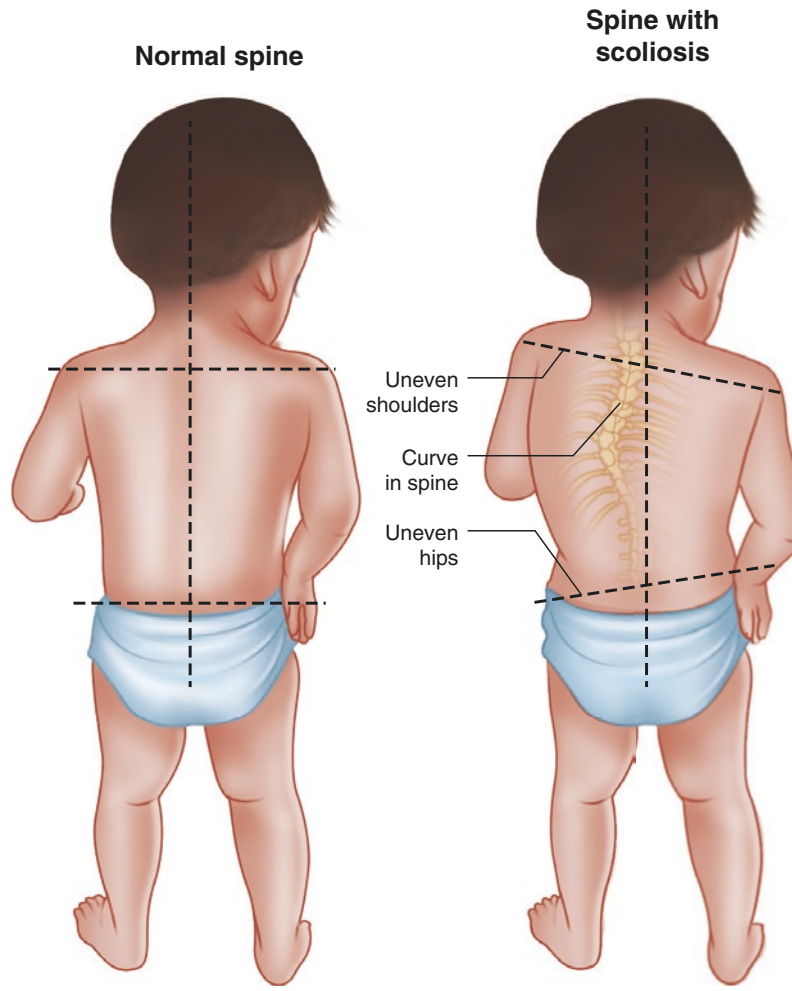


Fig. 17.1 Scoliosis characteristics

17.2 Clinical Examination

First inquire on the age of onset, family history of scoliosis, pain, and menarchal status.

The diagnosis of scoliosis is clinical. In case of unequal length of the lower limbs, the pelvis must be rebalanced with a block (Fig. 17.2).

17.2.1 Trunk Asymmetry

The child is standing arms alongside the body; there is an asymmetry in the distance between the arms and the trunk. This sign is more clearly visible in the lumbar region (Fig. 17.3).

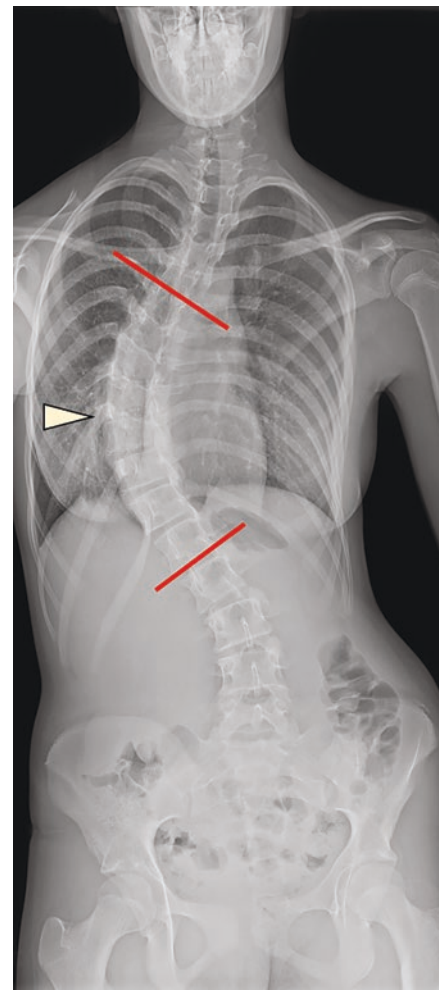
The balance of the trunk must be evaluated with a plumb line positioned at the spinous process of the seventh cervical vertebra. Normally, it projects at the intergluteal cleft. A trunk imbalance is a bad prognosis factor.

17.2.2 Rib Prominence

It signals the deformation of the spine in the horizontal plane. With the child bent forward, it can be measured using a scoliometer or a graduated ruler (Adam's test) (Fig. 17.2).

Fig. 17.2 Physical examination of the scoliosis. Black arrows are pointing the trunk asymmetry (horizontal one) and the rib prominence (vertical one)

Fig. 17.3 Radiograph of scoliosis. Arrow is the apex of the deformity. Red lines point to the limits of vertebrae and define the Cobb angle (70°)



17.2.3 Etiological Examination

The clinical examination must be supplemented by a neurological and skin examination in search of an etiology.

17.2.4 Growth Report

The child must be measured during each consultation. A pubertal staging is necessary to assess the remaining growth potential.

17.3 Radiographic Evaluation

X-ray is essential to confirm the diagnosis, carry out surveillance, and establish the prognosis and therapeutic indication. However, it should be done sparingly due to the deleterious effects of ionizing radiation in the long term. As far as possible, low-dose radiography should be preferred, which makes it possible to reduce the dose received by 6 or stereoradiography that allows three-dimensional reconstruction of the spine.

17.3.1 Cobb Angle

It corresponds to the angle formed by the parallels to the vertebral end plates of the most tilted vertebrae (limit vertebrae) of the curvature (Fig. 17.3).

Flat back or inverted curves (thoracic lordosis) are a sign of bad prognosis.

17.3.2 Spinal Rotation

It is measured by the asymmetry of the pedicles and the lateralization of the spinous process, which is located in the concavity of the curvature. The vertebral rotation confirms the diagnosis of scoliosis. It is maximum at the apex of the deformation.

17.3.3 Assessment of Spinal Maturation

The analysis of the ossification of the iliac crests is a useful reflection of skeletal maturity in the pubertal period. It is classified into five stages according to the Risser classification. According to Risser, stage 4 would indicate the end of spinal maturation. Analysis of triradiate cartilage (visible or fused) and growth plate of the greater trochanter is also useful.

17.4 Etiology

Scoliosis might be a symptom: the primary origin of disease should be investigated. In 25% of cases, an etiology will be found and will require specific management.

In 75%, on the other hand, no etiology is found; it is then an idiopathic scoliosis.

17.4.1 Idiopathic Scoliosis

It is much more common in girls (80%). A genetic origin is established, but the mode of transmission remains unknown. It seems to be multifactorial. Family forms are frequent, hence the need to regularly monitor siblings and descendants. A central balance disorder is also suspected, for which certain neurotransmitters such as melatonin have been implicated. Other etiological tracks have been advanced. Idiopathic scoliosis would in fact group together several causes not yet identified.

When seated at the thoracic level, idiopathic thoracic scoliosis has the distinction of almost always having a right-side convexity. In case of thoracic curvature with left convexity, a syringomyelia must be suspected. In the lumbar region, on the other hand, the curvature can be either right or left. Idiopathic scoliosis is classified by Lenke in six different types, depending on the specific curve pattern. This classification is used to define the proper surgical management.

17.4.2 Secondary Scoliosis

Multiple causes can be responsible for scoliosis. Three major entities are detailed:

17.4.3 Neuromuscular Scoliosis

For certain cases, the diagnosis is already made, in front of a child presenting a cerebral palsy, an infantile spinal muscular atrophy, a myopathy, etc. The problem is not with the diagnosis but with the assumption of responsibility, which will often lead to a difficult surgical procedure involving vulnerable patients.

In other cases, scoliosis might be the first symptom of the neurological disease to be identified. A neurological examination is therefore systematic at first referral. Scoliosis is often the first sign of syringomyelia. Clinically, an asymmetry of the abdominal skin reflexes or a left thoracic localization of the gibbosity is a very suggestive sign. MRI scan is indicated and may display syringomyelic cavity and an Arnold-Chiari malformation. Scoliosis may occur in peripheral neuropathy such as Charcot-Marie-Tooth disease. Osteo-tendinous reflexes and feet examination must be performed.

17.4.4 Congenital or Malformative Scoliosis

There are two types of malformations: either a formation defect corresponding to a hemi-vertebra or a segmentation defect corresponding to a vertebral fusion. These two malformations can be associated, and there can be layered malformations which are often associated with costal fusions. These malformations occur early in the embryonic period, which explains the frequency of associated malformations sometimes integrated into a syndrome of VACTERL partial or complete: **V**ertebral anomaly, **A**nal imperforation, **C**ardiac malformation, **T**racheo-**E**sophageal fistula, **R**enal malformation, and **L**imb malformation.

17.4.5 Syndromic Scoliosis

Neurofibromatosis type 1 is easily diagnosed in the presence of "café au lait" skin spots. It is responsible for a short scoliosis secondary to a vertebral dystrophy (affects 3–4 vertebrae). Scoliosis is often very progressive and rarely controlled by nonoperative measures. Circumferential fusion of the dystrophic area is often indicated even before skeletal maturity. Marfan's disease easily evoked in front of the patient's morphology (tall individual, dolichostenomelia, arachnodactyly). Monitoring and management are similar to idiopathic scoliosis.

17.5 Differential Diagnosis

17.5.1 Limb-Length Discrepancy

It is easily detected. The deviation is only coronal, without vertebral rotation. After rebalancing the pelvis, the spine is straight. Gibbosity is absent.

17.5.2 Tumor

A spinal or cord tumor can generate spinal deviation which is often stiff and painful. The pain is in the foreground perfectly described by the child and often well localized. This differentiates it from the so-called growth pain or mechanical pain, which is episodic and poorly systematized. The pain is accompanied by stiffness in the spine, and the child can barely bend forward.

In this situation, we must first evoke a tumor which can be either medullary or bone localized at the level of a vertebra, benign or malignant. In all cases, it is necessary to carry out an assessment by MRI and bone scan. The most common bone tumors are osteoblastoma, osteoid osteoma, aneurysmal cyst, and Ewing's sarcoma.

17.6 Natural History of Idiopathic Scoliosis

Secondary scoliosis has own evolution dependent upon the underlying condition. We present only natural history of idiopathic scoliosis.

The natural evolution of scoliosis is progression with growth and then a stabilization from skeletal maturity. However, this should be adjusted according to the periods of slow or rapid growth and then during adulthood, to the extent and exact level of the curve(s).

17.6.1 Before 1 Year: Infant Scoliosis

Eighty percent of cases are related to a postural anomaly and will heal spontaneously within a few months. Twenty percent represent real early scoliosis, which can rapidly progress and therefore require early treatment. An asymmetric orientation of the ribs (Mehta index) is a good way to differentiate them.

17.6.2 1–4 Years Old: Childhood Scoliosis

These scolioses can affect boys as much as girls, and they are either right thoracic or left. They are relatively rare. As spinal growth is rapid at this age, they often evolve rapidly and require active treatment.

17.6.3 From 5 Years to 10: Juvenile Scoliosis

This period corresponds to a slow growth phase of the spine. Scoliosis can progress, most often with moderate angular increase, and therefore requires brace treatment. Scoliosis can also remain stable or even decrease spontaneously but temporarily (this phenomenon is called "honeymoon"); however, there is always a risk of restart at puberty. It is therefore essential to follow careful monitoring.

The Scoliosis Research Society (SRS) defines all the scoliosis presented before 10 years old as early-onset scoliosis (EOS).

17.6.4 At Puberty: Adolescent Idiopathic Scoliosis (AIS)

This period corresponds to the pubertal growth spurt. Scoliosis can progress quickly. The risk is greatest at first menarche, when the triradiated cartilage is open and at Risser 0. Then the risk will gradually decrease until skeletal maturity.

17.6.5 Growth Assessment

It makes it possible to determine the risk of progression during the pubertal period. It is multifactorial. Clinically, it is based on the child's height, both in sitting and standing positions, and the onset of menstruation for girls. Radiographically, the most used bone maturation criteria are the analysis of triradiate cartilage, cartilage of proximal growth of the femur, and ossification of the ilium (Risser classification).

17.6.6 Adulthood

A majority of scolioses will remain stable, but some will progress. Progression is then slower than during growth, a few degrees per year.

The risk factors for worsening are the angle of the scoliosis, imbalance of the trunk, and type of curvature (single or combined). Lumbar scoliosis above 35° and thoracic scoliosis above 45° are at high risk of progression. Combined thoracic and lumbar scoliosis or double-thoracic scoliosis is less progressive and usually better balanced due to the presence of the two curves.

As a general rule, it is necessary to monitor the patients even every 5–10 years depending on the type of curvature.

Progression in adulthood can lead to an imbalance of the spine, pain, respiratory consequences in the thoracic zone, and rotational dislocation in the lumbar zone, a source of intermittent claudication.

17.7 Non Operative Treatment

17.7.1 Goals

The main goal is to avoid progression deformity and reach skeletal maturity with mild balanced scoliosis, likely to remain stable throughout adulthood and to allow a normal, active life. The progression is defined as increasing the Cobb angle by 5° every 6 months.

A correction of the deformity is more difficult to obtain with nonoperative management. This is especially possible in young children, and much less in adolescence.

17.7.2 Physiotherapy

It has never been shown to be effective in correcting or stopping the progression of scoliosis. It is based on postural work, flexibility, muscle strengthening, and respiratory work, usually in association with bracing.

17.7.3 Bracing

It is the main treatment of scoliosis in children and adolescents. Many types of braces have been designed. Most are now produced by computer-assisted design (CAD). The Cheneau-Toulouse-Munster (CTM) brace is one of the most popular. International studies have shown that brace treatment decreases the risk of progression and the need for operative treatment. Braces need to be used at least for 12 hours per day. In case of AIS, brace is indicated for scoliosis between 20° and 45° in growing children.

17.7.4 Casting

EDF cast (elongation, derotation, flexion) is mainly used for infantile scoliosis not controlled by a brace. The plaster is changed every 1–2 months until correction of the vertebral torsion. It is sometimes necessary to make serial casting before switching to a full-time brace.

17.8 Operative Treatment

It consists of correcting the deformity and fusing the affected spinal segment to prevent recurrence and progression (Fig. 17.4). The metal instrumentation includes rods, screws, hooks, and sublaminar clamps. Biomechanical studies proved the superiority of screw implants in severe curve correction. The surgeon may decide between high- and low-density implants, depending on the number of screws/hooks/clamps per level fused. Several types of instrumentation and techniques are available. The procedure must be carried out under neuromonitoring to prevent neurological complications (damage to the spinal cord).

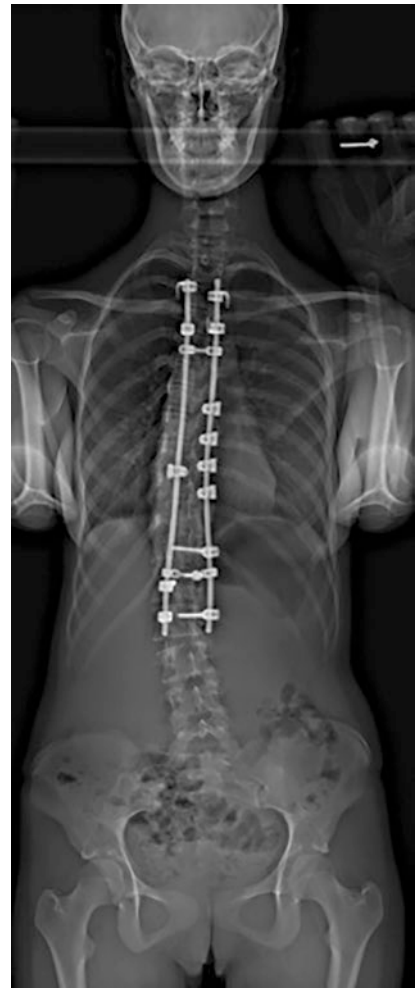


Fig. 17.4 Radiograph of scoliosis 2 years after correction and spinal fusion (same patient as in Fig. 17.2)

17.9 Indications for Treatment

Decision-making is multifactorial. It depends on the child's age (slow or fast growth phase), the type of curvature (single or multiple), the location of the scoliosis, the balance of the trunk, the Cobb angle, the amount of spinal rotation, and the worsening of scoliosis 6 months apart.

17.9.1 Childhood Scoliosis

A full-time brace or an EDF cast is proposed to correct the deformation.

17.9.2 Juvenile Scoliosis

At least clinical monitoring should be performed every 6 months. Bracing is indicated if the scoliosis progresses.

Nowadays, recent advantages in engineering have produced new devices for the treatment of EOS. In particular, during the past years, these scolioses were treated by standard growing rods. This technique involved multiple surgical procedures every 6 months to progressively lengthen the spine. Nowadays, the magnetic growing rods constitute a breakthrough in EOS surgery. With this implant, it is possible to perform only one or two surgeries and obtain the same spine lengthening in a noninvasive manner. In fact, with the use of a magnet, it is possible to lengthen the rods without touching the spine.

17.9.3 Adolescent Scoliosis

Bracing treatment is often proposed because of the significant risk of progression, especially at the onset of the puberty.

The indication for operative treatment is multifactorial and does not depend only on the severity of the curve. The psychological repercussions of the deformation must be considered in the indication. Surgery is offered in cases of progressive scoliosis not controlled by brace treatment or scoliosis presenting a significant risk of progression in adulthood. It can be discussed from 35° for lumbar scoliosis, from 40° for thoracic scoliosis, and above 45° for combined scoliosis.

Spinal fusion can be considered as soon as the triradiate cartilage is closed, without waiting for the complete skeletal maturity. Before puberty, spinal fusion is not recommended because it limits the development of the thorax. In rare cases, treatment with growing rods (nonfusion surgery) may be offered, but results are currently mixed, and the complications are frequent.

17.10 Sporting Activities

Sports activities are strongly recommended and must be maintained even in the event of brace treatment. Any kind of sport is allowed. There is no need to direct the child to a particular sport.

17.11 Monitoring and Screening During Growth

A spinal exam is necessary in all children at least once a year.

If scoliosis is diagnosed before puberty, monitoring every 6 months is necessary (at least clinically, and radiographically if there is any progression). In the pubertal phase, clinical and radiographic monitoring every 6 months is recommended.

It is important not to multiply the X-rays. A standing full spine anteroposterior radiograph is sufficient. It is preferable to use the stereoradiography, which is less irradiating and sufficient for monitoring.

Take-Home Message

- Scoliosis is a structural three-dimensional deformation of the spine with coronal Cobb angle greater than 10°.
- The diagnosis of scoliosis is clinical.
- Cobb angle, spinal rotation, and assessment of spinal maturation are evaluated by X-ray.
- The natural evolution of scoliosis is progression with growth and then a stabilization from skeletal maturity.
- The main goal is to avoid progression deformity and reach skeletal maturity with mild balanced scoliosis, likely to remain stable throughout adulthood and to allow a normal, active life.
- A correction of the deformity is more difficult to obtain with nonoperative management.

Summary

Scoliosis is a structural three-dimensional deformation of the spine with coronal Cobb angle greater than 10°. The coronal deviation is associated with a loss—or even an inversion—of the physiological sagittal curvatures and a vertebral rotation. Clinicians must indagate the age of onset, family history of scoliosis, pain, and menarchal status. The diagnosis of scoliosis is clinical. In case of unequal length of the lower limbs, the pelvis must be rebalanced with a block. Trunk asymmetry and rib prominence are common characteristics that could be observed during clinical examinations. The clinical examination must be supplemented by a neurological and skin examination in search of an etiology. A pubertal staging is necessary to assess the remaining growth potential. X-ray is essential to confirm the diagnosis, carry out surveillance, and establish the prognosis and therapeutic indication. However, it should be done sparingly due to the deleterious effects of ionizing radiation in the long term. Cobb angle, spinal rotation, and assessment of spinal maturation are evaluated by X-ray. Scoliosis might be a symptom: the primary origin or disease should be investigated. In 25% of cases, an etiology will be found and will require specific management. In 75%, no etiology is found; it is then an idiopathic scoliosis. Scoliosis could be idiopathic or caused by other conditions (secondary scoliosis) and could be defined as neuromuscular, congenital, malformative, or syndromic scoliosis. Limb-length discrepancy and presence of tumors should constitute the proper differential diagnosis. The natural evolution of scoliosis is progression with growth and then a stabilization from skeletal maturity. The main goal is to avoid progression deformity and reach skeletal maturity with mild balanced scoliosis, likely to remain stable throughout adulthood and to allow a normal, active life. The progression is defined as increasing the Cobb angle by 5° every 6 months. A correction of the deformity is more difficult to obtain with nonoperative management. Bracing is the main treatment of scoliosis in children and adolescents. EDF cast (elongation, derotation, flexion) is mainly used for infantile scoliosis not controlled by a brace. Operative treatment consists of correcting the deformity and fusing the affected spinal segment to prevent recurrence and progression. Indications for surgery depend on the child's age (slow or fast growth phase), the type of curvature (single or multiple), the location of the scoliosis, the balance of the trunk, the Cobb angle, and the amount of spinal rotation. Surgery is offered in cases of progressive scoliosis not controlled by brace treatment or scoliosis presenting a significant risk of progression in adulthood. It can be discussed from 35° for lumbar scoliosis, from 40° for thoracic scoliosis, and above 45° for combined scoliosis.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. The Risser test is used to evaluate:
   (a) The prognosis of a scoliosis
   (b) The prognosis of a gonarthrosis
   (c) The prognosis of a coxarthrosis
   (d) The prognosis of an epiphysiolysis

2. Surgical treatment of scoliosis is indicated:
   (a) In scoliotic posture
   (b) In scoliosis greater than 50° without rotation
   (c) In scoliosis greater than 50° with rotation
   (d) In scoliosis greater than 50° regardless of the degree of rotation
3. The following are the cause of congenital scoliosis:
   (a) Hemi-vertebra
   (b) Obstetrical paralysis
   (c) Congenital hip dysplasia
   (d) Congenital clubfoot
4. In the treatment of severe scoliosis, how many hours per day should the corset be worn initially?
   (a) 2–3 h per day
   (b) About 20 h per day
   (c) Daytime only
   (d) Only at night
5. The Milwaukee-type corset is indicated:
   (a) In dorsal vertebral fractures
   (b) In lumbar vertebral fractures
   (c) In dorsal scoliosis
   (d) In lumbar scoliosis

Further Reading

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Osteochondroses

18

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Overview
The osteochondrosis is a disease that involves the epiphyses or epiphyseal equivalents during the growth of the child and usually requires conservative treatment.

18.1 Definition

Osteochondroses are a group of orthopedic disease that involves the epiphyses or epiphyseal equivalents (carpal and tarsal bones, apophyses) during the growth of the child. The term osteochondrosis is used to describe a group of conditions with similar radiological images, clinical characteristics, and natural course. This condition is usually self-limited, does not necessitate surgical treatment, and concludes when the skeletal maturity is achieved. Osteochondrosis could affect any sites of the body. It is generally derived from abnormal development, injury, or overuse of the growth plate. The terms "osteochondritis" and "osteochondrosis" are often used reciprocally, creating uncertainty. Osteochondritis dissecans is an inflammatory disease of the bone and the articular cartilage that affects patients during the growing age. The most common symptom is pain after physical activity, but unlike osteochondrosis, joint locking could be present. Differently from osteochondrosis, the osteochondritis dissecans may not resolve with nonsurgical treatment or when the skeletal maturity is reached.

18.2 Epidemiology

Males are usually more affected than females, and the mean age is between 10 and 14 years. Boys are generally more involved in high-level physical activities and tend to experience symptoms more often compared to girls. The epidemiology depends on the specific type of osteochondrosis. The incidence of osteochondroses is reported in Table 18.1.

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Table 18.1 Summary of the most common osteochondrosis. *NSAIDs* Nonsteroidal anti-inflammatory drugs

Osteochondrosis	Incidence	Age	Site	Clinical findings	Imaging	Treatment
Legg-Calvé-Perthes	4–32 (× 100,000)	4–8	Hip	Limping; hip and knee pain	Sclerosis, flattening, and fragmentation of proximal femur growth center	Nonoperative (non-weight-bearing) or surgical treatment
Osgood-Schlatter	20,000 (× 100,000)	10–15	Tibial tuberosity	Localized pain after activity or kneeling	Soft tissue swelling and fragmentation	Rest; acetaminophen or NSAIDs
Sinding-Larsen-Johansson	–	9–13	Patella inferior pole	Pain after activity or kneeling	Soft tissue swelling and calcification	Rest; acetaminophen or NSAIDs
Blount	–	2–5	Proximal medial tibia epiphysis	Varus, procurvatum, internal tibial rotation, limb-length discrepancy	X-ray and MRI confirm the diagnosis	Bracing, hemiepiphysiodesis of the lateral epiphysis or osteotomy
Sever	370 (× 100,000)	12–15	Calcaneal apophysis	Heel pain after exercise or shoe wear	Normal X-ray	Rest and shoe modifications; heel cups; calf stretches; acetaminophen or NSAIDs
Freiberg	–	12–18	II and III metatarsal head	Pain and swelling	Sclerosis, flattening, and fragmentation	Rest; metatarsal pads; well-padded shoes
Köhler	2% of all children	2–8	Tarsal navicular	Mid-foot pain and limp	Sclerosis, flattening, and fragmentation	Cast immobilization for up to 8 weeks
Medial epicondyle apophysitis	26% of young baseball players	9–12	Medial epicondyle	Pain over medial epicondyle with pitching	Fragmentation or widening	Rest; acetaminophen or NSAIDs
Panner	–	5–10	Humerus capitellum	Lateral elbow pain	Flattening and fragmentation	Rest; acetaminophen or NSAIDs
Scheuermann	1–8% of the general population	10–12	Spine	Back pain and structured deformity	Anterior wedging of three adjacent vertebrae and endplate irregularities	Physical activity, bracing, or surgery

18.3 Pathogenesis

The etiology is idiopathic, but multiple risk factors are associated, as an endocrine imbalance, genetic disease, mechanical factors, rapid growth, trauma, and vascular anomalies. The damage of the blood supply leads to vascular ischemia to the growth cartilage of the endochondral ossification center. The result of this mechanism of injury is the avascular necrosis of the epiphysis. This condition usually solves spontaneously without surgical treatment. The radiological imaging of the osteochondrosis is similar for all the sites involved and differs depending on the phase of the disease. In the first time, bone and cartilage necrosis are present. Suddenly, it is possible to observe a revascularization process and a granulation tissue deposit. In this phase, the number of osteoclasts increases in the site of injury and resorption of the necrotic bone occurs. The subsequent process is the layering of mature lamellar bone by osteoid.

The most studied osteochondrosis is Legg-Calvè-Perthes disease (LCP). This condition has been evaluated by X-ray and magnetic resonance imaging (MRI) by many authors. The phases of the disease described for the LCP establish a model for the other osteochondroses. In the first time of the disease, X-ray could be nondiagnostic; hence, in suspected patients, it is recommended to perform an MRI. When the blood supply is injured, the necrotic bone may collapse due to repetitive mechanical stress. In further phases, the revascularization process begins, healing the bone with a remodeling of the normal architecture. In this phase, the MRI signal intensity returns to the standard value.

18.4 Classifications

Osteochondroses are classified according to their site of origin. All the osteochondroses are reported in Table 18.1.

18.5 Diagnosis and Treatment

18.5.1 Hip

18.5.1.1 Legg-Calvé-Perthes

Legg-Calvè-Perthes disease (LCP) is a pediatric disorder in which the blood supply of the femoral head is interrupted. This condition could lead to osteonecrosis and chondronecrosis with a resulting gradual deformity of the femoral head. LCP was historically used to describe the pathogenic mechanism of osteochondroses. The incidence of this condition differs between races and countries, and the etiology is idiopathic. Some risk factors are involved as breech position, low birth weight, multiple birth, familiarity, maternal smoking, and mechanical overload. Many authors studied the etiology of osteonecrosis. Otherwise, it is not clear if it originates from a single or multiple episodes of infarction. In 68% of patients, an injury of the lateral epiphyseal artery could lead to LCP. The blood disruption leads to the formation of a necrotic area that is reabsorbed by osteoclasts. Then, osteoblasts deposit new bone replacing the fibrovascular tissue. The mechanical overload during the ischemia phase could damage and deform the femoral head. The typical patient is a 4–8-year-old boy with limping and hip pain. At the physical examination, it is possible to assess a decreased range of motion (ROM) of the hip (in particular limited internal rotation, abduction) and lower limb discrepancy. The clinical diagnosis needs to be verified by radiological images (X-ray and MRI). The most used classification system was developed by Herring (lateral pillar classification system) and could be used during the fragmentation stage of LCP. Another classification by Stulberg used the evaluation of the femoral head deformity. The contrast-enhanced MRI and the MRI perfusion index are related to femoral head deformity and could be used for an early diagnosis. The joint line is narrowed, and fragmentation, flattening, and sclerosis of the proximal growth center of the femur are demonstrated. LCP is a self-limited condition, but in case of delayed treatment, the risk of hip osteoarthritis is higher. In case of older age presentation (>6 years old), significant femoral head deformity, hip joint incongruity, and decreased ROM, the prognosis is worst.

The correct treatment and timing are still debated. Some factors as age presentation, amount of epiphyseal involvement, stage of LCP, and status of femoral head deformity may guide the surgeon's decision. Nonoperative treatment aims to avoid weight-bearing with the use of crutches, wheelchair, or orthoses for an extended period. Bisphosphonate therapy represents another treatment helpful for LCP. It aims to decrease bone resorption, limiting osteoclastogenesis. Surgical treatment proposes to restore the correct hip joint anatomy, minimizing femoral head deformity. Femoral and pelvic osteotomies are used when the femoral head is deformed or to prevent joint incongruity. Some authors found no differences in outcomes in patients younger than 6 years old treated surgically or nonsurgically. Otherwise, in children older than 6 years, femoral head osteotomy could reduce the rate of necrosis.

18.5.2 Knee

18.5.2.1 Osgood-Schlatter

Osgood-Schlatter (OS) disease is a common cause of anterior knee pain in children and adolescents. It involves the insertion of the patellar tendon on the tibial tubercle. OS is caused by an inflammation of the ossification center of the tibial tubercle.

The typical clinical case is a 10–14-year-old male who reports anterior knee pain after physical exercise. In one-third of patients, knees are bilaterally involved, and the pain increases during jumping activity or kneeling. The direct pressure over the tibial tubercule evokes pain. The examiner could observe swelling, tenderness, and increased bump of the tibial apophysis.

The diagnosis is clinical, but imaging could be helpful to dismiss other conditions like fractures, tumors, or osteomyelitis. The X-ray could highlight the typical osteochondrosis pattern (fragmentation of the growing ossification center). MRI and ultrasonography are usually not adopted.

OS usually has an excellent prognosis, with complete functional recovery after conservative treatment (ice, rest, analgesics). Physical therapy and flexibility exercises are practiced to increase the ROM and reduce symptoms in the first phases of the disease. In the rare case of failure of conservative treatment or mature skeleton patients, surgical excision of the fragment could be considered.

18.5.2.2 Sinding-Larsen-Johansson

Sinding-Larsen-Johansson disease is a comparable condition to OS disease but involves the inferior pole of the patella. A cartilaginous fragment of the distal pole of the patella may ossify inside the tendon thickness. The typical patient is a 10–13-year-old boy with anterior knee pain after physical activity. The pain could be provoked by direct pressure on the inferior pole of the patella, by jumping or kneeling. The X-ray could display fragmentation of a small portion of the inferior pole of the patella. Moreover, thickening of the proximal patellar tendon and swelling could be present. MRI may confirm the edema of the patellar tendon and reveals infrapatellar bursitis. The prognosis is excellent, as other osteochondroses. The treatment includes ice, rest, acetaminophen, and analgesics. Stretching exercises of the quadriceps and hamstrings could reduce the pain. Patients with Sinding-Larsen-Johansson usually heal within 1 year. In case of failure of the conservative treatment or mature skeleton patients, surgical excision of the fragment could be performed.

18.5.2.3 Blount Disease

Blount disease is an osteochondrosis of the proximal medial tibia epiphysis that leads to a tibia vara. The higher incidence of this condition is in early childhood and adolescence. Only the former is considered real osteochondrosis because the latter is due to premature closure of the medial tibial physis. Child obesity and genetic familiarity are the main risk factors. X-ray and MRI confirmed the clinical suspect, also showing the stress in the entire knee structures (femoral epiphysis and medial meniscus).

18.5.3 Foot

18.5.3.1 Sever-Blanke

Sever-Blanke disease affects the calcaneal apophysis (usually bilaterally), causing heel pain in young athletes. This condition is caused by repeated Achilles tendon traction or by frequent axial loading. The typical clinical case is a 12–15-year-old soccer player with posterior heel pain after sport. Risk factors are obesity and high levels of physical activity. Some type of shoes could increase pain (soccer cleats) due to the absence of heel support. The examiner could observe a limp during gait, tenderness at the insertion of the Achilles tendon, tight heel cords, and pain after direct compression of the medial-lateral part of the heel. The X-ray may exclude calcaneal stress fractures and reveal the typical signs of osteochondrosis (fragmentation and

sclerosis of the apophysis). MRI could demonstrate edema of the calcaneal apophysis and could be used in patients with nondiagnostic X-ray images. Prognosis is excellent, with a fast recovery within 2 weeks to 2 months. The conservative treatment (ice, rest, restricted weight-bearing, and analgesics) is indicated. In case of persistence of symptoms, a cast is used for immobilization.

18.5.3.2 Freiberg

This condition also named as "Freiberg infraction" is a painful disease of the forefoot that affects adolescent dancers. The typical case is a 16-year-old girl who feels forefoot pain after dancing. The etiology is idiopathic, but traumas, vascular deficiency, familiarity, and mechanical overloads are considered as risk factors. It is characterized by a disordered ossification of the II and III metatarsal head. This condition is usually unilateral, but the bilateral presentation is reported in up to 10% of cases. The examiner could describe localized pain, which worsens by weight-bearing, tenderness, and swelling of the forefoot over the affected metatarsal heads. X-rays could show a widening of the metatarsophalangeal joint. Typical signs of osteochondrosis as cortical irregularity, sclerosis, and flattening of the articular surface could be present. MRI is more sensitive in the first stages of the disease, highlighting hypointensity of the marrow on T1-weighted images and low/high signal on T2-weighted images. The standard treatment is conservative (rest, metatarsal pad, and unloading of the affected foot). Early treatment could solve the symptoms; otherwise, the normal head morphology on X-ray appears evident after 2–3 years.

18.5.3.3 Köhler

Köhler disease affects the navicular bone in the foot. Boys are more commonly affected than girls (3–5 times higher). The typical case is a 2–8-year-old boy with foot pain after physical activity. The etiology is idiopathic, and no history of trauma is usually present. The examiner could observe mild symptoms and point tenderness over the medial part of the mid-foot. Limping is present, and patients typically avoid weight-bearing on the medial part of the foot. The diagnosis is clinical, but X-rays could confirm the hypothesis. Radiographs show the typical signs of osteochondrosis (sclerosis, flattening, and fragmentation of the tarsal navicular). The prognosis is excellent with a full recovery within a few months. It has been demonstrated that a short leg cast for 8 weeks could accelerate symptom resolution.

18.5.4 Elbow

18.5.4.1 Medial Epicondyle Apophysitis

Also known as "little leaguer's elbow", it usually affects young pitchers. The repetitive throwing could lead to frequent stress of the epicondyle growth plate. The typical case is a 9–12-year-old boy who practice baseball or volleyball with pain over the medial epicondyle. The examiner could observe mild tenderness in early stages, but in the case of an avulsion fracture, severe tenderness is present. This condition is usually related to other shoulder conditions; therefore, a clinical assessment of this joint is required. X-rays may confirm the clinical diagnosis, showing sclerosis and fragmentation of the medial epicondyle. Conservative treatment involves the collaboration of the patient. Some kind of throwing that requires elbow rotates is avoided (curveball and sliders). The standard conservative therapy solves the symptoms in a few weeks (ice, rest, and analgesics). The surgical excision of the fragments could be necessary in case of avulsion fractures.

18.5.4.2 Panner

Panner disease is an osteochondrosis of the humeral capitellum and represents the most common cause of lateral elbow pain in children younger than 10 years. The typical clinical case is a 5–10-year-old boy with pain over the lateral part of the elbow without a history of repetitive trauma. The clinical exam could report mild, lat-

eral sided elbow pain without localized tenderness. The X-rays could show a degenerative process of the distal humeral ossification center of the capitellum. It is also possible to observe other classical signs of osteochondrosis (fragmentation and fissuring of the humeral capitellum). Panner disease is self-limited, and the prognosis is excellent. Rest, conservative therapy with ice, and analgesics usually solve symptoms.

18.5.5 Spine

18.5.5.1 Scheuermann

Scheuermann disease is an osteochondrosis of the anterior part of the thoracic vertebral growing plate. It is a common cause of back pain in adolescents and could present with a rigid kyphosis or humpback deformity. Osteochondrosis causes an anterior deformity of the vertebral body with a resultant wedging of the vertebra and a kyphosis during a growth spurt. The etiology is unknown, but an autosomal dominant inheritance has been reported. The typical patient is a 12–15-year-old male with a family history of Scheuermann and occasional back pain centered at the curve apex. The examiner could demonstrate the presence of a rigid humpback deformity that does not change with extension. X-rays confirm the clinical hypothesis showing the radiologic findings for the diagnosis of this disease: thoracic kyphosis greater than 45° and at least one vertebral body wedging >5°. Schmorl's nodes and narrowed intervertebral disks are common associated findings. The thoracic presentation of Scheuermann is defined as type 1; differently, type 2 defines the lumbar form of this disease (rare). In the case of deformity <50°, the treatment is conservative (physical activity). In symptomatic patients between 50° and 65°, bracing could be used until skeletal maturity is reached. In deformities higher than 70°, unresponsive to conservative treatment or in the presence of neurologic deficit, surgical therapy with posterior fixation is indicated.

Take-Home Message

- The osteochondrosis is an anomaly of the epiphyses or epiphyseal equivalents.
- Males are usually more affected than females.
- The clinical presentation usually appears between 10 and 14 years of age.
- Legg-Calvè-Perthes was used as a model to describe the pathogenesis of all osteochondroses.
- The etiology is idiopathic, but multiple risk factors are involved (endocrine imbalance or genetic disease, mechanical factors, rapid growth, trauma, and vascular anomalies).
- This condition usually resolves spontaneously without surgical treatment.

Summary

The osteochondrosis is an anomaly of the epiphyses or epiphyseal equivalents during the growth of the child. The term osteochondrosis is used to describe a group of conditions with similar radiological images, clinical characteristics, and natural course. Males are usually more affected than females, and the clinical presentation appears between 10 and 14 years of age. The etiology is idiopathic, but multiple risk factors are involved. The radiological imaging of the osteochondrosis is similar for all the sites involved and varies depending on the stage of the disease. Osteochondrosis is classified according to their site of origin. This condition usually resolves spontaneously without surgical treatment.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Twelve-year-old boy complains of pain in his left knee after physical activity. The examiner can localize pain by direct pressure over the tibial tuberosity. Which is the right diagnostic algorithm?

(a) X-ray to assess the presence of Osgood-Schlatter disease; ice, rest, and analgesics
(b) MRI to assess the presence of Osgood-Schlatter disease; surgical treatment
(c) X-ray to assess the presence of slipped capital epiphysiolysis of the femur; surgical treatment
(d) Ultrasonography assessment to evaluate the presence of Sinding-Larsen-Johansson; conservative treatment

2. What is the main difference between medial epicondyle apophysitis and Panner disease?
   (a) In the former, the pain is localized over the medial part of the elbow, and in the latter over the lateral part.
   (b) In the former, the pain is localized over the lateral part of the elbow, and in the latter over the medial part.
   (c) In the former, only surgical treatment is required.
   (d) In the latter, only surgical treatment is required.
   (e) In the latter, the prognosis is worse than in the former.
3. Which are the most common radiographic classifications for LCP?
   (a) Herring and Stulberg classifications
   (b) Herring classification
   (c) Stulberg classification
   (d) Dennis classification
   (e) Jupiter classification
4. Fourteen-year-old male with painful kyphosis. The X-ray showed a value of thoracic kyphosis of 80° and T4–T9 anterior wedging. Which is the correct treatment?
   (a) Surgical correction with posterior instrumentation and osteotomies
   (b) Conservative treatment with rest and physical activity
   (c) Surgical correction with only posterior instrumentation
   (d) Surgical correction with anterior correction
   (e) Bracing until the symptoms resolve
5. Thirteen-year-old male with heel pain after activity or shoe wear. Which is your suspected diagnosis?
   (a) Sever disease
   (b) Freiberg disease
   (c) Köhler disease
   (d) Osgood-Schlatter disease
   (e) Blount disease

Further Reading

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Slipped Capital Femoral Epiphysis

19

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Overview

Slipped capital femoral epiphysis is the most common pediatric hip disease that affects patients of 10–14 years old. An early diagnosis and treatment could reduce the rate of complications and hip osteoarthritis.

19.1 Definition

Slipped capital femoral epiphysis (also known as epiphysiolysis of the femoral head, SCFE) is the most common pediatric hip disease that affects patients of 10–14 years old. It is defined by posterior and inferior displacement (through the epiphyseal plate) of the proximal femoral epiphysis in relation to the metaphysis (Fig. 19.1).

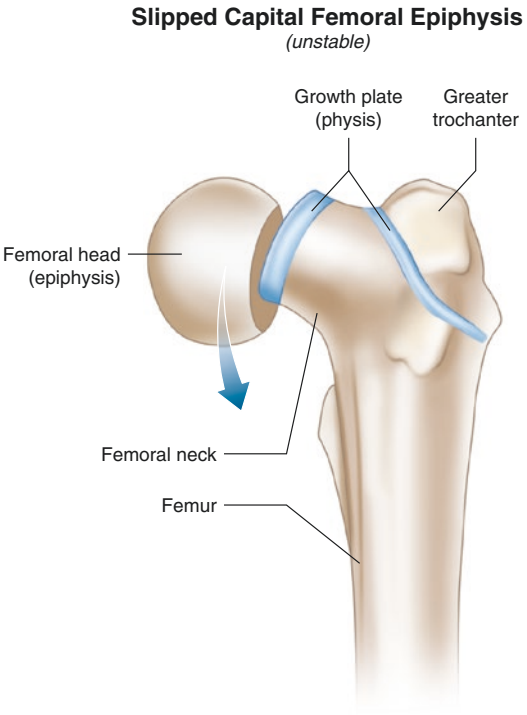

Fig. 19.1 Slipped capital femoral epiphysis

19.2 Epidemiology

Each year in the USA, approximately 10.8 cases per 100,000 children of SCFE occur. This condition is more frequent in males than in females. The mean age at diagnosis ranges from 12 years (females) to 13.5 years (males). The bilateral pre-

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sentation is frequent, ranging from 18% to 50% of patients. In some cases, the second SCFE occurs after 1–2 years since the first episode. In the North of the USA, there is a seasonal variation in the rate of incidence. The number of hospitalizations for SCFE increases in late summer and fall. This variation could be caused by a low vitamin D synthesis (due to the northern latitude) and the increased level of physical activity during summer.

19.3 Pathogenesis

The etiology of SCFE is idiopathic. Some risk factors involved are endocrine disorders, growth spurs, and obesity. Children with SCFE in 63% of cases have a weight over the 90th percentile. Hypogonadism, growth hormone deficiency, hypothyroidism, and panhypopituitarism are related to SCFE. Therefore, in case of uncommon presentation of SCFE (younger than 8 years and older than 15 years; underweight; short stature), it is recommended to assess the endocrine profile of the patient. History of trauma to the hip is uncommon.

19.4 Classifications

SCFE is classified into "acute," "acute on chronic," and "chronic forms," based on symptom duration. If symptoms present within 3 weeks, it is considered acute (10–15% of all cases). The acute form is related to the higher incidence of avascular necrosis (AVN). If the symptoms last for more than 3 weeks, it is considered as a chronic form (85% of cases). In patients with previous symptoms of pain and limping, a clinical worsening could be due to an "acute on chronic" form. Otherwise, the limit of this classification is the lack of a prognostic value.

The Loder classification divides SCFE into stable or unstable form, based on the patient's capacity to bear weight, with or without crutches. In case of stable SCFE, the risk of AVN within 6–18 months is 0%; in unstable forms, it ranges from 18% to 29%.

Lastly, it is possible to define the grade of SCFE by radiographical parameters. In the anteroposterior (AP) view, the epiphyseal displacement could be evaluated as a fraction of the total physeal diameter. The SCFE could be defined as mild (<33%), moderate (33–50%), or severe (>50%). The same analysis could be performed in frog leg view (Wilson method), defining the slippage as mild, moderate, or severe, dividing the physeal into thirds (1/3, 2/3, and 3/3, respectively). The most common radiological method, used with frog leg view, was developed by Southwick. It is a comparative approach, in which the difference in the angle subtended by the proximal femoral physis and the ipsilateral femoral shaft is compared to the healthy femur. Southwick defines the slippage as mild (<30), moderate (30–50), and severe (>50). Otherwise, this method may be invalidated by the femoral rotation or incorrect X-ray views.

19.5 Diagnosis (Clinical and Imaging)

The classical clinical case is a 12-year-old obese boy, with pain and limping to the hip. The most common symptoms are pain and limping localized to the hip, groin, thigh, or knee. The latter is present in 15–50% of cases and is due to a reflex arc of the somatic sensor nerves that ends at the same spinal level. A differential diagnosis with irritation of obturator nerve branches is mandatory. In this case, the pain is more localized to the medial knee. Few cases of SCFE are entirely asymptomatic. A precise and rapid diagnosis is challenging due to the differential causes of hip pain in young patients (apophyseal avulsion fracture of the anterosuperior and anteroinferior iliac spine; apophysitis of the anterosuperior and anteroinferior iliac spine; transient synovitis; fracture; Legg-Calvè-Perthes disease; septic arthritis; adductor muscle strain). A delayed diagnosis could avoid short- and long-term complications as AVN of the femoral head and hip osteoarthritis.

The examiner needs to assess the gait of the patient. Antalgic or Trendelenburg gait with the

Table 19.1 Differential diagnosis of SCFE

Disease	Age	Clinical presentation	Diagnosis
Apophyseal avulsion fracture of the anterosuperior and anteroinferior iliac spine	12–25	Pain after forceful movement	History of trauma; X-ray
Apophysitis of the anterosuperior and anteroinferior iliac spine	12–25	Activity-related hip pain	History of overuse; X-ray to exclude fractures
Transient synovitis	<10	Limping or hip pain	X-ray, laboratory testing, ultrasonography
Fracture	All	Pain after a traumatic event	History of trauma; X-ray
Legg-Calvé-Perthes disease	4–9	Diffused hip pain, decreased internal ROM	Hip X-ray or MRI
Septic arthritis	All	Fever, limping, hip pain	X-ray; laboratory testing; joint aspiration
Adductor muscle strain	12–20	Groin pain after activity	X-ray to exclude fracture; clinical examination

femur in the external rotation could be present. Active and passive range of motion (ROM) of the affected hip is reduced if compared bilaterally. The "Drehmann sign" (hip flexion obliges to the external rotation of the femur) is often present. The clinical diagnosis needs to be confirmed by a radiographic evaluation with AP and frog leg views. In the AP view, it is possible to identify the Klein line. It is a virtual line drawn parallel to the upper edge of the femoral neck. If the line does not intersect with the outermost part of the femoral head's, the diagnosis of SCFE is confirmed ("Trethowan sign"). Otherwise, the sensitivity of this sign is debated. Sixty-one percent of cases of missing SCFE were found using this sign, demonstrating the low sensitivity of the Klein line. Green proposed to compare the amount of epiphysis that is lateral to the Klein line with the healthy hip. Following this method, a 2 mm difference could be suspicious of SCFE. Although numerous other radiographic findings on AP view are useful to assess SCFE, frog view is usually more sensitive in the earliest phases of the disease.

In the case of chronic SCFE, resorption of the superior metaphysis, with periosteal reaction and bone formation could be present. Second-stage imaging as computed tomography (CT) and magnetic resonance imaging (MRI) is controversial. CT is useful for preoperative planning in case of complex osteotomy. Otherwise, MRI can be adopted to assess the vascularization of the femoral head. Differential diagnosis of SCFE is reported in Table 19.1.

19.6 Treatment

SCFE often requires surgical treatment to prevent further complication and joint deformity. The initial step in the treatment of SCFE is to place the patient on non-weight-bearing crutches or in a wheelchair. It is mandatory to prevent slip progression and insurgence of complications. A closed reduction with casting should not be attempted because it can result in AVN. Otherwise, some authors reported a low rate of AVN if the reduction is gently performed within the first 24 h. After this period, a contracture and shortening of the ascending cervical branches of the medial circumflex femoral artery are described. Otherwise, patients treated by closed reduction and spica casting resulted in higher rates of hip surgery and osteoarthrosis.

Some authors recommend the prophylactic treatment of the contralateral hip, but there is no consensus on this topic. This treatment is usually reserved to an unusual patient (younger than 10 years, older than 16 years, with an endocrine disease).

19.6.1 Stable SCFE

The optimal treatment and timing are debated. The standard procedure is in situ closed fixation with a single screw. The correct entry point for the screw is perpendicular to the epiphysis, in the geometrical center of the physis. There are no

biomechanical advantages concerning the use of multiple versus one single screw. In case of severe displacement, the femoral head is more posterior; therefore, the correct entry point is more anterior. The most significant error to avoid is the intra-articular penetration of the screw that could lead to an acute loss of cartilages (chondrolysis). Screws placed closer than 4 mm to the subchondral bone on lateral view may penetrate the joint.

19.6.2 Unstable SCFE

The unstable SCFE is related to a higher risk of osteonecrosis, and the standard treatment is in situ open fixation. However, there is no consensus in the timing of surgery and the technique of reduction. The most commonly used method is the modified Dunn procedure, which restores the alignment of the proximal femur, decreasing the risk of femoroacetabular impingement (FAI). This procedure consists of an intracapsular osteotomy of the femoral neck and a reduction of the head, preserving the periosteum and the blood supply of the head. The aim is to realign the femoral head, minimize the FAI, and improve the ROM. The contracture of the blood vessel due to the chronic slip could require a shortening of the femoral neck, to avoid AVN. Therefore, it is possible to perform a neck or head osteotomy. Intertrochanteric, basicervical, or subcapital osteotomy could represent alternative methods.

19.7 Complications

The most relevant complications of SCFE are AVN (0% in stable SCFE, 23.9% in unstable SCFE); degenerative osteoarthritis; chondrolysis (reported after SCFE surgery or in untreated SCFE); and FAI. Chondrolysis could be suspected when there is a loss of joint space and osteopenia compared to the contralateral part.

19.7.1 Avascular Necrosis

In unstable SCFE, the contracture of the blood vessel due to the chronic slip could lead to AVN in 18–29% of cases. The formation of hematoma or the kinking of ascending cervical branches of the medial circumflex femoral artery damages the blood supply of the femoral head. AVN could lead to degenerative hip osteoarthrosis.

19.7.2 Chondrolysis

The use of spica cast or the presence of intra-articular screws could cause an acute loss of cartilage.

Chondrolysis may disrupt the joint and produce hip osteoarthrosis. Nowadays, the improvement in technology has decreased the rate of chondrolysis from 7% to 1%.

19.7.3 Femoroacetabular Impingement

FAI was observed as a long-term complication in patients treated for SCFE. It consists of an abnormal shape of the components of the hip joint that lead to early osteoarthritis. Some authors demonstrated that a subtrochanteric osteotomy might prevent FAI.

19.8 Rehabilitation

No high-quality studies focus on postoperative rehabilitation protocols. The most common protocol consists of a five-step method. In step one, the aim is to reduce the joint inflammation and improve muscle activation and ROM. In phase two, the patient can discharge crutches if he/she can walk pain-free and to perform a leg-raise abduction. In steps three and four, the aim is to improve strength with aerobic exercises and increase ROM. In the last step, the objective is to restore the strength for return to daily activities.

Take-Home Message

- SCFE is the most common pediatric hip disease that affects patients of 10–14 years old.
- An obese adolescent boy with hip pain and limping is suspicious of SCFE.
- AP and frog leg X-ray view may be compared with contralateral hip.
- Dunn procedure is the most indicated treatment for the unstable form.
- Bilateral fixation is recommended in an unusual form.

Summary

Slipped capital femoral epiphysis (SCFE) is the most common pediatric hip disease that affects patients of 10–14 years old. It is defined by posterior and inferior displacement. The etiology of SCFE is multifactorial. SCFE could be divided into acute, acute on chronic, and chronic forms, based on symptom duration. Another classification divides SCFE into stable or unstable form. The most common symptoms are pain and limping localized to the hip, groin, thigh, or knee. The diagnosis is clinical and radiological. In stable SCFE, the standard procedure is in situ closed fixation with a single screw. In unstable SCFE, the standard treatment is in situ open fixation. The most commonly used method is the modified Dunn procedure.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Twelve-year-old boy complains of pain in his left knee. He has been experiencing this for the past few weeks. Earlier that week, his mother started to notice that he was limping. His vitals are within normal limits. A physical exam reveals an obese boy that is not in any distress or pain. His left leg is turned inward, and his knee and toes point toward his right leg. His affected knee is unremarkable, without any swelling or effusion. There is a limited ROM at the left hip. As the hip is flexed, the leg externally rotates and locks in place. What is the likely diagnosis?
   (a) SCFE
   (b) Perthes disease
   (c) Femoral head fracture
   (d) L3 radiculopathy
   (e) Rheumatologic disease
2. What are the differences between stable and unstable forms of SCFE?
   (a) In the unstable form, the patient is not able to bear weight, with or without crutches.
   (b) In stable forms, it is ever necessary to perform bilateral surgery.
   (c) In unstable forms, it is ever necessary to perform bilateral surgery.
   (d) In stable forms, the disease could never worsen.
   (e) In unstable forms, the disease could never worsen.
3. What is the Drehmann sign?
   (a) During the passive hip flexion, the femur is obliged in external rotation.
   (b) When standing on one leg (the "stance leg"), the pelvis drops on the side opposite to the stance leg.
   (c) During the active hip flexion, the femur is obliged in external rotation.
   (d) During the passive hip flexion, the femur is obliged in internal rotation.
   (e) During the active and passive hip flexion, the femur is obliged in external rotation.
4. What is the Trethowan sign?
   (a) It could be observed when the Klein line does not intersect with the outermost part of the femoral head. It is diagnostic of SCFE.
   (b) During the passive hip flexion, the femur is obliged in external rotation.
   (c) When standing on one leg (the "stance leg"), the pelvis drops on the side opposite to the stance leg.
   (d) It is a virtual line drawn parallel to the upper edge of the femoral neck.
   (e) It is a virtual line drawn perpendicular to the upper edge of the femoral neck.

5. What is chondrolysis?
   (a) Chondrolysis is an acute loss of cartilages most commonly due to the intra-articular penetration of the screw. Chondrolysis could be suspected when there is a loss of joint space and osteopenia compared to the contralateral part.
   (b) Chondrolysis is a chronic loss of cartilages most commonly due to the intra-articular penetration of the screw. Chondrolysis could be suspected when there is a loss of joint space and osteopenia compared to the contralateral part.
   (c) Chondrolysis is a chronic loss of cartilages most commonly due to nonsurgical treatment.
   (d) Chondrolysis could be suspected when there is an increase of joint space and sclerosis of the acetabulum compared to the contralateral part.
   (e) Chondrolysis is a rheumatological disease.

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Pediatric Trauma Diseases

20

M. Esat Uygur

Overview

Trauma is more than a fracture. This chapter emphasizes the differences between pediatric fractures and adult fractures and the complications in children after trauma that may be overlooked.

20.1 Introduction and Epidemiology

Trauma affects not only the entire region, including the skin and the musculotendinous and neurovascular structures, but also the whole body. Therefore, a physician should keep in mind that fractures may cause both early and late complications.

The percentages of children under 16 years old sustaining at least one fracture are 42–60% for boys and 27–40% for girls. **The radius is the most commonly fractured bone during childhood**. It is followed by the humerus. In the lower extremity, tibial fractures are more common than femoral fractures. The incidence of fractures in children increases with age, and the peak age for a fracture is 11–12 years of age for girls, while it is 13–14 years of age for boys.

In every pediatric trauma case, **abuse should be considered** in the etiology of the injury. The most common findings for abuse-related injuries are multiple skin ecchymosis at different ages, multiple fractures at different ages, long bone fractures under ambulatory age, and posterior rib fractures. While recreational sports and accidents are the most common causes of fractures in children, the lack of proprioception and balance control often leads to falls in sedentary and obese children.

The pediatric population has special attributes in terms of dealing with trauma and is less affected than the adult population. First, the tissues of pediatric patients, including the fracture and the skin that covers it, are more capable of healing. Second, complications, which can be encountered after trauma, rarely occur. For instance, in a review article, authors declare that thrombosis, infectious multiorgan failure, or respiratory distress syndrome, which are quite common complications in adults, did not occur in any of the patients after a pelvic fracture. This can also be due to the treatment options chosen for children. While open reduction and internal fixation are required in adult patients, a nonoperative treatment using a cast or a minimally invasive treatment using a Kirschner wire (K-wire) is adequate to get the fracture line united. Therefore,

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less invasive methods are popular treatment options for children. **The periosteum is thicker in children**, so there is a higher potential for fracture healing than in adults due to better bone nutrition. In addition, in the case of a deformity occurring after a fracture (malunion) in a growing child, **the deformity can be corrected over time during normal growth**. If the metaphyseal deformity is in the same plane as the closest joint's range of motion, the malunited deformity is corrected more over time. For example, if a flexion and varus deformity occurred in a growing child after a supracondylar femoral fracture, the flexion deformity would lessen over time because the flexion deformity is in the same plane of range of motion as the knee joint, which mainly does flexion and extension. However, a varus-valgus deformity does not have the same chance as a flexion-extension deformity. This also means that the fractures around the joints that have multidirectional movements (e.g., the shoulder and the hip) have a wider acceptance range for nonoperative treatment. For example, excessive deformity (i.e., varus-valgus, flexion-extension, or translation deformities in any plane) at the proximal humerus or the proximal femur can be healed over time without a major sequela.

20.2 Physeal Injuries

Undoubtedly, the most important complication, which is unique for the pediatric fractures, is **physeal injuries**. Fractures affecting the epiphysis may impair growth by causing or not causing physeal arrest. In the lower extremity, the growth mostly occurs around the knee (i.e., the distal femur and the proximal tibia), while the upper extremity grows longer mostly from the distal radius and the proximal humerus epiphysis. Therefore, fractures around the knee may contribute to **limb-length discrepancy**. On the other hand, ankle fractures may cause alignment impairments instead of shortening. Similarly, gunstock deformity may occur in children after elbow fractures. In these cases, some parts of the epiphysis have been affected because of the trauma; however, other parts have not been influenced. The growth at the affected part is impaired while it keeps growing at the unaffected side. This mechanism identifies the varus and valgus deformities occurring after fractures in those of growing age. In contrast, **Cozen's fracture** is identified for genu valgum deformity after proximal medial tibial fractures. The hypothesis in these cases is based on the increased blood flow at the epiphysis during the fracture healing process.

20.3 Torus Fractures

The **torus fractures** are specific for pediatric age. The cortex of the bone is bent instead of a complete fracture. This happens due to elasticity of the cortex of the child's bone.

Greenstick fractures also occur because of the plastic deformation of the bone. However, at this time, the long bone is bent entirely and an incomplete fracture line can be observed on the radiographs. Therefore, while we see the greenstick fracture in the middle part of the diaphysis of a long bone, torus fractures are seen at the metaphysis. In specific, a torus fracture results from a compressive force acting on the metaphysis of the bone, which is a point of decreased strength. Instead, greenstick fractures occur in the same way, but more severe forces are applied, and the disruption of one cortex occurs while the other is bent.

The **toddler fracture** is another children-specific fracture seen at the tibia, mostly in 4–6-year-old children. This is a type of incomplete spiral oblique fracture that occurs after rotational movements.

20.4 Physeal Fractures

Physeal fractures at the ankle joint have their own characteristics. It can occur as **triplanar fractures** consisting of the typical **Thurstan Holland fragment**, epiphyseal medial malleolar fracture, and epiphyseal Tillaux fracture. In adolescents, a

juvenile Tillaux fracture occurs at the anterolateral part of the distal tibial epiphysis. It is related to the order of the closing of the epiphysis during growth. Closure of the distal tibial epiphysis begins centrally and then extends anteromedially, posteromedially, and laterally, respectively. Therefore, a Tillaux fracture occurs as an avulsion of the anterior inferior tibiofibular ligament.

Osteomyelitis of the epiphysis is another devastating complication. It usually occurs after open fractures or spontaneous osteomyelitis, which typically occurs at metaphyseal regions. Eventually, the growth is impaired because of the lytic effect of the infection and additional trauma if it is an open fracture. A **Seymour fracture** is an open fracture of the distal phalanx in which the nail bed is stuck into the fracture line and causes osteomyelitis and nonunion of the physis.

Another fracture pattern, which is more specific to the pediatric population, is an **avulsion fracture**. Pelvic avulsion fractures are especially seen in this age group. As the bone and epiphyseal cartilage are relatively weak compared to the adjacent ligaments, the ligaments cause a pullout type of fracture.

Although there are specific classifications for specific fractures, the Arbeitsgemeinschaft für Osteosynthesefragen (AO) expert group made a general classification system for pediatric fractures. This classification is used for fractures of the long bones, including the humerus, the radius and ulna, the femur, and the tibia and fibula. It is based on three segments (proximal, shaft, and distal) and three subsegments (epiphyseal, metaphyseal, and diaphyseal). Additionally, there are other classifications specific to fracture sites. For example, in Gartland classification, supracondylar humerus fractures are classified according to the injury mechanism (extension/flexion) and displacement based on the integrity of the posterior cortex. Radial neck fractures are classified according to the angulation of the fracture. A Monteggia fracture-dislocation is a radial dislocation with an ulnar fracture. It is based on the direction of the radial dislocation (anterior/posterior/lateral) and the presence of a radius fracture, which is called Bado classification. Epiphyseal involvement of a pediatric fracture is evaluated according to **Salter-Harris classification**, which depends on the extension of the fracture line to the epiphysis and/or the metaphysis. These classifications aid in the planning of treatment; however, these are not adequate to preoperatively determine clinical outcomes and surgical techniques.

These are not specific to pediatric trauma; however, other complications after trauma include **synostosis and heterotopic ossification**. Synostosis is defined as ossification between two bones, which can be seen at the forearm and cruris. Of course, synostosis limits supination-pronation movements at the forearm; however, it usually does not affect the patient when it occurs between the tibia and the fibula. The risk factors for synostosis are high-velocity trauma, open fractures, open surgery, and blunt head trauma. Heterotopic ossification is defined as calcification occurring at an abnormal location, which is usually within the soft tissue around the fracture. As it is related to bleeding around the soft tissue, the same risk factors with synostosis are also relevant with heterotopic ossification.

20.5 Complications

Some complications are related to cast applications. Fractures in the pediatric population are usually healed nonoperatively or by minimally invasive surgeries. Therefore, cast applications are widely used. If a short leg cast has been applied longer than needed, it is inevitable to irritate the posterior knee joint when the knee is flexed. This is similar for a short arm cast and the elbow. The most common complication after cast application is **rigidity or stiffness**, which is realized after cast removal. Although it is common, children have a great chance of handling this. If the fracture has healed anatomically, rigidity will resolve within a few weeks. Disuse osteopenia is another complication of cast application, especially when it has been used for a long time. Due to these complications, an orthopedic surgeon

should not only have advanced surgical capabilities but also be able to apply improved casting techniques.

As most casts (e.g., plaster of Paris and fiberglass) harden through exothermic reactions, there is a risk for **thermal cast complications**. Another common complication of casts is **localized pressure sores**. The posterior of the heel and the malleoli of the ankle are most susceptible for the pressure sores. However, the most catastrophic complication of cast usage is ischemic problems. A tightly applied cast, cast application on a swelling extremity, or not obeying the elevation rule can lead to increasing pressure in the compartments. Children—especially those younger than age 3—may not express themselves, and parents may think that the pain their child is suffering is due to the fracture instead of compartment syndrome. A **Volkmann's ischemic contracture** can be seen by this mechanism after elbow trauma such as a supracondylar humeral fracture. Therefore, the cast should be applied properly, the family should be informed of possible emergency situations, and the child should be discharged from the hospital after being under observation for 1–2 h following the cast application. Another problem is the cast "removal issue." The noise of the machine or the size of the scissors may cause the removal of a cast to be a real problem for children, especially younger ones.

20.6 Imaging

Direct radiographs are usually adequate for the diagnosis of the fracture pattern. However, fractures close to the joint should be evaluated with computerized tomography. Magnetic resonance imaging would be useful to indicate the effect of the injury on the epiphysis in the fractures adjacent to the physis. According to the Salter-Harris classification system, type I and type V epiphyseal fractures are seen to be similar at direct radiographs.

Families should be informed about the possibility of emergency situations after cast applications. If the fracture is adjacent to the epiphysis, the family should also be informed of possible limb-length discrepancy and the possibility of deformity occurrence a few years after the injury. The most common age for children to have a deformity after ankle-physeal fracture is 11–13 years. These types of fractures are called triplanar fractures, and they are a combination of both Salter II and Salter V physeal injuries.

20.7 Treatment

The treatment of pediatric fractures is generally nonoperative due to the remodeling ability of the bones. Treatment is decided according to the patient's age, presence of open fracture, and fracture characteristics such as location, soft tissue degree of angulation, and displacement.

Surgery is indicated in multiple fractures, open fractures, neurovascular injuries, and some pathologic fractures. Reduction loss during conservative treatment is another surgical indication. **Displaced metaphyseal fracture at the distal radius is typically unstable** in cast immobilization. Care should be taken, and weekly radiographs should be performed to follow the loss of reduction. Physeal locations, femoral neck fractures, and displaced elbow fractures need K-wire or screw fixation. As a patient ages, remodeling capacity decreases. Therefore, femur, tibia, and forearm fractures as well as unstable pelvic injuries require surgical treatment, especially in patients older than 10 years of age.

Fractures close to the epiphysis can result in growth disturbances. In these conditions, various strategies such as physeal bar resection, epiphysiodesis, and osteotomies for correction of the deformity can be done. The primary aim of the treatment is to reach the remaining potential for growth. The development of deformities depends on the location of the affected physis, bone maturity, and duration of time between injury and treatment. Epiphysiodesis is helpful in preventing the occurrence of deformity and is applied to the contralateral side. If a deformity has occurred, corrective osteotomies are needed.

Take-Home Message

- Pediatric trauma requires a detailed evaluation by emergency caregivers. After evaluation of life-threatening possibilities, attention is given to musculoskeletal injuries.
- Abuse should be considered in the etiology of pediatric injuries.
- The mechanism of injury, status of the soft tissue, age of the patient, and fracture characteristics are important in the treatment strategy.
- Due to remodeling capacity, most of the pediatric fractures are treated nonoperatively; however, there are some indications for surgical fixation. These include multiple fractures; open fractures; physeal injuries; neurovascular injuries; some pathologic fractures; reduction loss during conservative treatment; femoral neck fractures; displaced elbow fractures; femur, tibia, and forearm fractures; and unstable pelvic fractures in children older than 10 years of age.
- As patients age, the remodeling capacity decreases.
- After physeal injuries, development of deformities depends on the location of the affected physis, bone maturity, and duration of time between injury and treatment.
- Cast treatment requires meticulous neurovascular examination.

Summary

The management of pediatric fracture care differs from that of adulthood in terms of anatomic variations. Due to a high remodeling capacity, most of these fractures are treated conservatively. However, surgery would be necessary for particular types of fractures, especially in adolescents and in those of older age. Unstable fractures, open fractures, displaced epiphyseal fractures, and multiple fractures are some of the surgical indications. Both the surgical and nonoperative treatments are not without complications. Parents should be informed about cast- and fracture-specific complications such as compartment syndrome, wound problems, growth disturbances, deformity, and restricted range of motion.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Which bone is the most commonly fractured during childhood?
   (a) The radius, especially the distal radius, is the most common.
   (b) The ulna, especially the distal ulna, is the most common.
   (c) The femur, especially the distal femur, is the most common.
   (d) The femur, especially the proximal femur, is the most common.
2. A 10-month-old female patient is admitted to the outpatient clinic. Her parents are telling about a fall that happened while she was crawling. There is tenderness on her left thigh. What should be the most probable diagnosis?
   (a) Child abuse should be considered for those under ambulatory age
   (b) Torus fracture
   (c) Proximal radius fracture
   (d) Toddler fracture
3. Typical growth of the upper extremity mostly occurs at:
   (a) The proximal humerus and distal radius epiphysis
   (b) The proximal radius and distal ulnar epiphysis
   (c) The distal humerus and distal radius epiphysis
   (d) The distal humerus and proximal radius epiphysis
4. Typical growth of the lower extremity mostly occurs at:
   (a) The distal femur and proximal tibia epiphysis
   (b) The proximal femur and distal tibia epiphysis
   (c) The distal talus and proximal tibia epiphysis

(d) The distal tibia and proximal tibia epiphysis

5. Which type of fracture does the Thurstan Holland fragment refer to according to the Salter-Harris classification system?
   (a) The Thurstan Holland fragment defines an epiphyseal fracture extending to the metaphysis, so it is a type II epiphyseal fracture according to the Salter-Harris classification system.
   (b) The Thurstan Holland fragment defines an epiphyseal fracture extending to the metaphysis, so it is a type I epiphyseal fracture according to the Salter-Harris classification system.
   (c) The Thurstan Holland fragment defines an epiphyseal fracture extending to the metaphysis, so it is a type III epiphyseal fracture according to the Salter-Harris classification system. However, in some cases, the epiphysis may be influenced entirely as compression according to the velocity of the trauma, and a combination of both type II and V can also be considered.
   (d) The Thurstan Holland fragment defines an epiphyseal fracture extending to the metaphysis, so it is a type IV epiphyseal fracture according to the Salter-Harris classification system.

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Part V

Osteoarthritis

Osteoarthritis

21

Jawad H. Abidi, Flavia M. Cicuttini, and Anita E. Wluka

Overview

Osteoarthritis can affect any synovial joint, and it is characterised by an abnormal response to tissue injury in cellular and extracellular pathways, including activation of the inflammatory response via the innate immune system.

21.1 Definition

Osteoarthritis can affect any synovial joint, where it may involve all the tissues in the joint. It is characterised by an abnormal response to tissue injury in cellular and extracellular pathways, including activation of the inflammatory response via the innate immune system. The resulting abnormalities in tissue metabolism lead to anatomical and physiologic derangements (described below). These changes result in osteoarthritis.

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21.2 Epidemiology

Osteoarthritis is the most common form of arthritis in the world. It affects 10–15% of the adult population, or about 250 million people worldwide. It causes significant morbidity. Over the last half century, the proportion of the population affected by osteoarthritis in developed countries has more than doubled, related to increases in life expectancy and steadily increasing body mass index (BMI). The incidence and prevalence of osteoarthritis have historically been lower in low-income compared to high-income countries although it is likely to change with the ever-ageing and increasingly obese international population.

21.3 Risk Factors

Osteoarthritis is a heterogeneous condition, with both modifiable and non-modifiable risk factors implicated in its pathogenesis. The relative importance of each risk factor differs in an individual and the affected joint.

Age is strongly associated with osteoarthritis, with 10–13% of adults affected by the age of 60. The incidence of osteoarthritis increases with age, with the prevalence of symptomatic osteoarthritis (of the knees, hands or hips) in the mid-80s being estimated to be 45–55% in the United States.

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Females are more likely to experience osteoarthritis particularly after the age of 50, although prior to that OA is more common to men. Where present in women, OA may have a more severe trajectory, with worse pain and functional deficits than men. Given the sexually dimorphic distribution of OA, a hormonal component to the pathogenesis has been proposed. Although there is evidence for endogenous hormones and the menopause to be associated with new diagnoses of OA, the role of exogenous hormones is less clear.

There is likely to be a **genetic component**, with a positive family history of osteoarthritis conferring increased risk. Having a grandparent, parent or sibling with OA presents increased risk of generalised OA, spinal OA, hip OA and hand OA (generalised and hand OA being most implicated). Multiple genetic markers have been postulated, although none individually pose significantly increased risk.

Overweight/obesity increases the risk of osteoarthritis most strongly at the knee, but also to a lesser degree at the hips and hands. The effect of obesity is likely due to a combination of mechanical and systemic factors related to meta-inflammation.

Previous joint injury increases the risk of OA. Injury to the anterior cruciate ligament injury in the late teens and 20s has been shown to be associated with developing symptomatic and radiological knee OA within 7–15 years. Both mechanical and inflammatory factors have been implicated. Joint injury increases the risk of OA to the affected joint.

Occupational factors/strain, including lifting, significant labour-related work and farm work, in particular, has been associated with increased risk of developing osteoarthritis, particularly of the knee and hips.

21.4 Pathogenesis

Osteoarthritis can affect any synovial joint, ultimately involving all tissues within the joint, resulting in joint failure. The underlying mechanisms by which its risk factors work have not been fully elucidated. Despite the varied initial insults, the pathological findings are similar in all joints affected by osteoarthritis.

Articular hyaline cartilage has been viewed as the target tissue of OA. Initially, cartilage swells due to increased hydration associated with glycosaminoglycan loss, with anabolic changes. With the resultant changes to tissue composition, collagen type II breakdown occurs, leading to cartilage compression and increased tissue pressure, with catabolic effects becoming more prominent, with inflammatory change and cartilage degradation. The hyaline cartilage fibrillates and develops fissures, resulting in erosions and subsequent cartilage loss.

Bone changes are notable from early in disease. Increased subchondral bone turnover is seen in early disease, associated with the extension of blood vessels into the previously avascular cartilage. These subchondral changes are associated with the development of bone marrow lesions, seen on MRI. Bone marrow lesions are evidence of the tight link between bone and cartilage metabolism in OA, as their presence is associated with increased cartilage loss. Both metabolic and mechanical factors contribute to bone changes in osteoarthritis, with progressive disease associated with increased joint size, manifesting clinically as bony hypertrophy and radiographically as subchondral bone sclerosis, subchondral cyst and osteophytes. Increased severity of OA has been defined by the formation and growth of osteophytes, the growth of which is affected by metabolic and biomechanical factors.

Synovial inflammation, seen as synovitis with or without effusion, is often present in osteoarthritis, despite it being viewed as a non-inflammatory condition. Synovial inflammation may be seen in early osteoarthritis, characterised by lining cell hyperplasia, fibrosis and increased vascularity. The inflammatory cells that are present predominantly originate from synovial macrophages, with a contribution from synoviocytes and chondrocytes. Traditionally, OA is not considered an inflammatory arthritis; less aggressive synovitis is often present than is seen in rheumatoid arthritis.

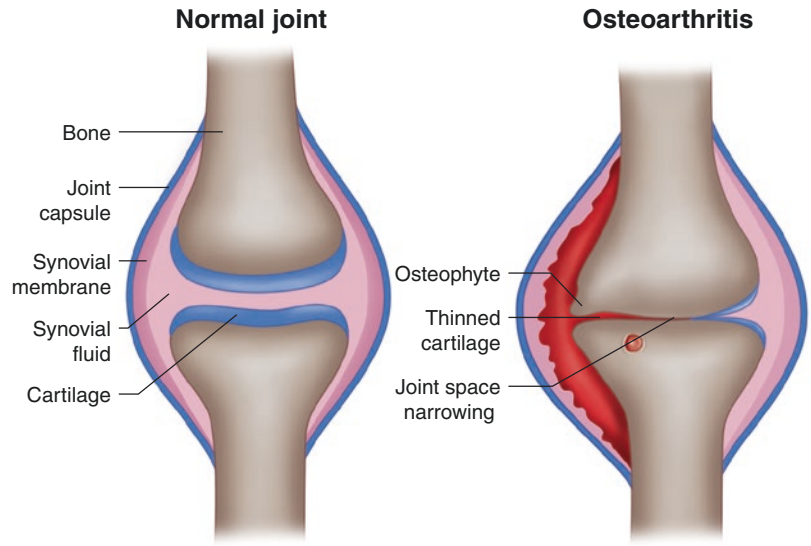

Fig. 21.1 Typical changes seen in osteoarthritis. On the left-hand side of the joint, structures are normal

Meniscal and ligamentous pathology is often present in OA. The pathological changes seen in these tissues are similar to those seen in cartilage with disruption of the matrix, fibrillation, calcification tissue, cell death and ligamentous tears being common. At the knee, meniscal degeneration is often present.

Muscle adjacent to joints affected by OA is often wasted. Joint pain causes muscle inhibition and wasting. However, there is some evidence that it may precede incident OA, with muscle loss resulting in poor joint mechanics, predisposing to loading injury and OA development.

Some of the changes seen in OA are demonstrated in Fig. 21.1.

21.5 Classifications of OA

OA may be classified as primary, where it occurs with no clear precipitant identifiable, or secondary, where its presence relates to another factor. Secondary causes of OA include significant joint injury or instrumentation, obesity, previous sepsis of the joint, metabolic defects (haemochromatosis and Wilson's disease), avascular necrosis of subchondral bone or response to other forms of arthritis that damage the joint initiating the pathogenesis of OA.

Osteoarthritis has also been classified according to the distribution of affected joints, as either local or generalised osteoarthritis. In local arthritis, a single joint is involved. In generalised osteoarthritis, usually hands, particularly first CMC and DIP joints are affected in association with knee OA.

21.6 Clinical Features of OA

The clinical features of osteoarthritis include use-related pain, limited (<30 min) morning stiffness, short duration of gel phenomenon (<5 min), muscle wasting and loss of function. Joint pain tends to wax and wane, with periods of pain, which may last for hours to weeks, interspersed with less severe symptoms. Some features are common to any affected joint, including pain that is worse on movement and in the evening, joint-line tenderness, bony deformity, instability, crepitus, limited movement and function. Joint-specific manifestations are as follows.

Hand OA may be associated with difficulty completing activities of daily living, loss of fine motor skills and pain that is worse on movement. On examination, the following features are seen: wasting of the intrinsic muscles of the hand, osteophyte formation presenting clinically as Heberden's

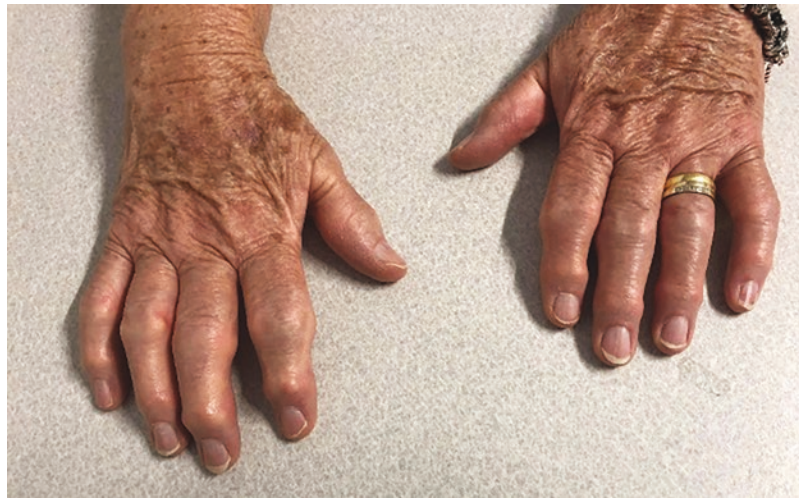

Fig. 21.2 The typical changes seen in hand OA. Note the presence of Heberden's and Bouchard's nodes

(distal interphalangeal joints) and Bouchard's nodes (proximal interphalangeal joints), base-of-thumb involvement and carpometacarpal joint involvement. Figure 21.2 demonstrates these changes.

Knee OA is characterised by pain that is worse particularly at the end of the day and on movement, crepitus, a sensation of instability and giving way. On examination, findings may include valgus or varus deformity, antalgic gait and quadriceps muscle wasting. The joint line may be tender, with bony expansion, soft tissue swelling, joint effusion and swelling in the popliteal fossa (Baker's cyst).

Hip OA results in inguinal, buttock or knee pain, related to innervation of the hip by the femoral and sciatic nerves. An antalgic gait and shortening of affected limb may be observed, with pain on internal rotation of affected hip and loss of hip flexion.

21.7 Diagnosis

For a diagnosis of osteoarthritis to be made, the presence of the following clinical features are often sufficient:

- Joint pain exacerbated by movement
- Age greater than 45 years of age
- Morning stiffness less than 30 min
- No other systemic inflammatory features are present

Investigations may be needed to exclude other conditions.

The radiological changes in OA reflect the pathological changes—joint space narrowing, osteophyte formation, subchondral cysts and bony sclerosis. Some joint-specific changes are presented in Table 21.1.

Table 21.1 Some specific features of osteoarthritis of various joints seen on X-ray and MRI

Imaging modality	Changes seen
X-ray	• Hand: Involvement of carpometacarpal joints, base of thumb, proximal and distal interphalangeal joints. Involvement of the second and third metacarpophalangeal joints raises the possibility of haemochromatosis • Hip: Cam and pincer deformity, dysplasia of the femoral head (Fig. 21.3a) • Knee: Narrowing of the patella-femoral (on skyline view) and patella-tibial joint space, and valgus and varus deformity, may be seen depending on the specific compartment involved (Fig. 21.3b)
MRI	Bone expansion, cartilage loss with thinning and cartilage defects, meniscal tear, effusion, synovitis, subchondral bone marrow lesions and subchondral bone cysts

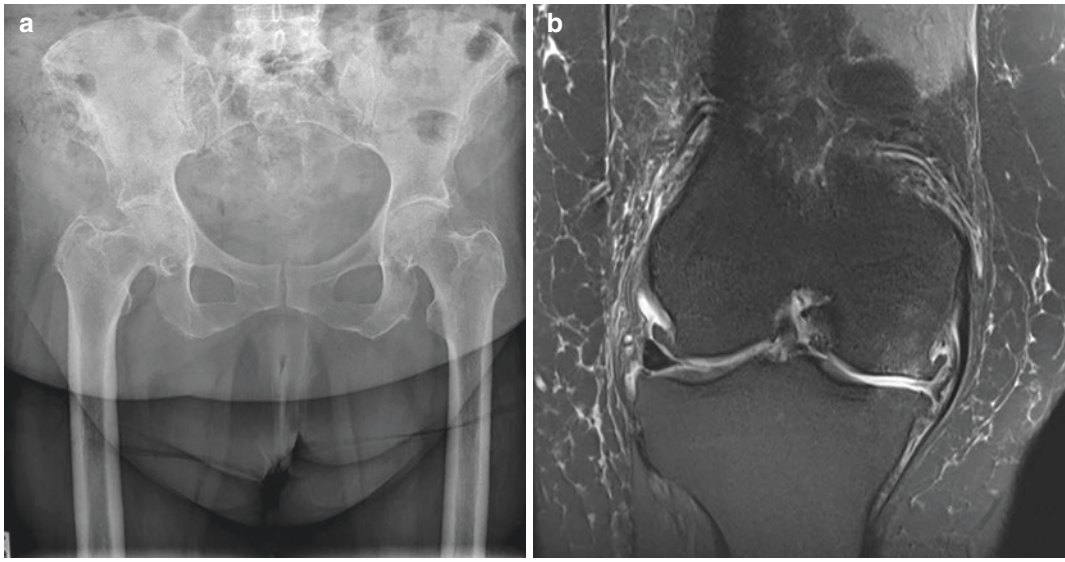

Fig. 21.3 (**a**) A plain X-ray of bilateral hips, demonstrating severe osteoarthritis of the right hip, with loss of joint space, subchondral sclerosis and osteophytes. (**b**) A knee MRI demonstrating changes seen in OA. Note the cartilage thinning, osteophytes, subchondral bone oedema and a synovial effusion

21.8 Management

Optimal management of osteoarthritis is holistic and may require multidisciplinary interventions. It requires a caring therapeutic approach, as patients often feel as if their condition is hopeless. Patients want and need to understand their condition, its prognosis, self-management and prevention. The fundamental components of care relate to weight management and physical activity and/or exercise: these are the only two factors that have been shown to reduce progression. Patients' highest priority is pain control. To achieve the above, the Osteoarthritis Research Society International (OARSI) has recently developed new management guidelines.

21.9 Non-pharmacological Management

Exercise and weight management (with weight loss in those who are overweight) are key. Land- and water-based exercise with or without weight loss has been demonstrated to be of benefit in reducing OA-related pain. Land-based exercise has short-intermediate-term improvements in pain scores even when ceased. However, any weight loss has been shown to reduce the progression of OA. Approximately 5 kg of weight loss reduces the risk of developing knee OA in one study. In contrast, weight gain has been shown to increase pain, reinforcing the need to promote weight maintenance.

When weight loss and exercise are inadequate, involvement of a physiotherapist to improve muscle strength and minimise pain may be considered. Cognitive behavioural therapy has been shown to have some efficacy at addressing associated depression and/or complex pain syndromes in patients afflicted with OA.

Gait aids can also be used in the management of osteoarthritic pain pending more definitive management. Use of a single-point stick for example on the contralateral side in unilateral osteoarthritis of the knee/hip has been shown to reduce pain.

21.10 Pharmacological Management

First-line pharmacological therapy includes topical therapy with non-steroidal anti-inflammatories (NSAIDs) and capsaicin, which can be used as needed. Topical NSAIDs' adverse effects are minimal (local skin reactions), and these medications are absorbed minimally into the systemic circulation and are thus safer than their oral counterparts.

Second-line agents which can be used additionally include oral non-selective NSAIDs (with proton pump inhibitors) and paracetamol as needed. However, consideration must be made to the patient's comorbidities. If the patient has hepatic impairment, regular paracetamol would then be discouraged; patients with Barrett's oesophagus, gastritis and peptic ulcer disease may be better suited to COX-2-specific inhibitors (e.g. celecoxib). Patients with significant renal and/or cardiovascular disease should not be placed on oral NSAIDs for risk of acute deterioration.

Specifically for knee OA, glucocorticoid (IAGC) or hyaluronan (IAHA) can be injected into the joint, although these offer modest short-term therapeutic benefit at best. Note that intra-articular glucocorticoid may result in elevated blood sugar levels in patients with impaired glucose metabolism or diabetes mellitus, which may require additional care.

In patients with overlying depression, complex pain syndromes (with allodynia or spreading of pain) or other significant comorbidities which preclude the use of oral NSAIDs, duloxetine may be considered.

As there is no long-term evidence of benefit from opiates, and their use has significant adverse consequences, their use should be strongly discouraged. Although the use of complementary medicines is widespread, used by more than approximately 40% of patients, there is little strong data to support their use.

There is increasing interest in targeting patient subgroups to tailor therapies to those who are most likely to benefit. A recent example was the finding that turmeric (*Curcuma longa* extract) reduces knee pain in those with underlying synovitis identified via MRI or ultrasound. These results highlight the importance of targeted therapy, although this is still quite a novel area.

21.11 Surgical Management

End-stage joint disease in OA can be treated with joint replacement, particularly in the case of knee and hip OA. Care must be taken in selecting the timing for joint replacement as they have a limited duration of function, and results for revision surgery are poor compared to primary joint replacement. This may be considered if non-pharmacological management is not improving function appropriately and there is ongoing patient dissatisfaction and reduction in quality of life. Care should be taken to assess and treat pain sensitisation and affective disorders prior to considering surgery, as both of these increase the likelihood of poor joint replacement outcomes.

Take-Home Message

- Osteoarthritis is a common condition which is becoming more prevalent in increasingly aged and obese world population.
- The clinical picture is often enough to make the diagnosis; radiological imaging is not always required.
- The typical pathological/radiological features seen in osteoarthritis are osteophytosis, joint space narrowing, subchondral sclerosis and subchondral cyst formation.
- Weight management, physical activity and exercise and analgesic form the foundation of care for osteoarthritis.
- The management of osteoarthritis requires a caring therapeutic relationship with the patients, as it has the potential to affect all facets of their life.

Summary

Osteoarthritis is the most common form of arthritis in the world. It affects 10–15% of the adult population, or about 250 million people worldwide. Osteoarthritis can affect any synovial joint, where it may involve all the tissues in the joint, ultimately resulting in joint failure. It is characterised by an abnormal response to tissue injury in cellular and extracellular pathways, including activation of the inflammatory response via the innate immune system. The resulting abnormalities in tissue metabolism lead to anatomical and physiologic derangements. Osteoarthritis is a heterogeneous condition, with both modifiable and non-modifiable risk factors implicated in its pathogenesis. The underlying mechanisms by which its risk factors work have not been fully elucidated. OA may be classified as primary, where it occurs with no clear precipitant identifiable, or secondary, where its presence relates to another factor. The clinical features of osteoarthritis include use-related pain, limited (<30 min) morning stiffness, short durations of gel phenomenon (<5 min), muscle wasting and loss of function. The radiological changes in OA reflect the pathological changes—joint space narrowing, osteophyte formation, subchondral cysts and bony sclerosis. Optimal management of osteoarthritis is holistic and may require multidisciplinary interventions. Exercise and weight management (with weight loss in those who are overweight) are key. First-line pharmacological therapy includes topical therapy with non-steroidal anti-inflammatories (NSAIDs), which can be used as needed. End-stage joint disease in OA can be treated with joint replacement, particularly in the case of knee and hip OA.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. What are the main modifiable and non-modifiable risk factors implicated in osteoarthritis?
   (a) Modifiable: body weight, joint injury, occupation strain, alcohol consumption; non-modifiable: age, female gender, family history/genetics, menopause
   (b) Only modifiable: body weight, joint injury, occupation strain, alcohol consumption
   (c) Only non-modifiable: age, female gender, family history/genetics, menopause
   (d) Modifiable: age, gender, joint injury, occupation strain, alcohol consumption; non-modifiable: smoking, family history/genetics, menopause
2. What are the main clinical features that differentiate osteoarthritis from other arthropathies?
   (a) OA is characterised by pain worse after joint use, <0.5 h of morning stiffness, <5 min of gel phenomenon, muscle wasting and loss of function. On examination, there will be joint-line tenderness, bony deformity and limited range of motion
   (b) OA is characterised by pain worse after joint use, <0.5 h of night stiffness, <5 min of gel phenomenon, muscle wasting and loss of function. On examination, there will be joint-line tenderness, bony deformity and limited range of motion

(c) OA is characterised by night pain worse after joint use. On examination, there will be limited range of motion without joint deformities
(d) OA is characterised by pain present during the whole day. On examination, there will be joint-line tenderness, bony deformity and limited range of motion

3. What are the radiological features seen on plain film that characterise osteoarthritis?
   (a) Osteophytosis, joint space narrowing, subchondral sclerosis and subchondral cyst formation
   (b) Osteoporosis, joint space narrowing and subchondral cyst formation
   (c) Osteoporosis, joint space widening, subchondral sclerosis and subchondral cyst formation
   (d) Joint space widening and subchondral cyst formation
4. What are the main management principles when treating patients with osteoarthritis?
   (a) Via a multidisciplinary approach, minimising pain, maximising and preserving function, engagement with activities of daily living, exercise and weight maintenance
   (b) Via a symptomatic approach, with NSAIDs
   (c) Via a surgical approach. Joint replacement surgery is indicated in each case of osteoarthrosis
   (d) Ice therapy, rest and immobilisation
5. What is the role of opioid therapy in osteoarthritis?
   (a) No role for opioid treatment outside of acute periods perioperatively post-joint replacement
   (b) Opioids are useful in the early stages of the disease
   (c) Opioids should be considered as first-line therapy
   (d) Opioids are maintained 3 months after surgery

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Part VI

Tumors

General Concepts

22

Ryan T. Voskuil and Joel L. Mayerson

Overview
This chapter focuses on the general concepts of musculoskeletal tumors.

22.1 Definition

Musculoskeletal tumors comprise benign and malignant varieties of both the bone and soft tissues. The general concepts of these tumors include the epidemiology, causes, workup, and treatment.

22.2 Epidemiology

Malignant bone and soft tissue tumors are rare with roughly 2500 bone and 12,000 soft tissue sarcomas occurring in the United States yearly. Benign bone and soft tissue masses are much more common. Age is important to consider. When it comes to primary malignant bone tumors, osteosarcoma and Ewing's sarcoma typically present in patients less than 20 years of age while chondrosarcoma is more commonly seen in those over 40. Malignant bone lesions are logarithmically more likely to represent metastatic disease, multiple myeloma, or lymphoma versus a primary bone sarcoma in patients over 40 years old.

22.3 Etiology

For the majority, there are no identifiable genetic or environmental factors to date that lead to the development of a sarcoma, as most mutations identified are somatic. This is due in large part to the rarity of the disease and the fact that there are many subtypes of sarcoma that are studied uniformly in a retrospective manner. We do know that some bone sarcomas rarely can arise secondarily from an area of radiation, osteonecrosis of bone, or other benign bone tumors. There are some inherited disorders associated with an increased risk of developing a sarcoma.

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These include Li-Fraumeni (p53 gene mutation), neurofibromatosis (NF-1 alterations), and Rothmund-Thomson syndrome (RECQL4 gene mutations).

22.4 Classifications

There are a few classifications frequently used in musculoskeletal oncology that are worth mentioning. Dr. Enneking developed a classification for both benign and malignant bone tumors in the 1980s that was adopted by the Musculoskeletal Tumor Society and is the most commonly used for staging and direction of treatment. Benign bone tumors are broken down into latent, active, and aggressive. Latent benign bone tumors are those that do not grow much and have a limited natural history like a non-ossifying fibroma (NOF). Active lesions, like a chondroblastoma, continue to grow but do not expand normal bone and respect the cortical boundary. Aggressive lesions, like giant cell tumors, often appear locally malignant and can destroy cortical bone and expand the normal architecture. The malignant bone tumor classification is broken down into low and high grade, intra- and extracompartmental, and presence of metastatic disease whether that be skip, lymph node, or distant. There is no classification for benign soft tissue masses, and the American Joint Commission on Cancer (AJCC) soft tissue sarcoma staging system is the most popular and is now in its eighth edition. This system is more involved and considers grade, size, and depth, along with nodal and distant metastatic status. A summary of the characteristics of benign and malignant tumor is reported in Fig. 22.1.

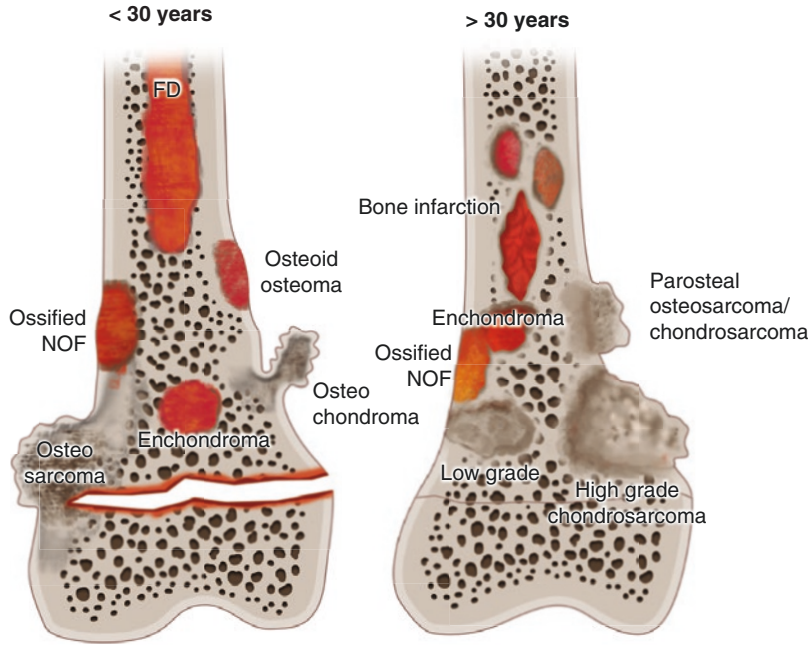

Fig. 22.1 Summary of the most important characteristics of benign and malignant tumors

22.5 Diagnosis

22.5.1 History

A thorough history is important when evaluating a person with a suspected bone or soft tissue tumor. Onset, location, duration of symptoms, character of pain, associated factors, timing of pain pattern, and severity are foundational questions. Medical, personal, and family cancer history should also be addressed. Soft tissue tumors are often painless, unless they grow to compress surrounding neurovascular structures, while bone tumors typically present with a constant, dull, pain pattern often present at night. Bone tumors can be at high risk for pathologic fracture, so this should always be carefully assessed as well.

22.5.2 Physical Exam

The physical exam should focus on assessing gait, location of pain, swelling, and mass characteristics. Concerning masses are often deep, firm, well circumscribed, and fixed, although they can be present superficial to fascia as well. The involved extremity should be evaluated for neurovascular status, joint motion, skin changes, swelling, and locoregional lymph node involvement with comparison to the contralateral arm or leg.

22.5.3 Imaging

22.5.3.1 Local Imaging

Plain radiographs of the entire involved extremity with inclusion of the joint above and below should always be the first imaging study ordered during any tumor workup. In bone tumors, it is important to ask four questions developed by Dr. Enneking, one of the fathers of musculoskeletal oncology: (1) age of patient, (2) location in bone, (3) what is tumor doing to bone, (4) what is bone doing to the tumor/how is it responding, and (5) hint of histology (bone, cartilage, or fibrous matrix forming). With soft tissue tumors, it is vital to look for soft tissue shadows, bone changes, and associated calcifications or bone formation within or around the soft tissue mass.

A similar but more involved diagnostic algorithm for bone tumors developed by Dr. Mayerson provides an excellent framework by answering five questions: type of matrix (bone, cartilage, fibrous, cystic calcium), type of bone (normal, reactive, tumor produced), pattern of destruction (geographic, moth-eaten, permeative), age (<20, 21–40, >40), and location in bone (epiphysis, metaphysis, diaphysis).

Magnetic resonance imaging (MRI) is usually the next necessary imaging study ordered. In bone tumors, the T1 sequence helps delineate its true location and relation to surrounding structures including nerves and vessels. It is vital to scan the entire bone to evaluate for skip metastases. MRI is important for soft tissue tumors as well for the same reasons. Contrast sequences help characterize the mass and delineate cystic versus solid lesions, with heterogeneity seen on T1 and T2 sequences concerning for malignancy. In general, tumors with peri-tumoral edema, periosteal reaction, cortical destruction, and a soft tissue mass are malignant until proven otherwise.

22.5.3.2 Staging Studies

Staging studies are an important aspect of the imaging workup with particular imaging modalities employed to evaluate for skip, lymph node, and distant metastatic spread. These studies are vital from both a treatment planning standpoint and overall prognosis. Whole-body bone scans are used to evaluate other sites of bone disease in malignant bone tumors. Multiple myeloma can be cold on a bone scan, so a skeletal survey is often employed in this scenario. Computed tomography (CT) scans of the chest and sometimes the abdomen and pelvis are critical in both malignant bone and soft tissue tumors to evaluate for metastatic disease as the lungs are by far the most common location of metastatic disease in sarcomas. PET/CT scans are gaining popularity for staging both bone and soft tissue malignancies.

22.5.4 Labs

Labs are rarely diagnostic, but they aid in painting a diagnostic picture. A complete blood count (CBC), chemistry (CMP), and inflammatory markers (C-reactive protein {CRP}) are a great place to start. Alkaline phosphatase and lactate dehydrogenase are helpful with suspected primary bone tumors. The metastatic carcinoma workup should include a calcium and protein electrophoresis in addition to a CBC, CMP, and CRP. Hypercalcemia when present can be life threatening and should be quickly addressed.

22.6 Differential Diagnostic Considerations

22.6.1 The Biopsy

The biopsy is arguably the most important aspect of the diagnostic processes as a verification of histology is usually required before initiation of treatment (Table 22.1). Open biopsy has long been the gold standard and is most often utilized in primary bone tumors without a soft tissue mass, to confirm metastatic disease prior to prophylactic stabilization, and when other techniques are nondiagnostic. Open biopsy in the setting of a bone sarcoma is prone to errors that can lead to bleeding and contamination with occasional loss of limb salvage ability. Complications of open biopsy can be avoided by making the incision in a longitudinal manner in line with the eventual incision for definitive surgery, through one compartment, avoiding neurovascular structures, and with meticulous hemostasis. The open biopsy tract should be excised during the definitive surgery to minimize the risk of local recurrence in that location. Core needle biopsy (CNB) has gained popularity in many centers due to its versatility without the need for biopsy tract excision. CNB can be utilized on palpable soft tissue masses in the office or with image guidance when gathering tissue samples from difficult locations around neurovascular structures or in the deep pelvis. Fine needle aspiration (FNA) is an office-based biopsy technique that has demonstrated significant benefits in centers with pathologists experienced with this method. The FNA can be used on any palpable soft tissue mass in the office without the need for local anesthetic and the least risk for bleeding or contamination among the three techniques. A preliminary diagnosis can often be delivered to the patient within 30 min of the procedure while minimizing cost and maximizing staff and resource utilization.

Table 22.1 Surgical principles of open biopsy

Incision should be in line with planned incision for limb salvage, if the surgeon does not know what this is, the biopsy should not be done. Transverse incisions should be avoided
The most direct route should be taken through a single compartment with avoidance of contamination of the neurovascular bundle and other compartments
Strict hemostasis and hematomas are contaminated with tumor cells
If a tourniquet is applied, gravity exsanguination should only be used
If a drain is used, it should be placed in line with the incision
Open biopsy of high-risk areas like the popliteal fossa, axilla, and carpal tunnel should be avoided as contamination of these areas could lead to amputation
Arthroscopy should not be employed as a primary method of biopsy for an intra-articular mass

22.7 Treatment

22.7.1 Surgical Management

The surgical management of musculoskeletal tumors demands a thoughtful and thorough plan. The primary goals of sarcoma surgery are twofold: oncologic and functional. The oncologic goal is ultimately the most important. Oncologic resections can be classified as intralesional, marginal, wide, and radical excision (Fig. 22.2). An intralesional excision is typically performed by entering the substance of the tumor and removing it in a piecemeal fashion. This is often utilized on benign cysts. Marginal excision involves removal of the tumor through the reactive zone, or the thin capsule surrounding many lesions where signal change is often seen on MRI. This technique is

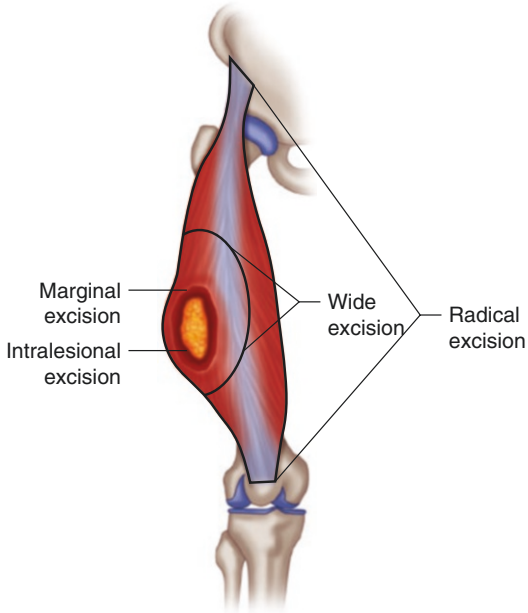

Fig. 22.2 Surgical resection margins as accepted by the musculoskeletal tumor society

performed on benign soft tissue masses like a lipoma. A marginal excision is not acceptable for resection of sarcomas as tumor cells are often present in the reactive zone. This is the reason why a wide excision is performed on primary bone and soft tissue malignancies. This method involves removing a tumor without ever seeing the tumor itself, by leaving a cuff of normal tissue around the mass as the margin, thus ensuring complete removal of all cancer cells. Radical resection refers to a removal of the entire involved compartment. This is rarely required and is usually performed when a wide excision would leave the compartment nonfunctional. Primary bone and soft tissue malignancies are usually treated with a curative wide excision in an attempt to remove all disease. When metastatic carcinoma, myeloma, or lymphoma involves the skeletal system and there is an impending or complete pathologic fracture, the bone affected usually requires stabilization for palliation.

The functional goal is a close second to the oncologic goal but is often more important to the patient. Curative surgery for sarcoma has swung from amputation to limb salvage with vast improvements seen in the latter. Over 90% of patients with extremity sarcomas can be treated with limb salvage. Amputation is now reserved for cases in which a wide excision with a negative margin cannot be achieved or when amputation would result in better function.

22.7.2 Adjuvant Treatments

Adjuvant therapy often includes chemotherapy, external beam radiation therapy, or both that have transformed the treatment of primary musculoskeletal tumors with vast improvements seen in rates of local recurrence, metastatic disease, and overall survival. Sarcoma treatment is usually broken down into three separate buckets when explaining options with the patient, with radiation and surgery addressing local tumor control while chemo represents the only systemic modality. Chemotherapy is utilized on all high-grade primary bone tumors with the exception of chondrosarcoma, which is not chemosensitive. It is often given in a standard protocol in a neoadjuvant (before surgery) fashion and then given after surgery adjuvantly. This allows some evaluation of the treatment effect in the middle of the chemotherapy, providing both prognostic and therapeutic information. Soft tissue sarcomas have traditionally not been treated with chemo, but many centers are utilizing certain regimens on younger patients with large tumors that are at high risk for metastasis or have metastatic disease at presentation.

Radiation therapy is typically applied to all high-grade deep soft tissue sarcomas and can be given either pre- or postoperatively. Preoperative radiation increases the risk of wound complications but improves the ability to achieve a negative margin. Postoperative radiation mitigates the wound complication issue but has more long-term side effects that include fibrosis, lymphedema, fracture, and an increased risk of radiation-induced sarcoma since the radiation field and dose are higher. Radiation and surgery together have demonstrated better local control of high-grade soft tissue sarcomas with a controversial effect on overall survival. The future of

cancer treatment rests in the evolving discovery of molecular and genetic markers that are specific to cancer cells and targetable. This would individualize the systemic treatment and eventually replace the shotgun approach of traditional chemo. Currently, certain sarcoma subtypes are showing promising response to targeted therapy with a minimal side effect profile.

Take-Home Message

Musculoskeletal tumors encompass a wide array of benign and malignant bone and soft tissue pathologies. The musculoskeletal oncology principles discussed in this overview can be applied to all tumors and aid in diagnosis and treatment. It is important that all medical providers consider sarcoma in their differential and follow the principles to avoid potential errors that could compromise limb salvage or even lead to a decrease in overall survival when evaluating bone and soft tissue lesions.

Summary

In summary, benign and malignant tumors cover a heterogenous group of pathologies. Adherence to these general concepts will assist with diagnosis and provide a treatment framework. Inappropriate workup can lead to substantial delays in diagnosis and increased morbidity and mortality. Centers that specialize in sarcoma treatment are best equipped to handle these complex problems.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Which of the following is not a principle of open biopsy?
   (a) Meticulous hemostasis
   (b) Transverse incision
   (c) Incision in line with definitive surgical approach
   (d) Single compartment
2. What is the single best imaging study when evaluating a soft tissue mass?
   (a) MRI with and without contrast
   (b) CT scan
   (c) Plain X-ray
   (d) MRI without contrast
3. What is the best initial imaging study when evaluating a bone lesion?
   (a) MRI with and without contrast
   (b) CT scan
   (c) Bone scan
   (d) Plain X-ray
4. What is the most common presentation of a soft tissue sarcoma?
   (a) Painful superficial, mobile, soft mass
   (b) Painless deep, fixed, firm mass
   (c) Painless superficial, mobile, soft mass
   (d) Painful deep, fixed, soft mass
5. What is the most common presentation of a malignant bone tumor?
   (a) Painless incidental finding on plain X-ray
   (b) Constant, dull, achy pain present at rest and at night
   (c) Constant, sharp pain present more with activity that goes away with rest
   (d) Intermittent shooting pain worse with certain positions

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Benign Tumors

23

R. Lor Randall

Overview
This chapter focuses on the general concepts of primary benign musculoskeletal tumors.

23.1 Definition

Benign musculoskeletal tumors have a broad spectrum of clinical presentation and radiographic findings that often correlate with the underlying biologic behavior. They may present as clinically asymptomatic incidental findings or with rapidly progressive symptoms with corresponding aggressive bony destruction. Thorough history and physical and systematic evaluation of radiographs are often sufficient to establish a differential diagnosis. Selective use of advanced imaging will often confirm a diagnosis though biopsy is sometimes needed to establish a definitive diagnosis and rule out a malignancy.

Accurate and timely diagnosis of benign bone lesions is important for a multitude of reasons. First, in aggregate, they are very common, often found incidentally by practitioners without expertise in interpreting the imaging findings, and until a definitive diagnosis is rendered, this can cause significant distress to patients and families. Accurate diagnosis and monitoring with radiographs are a simple way to provide reassurance and avoid complex, expensive, and invasive workup for benign latent incidental findings. Secondly, there are benign bone lesions that behave in a biologically aggressive manner and can represent limb-threatening clinical problems that need skilled subspecialty treatment. Lastly, there is significant overlap between the clinical and radiographic presentation of benign and malignant bone tumors, and efficient accurate diagnosis and referral to a treating center are essential in preventing the life- and limb-threatening complications that can be associated with delayed diagnosis or early mismanagement of malignancy.

23.2 Clinical Evaluation

Readily available information such as the age of the patient and the location of the primary complaint can form the basis of an initial differential diagnosis, which is narrowed further with thorough history, physical evaluation, and imaging. These two simple questions, (1) what is the age of the patient and (2) where is the lesion, can help to narrow the number of possibilities and focus

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Table 23.1 Diagnoses by age

Age	Benign	Malignant
0–5 years	Langerhans cell histiocytosis Infection	Leukemia Metastatic neuroblastoma Metastatic rhabdomyosarcoma
5–20 years	Non-ossifying fibroma Fibrous dysplasia Aneurysmal bone cyst Simple bone cyst Osteoid osteoma Osteochondroma Giant cell tumor Chondroblastoma	Osteosarcoma Ewing sarcoma Leukemia
20–40 years	Giant cell tumor Fibrous dysplasia Enchondroma	Lymphoma Ewing sarcoma
>40 years	Enchondroma Bone infarct Infection Paget's Brown tumor	Metastatic carcinoma Lymphoma Multiple myeloma/plasmacytoma Chondrosarcoma Secondary osteosarcoma (Paget's and radiation)

on the most likely pathologic entities. The location of the lesion refers to both where in the body, i.e., axial skeleton or appendicular skeleton, or long bone or flat bone, and where within the bone the lesion is centered, as most lesions will have a propensity to be diaphyseal, metaphyseal, or epiphyseal or cortically or medullary based. Tables 23.1 and 23.2 provide examples of common benign and malignant entities categorized by age and by location.

The presence, absence, character, and timing of pain as well as other associated symptoms can be helpful in narrowing the differential diagnosis. Most benign latent tumors that do not cause significant mechanical cortical compromise will not be associated with pain, and therefore pain that leads to imaging should be carefully examined to elucidate whether there is a separate trauma, overuse, or degenerative related cause with the lesion being incidental.

A specific pain pattern that is important to recognize is functional or weight-bearing pain, which can be indicative of mechanical instability of affected bone. Recollection of antecedent pain prior to a pathologic fracture should prompt scrutiny and indicates evaluation for benign aggressive or malignant bone lesion. Another specific pain pattern that is noteworthy is the prostaglandin-driven pain worsened by activity, persistent at night and highly responsive to NSAIDs that should prompt the consideration of osteoid osteoma or osteoblastoma.

Aside from pain, other associated systemic symptoms such as fatigue, fevers, or weight loss should prompt consideration of malignancy or infection over benign bone lesion. Langerhans cell histiocytosis is an exception to this as with systemic manifestations, which can present with fever, rash, headaches, polydipsia, and polyurea depending on the organ systems involved. Other entities such as fibrous dysplasia and its association with McCune-Albright syndrome, enchondromas with Ollier's disease and Maffucci's syndrome, and osteochondromas with MHE emphasize the importance of thorough skin and skeletal exam as well as developmental and family history in the workup of bone lesions.

Once the patient demographics and clinical presentation have been used to focus on the most likely entities, imaging plays an essential role in further narrowing the differential and confirming the diagnosis.

Table 23.2 Diagnoses by skeletal site and location

Site	Benign	Malignant
Pelvis	Giant cell tumor Aneurysmal bone cyst Simple bone cyst	Chondrosarcoma Metastatic bone disease Multiple myeloma Chordoma
Spine (vertebral body)	Hemangioma Infection Langerhans cell histiocytosis Giant cell tumor	Metastatic bone disease Lymphoma Ewing sarcoma Chordoma
Spine (posterior elements)	Aneurysmal bone cyst Osteoid osteoma Osteoblastoma	Metastatic bone disease
Diaphyseal	Fibrous dysplasia Histiocytosis Osteoid osteoma	Ewing sarcoma Adamantinoma Lymphoma Metastatic bone disease
Metaphyseal	Non-ossifying fibroma Aneurysmal bone cyst Simple bone cyst Enchondroma Osteochondroma	Osteosarcoma Chondrosarcoma Metastatic bone disease
Epiphyseal	Chondroblastoma Giant cell tumor Aneurysmal bone cyst Infection	Clear cell chondrosarcoma Metastatic bone disease
Eccentric/cortical	Non-ossifying fibroma Osteoid osteoma Osteofibrous dysplasia Chondromyxoid fibroma Adamantinoma Periosteal chondroma	Parosteal osteosarcoma Periosteal osteosarcoma
Central/medullary	Fibrous dysplasia Enchondroma Bone infarct Aneurysmal bone cyst Simple bone cyst	Osteosarcoma Chondrosarcoma Metastatic bone disease

23.3 Radiographic Evaluation

Accurate characterization, diagnosis monitoring, and treatment of benign bone lesions rely on the full spectrum of imaging modalities including radiographs, CT, MRI, skeletal scintigraphy, and occasionally PET-CT. Biplanar radiographs remain the cornerstone of initial imaging in most benign bone tumors, and in some cases non-ossifying fibroma and unicameral or simple bone cyst can be definitively diagnostic. Advanced imaging with CT and MRI provides further information by elucidating tissue content, lesional architecture, relevant anatomy, and biologic activity when contrast is used. Certain lesions have extremely recognizable patterns on advanced imaging such as the radiolucent nidus of osteoid osteoma on CT scan and the fluid/fluid levels of aneurysmal bone cyst on MRI.

Systematic evaluation of radiographs can provide a great deal of diagnostic information by allowing inference into the tissue content and biologic behavior of a lesion. Initially, the radiograph demonstrates where the lesion is, within the body and within the bone, to begin to narrow the differential diagnosis. Next, by analyzing the radiographic sequalae of the effect that the lesion has on the bone and how the host bone reacts to the lesion as well as looking for any clues to what tissue the lesion is producing or contains, we can

begin to infer the nature of the lesion as well as how the lesion may behave biologically. These steps of systematic analysis are laid out in the four essential questions attributed to Dr. William Enneking. Where is the tumor? What is the tumor doing to the bone? What is the bone doing to the tumor? And are there any clues to the histology, i.e., what is the matrix of the lesion?

There is a vast and overlapping vocabulary that describes what is observed on radiographs and what inferences of behavior and biology can be made based on these observations. In regard to what the tumor is doing to the bone: Lytic describes a process of focal loss of bone density often due to replacement by tumor. Expansion refers to a dilatation of the normal diaphysis or metaphysis as is seen in aneurysmal bone cyst, usually defined as expansion to a width greater than that of the adjacent epiphysis. The zone of transition or margin describes how clearly you can delineate the edge of the tumor where it interfaces with the bone. A defined lesion border or narrow zone of transition is characteristic of benign lesions, whereas a more ill-defined margin is more likely to represent an aggressive or rapidly growing lesion. In regard to host bone response to a lesion, a sclerotic rim which represents bone laid down at the periphery of a lesion such as in NOF is demonstrative of a slower growing lesion where the host bone is easily able to wall off and contain the lesion. On the other hand, a fast-growing lesion will more likely have no reactive rim of sclerotic bone. Other reactions that the bone can have to fast-growing lesions that should raise the concern for a malignant or aggressive lesion include the Codman triangle, sunburst, and onionskin appearance derived from periosteal new bone formation in reaction to a rapidly expanding soft tissue mass expanding through bone and lifting periosteum.

By using demographics and location to develop a differential diagnosis, understanding the relevance of clinical presentation, and systematic interpretation of imaging, one can begin to focus on the specific pathologic entities that make up the broad spectrum of benign bone tumors.

Benign bone tumors are commonly divided into histopathologic and imaging subcategories. These include osteoblastic, cartilaginous, cystic, fibrous, and histiocytic tumors and giant cell tumor of bone. This is not an exhaustive list but aims to highlight key elements of the more commonly encountered benign bone lesions.

23.3.1 Benign Osteoblastic Tumors

- Osteoid osteoma
- Osteoblastoma
- Osteoma
- Enostosis (bone island)
- Osteopoikilosis
- Melorheostosis

23.3.1.1 Osteoid Osteoma

Osteoid osteomas are relatively common lesions with peak incidence in the second decade of life. They are most commonly found in diaphyseal/metaphyseal regions of long bones and are more commonly cortically than medullary based. They can be found in nearly any part of the axial and appendicular skeleton including small bones of the hand and foot and in the posterior elements of the spine at the concavity of a painful short-segment scoliosis as well as in periarticular locations near the hip, elbow, and ankle.

The clinical symptoms associated with osteoid osteoma are very distinctive and helpful in making the diagnosis. They most often present with increasing severity of pain that is worse at night and relieved by NSAIDs and aspirin. Both the responsiveness to NSAIDs and the large amounts of surrounding tissue edema on MRI, reactive sclerotic bone, and even skin swelling and redness in subcutaneous lesions are thought to be due to the high levels of proinflammatory prostaglandin E2 and prostacyclin present in the nidus.

Osteoid osteomas have very characteristic imaging findings with plain radiographs demonstrating a circular radiolucent nidus surrounded by dense sclerosis. Though often the plain radiographs are sufficient to make the diagnosis, a

thin-cut CT scan can be confirmatory demonstrating a central hazy nidus surrounded by a radiolucent halo imbedded in sclerotic bone. The most obvious feature is the solid fusiform periosteal reaction on plain radiographs and CT. On MRI, there can be extensive edema and enhancement concerning for malignancy, but the solid periosteal reaction present in osteoid osteoma is distinct from the layered appearance of the rapidly expanding soft tissue masses of malignant bone tumors.

Histologically, the nidus is characterized by thin, uniform osteoid trabeculae lined with osteoblasts, often with multinucleated giant cells.

The differential diagnosis for osteoid osteoma can include infection as well as malignant bone tumors due to the extensive inflammation and reactive bone formation. The solid periosteal reaction must be differentiated from stress fracture, which can have a similar appearance. Osteoblastoma can have a very similar appearance and clinical presentation but has a large nidus than the 1.0–1.5 mm typical of osteoid osteoma.

23.3.1.2 Osteoblastoma

Osteoblastomas are rare tumors related to osteoid osteoma sharing a peak incidence in the second decade of life, but with much greater growth potential beginning with a nidus size greater than 1.5 cm. There are two distinct forms of osteoblastoma with different histologic appearance and biologic behavior. Benign osteoblastomas are usually 1.5–4 cm, and aggressive osteoblastomas are usually 4 cm or greater. Osteoblastomas can be found in similar locations to osteoid osteomas; however, they have a predilection for the posterior elements of the spine and axial skeleton. Due to their significant growth, they can present with significant symptoms but are more variable in terms of the characteristic pattern of pain worse at night relived by NSAIDs seen in osteoid osteoma.

Imaging findings of osteoblastoma consist of a central radiolucent region greater than 1.5 cm with surrounding reactive zone. The osteoid matrix and calcification pattern are much larger and the reactive zone more heterogeneous than with osteoid osteoma. Osteoblastomas can have secondary ABC components, which can complicate the diagnosis radiographically.

Histologically, benign osteoid osteomas closely resemble osteoid osteomas. Aggressive osteoblastomas are uniquely characterized by the presence of epithelioid osteoblasts rimming the trabeculae.

The differential diagnosis for benign osteoblastomas primarily includes osteoid osteoma with the distinction being size. The differential for aggressive osteoblastoma is primarily osteosarcoma with distinguishing features being increased number of mitoses and distinct cellular atypia in osteosarcoma.

23.3.1.3 Enostosis (Bone Island)

Enostoses are common asymptomatic incidental findings in patients of all ages in both the axial and appendicular skeleton. There are no associated symptoms. Radiographically, they are dense sclerotic regularly shaped lesions surrounded by normal cancellous bone. Histologically, they demonstrate structurally normal lamellar bone. The differential diagnosis for enostoses includes osteoblastic metastases or the osteoblastic component of osteosarcoma, both of which should be associated with pain and potentially expansile or lytic components of the lesion.

23.3.1.4 Osteopoikilosis

Osteopoikilosis is a benign often incidentally found condition of multiple punctate regularly shaped osteoblastic lesions often symmetrically distributed in the metaphysis and epiphysis of long bones in patients of any age. Both enostoses and osteopoikilosis can be differentiated from osteoblastic metastasis as they will have little to no uptake on skeletal scintigraphy.

23.3.1.5 Melorheostosis

Melorheostosis is a rare benign sclerosing bone lesion that occurs most commonly in adolescents and young adults. It is often associated with progressive or relapsing pain and occasionally contractures. Radiographically, there is a very

characteristic cortical and medullary sclerosis described as a "dripping candle wax" appearance. This can occur in a sclerotomal distribution, affecting adjacent cortices of different bones or the same cortex of multiple bones across joints of a given limb. Radiographically, it must be differentiated from osteoid osteoma and parosteal osteosarcoma.

23.3.2 Benign Cartilage Tumors

- Enchondroma
- Osteochondroma
- Periosteal chondroma
- Chondroblastoma
- Chondromyxoid fibroma

23.3.2.1 Enchondroma

Enchondromas are common benign cartilaginous neoplasms composed of well-differentiated cartilage often discovered incidentally with a peak incidence in the third and fourth decades of life. Most common location for enchondromas is the small bones of the hand and foot followed by metaphyseal distal and proximal femur and proximal humerus. Most commonly, enchondromas present as asymptomatic incidental findings following workup of pain from trauma, tendinopathy, or arthritis. Pain clearly attributable to an enchondroma in a long bone should prompt further investigation and cross-sectional imaging. In the hand however, enchondromas can be expansile and present with painful swelling or pathologic fracture without similar concern for malignancy.

Radiographically, enchondromas present as primarily radiolucent lesions with variable amounts of stippled or "ring and arc" calcification generally centrally located in the metaphysis.

Histologically, enchondromas show nodules of mature cartilage that is hypocellularly interspersed with normal cancellous bone.

The differential diagnosis for enchondroma includes tumorlike conditions such as bone infarct, but most importantly it must be distinguished from chondrosarcoma. Clinically, chondrosarcoma of a long bone is more likely to present with pain; radiographically, there may be scalloping or erosion of the cortex or frank soft tissue mass on cross-sectional imaging. Histologically, the distinction can be very difficult, but increasing cellularity of the cartilage tissue, cellular atypia, and bony entrapment should raise concern for the presence of a chondrosarcoma.

Two distinct syndromes associated with multiple enchondromas with the risk for malignant transformation exist, Ollier's disease and Maffucci's syndrome. Both are associated with IDH1 mutations and present with multiple enchondromas throughout the skeleton and carry significant risk of malignant transformation of the enchondromas in addition to the risk of other visceral malignancies.

23.3.2.2 Osteochondroma

Osteochondroma is a very common benign cartilaginous lesion of bone characterized by an osseous stalk with corticomedullary continuity and a cartilaginous cap presenting with a firm deep fixed mass usually in the first or second decade of life. They can be either painless or painful in association with mechanical irritation of overlying structures. Morphologically, they can present as either sessile or pedunculated. They are located most frequently on the metaphyseal portion of long bones with the majority arising from the distal femur or proximal tibia, though flat bones including scapula and pelvis are also common.

Radiographically, they appear as a bony outgrowth with either a broad or a thin stalk depending on the sessile or pedunculated morphology. The lesion projects away from the nearest joint/epiphysis. CT can delineate the corticomedullary continuity if it is not clearly in plane on orthogonal radiographs. MRI is used to delineate the thickness of the cartilaginous cap.

Histologically, osteochondromas demonstrate three distinct layers: underlying bone, a middle layer of cartilage cap of variable thickness, and a perichondrium that is in continuity with the normal periosteum.

The differential diagnosis for osteochondroma includes surface osteosarcomas, particu-

larly for the sessile form. Additionally, it is important to consider malignant transformation osteochondromas if they present with significant growth or changing architecture after skeletal maturity. Lesions with cartilage cap >2 cm in thickness are considered higher risk malignant transformation.

Multiple hereditary exostosis is an autosomal dominant condition associated with multiple osteochondromas and skeletal deformities including radial bowing, short stature, and angular deformities among others. Osteochondromas tend to be larger and more sessile and have more complex caps. MHE is associated with EXT1 and EXT2 gene mutations. The actual risk of malignant transformation is thought to be between 5% and 25% in MHE as opposed to 1% or less in solitary lesions, with the peak age of ~30.

23.3.2.3 Periosteal Chondroma

Periosteal chondroma, also referred to as juxtacortical chondroma, is a rare lesion arising most commonly on the surface of the humerus and hands of patients in the second and third decades of life. Axial locations and toes can also occur. They can present as incidental findings and painless palpable masses or with pain due to mechanical irritation of overlying structures.

Periosteal chondromas lie beneath the periosteum and lack corticomedullary continuity. Therefore, radiographically, they appear as raised cortically based lesions surrounded by a solid periosteal reaction and sometimes with a sclerotic rim separating the lesion from the medullary canal. This can create a "saucerized" or crater-like appearance of the lesion at the cortex with varying degrees of calcification within the cartilaginous portion of the lesion.

Histologically, they contain primarily bland cartilage with slightly more atypia and cellularity than an enchondroma.

The differential diagnosis for periosteal chondroma includes juxtacortical chondrosarcoma and periosteal osteosarcoma. The solid periosteal reaction seen in periosteal chondroma would not be typical of periosteal osteosarcoma. The distinction between periosteal chondroma and juxtacortical chondrosarcoma is difficult to make. A size of >5 cm is suggestive of periosteal chondrosarcoma.

23.3.2.4 Chondroblastoma

Chondroblastoma is a rare benign bone tumor with cartilaginous matrix that presents in the epiphyses about the knee in skeletally immature patients most commonly in the second decade of life. However, older patients and locations including proximal humerus, proximal femur flat bones, and spine are possible.

Clinically, they present with significant, constant, and long-lasting pain. Given the epiphyseal location, the pain is often associated with a joint and can be accompanied by an effusion and limited motion.

Radiographically, chondroblastoma appears as a well-demarcated often round or ovoid lytic epiphyseal lesion with variable sclerotic rim and internal calcification. There is often a significant amount of associated edema surrounding chondroblastomas. The presence of secondary ABC can complicate the radiographic diagnosis.

Histologically, chondroblastoma has distinctive features with nodular areas of cartilage and sheets of polygonal chondroblasts creating a "cobblestone" appearance. The cells are bordered by a lacy network of calcification described as "chicken wire calcification." Reactive giant cells and secondary ABC components can be present.

The differential diagnosis can be quite broad including giant cell tumor, aneurysmal bone cyst, infection, LCH in younger patients, and clear cell chondrosarcoma in older patients. Subtle radiographic differences with each of these exist, but ultimately histology is often required to make the final distinction.

23.3.2.5 Chondromyxoid Fibroma

Chondromyxoid fibroma is a rare benign bone tumor arising eccentrically in the metaphyseal region of long bones, most frequently in the proximal tibia in patients in the second to third decades of life. Distal femur, ilium, and feet are also known sites for chondromyxoid fibroma.

Clinically, they often present with pain and localized swelling, especially given the subcutaneous nature of the proximal tibia.

Radiographically, they appear as an eccentric, lobulated primarily lytic lesion with a narrow zone of transition and potentially a sclerotic rim on the medullary side. The cortical side is often completely replaced with no matrix or some calcification. There is no significant periosteal reaction. Secondary ABC can complicate the radiographic diagnosis. MRI can be useful in that the T1 and fluid-sensitive sequences of the fibrous and myxoid components can be distinctive.

Histologically, they have a pseudo-lobulated architecture with myxoid regions with stellate cells and chondroid and hypercellular areas, which can have nuclear atypia but lack mitoses.

Differential diagnosis includes periosteal chondroma, and chondroblastoma in the extremity. The solid periosteal reaction of periosteal chondroma should help distinguish this from chondromyxoid fibroma. Eccentric metaphyseal location of CMF should help distinguish this from chondroblastoma in the epiphysis, but when there is radiographic overlap, they can be difficult to distinguish histologically. In the pelvis and in older patients, CMF can have a more mineralized matrix and can have imaging findings that overlap with chondrosarcoma.

23.3.3 Cystic and Radiolucent Bone Lesions

- Simple bone cyst
- Aneurysmal bone cyst
- Intraosseous ganglion
- Calcaneal cyst
- Liposclerosing myxofibrous tumor (LSMFT)

23.3.3.1 Simple Bone Cyst

Simple bone cyst, also called unicameral bone cyst or solitary bone cyst, is a very common lesion most often located in the proximal femoral and humeral metaphysis of skeletally immature patients below 15 years of age.

Most frequently, they are found incidentally or with pain due to pathologic fracture or impending pathologic fracture.

Radiographically, they most commonly present as a central lucent lesion in the medullary cavity with thinning of the cortex with mild-to-moderate expansion of the cortex. MRI demonstrates a fluid-filled cavity without blood and with peripheral enhancement. There can frequently be multiple septations and chambers emphasizing the more appropriate moniker of simple bone cyst as opposed to unicameral bone cyst. As a patient grows, the lesions have a tendency to resolve and the lesion grows away from the physis. A unique finding associated with simple bone cyst is the "fallen leaf sign", which is considered pathognomonic and represents a portion of the cortex that breaks off during pathologic fracture and settles perpendicular to the cortex within the cyst cavity.

Histologically, the lesion is characterized by a thin fibrous lining lacking endo- or epithelial components and can contain inflammatory and giant cells.

The differential diagnosis for solitary bone cyst is primarily aneurysmal bone cyst. The differentiation can usually be made based on the radiographic appearance or biologic behavior, though MRI is also helpful if questions remain.

23.3.3.2 Aneurysmal Bone Cyst

Aneurysmal bone cyst is a benign bone lesion with expansile appearance and aggressive growth pattern most commonly in the metaphysis of long bones, but also in the sacrum and ilium and posterior elements of the spine in patients in the second decade of life. Given that ABCs can be primary lesions or secondary to other primary lesions, their radiographic appearance can be highly varied and they deserve to be considered in the differential diagnosis for many benign and malignant tumors.

Clinically, they often present with moderate pain related to swelling and expansion or pathologic fracture as they do not respect the natural bony barriers to growth. In the spine, this can lead to path fracture and neurologic and radicular symptoms.

Radiographically, they appear as aggressive expansile. Expansion can be beyond the physis, which distinguishes them from simple bone cyst. Despite the expansile nature, it is almost always bound by a thin rim of bone peripherally.

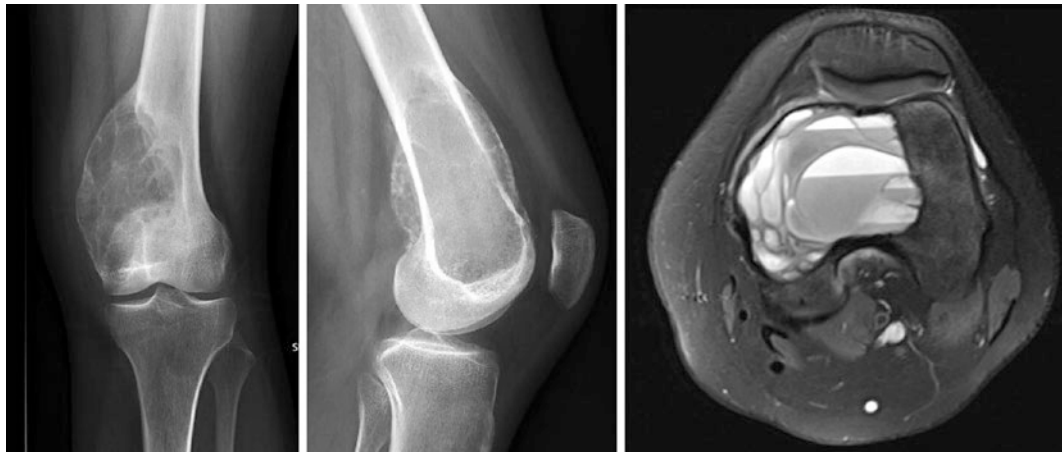

Fig. 23.1 AP and lateral radiographs and STIR MRI of distal femur aneurysmal bone cyst demonstrating expansile metaphyseal-epiphyseal lesion with fluid-fluid levels

Aneurysmal bone cyst is most frequently eccentric, but can be central and is often multiloculated. MRI demonstrates a characteristic pattern of multiple chambers with fluid/fluid levels (Fig. 23.1).

Histologically, ABCs show a thin fibrous lining with hemorrhage and hemosiderin-laden macrophages, chronic inflammatory changes, and giant cells with blood-filled cavities termed "blood lakes." There is a molecular finding of USP6 gene mutations that can be helpful in confirming the diagnosis, though it is not uniformly detected in all cases.

The differential diagnosis for ABC is very broad because of its appearance as a secondary lesion and varied radiographic appearance. As an expansile lesion in periarticular locations, it must be differentiated histologically from giant cell tumor. Telangiectatic osteosarcoma is an expansile highly vascular variant of osteosarcoma that can have clinical appearance that overlaps significantly with ABC. ABC should have a thin rim of bone which may be absent in telangiectatic osteosarcoma, and histologically osteosarcoma will have atypical mitosis and tumor-produced osteoid that should be absent in ABC.

23.3.3.3 Intraosseous Ganglion

Intraosseous ganglion, also known as degenerative cyst or geode, is usually a subchondral well-circumscribed lesion with a sclerotic rim often found in periarticular regions incidentally in patients with degenerative joint disease though this is not an essential precursor. The lesions can become quite large and experience pathologic fracture. Advanced imaging demonstrates a fluid-filled cyst and may show communication with the intra-articular space. They must be differentiated from other epiphyseal lytic lesions.

23.3.3.4 Calcaneal Cyst

Calcaneal cysts are fairly common and represent a variant of simple bone cyst, but also are a fairly common location for intraosseous lipoma which has very similar findings on plain radiographs with a lack of matrix or trabeculae. These lesions have a propensity for the anterior portion of the calcaneus beneath the calcaneal sulcus middle facet. They are often found incidentally and in this situation are not thought to have a significant risk for pathologic fracture.

23.3.3.5 Liposclerosing Myxofibrous Tumor (LSMFT)

Liposclerosing myxofibrous tumor is a benign bone lesion with a strong propensity for the intertrochanteric region of the proximal femur. These lesions are usually discovered incidentally in patients in the third to fourth decades of life, though they can present with pain and risk for pathologic fracture. Imaging findings demonstrate a sclerotic heterogenous or multicystic

lesion. MRI demonstrates areas of predominant T1 signal isointense to muscle and areas of T2 myxoid signal. Despite the nomenclature, macroscopic fat within the lesion is not usually seen. The differential diagnosis includes fibrous dysplasia, interosseous lipoma, and ABC. LSMFT is unique in that malignant transformation to osteosarcoma or other high-grade undifferentiated sarcomas has been reported in up to 10% of lesions.

23.3.4 Benign Fibrous and Histiocytic Tumors

- Non-ossifying fibroma (NOF)
- Fibrous dysplasia
- Osteofibrous dysplasia (OFD)
- Langerhans cell histiocytosis (LCH)

23.3.4.1 Non-ossifying Fibroma (NOF)

Non-ossifying fibroma, also called fibrous cortical defect or metaphyseal fibrous defect, is a very common benign bone lesion found eccentrically in the cortex of the metaphyseal region of long bones, most frequently discovered in the distal femur and proximal tibia in the second decade of life. They are reported to be present in up to 30–40% of children.

Clinical presentation is usually as an incidental finding, though some larger more expansile lesions can present with pain associated with pathologic fracture.

Radiographically, it has a classically lobulated, "bubbly" eccentric, cortically based metaphyseal lesion with a sclerotic margin. Advanced imaging is not necessary to make the diagnosis, but MRI demonstrates T2 hyperintense areas associated with the fibrous tissue.

Histologically, NOF demonstrates sheets of fibroblastic cells in a storiform or whirled pattern with areas of hemosiderin depositions. Two genetic syndromes are associated with multiple NOFs. This includes neurofibromatosis type 1 and Jaffe-Campanacci syndrome, both of which include café au lait spots among other manifestations.

Differential diagnosis includes other cortically based metaphyseal lesions such as osteofibrous dysplasia, chondromyxoid fibroma, and aneurysmal bone cyst.

23.3.4.2 Fibrous Dysplasia

Fibrous dysplasia is more appropriately a developmental abnormality of bone than a bone tumor. It is a process in which normal cancellous bone is replaced by pathologic fibro-osseous tissue, which can appear expansile and result in deformity of the bone. It is generally diagnosed in patients in the first three decades of life most commonly presenting in the femur, tibia, and pelvis. It can be present in one isolated location (monostotic) or polyostotic, affecting multiple bones in a single extremity or more broadly. Two syndromes are associated with fibrous dysplasia. McCune-Albright syndrome presents with café au lait areas, polyostotic fibrous dysplasia, and precocious puberty (among other endocrine abnormalities). Mazabraud syndrome is a rare syndrome presenting with fibrous dysplasia and multiple intramuscular myxomas.

Fibrous dysplasia may present as an incidental finding or with pain associated with chronic stress fracture, remodeling, and deformity, classically described as "shepherd's crook" deformity in the proximal femur.

Radiographically, fibrous dysplasia appears as a replacement of the normal cancellous bone that can be focal or distributed over the length of a bone. The matrix of fibrous dysplasia is classically described as a hazy "ground glass" appearance. It can be expansile but should not have soft tissue extension or periosteal reaction in the absence of pathologic fracture. MRI demonstrates homogenous areas of T1 signal isointense to muscle that is also hyperintense on T2 and enhances with contrast.

Histologically, the tissue has a fibrous stromal background of low cellularity with scattered bone spicules described as an "alphabet soup pattern." There is genetic association with GNAS mutation activating GS alpha and abnormal cAMP signaling.

The differential diagnosis for fibrous dysplasia is quite broad. In older patients, it must be differentiated from Paget's disease of bone. In children, it must be differentiated from osteofibrous dysplasia, which has similar appearance but is almost always in the anterior tibial in children and histologically will have rimming osteoblasts on the bone spicules.

23.3.4.3 Osteofibrous Dysplasia (OFD)

Osteofibrous dysplasia is a rare benign bone lesion occurring most commonly in the anterior proximal tibia of patients in the first decade of life, usually under 5 years of age. Clinically, this presents either incidentally or with anterior bowing deformity or pain due to mechanical compromise and pathologic fracture. Radiographically, it appears as a cortically based expansile multiloculated lesion in the midportion of the tibia. Histologically, OFD demonstrates a fibrous background with bony trabeculae surrounded by rimming osteoblasts which differentiate this from non-ossifying fibroma histologically. Adamantinoma has a similar radiographic appearance radiographically but is more common in adults and can be differentiated histologically.

23.3.4.4 Langerhans Cell Histiocytosis (LCH)

Langerhans cell histiocytosis represents a spectrum of pathologies characterized by abnormal proliferation of histiocytes ranging from isolated bony disease often called eosinophilic granuloma to severe systemic forms including Hand-Schüller-Christian syndrome and Letterer-Siwe disease. There is ongoing debate as to how to classify this spectrum of diseases. It is not clearly an immunologic process nor a malignancy. It has self-limiting forms that resolve with observation alone as well as life-threatening systemic forms that are treated with chemotherapy, which emphasizes the importance of thorough and timely workup.

The bony lesions of LCH can occur anywhere in the extremities, skull, axial skeleton, and ribs usually in the first two decades of life. They can have highly variable radiographic appearance that mimics the presentation of many other benign and malignant lesions, commonly presenting in the diaphysis or metaphysis of long bones and the vertebral body in the spine. They will often present with pain and swelling, a limp, or inability to bear weight in a younger child or with pathologic fracture. As opposed to other benign bone lesions, they can present with persistent aching pain that is worse at night as well as with systemic symptoms including low-grade fevers. Examination for skin lesions, exophthalmos, and splenomegaly, and questioning to elucidate symptoms of diabetes insipidus and headaches, should be included if LCH is suspected and a skeletal survey should be performed to identify other potential bony sites of involvement.

Radiographically, long bone lesions can be aggressive-appearing lytic expansile lesions with variable margins, sclerosis, and periosteal reactions. MRI shows a relatively heterogeneous intramedullary lesion with significant surrounding edema and even soft tissue mass that can generate concern for malignancy. In the spine, lesions often involve the vertebral body, sparing the disk spaces, but can have significant symmetrical vertebral height loss leading to findings of "vertebra plana."

Histologically, distinctive large Langerhans cells with granular pink cytoplasm and grooved or "coffee bean"-shaped nuclei are present in addition to eosinophils.

Differential diagnosis for eosinophilic granuloma is very broad and includes malignancies including Ewing and osteosarcoma. A biopsy is often necessary to make the definitive diagnosis.

23.4 Giant Cell Tumor of Bone

Giant cell tumor of bone is a benign aggressive lesion seen most commonly in the third to fourth decades of life and can affect nearly any skeletal site including the spine, sacrum, and pelvis, though metaphyseal/epiphyseal distal femur, proximal tibia, and distal radius are the most common sites.

Clinically, giant cell tumor most frequently presents with insidious onset of pain with variable soft tissue mass or swelling due to focal expansion and not infrequently with pathologic fracture in periarticular locations.

Radiographically, giant cell tumors are typically eccentric metaphyseal-epiphyseal lesions that are often eccentric, without a sclerotic rim or discernable matrix and variable degree of bony expansion. MRI is often used to define the soft tissue extent and can also show subtle subchondral fracturing and delineate bony rim for treatment planning. Typical signal characteristics include homogeneously low T1 signal with heterogeneously hyperintense T2 signal and internal enhancement on postcontrast sequences.

The differential diagnosis for giant cell tumor radiographically can include entities such as aneurysmal bone cyst and telangiectatic osteosarcoma due to the eccentric and sometimes expansile nature. Chondroblastoma has some overlapping features due to the periarticular presentation of both entities. Brown tumors of hyperparathyroidism can have similar appearance on plain radiographs with a lytic lesion without sclerotic borders or defined matrix.

Histologically, the neoplastic cell of GCT of bone is the ovoid plump mononuclear cell that predominates the stroma. These are interspersed with the multinucleated osteoclast-like giant cells for which the tumor is named. However, these are also present in many other entities from which GCT must be differentiated making imaging and other elements of the histology important for differentiating them from GCT. The uniquely high RANKL expression of the multinucleated giant cells had created a novel opportunity for the use of inhibitors in the form of denosumab, which is now used in both the neoadjuvant and refractory unresectable disease setting to slow bony destruction and allow new bone formation though controversy exists over the role of denosumab and local recurrence.

Giant cell tumor of bone is unique in that it is considered a benign entity with the potential to spread to the lung. Though often termed metastasis, the spread of disease to the lung often represents disease that is histologically identical to the primary site, so the term tumor embolization may be more appropriate. Chest imaging is an important part of staging and surveillance of GCT. In addition to spread to the lungs, malignant transformation has been observed in GCT; therefore, particularly in cases of rapid, aggressive recurrence or GCT treated with radiation, repeat biopsy and close examination of histology to rule out malignancy are essential.

Take-Home Message

- Osteoid osteomas are most commonly found in diaphyseal/metaphyseal regions of long bones and are more commonly cortically than medullary based.
- Osteoblastomas are rare tumors related to osteoid osteoma sharing a peak incidence in the second decade of life.
- Enostoses are common asymptomatic incidental findings in patients of all ages in both the axial and appendicular skeleton. There are no associated symptoms.
- Simple bone cyst is a very common lesion most often located in the proximal femoral and humeral metaphysis of skeletally immature patients below 15 years of age.
- Aneurysmal bone cyst is a benign bone lesion with expansile appearance and aggressive growth pattern most commonly in the metaphysis of long bones, but also in the sacrum and ilium and posterior elements of the spine in patients in the second decade of life.
- Non-ossifying fibroma, also called fibrous cortical defect or metaphyseal fibrous defect, is a very common benign bone lesion found eccentrically in the cortex of the metaphyseal region of long bones.

Summary

- Osteoid osteomas are relatively common lesions with peak incidence in the second decade of life. They are most commonly found in diaphyseal/metaphyseal regions of long bones and are more commonly cortically than medullary based.
- Osteoblastomas are rare tumors related to osteoid osteoma sharing a peak incidence in the second decade of life, but with much greater growth potential beginning with a nidus size greater than 1.5 cm.
- Enostoses are common asymptomatic incidental findings in patients of all ages in both the axial and appendicular skeleton. There are no associated symptoms.
- Enchondromas are common benign cartilaginous neoplasms composed of well-differentiated cartilage often discovered incidentally with a peak incidence in the third and fourth decades of life.
- Osteochondroma is a very common benign cartilaginous lesion of bone characterized by an osseous stalk with corticomedullary continuity and a cartilaginous cap presenting with a firm deep fixed mass usually in the first or second decade of life.
- Periosteal chondroma, also referred to as juxtacortical chondroma, is a rare lesion arising most commonly on the surface of the humerus and hands of patients in the second and third decades of life.
- Chondroblastoma is a rare benign bone tumor with cartilaginous matrix that presents in the epiphyses about the knee in skeletally immature patients most commonly in the second decade of life.
- Chondromyxoid fibroma is a rare benign bone tumor arising eccentrically in the metaphyseal region of long bones, most frequently in the proximal tibia in patients in the second to third decades of life.
- Simple bone cyst, also called unicameral bone cyst or solitary bone cyst, is a very common lesion most often located in the proximal femoral and humeral metaphysis of skeletally immature patients below 15 years of age.
- Aneurysmal bone cyst is a benign bone lesion with expansile appearance and aggressive growth pattern most commonly in the metaphysis of long bones, but also in the sacrum and ilium and posterior elements of the spine in patients in the second decade of life.
- Non-ossifying fibroma, also called fibrous cortical defect or metaphyseal fibrous defect, is a very common benign bone lesion found eccentrically in the cortex of the metaphyseal region of long bones, most frequently discovered in the distal femur and proximal tibia in the second decade of life.
- Fibrous dysplasia is more appropriately a developmental abnormality of bone than a bone tumor.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. A healthy 12-year-old nonathletic patient presented with predominantly nocturnal pain in the right tibia. The patient is able to precisely indicate the origin of the pain with a finger. The skin apears normal but there is tenderness to palpation. The patient is able to precisely indicate the origin of the pains with one finger. The skin shows sweating and a local increase in temperature. In order to better orientate the diagnosis, the orthopedic surgeon asks the patient if the pain goes away after:
   (a) The intake of acetylsalicylic acid
   (b) Intake of antibiotics
   (c) A long walk
   (d) Sporting activity
2. With reference to the previous question, the possible diagnosis will be:
   (a) Osteoid osteoma
   (b) Enchondroma
   (c) Stress fracture
   (d) Superficial tibial nerve neuropathy
3. In reference to the previous question, the report of the radiographic examination states: “consistent fusiform thickening of the bony cortical of the lateral side of the III medium of the right tibia. There are no macroscopic signs

of bone infiltration or macroscopic soft tissue lesions." What further investigation is required?
   (a) Computed tomography
   (b) Magnetic resonance imaging
   (c) Positron-emission tomography
   (d) Ultrasound
4. Chondroblastoma is mainly located:
   (a) In the flat bones
   (b) In the short bones
   (c) In the epiphyses of the long bones
   (d) In the metaphyses of the long bones
5. Which of these tumor lesions is most prone to local recurrence?
   (a) Osteoid osteoma
   (b) Aneurysmal cyst
   (c) Juvenile bone cyst
   (d) Giant cell tumor

Further Reading

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Malignant Tumors

24

Ryan T. Voskuil and Joel L. Mayerson

Overview
This chapter focuses on the general concepts of primary malignant musculoskeletal tumors.

24.1 Definition

Musculoskeletal tumors comprise benign and malignant varieties of both the bone and soft tissues. Primary malignant bone tumors are called sarcomas; they begin in the bone, where they behave aggressively and have the potential to spread to other anatomic locations.

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24.2 Epidemiology

Malignant bone and soft tissue tumors are rare with roughly 2500 bone and 12,000 soft tissue sarcomas occurring in the United States yearly. Benign bone and soft tissue masses are much more common. Age is important to consider. When it comes to primary malignant bone tumors, osteosarcoma and Ewing sarcoma typically present in patients less than 20 years of age while chondrosarcoma is more commonly seen in those over 40. There are some secondary bone sarcomas that can arise later in life from radiation, Paget's, or transformation of benign bone lesions like an enchondroma or osteochondroma. There are a few other primary sarcomas of bone that include malignant fibrous histiocytoma/undifferentiated pleomorphic sarcoma, fibrosarcoma, leiomyosarcoma, epithelioid hemangioendothelioma, angiosarcoma, and adamantinoma, all of which are exquisitely rare. Malignant bone lesions in general are logarithmically more likely to represent metastatic bone disease from carcinoma, multiple myeloma, or lymphoma versus a primary bone sarcoma in patients over 40 years old.

24.3 Etiology/Pathogenesis

For the majority, there are no identifiable genetic or environmental factors to date that lead to the development of a sarcoma, as most mutations

UG. Longo, V. Denaro (eds.), *Textbook of Musculoskeletal Disorders*,
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identified are somatic. This is due in large part to the rarity of the disease and the fact that there are several subtypes of sarcoma that are studied uniformly in a retrospective manner. We do know that some bone sarcomas rarely can arise secondarily from an area of prior radiation, osteonecrosis of bone, or other benign bone tumors like Paget's disease of bone. There are some inherited disorders that have been identified to be associated with an increased risk of developing an osteosarcoma. These include Li-Fraumeni (germline p53 mutation), retinoblastoma syndrome (germline Rb mutation), and Rothmund-Thomson syndrome (RECQL4 gene mutations).

24.4 Classifications

There are a few classifications frequently used in musculoskeletal oncology that are worth mentioning. Dr. Enneking developed a classification for both benign and malignant bone tumors in the 1980s that was adopted by the Musculoskeletal Tumor Society and is the most commonly used for staging and direction of treatment. The malignant bone tumor classification is broken into low and high grade, intra- and extra-compartmental, and presence of metastatic disease whether that be skip, lymph node, or distant. This classification is easy to use and familiar to most orthopedic oncologists, but has not changed in 30 years. The American Joint Commission on Cancer (AJCC) bone sarcoma staging system is more popular among medical and radiation oncologists and is now in its eighth edition. This system is more involved and considers grade, size, and depth, along with nodal and distant metastatic status.

24.5 Diagnosis

24.5.1 History

A thorough history is important when evaluating a person with a suspected bone tumor. Onset, location, duration of symptoms, character of pain, associated factors, timing of pain pattern, and severity are foundational questions. Medical, personal, and family cancer history should also be addressed. Questions regarding prior radiation, areas of bone infarct, or other known benign bone tumors should be explored. Bone tumors typically present with a constant, dull, pain pattern often present at night. Night pain that wakes people from sleep and present for more than 3–5 days should be a red flag. Bone tumors can be at high risk for pathologic fracture, so this should always be carefully assessed as well when patients present with low-energy mechanisms for long bone fractures.

24.5.2 Physical Exam

The physical exam should focus on assessing gait, location of pain, swelling, and mass characteristics. Concerning masses are often deep, firm, well circumscribed, and fixed. The involved extremity should be evaluated for neurovascular status, joint motion, skin changes, swelling, and locoregional lymph node involvement with comparison to the contralateral arm or leg. Axial loading and twisting of the involved extremity should be done to assess for mechanical weakening or functional pain.

24.5.3 Imaging

24.5.3.1 Local Imaging

Plain radiographs of the entire involved extremity with inclusion of the joint above and below should always be the first imaging study ordered during any tumor workup. In malignant bone tumors, it is important to ask five questions developed by Dr. Enneking, one of the fathers of musculoskeletal oncology: (1) age of the patient, (2) location in bone, (3) what is tumor doing to bone, (4) what is bone doing to the tumor/how is it responding, and (5) hint of histology (bone, cartilage, or fibrous matrix forming). It is important to pay attention to soft tissue shadows and look at the periosteum as well. Malignant bone tumors tend to grow faster than the body has a chance to respond to. This causes cortical destruction and breakthrough, and the periosteum attempts to

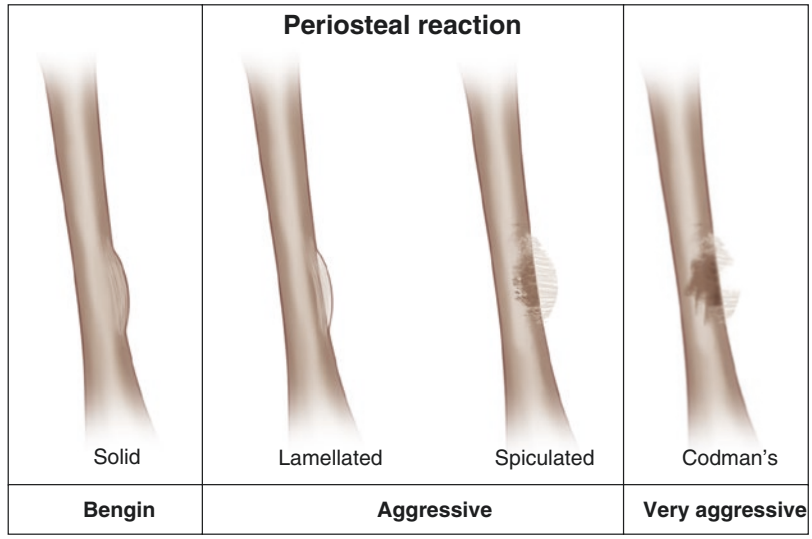

Fig. 24.1 Periosteal reaction types

repair this area but cannot keep up. This leads to several pathognomonic periosteal reactions that are usually only seen in malignant bone tumors and include onion skinning, sunburst pattern, and Codman triangle (Figs. 24.1 and 24.2).

A similar but more involved diagnostic algorithm for bone tumors developed by Dr. Mayerson provides an excellent framework by answering five questions: type of matrix (bone, cartilage, fibrous, cystic calcium), type of bone (normal, reactive, tumor produced), pattern of destruction (geographic, moth-eaten, permeative), age (<20, 21–40, >40), and location in bone (epiphysis, metaphysis, diaphysis).

Magnetic resonance imaging (MRI) is usually the next necessary imaging study ordered. In bone tumors, the T1 sequence helps delineate its true location and relation to surrounding structures including nerves and vessels. It is vital to scan the entire bone to evaluate for skip metastases. In general, tumors with peri-tumoral edema, periosteal reaction, cortical destruction, and a soft tissue mass are malignant until proven otherwise.

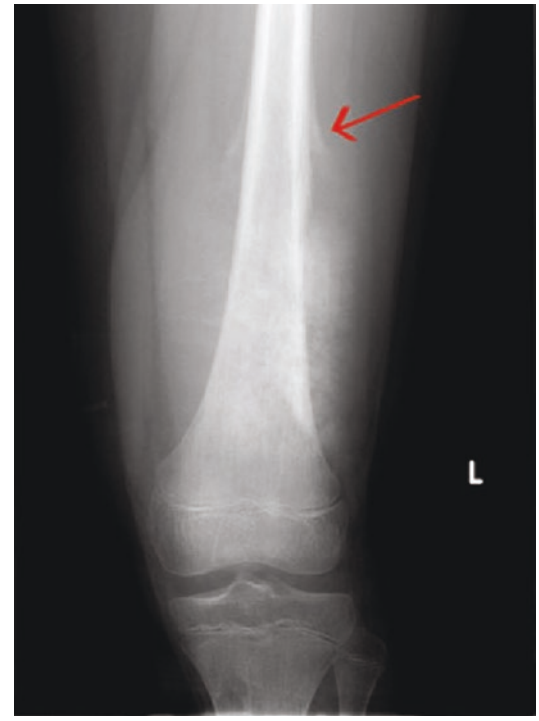

Fig. 24.2 Distal femur osteosarcoma with periosteal reaction showing both the classic sunburst appearance and Codman triangle (arrow)

24.5.3.2 Staging Studies

Staging studies are an important aspect of the imaging workup with particular imaging modalities employed to evaluate for skip, lymph node, and distant metastatic spread. These studies are vital from both a treatment planning standpoint and overall prognosis. Whole-body bone scans are used to evaluate other sites of bone disease in malignant bone tumors. Computed tomography

(CT) scans of the chest and sometimes the abdomen and pelvis are critical in both malignant bone and soft tissue tumors to evaluate for metastatic disease as the lungs are by far the most common location of metastatic disease in sarcomas. PET/CT scans are gaining popularity for staging both bone and soft tissue malignancies and have shown that they can be predictive of response to chemotherapy and percent necrosis of the tumor.

24.5.4 Labs

Labs are rarely diagnostic, but they aid in painting a diagnostic picture. A complete blood count (CBC), chemistry (CMP), and inflammatory markers (C-reactive protein {CRP}) are a great place to start. Alkaline phosphatase and lactate dehydrogenase are helpful with suspected primary bone tumors and can be poor prognostic indicators if elevated. Ewing sarcoma can drastically mimic infection and can have several elevated inflammatory markers.

24.6 Differential Diagnostic Considerations

24.6.1 The Biopsy

The biopsy is arguably the most important aspect of the diagnostic processes as a verification of histology is usually required before initiation of treatment. Open biopsy has long been the gold standard and is most often utilized in primary bone tumors without a soft tissue mass and when other techniques are nondiagnostic. Open biopsy in the setting of a bone sarcoma is prone to errors that can lead to bleeding and contamination with occasional loss of limb salvage ability. Complications of open biopsy can be avoided by making the incision in a longitudinal manner in line with the eventual incision for definitive surgery, through one compartment, avoiding neurovascular structures, and with meticulous hemostasis. If a drain is placed, it should be in line with the skin incision. The open biopsy tract should be excised during the definitive surgery to minimize the risk of local recurrence in that location. Core needle biopsy (CNB) has gained popularity in many centers due to its versatility without the need for biopsy tract excision. CNB can be utilized when a palpable soft tissue mass is present in the office or with image guidance when gathering tissue samples from difficult locations around neurovascular structures or in the deep pelvis. Fine needle aspiration (FNA) is an office-based biopsy technique that has demonstrated significant benefits in centers with pathologists experienced with this method. The FNA can be used on any palpable soft tissue mass in the office without the need for local anesthetic and the least risk for bleeding or contamination among the three techniques. A preliminary diagnosis can often be delivered to the patient within 30 min of the procedure while minimizing cost and maximizing staff and resource utilization.

24.7 Treatment

24.7.1 Surgical Management

The surgical management of musculoskeletal tumors demands a thoughtful and thorough plan. The primary goals of sarcoma surgery are twofold: oncologic and functional. The oncologic goal is ultimately the most important. Oncologic resections can be classified as intralesional, marginal, wide, and radical excision. For high-grade bone tumors, wide resection is the best surgical treatment. An intralesional excision is typically performed by entering the substance of the tumor and removing it in a piecemeal fashion. This is often utilized on benign cysts. Marginal excision involves removal of the tumor through the reactive zone, or the thin capsule surrounding many lesions where signal change is often seen on MRI. This technique is performed on benign soft tissue masses like a lipoma. A marginal excision is not acceptable for resection of sarcomas as microscopic tumor cells are often present in the reactive zone. This is the reason why a wide excision is performed on primary bone malignancies. This method involves removing a tumor without

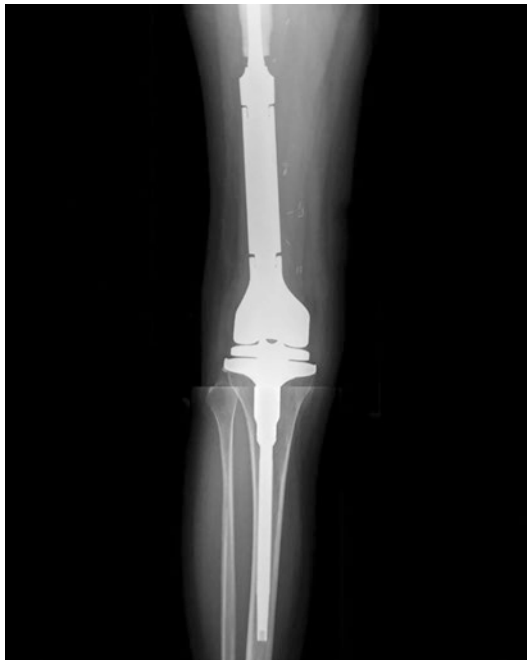

Fig. 24.3 Distal femur sarcoma treated with distal femur replacement

ever seeing the tumor itself, by leaving a cuff of normal tissue around the mass as the margin, thus ensuring complete removal of all cancer cells and a better chance at survival. Radical resection refers to a removal of the entire involved compartment. This is rarely required and is usually performed when a wide excision would leave the compartment nonfunctional. Primary bone and soft tissue malignancies are usually treated with a curative wide excision in an attempt to remove all diseases (Fig. 24.3). Bone margins are typically planned to be around 2 cm from the known end of tumor, and frozen marrow margin is often assessed before proceeding to be sure enough that bone was removed. The resected specimen is then orientated with stitches so that margins can be assessed by the pathology department.

The functional goal is a close second to the oncologic goal but is often more important to the patient. As technology in the worlds of endoprosthetics, allografts, and 3D printing explodes, function continues to improve. Pediatric bone sarcoma resections provide their own sets of challenges as the restoration and maintenance of equal leg lengths as the patient grows must be accounted for. This can be addressed with growing prostheses, bone transport, and epiphysiodesis or a combination of techniques. Curative surgery for sarcoma has swung from amputation to limb salvage with vast improvements seen in the latter. Over 90% of patients with extremity sarcomas can be treated with limb salvage. Amputation is now reserved for cases in which a wide excision with a negative margin cannot be achieved or when amputation would result in better function.

24.7.2 Adjuvant Treatments

Adjuvant therapy often includes chemotherapy, external beam radiation therapy, or both, which have transformed the treatment of primary musculoskeletal tumors with vast improvements seen in their rates of local recurrence, metastatic disease, and overall survival. Sarcoma treatment is usually broken down into three separate buckets when explaining options with the patient, with radiation and surgery addressing local tumor control while chemo representing the only systemic modality. Chemotherapy is utilized on all high-grade primary bone tumors with the exception of chondrosarcoma, which is not chemosensitive. It is often given in a standard protocol in a neoadjuvant (before surgery) fashion and then given after surgery adjuvantly. This allows some evaluation of the treatment effect in the middle of chemotherapy, providing both prognostic and therapeutic information. Radiation therapy can be utilized in Ewing sarcoma. Wide resection has proven to be better for local recurrence, but in some morbid locations, radiation can be the better choice. There has been some evidence that pelvic Ewing sarcoma may benefit from both radiation and surgery. Chondrosarcoma and osteosarcoma are not radiosensitive. The future of cancer treatment rests in the evolving discovery of molecular and genetic markers that are specific to cancer cells and are targetable. This would individualize the systemic treatment and eventually replace the shotgun approach of traditional chemo.

24.8 Specific Malignant Bone Tumors

24.8.1 Osteosarcoma

Osteosarcoma is the most common primary bone sarcoma and presents most typically in the second decade during periods of peak growth, with a second peak in the seventh and eighth decades. Conventional osteosarcoma is high grade and intramedullary 90% of the time. There are several other low-grade and/or surface-based variants that are much less common. Osteosarcoma presents most commonly in the metaphysis of a long bone with 50% occurring around the knee. Germline p53 and RB1 mutations predispose to osteosarcoma; however, the genomic profile is chaotic and no consistent mutation has been identified as most mutations are somatic and highly diverse. Histology from the biopsy reveals malignant spindle cells with immature osteoid production. Roughly 80% of patients present with localized disease with no clinically detectable metastatic sites; however, all are expected to have micrometastases. Osteosarcoma is treated with preoperative chemotherapy followed by wide surgical resection for local control and ending with postoperative chemotherapy. The most common systemic agents used in a multiagent fashion are high-dose methotrexate, doxorubicin, and cisplatin. Patients with localized disease are expected to achieve a long-term disease-free survival of around 70% with this approach. This drops to 20–30% in patients presenting with detectable metastasis or who have a relapse of disease on or after treatment. Our treatment regimens have not changed much in 30 years, and a continued, coordinated, global partnership is needed to push research for improved targeted agents forward.

24.8.2 Ewing Sarcoma

Ewing sarcoma is the second most common primary bone sarcoma in children and adolescents after osteosarcoma and is the most common tumor in the family of small round blue cell tumors. Ewing sarcoma typically presents in the first or second decade but can present later in life. There is a predilection for the lower extremity and pelvis and can occur in almost any bone including diaphyseal locations. Unlike osteosarcoma, Ewing sarcoma has a well-known translocation involving the EWS gene on chromosome 22 and the ETS family of transcription factors, most typically FLI1 on chromosome 11. Histology is consistent with sheets of small round blue cell, but immunohistochemical stains for CD99 and MIC2 help the diagnosis. Multiagent chemotherapy is used in the preoperative and postoperative setting. Ewing sarcoma is radiosensitive, but local control is best achieved with a wide resection. However, certain situations may dictate the use of radiation alone or in combination with surgery to achieve local tumor control. Since Ewing sarcoma has a defined translocation, much research has been poured into the downstream effects of this and the targetable inducers of disease.

24.8.3 Chondrosarcoma

Chondrosarcoma is the second most common primary bone sarcoma and presents most commonly in people in the third–eighth decades. Chondrosarcoma is a malignant cartilaginous tumor that has a predilection for the axial skeleton or the proximal regions of the appendicular skeleton. Unlike osteosarcoma and Ewing sarcoma that usually always present as high grade, chondrosarcoma presents in more of a spectrum of diseases from borderline benign to high grade. Conventional chondrosarcoma is typically intramedullary, central, and can present as a low-, intermediate-, or high-grade tumor. There are several other variants of chondrosarcoma that include secondary, juxtacortical, clear cell, mesenchymal, dedifferentiated, and extraskeletal mesenchymal. It is important to understand that the diagnosis of conventional chondrosarcoma involves the integration of clinical, radiographic, and histologic features. Secondary chondrosarcomas can develop from prior benign cartilage lesions like osteochondroma or enchondroma. Chondrosarcoma is classically radio- and chemoinsensitive, and surgery is the gold standard for

treatment. It is important to tailor each patient's clinical, radiographic, and histologic information when formulating a surgical treatment plan.

Take-Home Message

Malignant bone tumors are rare sarcomas that arise in the bone primarily. It is important to work up with plain images, MRI of entire bone, and staging studies. The biopsy is the last piece of diagnostic information. Osteosarcoma is treated with chemotherapy, wide resection, and then chemotherapy. Ewing sarcoma is treated the same, and surgery can be substituted with radiation in some cases where surgery may not be possible. Chondrosarcoma is treated with wide resection alone. Negative margins are important to both local recurrence and overall survival. There are a variety of limb salvage techniques that include mega endoprosthetics and structural allografts.

Summary

In summary, malignant primary bone sarcomas are rare entities that require a detailed and algorithmic approach. Inappropriate workup can lead to substantial delays in diagnosis and increased morbidity and mortality. Centers that specialize in sarcoma treatment are best equipped to handle these complex problems.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Which of the following is not a principle of open biopsy?
   (a) Meticulous hemostasis
   (b) Transverse incision
   (c) Incision in line with definitive surgical approach
   (d) Single compartment
2. What is the single best imaging study when evaluating for skip metastases?
   (a) MRI with and without contrast
   (b) CT scan
   (c) Plain X-ray
   (d) MRI without contrast
3. What is the best initial imaging study when evaluating a bone lesion?
   (a) MRI with and without contrast
   (b) CT scan
   (c) Bone scan
   (d) Plain X-ray
4. What is the most common surgical resection for a bone sarcoma?
   (a) Intralesional resection
   (b) Wide resection
   (c) Marginal resection
   (d) Narrow resection
5. What is the most common presentation of a malignant bone tumor?
   (a) Painless incidental finding on plain X-ray
   (b) Constant, dull, achy pain present at rest and at night
   (c) Constant, sharp pain present more with activity that goes away with rest
   (d) Intermittent shooting pain worse with certain positions

Further Reading

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Soft Tissue Tumors

25

Recep Öztürk

Overview

The definition of soft tissue mass more commonly includes benign soft tissue tumors and reactive lesions, but also includes a rare and more serious group of soft tissue sarcomas.

Soft tissue tumors are rare and are a heterogeneous group of tumors consisting of many subgroups with different natural trends, biologies, and treatment responses. For this reason, despite the availability of immunohistochemistry markers and studies on genetic and biochemical new markers in recent years, defining histopathological subgroups may be difficult.

25.1 Definition

Soft tissue tumors include the group of mesenchymal tumors seen in non-epithelial tissues. These tumors are grouped mainly by their histogenesis, and cytogenetic and molecular genetic information is included in these examinations. The presence of more than 100 benign and malignant soft tissue tumors has been identified.

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25.2 Epidemiology

Soft tissue sarcomas are known to cover about 0.5% of all cancers, and benign/malignant ratio of soft tissue tumors is about 100:1. About 13,000 cases are reported annually in the USA, and in Europe, approximately 23,000 new cases of soft tissue sarcoma are reported.

These tumors can be seen at any age; 15% of cases occur in childhood. Men are more affected compared to women [M:F: 1.4:1]. About 40% of these tumors are located in the lower extremities, 20% in the upper extremities, 30% in the trunk and retroperitoneum, and 10% in the head and neck.

25.3 Etiology/Pathogenesis

Benign and malignant soft tissue tumors often develop de novo without identifiable risk factors or predisposing conditions.

In some cases, radiation, chemical burns, thermal burns, trauma, phenoxyherbicide exposure, and HHV8 virus (Kaposi's sarcoma) have been identified.

In addition, some genetic diseases (neurofibroma, malignant peripheral nerve sheath tumor in neurofibromatosis type 1; fibromatosis in Gardner's syndrome, etc.) have an increased risk of developing soft tissue tumors.

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25.4 Classifications

Soft tissue tumors are classified histologically by predominant cell type. There are hundreds of different soft tissue tumors, some of which are given in Table 25.1.

The most commonly used system for staging is the American Joint Committee on Cancer (AJCC) system (Table 25.2). The order of importance of prognostic factors is presence of metastasis (stage IV), grade (low grade—stage I, high grade—stage II), size (T1 ≤5 cm, T2 >5 cm), and location (superficial or deep) [3].

Table 25.1 Histological classification of soft tissue tumors

Histologic type	Benign	Malignant
Adipocytic	Lipoma	Well-differentiated liposarcoma
Fibrous	Nodular fasciitis	Fibrosarcoma
Nerve sheath	Schwannoma	MPNST
Skeletal muscle	Rhabdomyoma	Rhabdomyosarcoma
Smooth muscle	Leiomyoma	Leiomyosarcoma
Synovial	Focal PVNS	Malignant PVNS
Vascular	Hemangioma	Angiosarcoma

DFSP dermatofibrosarcoma protuberans, *MPSNT* malignant peripheral nerve sheath tumor, *PVNS* pigmented villonodular synovitis

Table 25.2 AJCC version 8 staging for soft tissue sarcomas

	Primary tumor (T)
Tx	Primary tumor cannot be assessed
T1	Tumor ≤5 cm
T2	5 cm < tumor ≤10 cm
T3	10 cm < tumor ≤15 cm
T4	Tumor >15 cm
	Regional lymph nodes (N)
N0	No regional lymph node metastasis or unknown
N1	Regional lymph node metastasis
	Distant metastasis (M)
M0	No distant metastasis
M1	Distant metastasis
	Staging
IA	T1; N0; M0; G1
Ib	T2, or T3, or T4; N0; M0; G1
II	T1; N0; M0; G2 or G3
IIIA	T2; N0; M0; G2 or G3
IIIB	T3, or T4; N0; M0; G2 or G3
IV	Any T; N1; M0; any G or any T; any N; M1; any G

25.5 Diagnosis

25.5.1 Story

The story includes current symptoms and related history/family history.

As current symptoms, pain, palpable mass presence, and growth rate of the mass should be evaluated. A soft tissue sarcoma may not cause any symptoms or signs in the early stages. As the tumor grows, it can cause a noticeable lump or swelling. Pain may occur when pressing on muscles or neural structures. Rapid growth may be an indicator of malignancy, but malignancy may also be considered in a long-standing and slow-growing soft tissue mass.

Related story/family story includes the presence of cancer or syndrome (like neurofibromatosis) in the patient or in the family, exposure to carcinogens, infection, or trauma (myositis ossificans).

25.5.2 Physical Examination

Mass presence (compressible lesions suggest benign tumors, and firm masses suggest sarcomas or desmoid tumors), range of motion (tumor-associated joint range of motion may decrease), muscle atrophy (painful lesion), and lymphadenopathy (lymph nodes may also grow due to infection or metastasis) are examined.

25.5.3 Imaging

Plain radiographs: Scattered calcifications (at 30%) in synovial sarcomas, peripheral mineralization in myositis ossificans, phleboliths in hemangiomas, and radiolucent lesions in lipomas can be seen on radiographs.

MRI: It is the gold standard imaging method in soft tissue imaging. The nature of some lesions can be determined by MRI (in lesions such as lipoma, hemangioma, ganglion, muscle damage), and definitive treatment can be given in these lesions. In masses where the nature of the lesion cannot be determined with MRI, biopsy should be performed first. In the majority of sarcomas, the diagnostic image is uncertain in terms of

malignancy, and biopsy is required. Even if a lesion is small and painless, when a specific diagnosis cannot be made, potential malignancy should be considered and not merely observed.

CT: Soft tissue sarcomas most often metastasize to the lung. Lung CT is used to detect lung metastases.

25.5.4 Biopsy

It is the most important step in the diagnosis of many soft tissue lesions. Biopsy should be done by a team that will perform the final surgery at a center specialized in tumors. Inappropriate biopsy can lead to misdiagnosis due to the intake of tissue that does not represent the mass, or complications such as infection and neurovascular damage. In addition, the unplanned biopsy location can harm the final surgery. Contamination or inappropriate incision line during biopsy can cause more complicated surgeries that require a flap/graft or lead to unnecessary amputations.

Needle biopsy (fine needle or true-cut), open incisional biopsy, and excisional biopsy options are available. The most common method is needle biopsy, which requires experienced pathologist. In open incisional biopsy, the rules of biopsy should be followed at the highest level, and very good hemostasis should be performed. Excisional biopsy can be performed if the surgeon is sure that the lesion is benign.

25.6 Treatment

Treatment in soft tissue tumors is planned according to the biological activity of the lesion. In benign tumors, inactive-latent lesions (superficial lipoma, etc.) are followed, inactive-symptomatic or active lesions (intramuscular myxoma, etc.) are treated with simple excision, and aggressive lesions (extra-abdominal fibromatosis, etc.) are treated with wide resection.

In the treatment of soft tissue sarcoma, wide resection or combination of resection with external beam radiotherapy is applied. Preoperative or postoperative radiotherapy can be administered. Amputation is applied to the tumor in cases where wide resection cannot be performed or a functional limb cannot be obtained after wide resection. Today, about 90% of patients can be operated with limb-sparing surgery. Surgical margins are shown in Fig. 25.1.

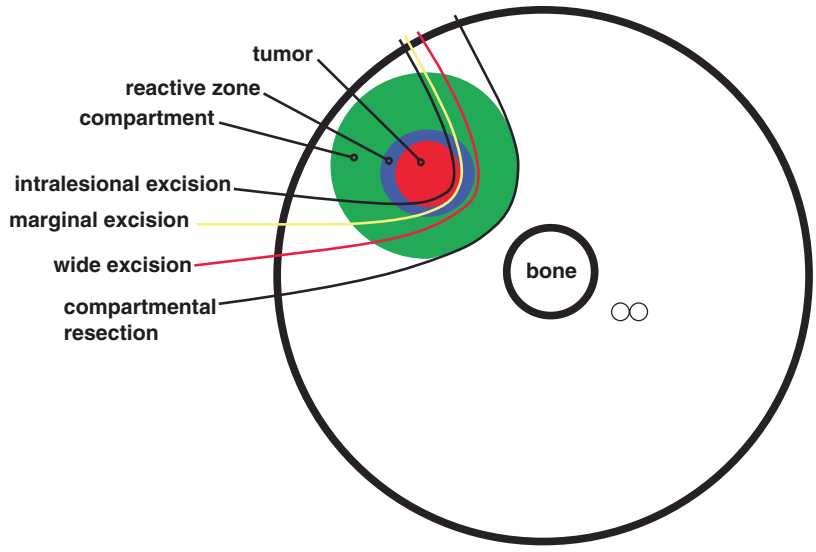

Fig. 25.1 Surgical margins: Intralesional excision is the method in which the surgical incision passes through the tumor and the macroscopic residual tumor is left behind. Marginal excision is the method where the surgical incision passes through the reactive region between the tumor and the pseudocapsule. Wide excision is the method where the surgical incision is outside the reactive region and pseudocapsule. The incision passes through normal tissue of all sizes. Compartmental excision is the excision of a tumor with an anatomical chamber containing the tumor

Take-Home Message

- Since soft tissue tumors are particularly rare and the majority of them are benign, there may be important problems in the diagnosis and treatment of malignant tumors.
- Soft tissue sarcomas are often painless masses, unlike bone sarcomas.
- Soft tissue sarcomas usually have well-defined margins, so it is difficult to distinguish them from benign soft tissue lesions in MRI.
- For lesions with malignant tumor characteristics (large and deep localization), STS should be kept in mind and biopsy should be performed.
- Unplanned biopsies and intralesional interventions can make extensive resection impossible or cause amputation.

Summary

Soft tissue tumors are divided into two main groups as benign and malignant. The most common treatment in benign STTs is simple excision. In soft tissue sarcomas, the basis of treatment is resection of the tumor with wide borders. In addition, RT is added to the treatment in appropriate cases.

The importance order of prognostic factors in soft tissue sarcomas is the presence of metastasis, grade, size, and localization. Soft tissue sarcomas most often metastasize to the lung. It is mainly detected by lung CT.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Which of the following is wrong with soft tissue tumor surgery?
   (a) Macroscopic tumor is left behind in intralesional excision
   (b) Marginal excision is the removal of the entire tumor
   (c) The preferred surgical treatment in soft tissue sarcomas is marginal excision
   (d) In wide resection, the surgical incision line passes through normal tissue
2. A 26-year-old male patient has a soft tissue mass on his left elbow. Figure A shows plain radiograph, and Fig. B and Fig. C show MRI images. What is the next most appropriate procedure for this patient?
   (a) Observation
   (b) Needle biopsy
   (c) Marginal excision
   (d) Wide excision
3. In which of the following, phleboliths can be seen on direct radiography?
   (a) Synovitis
   (b) Lipoma
   (c) Fibromatosis
   (d) Hemangioma
4. A 44-year-old woman has a painless mass on the posterior of the left knee. Figure A shows plain radiograph, and Fig. B and Fig. C show MRI images. What is the next most appropriate procedure for this patient?
   (a) Observation
   (b) Medical treatment
   (c) Needle biopsy
   (d) Marginal excision
5. After a simple excision of a 4 cm superficial mass on the right thigh of a 42-year-old male patient, the result of pathology was synovial sarcoma. No information was given about the surgical margins. What is the next most appropriate procedure for this patient?
   (a) Observation
   (b) Chemotherapy
   (c) Radiotherapy (to the tumor bed)
   (d) Wide resection of the tumor bed

Further Reading

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Metastasis

26

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Overview

Bone is the third most common site for metastasis in cancer. Bone metastasis occurs in most tumor types but is most prevalent in cancers of the breast, prostate, and lungs. The main purpose of the treatment for bone metastasis is to improve symptoms and prevent symptomatic skeletal events.

26.1 Introduction

Bone is the third most common site for metastasis in cancer. Bone metastasis occurs in most tumor types but is most prevalent in cancers of the breast, prostate, and lungs. The incidence of bone metastasis in prostate cancer is 65–90%, in breast cancer 65–75%, and in lung cancer 17–64%, and it is less frequent in thyroid, renal, and colorectal (10%) cancers. The prevalence of bone metastasis is estimated to be approximately 280,000 new cases per year in the USA and is expected to increase as medical management and surgery improve overall survival. Patients with bone metastasis may experience skeletal complications including bone pain, pathological fractures, spinal cord or nerve root compression, and hypercalcemia requiring orthopedic surgery and/or adjuvant treatments such as radiotherapy and chemotherapy. These events associated with an exacerbation of cancer-related pain have been described as "symptomatic skeletal events" (SSEs). The main purpose of the treatment for bone metastasis is to improve symptoms and prevent symptomatic skeletal events. The decision for surgical and/or medical treatments depends on the prognosis and expected survival for the cancer patients with bone metastasis. Less invasive and palliative procedures are performed for patients with poorer prognosis.

26.2 Pathogenesis

Paget's "seed and soil" hypothesis and Ewing's anatomical hypothesis have described the possible routes of migration and proliferation of cancer cells to bone. According to this hypothesis, cancer cells metastasize towards a favorable microenvironment or via the vessels. Bone is a rich source of laminin, collagen type I, osteopontin, and fibronectin; certain integrins expressed by malignant cells have specific receptors for these mole-

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cules. The bone-homing capacity of tumor cells is increased by the adhesive interaction between tumor cells and bone (mediated by the integrins). Once the surrounding environment becomes suitable for cancer cell proliferation, bone metastasis may arise. The type of interaction between the bone microenvironment and the tumor cells can potentially give rise to osteolytic or osteoblastic metastases. Osteoclastic activity is stimulated by factors secreted by malignant cells.

26.3 Long Bone Metastasis

Pathological long-bone fractures in bone metastasis cause difficulties to the most experienced orthopedic oncology surgeons. Prognosis, quality of life, and survival of the patients depend on their management. The incidence of a long-bone pathological fracture in bone metastasis has been reported to be between 10% and 29%. The most common primary tumor in long-bone metastasis is breast followed by lung, prostate, renal, and thyroid cancers, and the most commonly affected site is the femur. The surgical treatment is described in Chap. 26.

26.4 Acrometastasis

Acrometastasis is defined as bone metastasis distal to the elbow and the knee and accounts for approximately 0.1% of bone metastasis. Acrometastasis usually arises from lung (>50%) or renal cancer, followed by cancers of the colon, breast, and genitourinary tract. It may appear in patients of every age, with men being twice as likely as women to be affected. The prognosis of patients with acrometastasis is generally poor since it indicates a widespread disseminated disease in most cases. Acrometastasis may also appear as the first presentation of occult silent cancer in 10% of the cases and can mimic other benign conditions such as inflammatory lesions, cysts, gout, ganglia, osteomyelitis, tuberculous dactylitis, pyogenic granuloma, and primary skin tumors. As a result, it is essential not to misdiagnose such a lesion. Lung cancer is the main cause of hand acrometastasis. More specifically, the distal phalanx of the thumb in the dominant hand is more commonly affected while acrometastasis in the middle phalanx and the carpus is extremely rare. The tarsal bones are most commonly involved in foot acrometastasis. Lower circulatory flow in distal regions of the body such as the hand phalanges is beneficial for secondary tumor growth. It is also worth mentioning that foot acrometastasis is usually associated with subdiaphragmatic tumors most commonly originating in the gastrointestinal, vesicle, renal, and uterus areas. Clinical examination shows deep and intermittent pain, refractory to common analgesics, along with a palpable mass, an enlarging digit, or a mechanical dysfunction impairing daily activities. Pain is a result of the inflammatory process, which leads to swelling, erythema, ulceration, and bleeding. Differential diagnosis should include benign conditions such as osteomyelitis, rheumatoid arthritis, tenosynovitis, and gout. A destructive permeative lesion is usually shown in radiographs. Lytic solitary lesions in a single bone in the hand are usually due to lung and renal cell carcinomas, while breast cancer acrometastasis is mainly sclerotic, lytic or mixed, and often multiple. CT in distal extremities has a limited value due to the decreased resolution in these areas. However, MRI consists of a useful imaging study in evaluating marrow disease and extraosseous extension of the tumor. Radiographs of the lesion, chest, and abdomen CT along with bone scan are essential for complete staging of the patients with acrometastatic cancer since the extent of spread is the most important factor for the treatment approach and prognosis. Bone scan assesses possible multiplicity and leads to either false-negative or -positive results due to very aggressive tumors or previous trauma, respectively. Positron-emission tomography scan has increased sensitivity. Trocar biopsy tissue sampling is paramount for histological diagnosis and documentation of the acrometastasis, while incisional biopsy should be avoided. The surgical treatment is described in Chap. 26.

26.5 Spine Metastasis

The spinal column is the third most common site for bone metastasis (after the lungs and the liver) and the most common site for bone metastasis (especially the thoracic spine). More specifically,

10–15% of spine metastasis occurs in the cervical spine, 60–70% in the thoracic spine, and 20–25% in the lumbosacral spine. Multiple lesions occur in 17–30% of patients. It is estimated that 10% of patients with cancer develop symptomatic spinal metastases, while this percentage increases to 40% in those with actual metastatic bone disease. Interestingly, in postmortem cadaveric studies, the actual incidence of spine metastasis in cancer patients regardless of symptomatology has been up to 90%. The most common primary cancers are breast (16–37%), lung (12–15%), and prostate cancer (9–15%), while non-Hodgkin's lymphoma and renal cell cancer account for 5–10%; the remaining cases are due to gastrointestinal carcinomas, sarcomas, and unknown primary tumors. Batson's vertebral plexus, a valveless venous plexus of the spine, provides a route of metastasis from organs to axial structures. Bone lysis or sclerosis leads to cortical bone destruction that causes vertebral body collapse. Epidural spinal cord compression, an emergency complication of spine metastasis, may occur through epidural spread by an initial hematogenous metastasis to the vertebral body or less commonly from direct growth of a paravertebral tumor such as lymphomas into the spinal canal through an intervertebral foramen. Before establishment of the diagnosis, 83–95% of the patients experience back or neck pain that may be mechanical due to bone destruction or tumorigenicity (mostly nocturnal). Pain may be unilateral or bilateral in the spine; it is localized to the region of the spine that is affected by the metastasis; it is worse when the patients are recumbent due to lengthening of the spine and distension of the spinal epidural venous plexus and progressively increases in intensity over time. Metastatic epidural spinal pain is caused when the periosteum, paravertebral soft tissues, or nerves are invaded by the enlarging mass and also by spinal instability, pathological fractures, and inflammatory and nociceptor stimulating substances secreted by the cancer cells. Interestingly, pain may be absent with intradural or intramedullary metastasis. Radicular pain due to nerve invasion and compression is less common than mechanical pain and may be accompanied by depression of tendon reflexes, weakness, and sensory changes in the distribution of the roots injured by epidural metastasis along with bowel or bladder disorders. New onset of back or neck pain in cancer patients should be always evaluated in order to rule out spine metastasis. A detailed medical history and physical examination should be the first steps prior to any imaging studies. Vertebral abnormalities such as pedicle erosion, vertebral collapse, and bone dislocation are seen in radiographs in 70–80% of patients with spine metastasis. However, decreased sensitivity and specificity are a major drawback. Treatment of spine metastasis requires a multidisciplinary team approach by a neurologist, radiologist, oncologist, and orthopedic surgeon. The goals of treatment are the ability to walk, pain relief, spinal stability, and improved survival. However, in most cases, treatment is aimed for palliation.

Nonsurgical or minimally invasive palliative management is usually recommended for patients with very poor prognosis; radiotherapy, chemotherapy, vertebroplasty, and kyphoplasty have yielded satisfactory results for pain relief. If symptomatic metastatic spinal cord compression is confirmed, external beam radiation therapy and prompt administration of corticosteroids are standard options along with several analgesics and other supportive measures such as opioids controlled through a patient-controlled analgesia device, bisphosphonates, and spinal braces. Embolization should also be considered for pain palliation of patients with spine metastasis. Thorough patient evaluation should be performed prior to any treatment to minimize morbidity and increase therapy efficiency. Recently, a decision framework has been introduced which incorporates the basic principles that should be considered prior to the treatment of spine metastasis. These include the location of disease, with respect to the anterior and/or posterior columns of the spine and number of spinal levels involved (contiguous or noncontiguous), mechanical instability (as graded by Spine Instability Neoplastic Score—SINS), neurological manifestations and symptomatic epidural spinal cord compression, oncological properties and histopathologic diagnosis particularly with respect to radiosensitivity, and patient fitness, patient wishes, prognosis, and response to prior therapy. The surgical treatment is described in Chap. 26.

26.6 Pelvic Metastasis

The pelvis is the second most common site of bone metastasis after the spine. Clinical manifestations include pain, bone destruction causing mechanical instability, and pathological fractures. Metastatic lesions are always progressive and cause bone failure due to the tumor cell adhesive molecules that bind the tumor cells to the marrow stromal cells and bone matrix, allowing them to grow and produce angiogenic and bone-resorbing factors. In most cases, treatment is aimed at palliation since complete cure of the disease is not possible. Nevertheless, pelvic metastasis substantially decreases the quality of life of the patients and necessitates further treatment. The overall prognosis of the patients with pelvic metastasis extremely varies depending on the site of the lesion, type of primary cancer, and existence of associated metastases. Highly stressed anatomical sites are predisposed for pathological fractures. Enneking's classification divides the pelvic girdle into four different zones. Zones 1 and 3 are non-weight-bearing, while zone 2 is the periacetabular area. Periacetabular metastasis is at greater risk for mechanical failure with progressive destruction of the hip joint, while metastasis in zones 1 and 3 does not compromise the mechanical stability of the pelvic ring. Moreover, osteolytic lesions are considered more at risk for pathological fracture compared to osteoblastic or mixed lesions. Permeative pattern of osteolysis indicates the same risk of fracture as the more classic types that show a discrete area of osteolysis. Radiographs do not always reveal this permeative osteolysis; therefore, MRI is more sensitive to show the real extent of the disease. A prospective protocol containing guidelines on the indications for surgery and the type of procedure that should be undertaken was reported in 2001. Expected survival, type and stage of the tumor, visceral spread, time since detection of the primary tumor, risk for pathological fracture, and predictive sensitivity to chemotherapy, hormonal therapy, and radiotherapy were the accounted factors. According to this protocol, patients were categorized in four classes: class 1: patients with a single metastasis of a primary tumor with a good prognosis and an interval of >3 years from detection of the primary lesion to the development of bone metastasis: primary tumors with a favorable prognosis include well-differentiated thyroid, prostate, breast when sensitive to hormonal treatment or chemotherapy, clear-cell renal, and colorectal carcinoma; class 2: patients with a pathological fracture in a major long bone; class 3: patients with radiological and/or clinical signs of impending fracture in a major long bone or the periacetabular area; and class 4: patients with (a) osteoblastic lesions at all sites; (b) osteolytic or mixed lesions in non-weight-bearing bones such as the fibula, ribs, sternum, or clavicle; (c) osteolytic lesions in major bones with no impending fractures; and (d) lesions in the wing of the ilium, anterior pelvis, or scapula (excluding patients included in class 1). In addition, Harrington's classification has been used to indicate the acetabular destruction for patients in classes 2 and 3: Harrington group 1: integrity of medial and superior periacetabular bone; Harrington group 2: medial acetabular wall insufficiency; Harrington group 3: medial wall and supra-acetabular destruction; and Harrington group 4: total collapse of acetabulum. The surgical treatment is described in Chap. 26.

26.7 Palliative Treatments

Palliative treatment of metastasis in any site of the skeleton includes radiotherapy, radiopharmaceuticals, chemotherapy, embolization, thermal ablation techniques, electrochemotherapy, and high-intensity focused ultrasound (HIFU).

26.7.1 Radiotherapy

The primary goal of radiotherapy is to palliate painful metastasis, achieve local tumor control, and improve quality of life. Metastasis usually starts to get ossified in 3–6 weeks after treatment delivery, reaching the highest degree within 6 months. It has been reported that high partial response rates are approximately 60% and complete response rates

range from 10 to 25%. Single-fraction treatments have higher rates of retreatment.

External Beam Radiation Therapy: The exact mechanism of external beam radiation therapy is unknown. However, it is believed that tumor regression and cell death are implicated. Patients with repeat symptoms after their initial palliative radiotherapy and patients who receive single-fraction radiotherapy may require repeat radiotherapy.

Stereotactic Body Radiation Therapy: Stereotactic body radiation therapy is a highly focused form of radiotherapy designed to deliver high doses to targets while sparing normal surrounding tissue. It is performed using daily image guidance to deliver radiation precisely to the target. The complete response rates for pain and local control are improved because the biologically effective dose can range from 4 to 8 times that of conventional palliative radiotherapy.

26.7.2 Radiopharmaceuticals

Radiopharmaceuticals are now available for the palliation of metastatic bone pain, especially in patients with diffuse bone metastases for which external beam radiation alone is not adequate. Radionuclides, including beta-emitters such as strontium-89 (89Sr) and samarium-153 (153Sa), and alpha-emitters such as radium-223 (223Ra) can be selectively delivered towards bone to areas of amplified osteoblastic activity, sparing healthy organs from irradiation. Once target cells have been reached, radionucleotides induce DNA damage and apoptosis.

26.7.3 Chemotherapy

Systemic approaches to bone metastasis include specific anti-tumor treatments that are necessary to control disease progression at skeletal sites. The pathological type of the tumor is most important in selecting systemic treatment. Anticancer agents are used to control tumor progression, and bisphosphonates are used to prevent skeletal complications.

Bisphosphonates: Bisphosphonates are pyrophosphate analogues whose chemical structure is characterized by a P–C–P-containing central structure. Bisphosphonates cause osteoclast apoptosis by inhibiting farnesyl pyrophosphate synthase, which is essential for osteoclast survival and activity. They also reduce osteoclast activity indirectly through an effect on osteoblasts. Current guidelines suggest treatment with bisphosphonates for patients with bone metastasis. More specifically, zoledronic acid is approved for bone metastasis management in both solid tumors and multiple myeloma, while pamidronate can be used for breast cancer and multiple myeloma patients. Ibandronate is effective in breast cancer patients in both intravenous and oral formulations. Oral clodronate is another therapeutic option for the management of lytic skeletal metastases.

Denosumab: Denosumab is a human monoclonal antibody designed to target RANKL (RANK ligand), a protein that acts as the primary signal to promote bone loss. It inhibits the interaction between RANKL and RANK, to reduce osteoclast maturation and activity. General guidelines for denosumab use in bone metastasis are almost identical to bisphosphonates. Nevertheless, denosumab is not nephrotoxic, providing a valid treatment alternative in patients with kidney failure. Similarly to bisphosphonates, denosumab is generally well tolerated, with hypocalcemia and osteonecrosis of the jaw being the most common complications.

26.7.4 Embolization

Embolization is the selective occlusion of blood vessels that feed a tumor. Most metastatic lesions are hypervascular; some lesions such as renal and thyroid metastases are highly hypervascular. Embolization is a useful adjunctive procedure for the treatment of bone metastasis. Indication for embolization is control of hemorrhage, facilitation of subsequent surgery, inhibition of tumor growth, and relief of pain. Embolization can be single session or serial; repeat embolization is indicated when there is pain or imaging evidence of progressive disease. Pre-embolization, diagnostic

digital subtraction angiography must be performed in order to identify the feeding vessels. Embolization is technically successful when intravascular contrast material stops, or tumor's hypervascularity is completely eliminated, or when >80% of the tumor's vascularity is decreased in comparison with the initial angiography. It has been shown that embolization of a hypervascular bone metastasis reduces the intraoperative blood loss and the surgical time and that preoperative embolization has greater effectiveness in reducing intraoperative blood loss when surgery is performed at the same day of embolization. The post-embolization syndrome manifested with fever, pain, and malaise has been described in 18–86% of embolization cases. Embolization can also cause normal tissue loss and may be associated with nerve palsy, skin breakdown, subcutaneous or muscle necrosis, and infection.

26.7.5 Thermal Ablation

Ablative techniques allow for both tumor destruction and palliation of pain from bone metastasis. Thermal ablation was introduced as a palliative treatment of painful bone metastasis in the early 2000s. The aim of thermal ablation techniques is to induce coagulative necrosis with heat, ideally around 70 °C for bone lesions. Radiofrequency, cryotherapy, laser, and microwave thermal ablation are the mostly used techniques.

Radiofrequency ablation (RFA): Radiofrequency ablation (RFA) is the most frequently used thermal ablation technique, with high success rates for bone and soft tissue tumors for skeletal events and local tumor control. Radiofrequency ablation works by conducting an alternating current of high-frequency radio waves, which passes through a probe placed within the lesion.

Cryotherapy: Cryotherapy works by argon gas that is delivered through a partially insulated probe positioned at the center of the lesion. The low temperature causes loss of the cell membrane integrity and cell death. Cryotherapy has shown a greater reduction in analgesic doses and shorter hospitalization time after the procedure compared to radiofrequency ablation. Moreover, cryotherapy can be used for osteoblastic lesions since ice can penetrate bone.

26.7.6 Electrochemotherapy

Mir et al. first described electrochemotherapy (ECT) for the treatment of cutaneous nodules of head and neck malignant tumors, as the combination of permeabilizing electric pulses with intravenous infusion of a chemotherapeutic drug to which the cellular membranes are usually poor or non-permeant. The electric current pulses to the tumor tissue induce the opening of the transmembrane channels, therefore allowing the chemotherapeutic drugs to enter the cell, increasing a localized cytotoxic effect. Electrochemotherapy has proven effectiveness in the treatment of metastasis from solid tumors such as breast cancer and melanoma located in the skin or subcutaneous tissue.

26.7.7 High-Intensity Focused Ultrasound (HIFU)

High-intensity focused ultrasound is considered a safe and the only completely noninvasive and extracorporeal procedure to treat primary solid tumors and bone metastasis. High-intensity focused ultrasound causes a temperature rise in tissues inside the targeted area, resulting in coagulative necrosis at a thermal threshold of 65–85 °C, depending on the tissue absorption coefficient. Ultrasound beam focus results in high intensities only at a specific location within a small volume that minimizes the potential for thermal damage to tissue outside the focal region. Several lesions can be treated per session, and treatment may be repeated as many times as needed because there is no dose limit and no ionizing radiation from MRI and diagnostic ultrasound as opposed to

other systems that are guided by radiographs, and maintenance of the system is low. Other advantages are the low cost of the procedure as compared with traditional surgery, no remaining scars, and faster recovery, and if any hemorrhage occurs, ultrasound has the potential to stop it. The main limitation is that high-intensity focused ultrasound cannot be used to treat vertebral metastases.

Treatment decision in patients with bone metastases requires complete staging, biopsy, and oncological principles. Useful algorithms for decision-making and treatment have been proposed; pain should be first treated with analgesics, bisphosphonates, and denosumab, and secondly with radiotherapy, electrochemotherapy, radiofrequency ablation, embolization, high-intensity focused ultrasound, and chemotherapy, and finally, if there is no pain relief radiation, electrochemotherapy, radiofrequency ablation, embolization, and high-intensity focused ultrasound can be repeated.

26.7.8 Interventional Oncology Procedures and Bone Augmentation (IO)

Many of the above-described techniques can be now performed percutaneously, with minimal trauma, as they can be image-guided procedures (Figs. 26.1 and 26.2). Imaging techniques

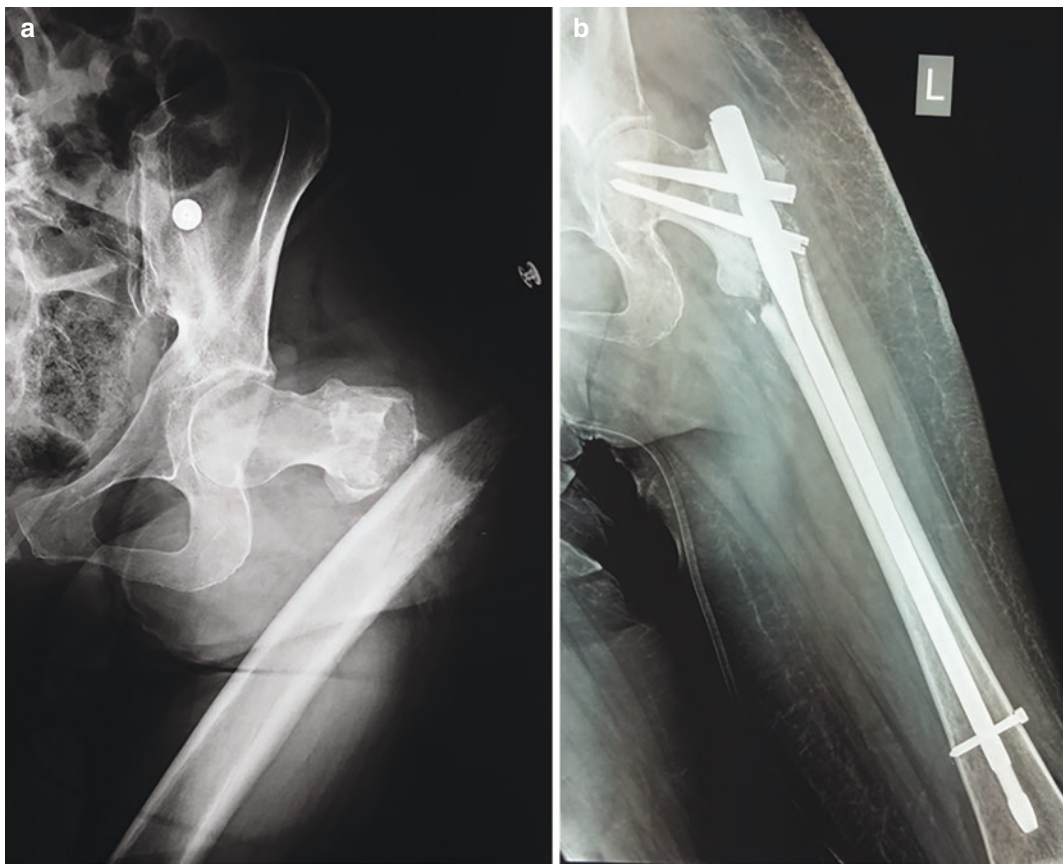

Fig. 26.1 (**a**) A female patient with a pathological subtrochanteric left-hip fracture; biopsy showed breast cancer. (**b**) Closed reduction and reconstruction-type intramedullary nail osteosynthesis were done

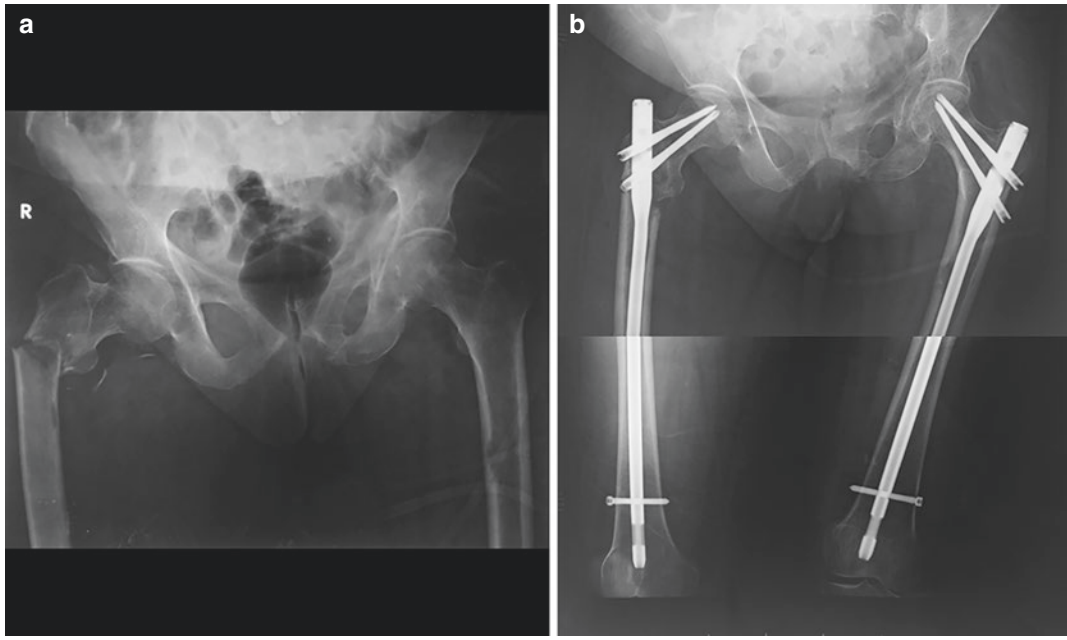

Fig. 26.2 (**a**) A female patient with a pathological subtrochanteric right-hip fracture and impending left-hip fracture; biopsy showed breast cancer. (**b**) Closed reduction of the right hip fracture and reconstruction-type intramedullary nail osteosynthesis of both femurs were done

vary from simple ultrasound guidance for soft tissue lesions to CT, MRI, and cone beam CT reconstructions with the use of C-arm or angiographic suites. Developments in this field include fusion imaging and needle guidance systems, which superimpose the needle track to the MPR reconstruction. The ablation techniques should usually be accompanied by stabilization procedures, especially in weight-bearing areas (Fig. 26.3), where the ablation affects the structural integrity of the bone. If the ablation occurs in non-weight-bearing areas, it can be a stand-alone procedure (Fig. 26.4). Otherwise, there is an increased risk of a secondary fracture and thus it should be accompanied by augmentation with intraosseous polymer injection (Fig. 26.5). This type of polymers could be percutaneously injected, independently, as a palliative procedure.

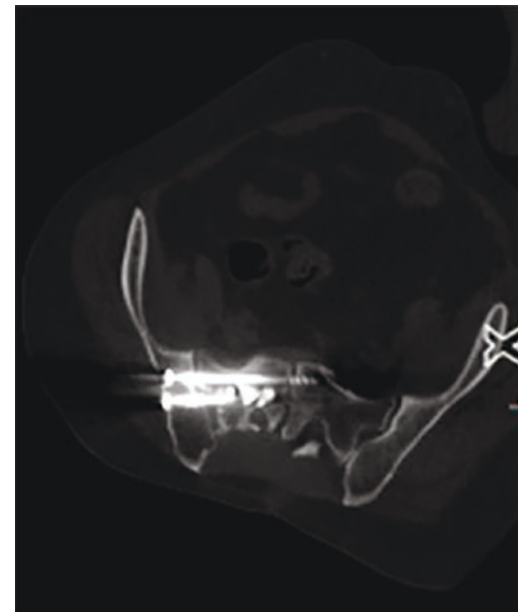

Fig. 26.3 A sarcoma male patient with residual tumor and a pathological fracture post-surgery and radiotherapy. The patient was treated with percutaneous microwave ablation combined to augment sacroplasty with cement injection and placement of two cannulated screws. Computed tomography axial reconstruction shows the cement and the cannulated screws in position

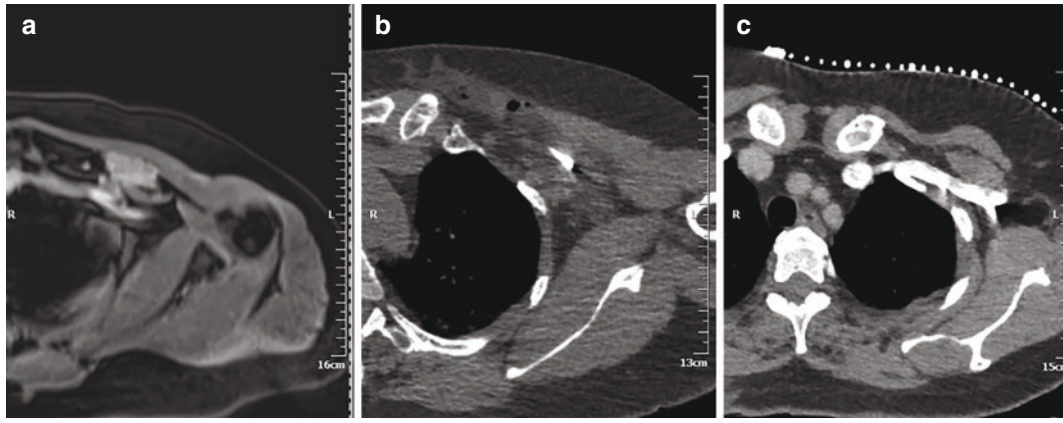

Fig. 26.4 A female breast cancer oligometastatic patient. (**a**) MRI post-contrast injection shows a solitary infraclavicular lymph node. The patient was treated with cryoablation under CT guidance. (**b**) Hypodense appearance of the ice around the needle. (**c**) Follow-up image at 6 months by contrast-enhanced CT shows disappearance of the lesion

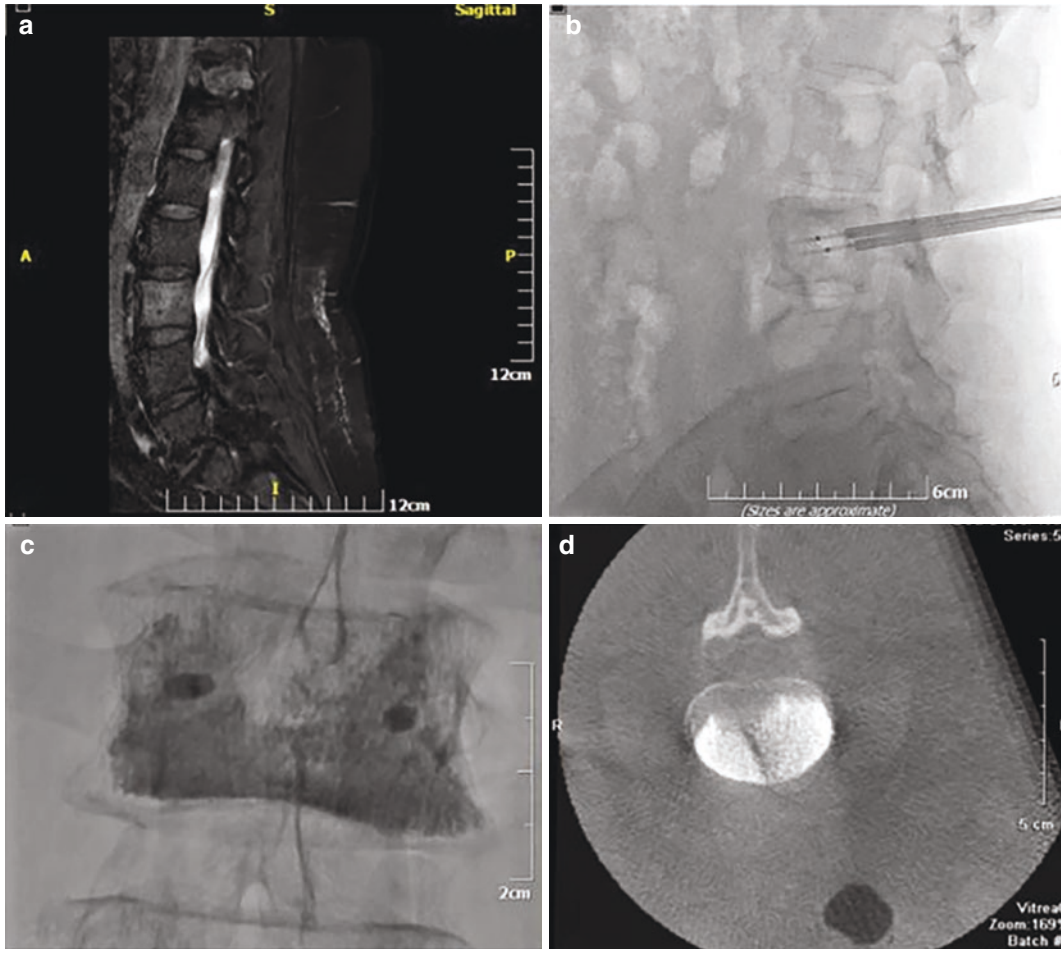

Fig. 26.5 A female breast cancer oligometastatic patient with a solitary L3 vertebral body metastasis. (**a**) T1-weighted sagittal fat suppression MRI shows lesion's uptake and enhancement. (**b**) Fluoroscopy lateral view shows two bipolar radiofrequency electrodes placed inside the lesion through percutaneous transpedicular approach. (**c**) Fluoroscopy posteroanterior view evaluates cement distribution following polymer injection. (**d**) Cone beam 3D CT reconstruction showing the filling of the vertebral body with the cement

Take-Home Message

- Bone metastasis occurs in most tumor types but is most prevalent in cancers of the breast, prostate, and lungs.
- Patients with bone metastasis may experience skeletal complications including bone pain, pathological fractures, spinal cord or nerve root compression, and hypercalcemia requiring orthopedic surgery and/or adjuvant treatments such as radiotherapy and chemotherapy.
- Acrometastasis is defined as bone metastasis distal to the elbow and the knee and accounts for approximately 0.1% of bone metastasis.
- Nonsurgical or minimally invasive palliative management is usually recommended for patients with very poor prognosis; radiotherapy, chemotherapy, vertebroplasty, and kyphoplasty have yielded satisfactory results for pain relief.

Summary

Bone is the third most common site for metastasis in cancer. Bone metastasis occurs in most tumor types but is most prevalent in cancers of the breast, prostate, and lungs. The incidence of bone metastasis in prostate cancer is 65–90%, in breast cancer 65–75%, and in lung cancer 17–64%, and it is less frequent in thyroid, renal, and colorectal (10%) cancers. Patients with bone metastasis may experience skeletal complications including bone pain, pathological fractures, spinal cord or nerve root compression, and hypercalcemia requiring orthopedic surgery and/or adjuvant treatments such as radiotherapy and chemotherapy. The main purpose of the treatment for bone metastasis is to improve symptoms and prevent symptomatic skeletal events. Paget's "seed and soil" hypothesis and Ewing's anatomical hypothesis have described the possible routes of migration and proliferation of cancer cells to bone. Pathological long-bone fractures in bone metastasis cause difficulties to the most experienced orthopedic oncology surgeons. Acrometastasis is defined as bone metastasis distal to the elbow and the knee and accounts for approximately 0.1% of bone metastasis. The prognosis of patients with acrometastasis is generally poor since it indicates a widespread disseminated disease in most cases. The spinal column is the third most common site for bone metastasis (after the lungs and the liver), and the most common site for bone metastasis; the thoracic spine is the most common spinal segment involved. Nonsurgical or minimally invasive palliative management is usually recommended for patients with very poor prognosis; radiotherapy, chemotherapy, vertebroplasty, and kyphoplasty have yielded satisfactory results for pain relief. The pelvis is the second most common site of bone metastasis after the spine. Clinical manifestations include pain, bone destruction causing mechanical instability, and pathological fractures. Palliative treatment of metastasis in any site of the skeleton includes radiotherapy, radiopharmaceuticals, chemotherapy, embolization, thermal ablation techniques, electrochemotherapy, and high-intensity focused ultrasound (HIFU).

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. The primary tumor that produces metastasis most frequently is:
   (a) Prostate cancer
   (b) Breast cancer
   (c) Lung cancer
   (d) Thyroid cancer
2. The seed and soil hypothesis stated that cancer cells metastasize:
   (a) Towards a favorable microenvironment or via the vessels
   (b) Per contiguity
   (c) At distance
   (d) Depending on the patient's characteristic
3. Acrometastasis accounts:
   (a) For 0.1% of bone metastasis
   (b) For 1% of bone metastasis

(c) For 10% of bone metastasis
(d) For 20% of bone metastasis

4. The most common site for bone metastasis is:
   (a) Spinal column
   (b) Femur
   (c) Distal radius
   (d) Knee
5. Bisphosphonates cause:
   (a) Osteoclast apoptosis
   (b) Osteoblastic generation
   (c) Osteoclast inhibition blocking RANK
   (d) Osteogenesis

Further Reading

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Surgical Management of Bone Metastasis

27

Vincenzo Denaro, Umile Giuseppe Longo, Sergio De Salvatore, and Luca Denaro

Overview

Surgical treatment of bone metastases aims to reduce pain, improve function, prevent tumour progression and decrease complications.

27.1 Introduction

The surgical treatment of bone metastases aims to reduce pain, improve function, prevent tumour progression and decrease complications.

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Table 27.1 Factors influencing the surgical strategy

Prognosis	• Good • Poor
Histotype	• Aggressive • Not aggressive
Chemo- and radio-sensitivity	• Responding • Not responding
Number of lesions	• Solitary • Multiple
Location	• Diaphysis • Meta-epiphysis
Type of pathological fracture	• Actual • Impending

Nowadays, the improvements in diagnostic technologies and therapy management have increased the life expectancy, granting the detection of metastases in the early stage of the disease. Generally, surgery is indicated only for patients with an expected survival of 3–6 months. Surgical indication is tailored to the patient, depending on the characteristic of the metastatic disease and prognostic factors of the patient (Table 27.1).

27.2 Epidemiology

The most frequent site for solid tumour metastases is the spine. In fact, 5–10% of patients with systemic cancer develop spinal metastases during their life. The most common cancers that metastasize in the spine are breast cancer, prostate cancer, cell lung carcinoma, renal cell carcinoma and thyroid cancer.

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The femur (60%), humerus and tibia are the most common sites affected by metastatic fractures. Regarding femur fractures, 50% involve the neck, 30% the subtrochanteric region and 15% the intertrochanteric region.

27.3 Surgical Management

Pain is persistent and more frequent during the night. The evaluation of mechanical stability is mandatory before considering the surgical option. Radiographs (static and dynamic), computed tomography (CT) and surgeon's experience are required to assess the mechanical stability of the lesion. The presence of structural integrity of the cortex or the absence of intracortical and cancellous bone alterations is a favourable prognostic factor. Magnetic resonance imaging (MRI) could assess the extent of the tumour and the possible soft tissue involvement. Moreover, MRI could detect spot lesions that are not accessible with X-rays.

A biopsy is mandatory to confirm bone metastasis, mainly when a bone lesion is solitary, and the primary tumour is unknown. In patients with an uncertain diagnosis, an open biopsy with frozen section should be performed before fixation.

Generally, a CT-guided biopsy is sufficient to obtain a specific diagnosis. However, in impending fractures or in case of uncertain diagnosis (with or without the risk of bone collapse), an open biopsy is preferred, followed by a surgical fixation.

The surgeon should not conclude the procedure if the pathology report has confirmed the metastatic disease. In uncertain frozen biopsy results, the surgery should be postponed until the definitive pathology diagnosis has been obtained.

Prophylactic embolization with angiography (24 h before surgery) is effective in 90% of hypervascular lesions (thyroid, clear cell kidney carcinoma, liver carcinomas or myeloma), reducing the intra- and post-operative bleeding and facilitating the surgical excision.

The parameters to consider before surgery are acceptable perioperative life risk and shorter recovery than residual life expectancy. Moreover, the surgical construct must guarantee a recovery of immediate functionality, mechanical resistance to disease progression and radiotherapy. Impending fractures could be pre-treated with shorter hospitalization and complications compared to actual fractures.

27.4 Long-Bone Metastasis Surgery

Pathological fractures and pain nonresponding to conservative treatment are the main indications for long-bone metastasis surgical treatment. The algorithm for long-bone metastasis management is reported in Figs. 27.1. Soft tissue sparing is fundamental to improve post-operative recovery and function, reducing the rate of infection. No specific

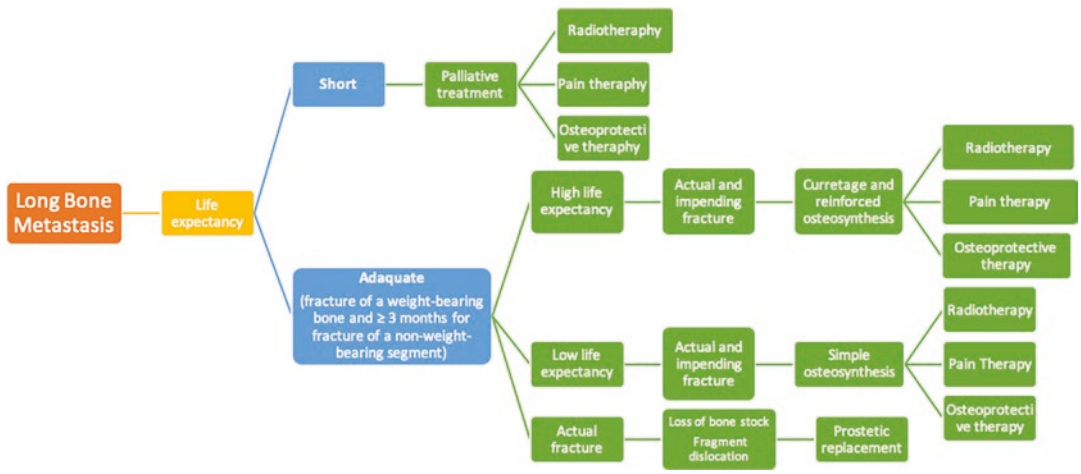

Fig. 27.1 Algorithm for long-bone metastasis management

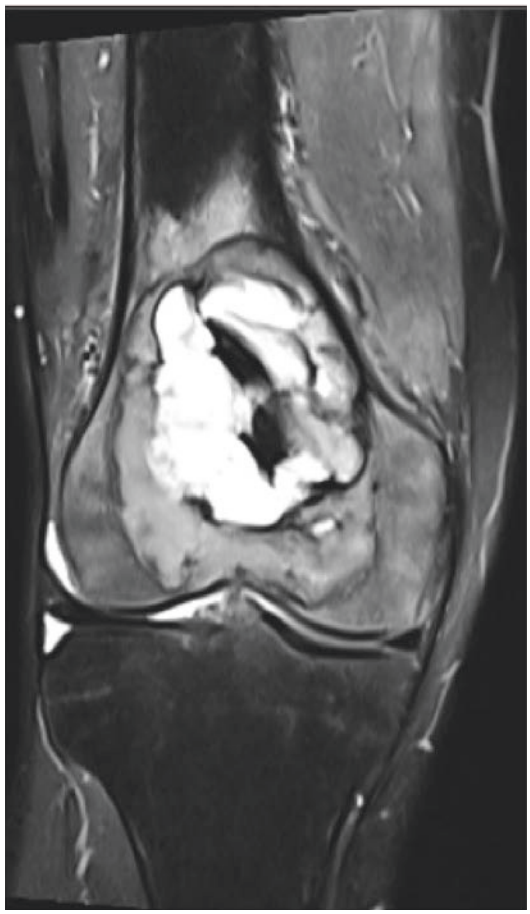

Fig. 27.2 MRI scan shows giant cell tumor of bone of the right distal femur

guidelines reported the precise parameters to perform a prophylactic fixation, but some studies provided clinical and radiographical indications. In particular, a cortical destruction >50%, lesions >25 mm and persistent pain after radiotherapy are indications to surgical fixations. The osteosynthesis aims to guarantee an early full weight-bearing for lower limb fractures and stabilize the fractures of the upper limb to allow daily activities. Titanium was traditionally used as a material for fixation implants, reducing the infection risk in patients eligible for post-operative radio- and chemotherapy. Examples of long-bone metastasis with surgical management and preoperative diagnosis are reported in Figs. 27.2, 27.3, 27.4, 27.5, 27.6, and 27.7.

Plating requires adequate cortical bone proximally and distally to the fracture to ensure the weight-bearing. Otherwise, in pathologic fractures, the bone stock is usually reduced. Therefore, plating could be adapted only for upper limb lesions and other segments that do not require weight-bearing or in sites where intramedullary nailing is difficult to be used.

Instead, intramedullary nailing (cemented or not) is the preferred fixation device for diaphyseal metastasis, due to the ease of insertion, reduced invasiveness, short operatory timing, reduced bleeding, low costs and mechanical properties. The surgeon should use a nail proximally and distally locked with static holes and interlocking screws to manage the torsion and distraction forces. If possible, it is better to use nails with the greater possible diameters, providing early functionality and weight-bearing. Closed surgery with nailing is performed in patients with actual or impending fractures with minimal bone destruction and fragment displacement. Nailing guarantees resistance to torsional forces and angular displacement, providing adequate stability and allowing an immediate post-operative weight-bearing. Otherwise, it is contraindicated in case of sclerotic lesions or if open surgery is required to reduce the fragments. In these cases, plating fixation is more adequate. After intramedullary fixation, early weight-bearing and walking are encouraged.

Fixation with cement augmentation is indicated in patients with good life expectancy or in case of inadequate response to adjuvant therapy. Cementing increases the mechanical stability, and the local control after debulking the lesion, but requires longer operating times, causes local bleeding and increases the wound healing process. The surgical indication for long-bone fracture fixation is reported in Table 27.2.

Pathological fractures with extensive bone loss require open surgery. Liquid nitrogen, alcohol, phenol and argon probes could be used as local adjuvant therapy, increasing the debulking of the tumour deposit and preventing local recurrences.

A prosthetic replacement is required in case of extensive intra-periarticular destructive lesions or in cases of inadequate bone stock at the metaphyseal site.

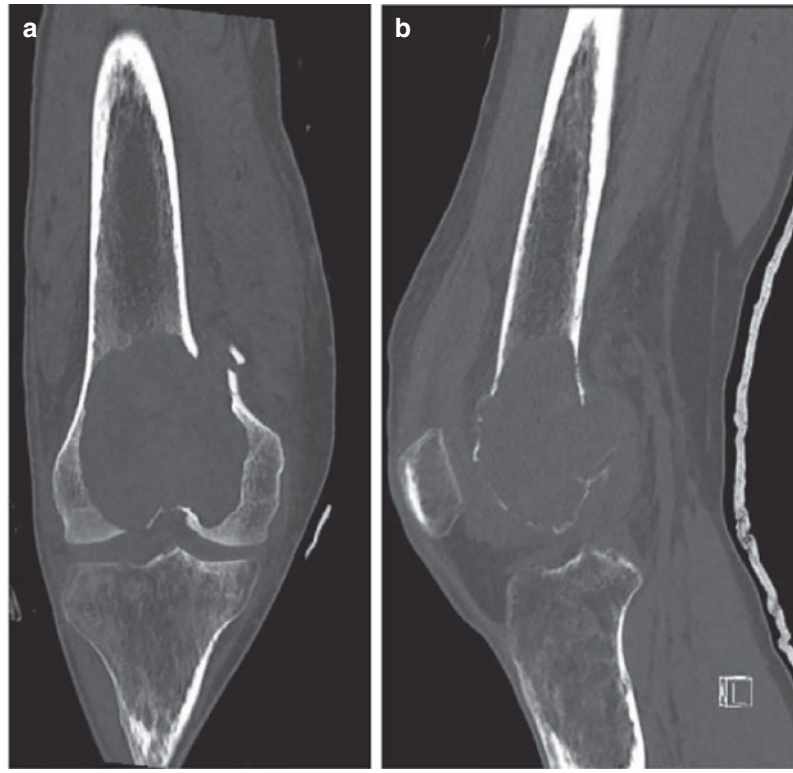

Fig. 27.3 CT scans at coronal (**a**) and sagittal (**b**) planes show pathologic fracture of the right distal femur

Fig. 27.4 Postoperative X-rays show correct placement of cemented custom-made total knee arthroplasty with a tumour prosthesis at the right knee

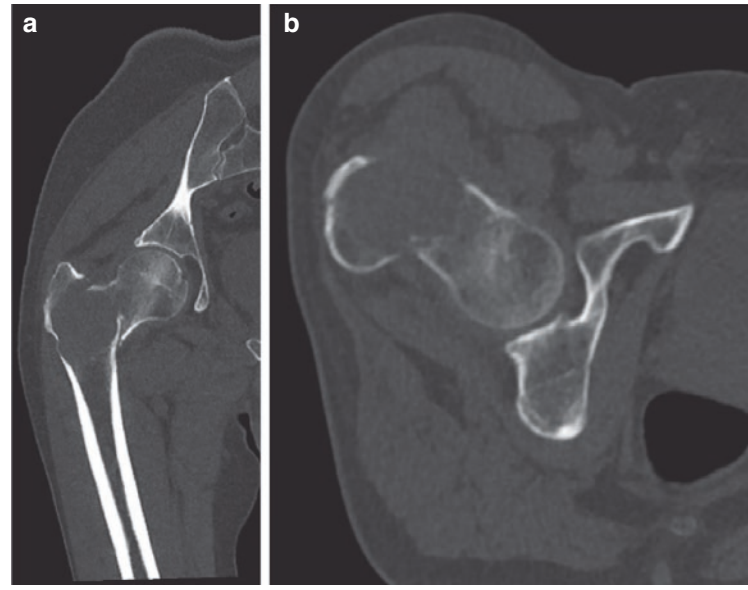

Fig. 27.5 CT scans at coronal (**a**) and axial (**b**) planes show pathologic fracture of the right proximal femur due to bone mesastasis from clear cell renal cell carcinoma

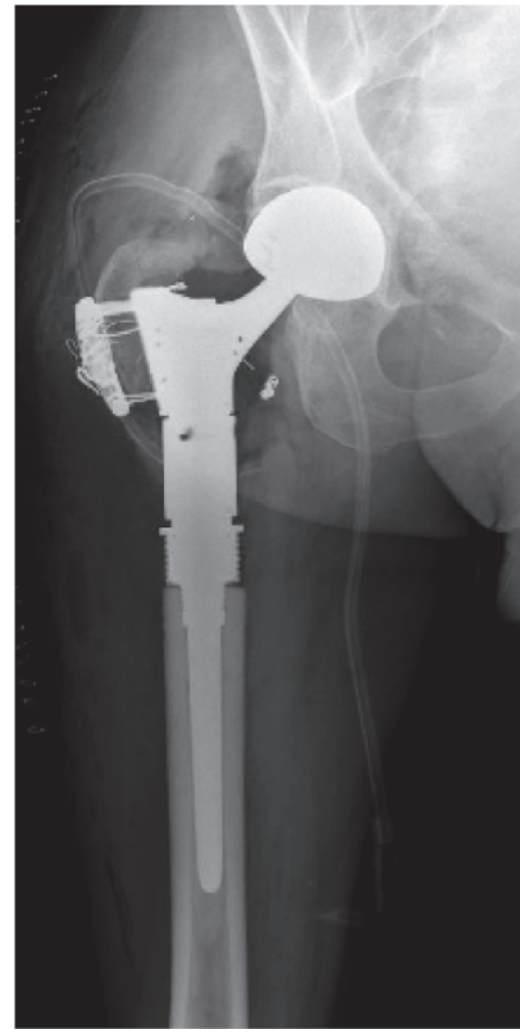

Fig. 27.6 Postoperative X-ray shows correct placement of cemented total hip arthroplasty with a tumour prosthesis at the right hip

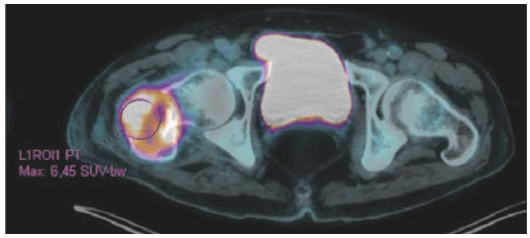

Fig. 27.7 Pet-CT shows FDG uptake at the level of the right proximal femur (SUV 6.45)

Table 27.2 Type of fixations for long-bone fractures

Surgical indication	Locations	Type of fixation
Specific sites	• Proximal humerus • Distal humerus (with <50% of diameter) • Distal femur • Distal tibia	Plating
	• Diaphyseal • Femoral neck • Trochanteric (impending fracture)	Nailing
Type of surgery	• Open • Curettage • Tumour excision • Cementing • Prosthesis • Revisions	Plating
Prognosis	• Poor	Nailing
	• Good	Cemented fixation (plate or nail)
Type of tumour	• Clear cell kidney carcinoma • Thyroid histotype • Chemo-radiotherapy resistance	Cemented fixation (plate or nail)

27.4.1 Complications

Plating and nailing fixations for long-bone fractures have 11% risk of complications. In these are included wound and deep tissue infection, implant breakage, nerve injuries and non-union. Implant long-term survival is determined by early implant breakage, tumour progression, stress surgery and improper fixation.

27.5 Spinal Metastasis Surgery

The most frequent site for solid tumour metastases is the spine. The dorsal spine (followed by lumbar and cervical) is the site involved during the early stage of the disease. The metastasis progressively produces structural destruction of the vertebral body, causing instability and mechanical pain. In the early stage of the disease, the posterior column of the vertebra is involved, while the anterior column is affected during the later stages. The periosteum stretching causes mechanical pain; instead, pathologic vertebral fractures cause pain in the later phases.

Neurological symptoms could be present in case of fracture or compression within the spinal canal. Conservative management, with corticosteroid, radio-chemotherapy and bracing, could be used in the early stages of the disease, in particular in radiosensitive tumours. However, surgical management is required in case of neurological symptoms or nonresponsive therapy.

Surgical management of spinal metastases has two different aims: palliative surgery or locally curative surgery. The former consists of the spinal canal or nerve decompression and stabilization, decreasing the local pain and preventing further damages to the neurological structures. The latter aims to completely remove the tumour and stabilize the segment involved. All the modern stabilization instrumentation devices are derived by the original system developed by Roy-Camille. These systems include laminar hooks and/or pedicular screws with titanium rods.

The proper management includes a combined approach as radiotherapy plus chemotherapy, surgery plus radiotherapy, hormone therapy and medical therapy.

Bracing is essential in patients who underwent radio- and chemotherapy and those who are not suitable for surgery.

In selected patients, with good long-term prognosis and single metastasis, complete excision of the tumoral mass could be per-

Table 27.3 Indications for management of spinal metastases

Intervention	Indication
Bracing	• Perioperative period • Supporting radio- and chemotherapy • Patients not suitable for surgery
Surgery and radiotherapy	• Asymptomatic metastases
Radiotherapy	• Radiosensitive tumour • Unclear segmental instability • Reduced life expectancy • Intact neurological status • Spinal cord compression lasting >48–72 h
Pain management	• Patients not operable • Tumour not sensitive to hormone and radiotherapy
Surgery	• Oligometastatic patient • Primary tumour with good prognosis (breast or prostate) • Nonresponsive pain • Neurological deficits • Myeloradicular compression • No visceral involvement • Good medium-long-term life expectancy

formed. Otherwise, the complete excision could require a complex surgery with high intra- and post-operative risks (vascular, neurological and visceral).

In other cases, the "debulking" of the mass with indirect decompression, followed by stabilization, is indicated. This surgery aims to decompress the spinal canal, reduce pain and improve the quality of life. The management of spinal metastases is summarized in Table 27.3.

Surgery with or without adjuvant radiotherapy is associated with better outcomes and 1-year survival rate compared to radiotherapy alone. Therefore, complete tumour excision is the chosen procedure in selected patients and histotype of tumours (Table 27.3).

Percutaneous approaches could treat nonresponsive pain and vertebral body fractures without neurological involvement. Vertebroplasty and kyphoplasty could be used in these patients, producing immediate pain relief and improving the functional capacity. Moreover, with these procedures, it is possible also to perform a biopsy. Otherwise, percutaneous procedures, particularly vertebroplasty, are related to a high rate of complications as leakage of the cement in the spinal canal or fat embolism.

27.5.1 Complications

Spinal metastasis surgery has a high risk of complication rate in the post-operative period, ranging from 5% to 76%. The perioperative mortality rate is 5.8%, and major and minor general complications (pneumonia, infection, dural tears, neurological deficit) occur in 21% of cases. Age, multilevel surgery, preoperative irradiation and myelopathy represent the main risk factors. Intraoperative neuromonitoring (IONM) represents a valid and sensitive device to detect neurological injury during tumour surgery. Reoperation rate reaches 20% of cases and is usually due to refracture, hematoma evacuation, local recurrence, breakage of the instrumentation and infections.

Take-Home Message

- Surgical treatment of bone metastases aims to reduce pain, improve function, prevent tumor progression, and decrease complications.
- Surgical indication is tailored to the patient based on prognostic factors, including prognosis, histotype, chemoand radio-sensitivity, number and location of lesions, and type of pathological fracture.
- In long-bone metastasis surgery, mechanical stability assessment is crucial, and options include plating or intramedullary nailing depending on the location and extent of the lesion. Cement augmentation may be considered in certain cases.
- Surgical management of spinal metastases aims at palliation or local cure, with

decompression and stabilization procedures. A combined approach involving surgery, radiotherapy, chemotherapy, and medical therapy may be necessary.
- Complications associated with bone metastasis surgery and spinal metastasis surgery are relatively commonand include infection, implant breakage, nerve injuries, non-union, and general complications. Intraoperative neuromonitoring is a valuable tool for detecting neurological injury during spinal tumor surgery.

Summary

- The surgical treatment of bone metastases aims to reduce pain, improve function, prevent tumour progression and decrease complications.
- Nowadays, the improvements in diagnostic technologies and therapy management have increased the life expectancy, granting the detection of metastases in the early stage of the disease.
- Surgery is indicated only for patients with an expected survival of 3–6 months. In other cases, the palliative treatment is preferred, aiming at pain management and complication prevention.
- When surgery is indicated for long-bone metastases, a wide or marginal excision or curettage is associated with osteosynthesis and cement or prosthetic reconstruction.
- The surgical indication is tailored to the patient, depending on the characteristic of the metastatic disease and prognostic factors of the patient.
- The evaluation of mechanical stability is mandatory before considering the surgical option.
- Long-bone metastases' osteosynthesis aims to guarantee an early full weight-bearing and stabilize the upper limbs to allow daily activities.
- Plating could be adapted only for upper limb lesions and other segments that do not require weight-bearing or in sites where intramedullary nailing is difficult to be used.
- Intramedullary nailing (cemented or not) is the preferred fixation device for diaphyseal metastasis, due to the ease of insertion, reduced invasiveness, short operatory timing, reduced bleeding, low costs and mechanical properties.
- The most frequent site for solid tumour metastases is the spine, with 5–10% of patients with systemic cancer who develop spinal metastases during their life.
- The dorsal spine (followed by lumbar and cervical) is the site involved during the early stage of the disease.
- Conservative management, with corticosteroid, radio-chemotherapy and bracing, could be used in the early stages of the disease.
- Surgical management is required in case of neurological symptoms or nonresponsive therapy.
- In selected patients, with good long-term prognosis and single metastasis, complete excision of the tumoral mass could be performed. The "debulking" of the mass with indirect decompression, followed by stabilization, is indicated in the other cases.
- This surgery aims to decompress the spinal canal, reduce pain and improve the quality of life.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Prophylactic fixation of long-bone metastases is required in case of:
   (a) >50% of cortical destruction
   (b) 20–50% of cortical destruction
   (c) >70% of cortical destruction
   (d) In any case of cortical destruction
2. Nailing is performed:
   (a) In all diaphyseal fractures
   (b) In proximal humerus fractures
   (c) In distal femur fractures
   (d) In distal tibia fractures

3. IONM:
   (a) Is a sensitive and valid tool for spinal metastasis surgery
   (b) Is a sensitive and valid tool for long-bone metastasis surgery
   (c) Is useful only in case of cervical fractures
   (d) Is useful only in case of lumbar fractures
4. Leakage of the cement in the spinal canal:
   (a) Is a complication of vertebroplasty and kyphoplasty
   (b) Is a complication of vertebroplasty and not kyphoplasty
   (c) Is a complication of kyphoplasty and not vertebroplasty
   (d) Occurs during open surgery
5. Bracing:
   (a) Is an adequate treatment for patients not suitable for surgery
   (b) Is the treatment of choice in patients eligible for surgery
   (c) Is not useful in any case
   (d) Is useful in case of nonresponding therapy

Further Reading

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Part VII

Fractures

Bone Fractures: Generalities

28

Francisco Forriol and Alessandro Mazzola

Overview

A bone fracture is defined as a medical condition in which a partial or complete interruption of the bone integrity occurs due to mechanical trauma. It happens when the mechanical stress on bone is greater than its deformability. Features of a specific fracture depend on two main factors: the magnitude, duration and direction of the injurious force (extrinsic factor) and the bone resistance (intrinsic factor).

28.1 Introduction

A bone fracture is a medical condition in which the continuity of the bone is broken. A significant percentage of bone fractures occurs because of high force impact or stress. The specific features of a bone fracture depend on extrinsic and intrinsic factors. Extrinsic factors include the type, duration and direction of the traumatic force. Intrinsic factors are the bone quality, its deformability and resistance. However, features of fractures are different in patients of different ages: during childhood, bones are more elastic, becoming more rigid with ageing.

Not all traumas produce a fracture. Running, walking and jumping cause stress forces that deform the bone. However, these forces are not as high to produce a fracture. Due to the properties of collagen, bone absorbs mechanical forces by deforming and then returns to its original state. A mechanical force applied to bone could produce different stresses (tensile, compressive or shear stress) depending on the type, direction and magnitude of the force itself. Biomechanics explains the different types of bone fractures. The bone is weaker when exposed to tensile forces and more resistant to compressive ones. When flexion of a long bone occurs, it fractures on the side under tension and then continues to the other cortex. Bone weakening for any cause increases the risk of skeletal fractures.

28.2 Classification of Fractures

The most part of bone fractures occurs when a high force impact breaks a healthy bone. However, two types of fractures have a different pathogenesis: pathological and stress fractures. Pathological fractures are seen in weakened bones for other conditions. Tumours, osteoporosis and other metabolic diseases may reduce bone

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strength, causing fractures also with minimal trauma. Stress fractures are tiny crack in weight-bearing bones of the lower limbs. They are caused by repetitive forces that overstress bones chronically and are frequent in military, athletes and walkers. Depending on the causative mechanism, fractures can be divided into direct or indirect. Direct fractures occur at the specific point the force acts. In this group, we include crushing (associated with soft tissue damage), bump (the damage is centred in a specific area) and penetrating (also called gunshot) fractures. In indirect traumas, the fracture occurs far from the force application point. These injures can be classified on the basis of the mechanical distribution of the forces: flexion, torsion, compression, traction or combined fractures.

Another classification criterium is based on the integrity of the skin: in closed fractures, there is no injury of soft tissue, and the damage site is not in communication with the external environment. In an open fracture, instead, the site of injury communicates with the external environment. Open fractures are classified according to the Gustilo and Anderson classification: it considers the amount of energy, the extent of soft tissue injury and the extent of contamination for determination of fracture severity.

The radiographic pattern may show complete or incomplete fractures. When the fracture line does not cover the entire diameter of the bone, it is an incomplete fracture. Greenstick (or torus) fractures, typical of children, are flexion fractures that do not break the two cortical bones. In this case, because of the thick periosteum, the bone bends and cracks, instead of breaking completely into separate pieces.

In complete fractures, both cortical bones are injured: they can also be divided into simple and displaced fractures. A simple fracture is a single line that divides the bone into two fragments, which remain in their anatomical position. In displaced fractures, the bone loses its alignment. The displacement may produce angulated fractures (the two ends of the broken bone are at an angle to each other), translated fractures (the ends of the bone have shifted out of alignment) and rotated fractures (the bone rotated during the rupture). Multifragmentary fractures have more than one fracture line and produce multiple fragments. They include wedge and complex fractures. A wedge fracture is a complex pattern with a third fragment in which, after reduction, there is a direct contact between the two main fracture fragments. In a complex fracture, one or more fragments are not in contact after reduction. Depressed fracture is an articular fracture in which there is only sinking of the joint surface without a split.

On the basis of the stability of the fragments, it is possible to distinguish stable and unstable patterns. In stable fractures, fragments do not move once reduced. In unstable patterns, fragments tend to displace after reduction. In general, all complex or oblique fractures greater than 45° are unstable, except for spiral fractures.

Fractures are articular when they involve the joint surface and extra-articular when the joint is preserved. It frequently happens in epiphyseal or metaphyseal-epiphyseal fractures of a long bone. In rare cases, fractures could be associated with joint dislocation (fracture-dislocation).

28.3 Fracture Classification Systems

Fracture classifications are usually anatomical and focused on the single fractured site. Many of these classifications are routinely adopted and are based on the location and number fragments of fracture. Some examples are the classification by Neer (proximal humerus), Schatzker (proximal metaphyseal fractures of the tibia), Pauwels or Garden (femoral neck fractures), Tyle (pelvis) or Denis (spine).

Many authors have attempted to define a general and univocal classification system in order to prevent errors. The two most widely used classification systems are the AO/ASIF for long bones and the Orthopaedic Trauma Association (OTA). The OTA classification, revised every 3 years, is based on the AO system, introducing the short bones, the pelvis and the spine.

In the AO classification, each bone (segment) fracture is divided into three types (A, B, C).

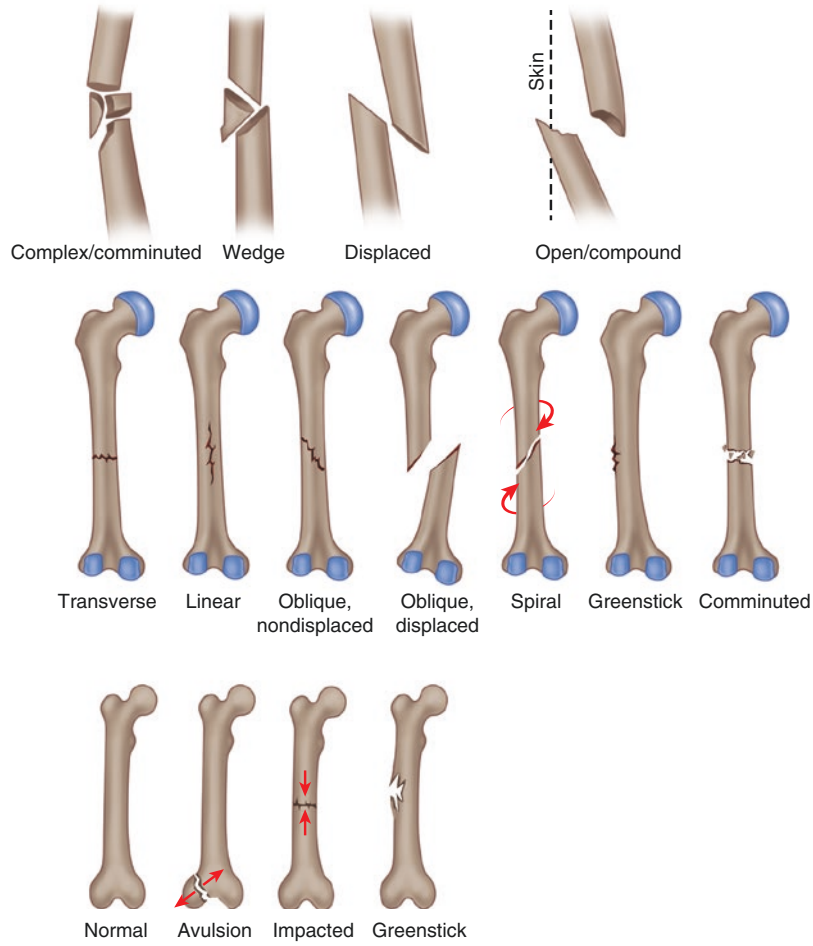

Fig. 28.1 Type of fractures

Furthermore, the three types are divided into three groups (1–3) and each of them into three subgroups.

In several cases, the definitive classification could be performed only during surgery, with the direct visualization of fragments. Each segment has a number that identifies it: 1 = humerus, 2 = ulna and radius, 3 = femur and 4 = tibia and fibula.

Each long bone has three portions: 1 = proximal, 2 = diaphyseal and 3 = distal. Diaphyseal fractures are divided into three groups: A = simple fracture, B = wedge fracture and C = complex fracture. A simple fracture (A) is divided into three types, A1 = spiral, A2 = oblique and A3 = transverse. Moreover, in joint involvement, fractures could be divided into A = extra-articular, B = partially articular and C = completely articular. The classification has some exceptions and special cases (Fig. 28.1).

28.4 Clinical Features of Fractures

Imaging is mandatory in order to properly classify and describe a fracture. However, some clinical signs and symptoms may suggest the presence of fractures. Clinicians should evaluate pain, disability, deformity, ecchymosis and bone crepitation of the injured site. The most frequent and constant symptom is pain, exacerbated by palpation and mobilization. Pain is usually accompanied by functional disability. Ecchymosis is frequently found hours or days after the trauma.

28.5 Bone Reparation

The repair process can be divided into three sequential phases: inflammation, consolidation and remodelling (Fig. 28.2).

28.5.1 Inflammation

The natural process of healing a fracture starts when the trauma occurs: the rupture of vessels causes extravasation of blood and hematoma formation between the broken fragments. Proinflammatory cytokines reach the fracture site through the blood and stimulate the repair process. Bone fragments release osteogenic molecular signals that contribute, together with the healing hematoma, to the formation of the granulation tissue. It is composed of proliferating fibroblasts, new thin-walled capillaries and infiltrated inflammatory cells in a loose extracellular matrix. The granulation tissue is then reabsorbed by macrophages. This constitutes the first step in bone fracture repair.

28.5.2 Consolidation

In this phase, new blood vessels form in the matrix of the blood clot. It brings phagocytes and fibroblasts to the affected area. Fragments of fracture become necrotic and are gradually reabsorbed by phagocytes; fibroblasts multiply and produce collagen fibres. In this way, the blood clot is replaced by a matrix of collagen. The pluripotential mesenchymal stem cells (MSCs) start the process of new tissue formation. In this phase, the temporary callus is formed by fibrous tissue, cartilage and immature bone, and it is called fibrocartilage callus. The callus progressively mineralizes, becoming stiffer and tougher and transforming into a bone callus. The function of the bone callus is to fix the fracture site, reducing the mobility between fragments. Bone proliferation begins far from the fracture site and slowly progresses toward fragments, producing a bone bridge.

28.5.3 Remodelling

In this phase, the repairing tissue matures and forms new bone tissue. The entire process could last from a few months to several years. The formation of new bone depends on vascularization and mechanical conditions of the affected area. Bone can be produced by endochondral ossification (from cartilage) or intramembranous ossification (from fibrous tissue).

A minimum range of movement is necessary to guarantee a proper repair process. In case of

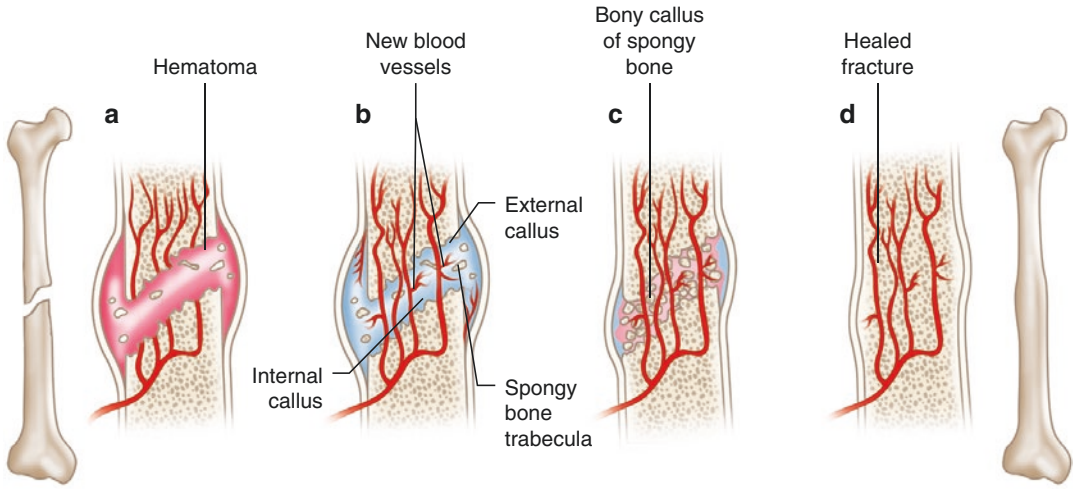

Fig. 28.2 Bone repair process (**a**, **b**) hematoma phase (**c**) fibrous tissue callus (**d**) bone remodelation

rigid fixation, the granulation tissue and the external callus are often insufficient as the high rigidity of the implant makes growth factors useless. The opposite occurs with elastic fixations.

Primary ossification occurs when there is direct contact between fragments and a rigid fixation that holds them together. Secondary ossification (more frequent) occurs when there is no direct contact between fragments and fixation is elastic. An exuberant callus is a biological response to reduce mobility between fragments. In case of excessive mobility between fragments, the bone callus cannot be formed and the fracture does not consolidate. It results in consolidation delay or non-union.

At this stage, fibroblasts of the collagen matrix begin to produce collagen monomers: they spontaneously assemble forming the bone matrix. In this way, the mineralization of the collagen matrix results in new bone formation.

28.6 Fracture Complications

Fracture complications may be associated with the trauma itself, with the patient's comorbidities (diabetes, corticosteroid therapy, smoking, etc.) or with an inadequate treatment. Complications can be classified into two groups: bone healing disorders and associated complications (localized or systemic).

28.7 Bone Healing Disorders

Bone healing is completed when the bone callus is sufficiently developed and mineralized so that it provides support and stability to the injured area. After 6 months of injury, in case of absence of bone union, it is possible to define a "consolidation delay" for well-reduced and fixed fractures. Radiographic findings continue to show the fracture line; it may be painful and present fracture instability. If there is no consolidation and a false joint is observed in the site of fracture after 9 months of injury (at this stage, the fracture will not heal anymore without other treatments), it is possible to define a non-union (or pseudoarthrosis) pattern. Non-union is characterized by the formation of permanent fibro-cartilaginous tissue between fracture fragments. Potential risk factors for bone healing disorders are distance between fragments, mobility between fragments, fracture-site infection and lesions of the periosteum.

There are several classifications of non-union. The most common is by Weber and Cech, distinguishing the radiographic patterns into two types: hypertrophic (or viable, capable of biological reaction with enlarged fragment extremities) and atrophic (or avascular, incapable of biological reaction with reabsorbed fragment extremities). Another classification divides the non-union into atrophic, hypertrophic and normotrophic (intermediate pattern). Open (or surgically) treated fractures may potentially become infected. In general, the infection itself prevents consolidation. When it occurs, the condition is called infected pseudoarthrosis.

Treatment of non-union is surgical and consists of the removal of the fibro-cartilaginous tissue, revascularization of the fracture site and creation of a new stable fixation of the bone fragments.

Vicious consolidation occurs when fracture fragments heal in an incorrect position, leading to angular or rotational deformities, shortening or lengthening of the injured bone, pain and functional limitation. Treatment is based on corrective osteotomies in which an iatrogenic fracture is created in order to fix the new fracture fragments in a correct position.

28.8 Associated Complications

Associated complications may affect the fracture site (localized) or the entire body (systemic). We distinguish early and late complications of fracture.

28.8.1 Early Complications

It includes complications occurring at the moment of trauma or in the days immediately following:

- Local acute complications: They include a wide range of cutaneous, capsular ligamentous, vascular or nervous lesions. A typical example is the radial nerve lesion in case of humeral diaphyseal fractures.
- Compartment syndrome: It is a serious condition caused by bleeding or swelling within an enclosed bundle of muscles (known as muscle compartment). The increasing pressure in the compartment determines poor blood flow through arteries and veins of the affected area. If not treated in a few hours, due to reduced perfusion, permanent muscle or nerve damage can result. Risk factors for this condition are related to the trauma itself (internal haemorrhage, local oedema, tissue inflammation) or to treatment (constricting bandages). Compartment syndrome is a medical emergency. It should be suspected in patients with a recent trauma presenting exaggerated pain, swelling and hypoaesthesia in the affected area. Apart from the typical signs and symptoms, measurement of intracompartmental pressure can also be important for the diagnosis. Treatment consists of the removal of any cause of external compression or performing a local fasciotomy. The Volkmann syndrome is a compartment syndrome that affects anterior nerves and muscles of the forearm.
- Thromboembolic disease: Venous thromboembolic disorders are a leading cause of morbidity and mortality in orthopaedic patients. Immobilization, inflammation and hypercoagulability are risk factors generally present in patients with bone fractures. It justifies the elevated incidence of these disorders. Deep vein thrombosis (DVT) is the formation of a blood clot in a deep vein, most commonly of the legs. Symptoms include pain, swelling, redness and enlarged veins in the affected area, but it may also be clinically silent. If the clot of a DVT dislodges and travels to the lungs, to the pulmonary arteries, it is called a pulmonary embolism (PE), which in some cases may be fatal. Symptoms of PE may include shortness of breath, chest pain and haemoptysis. In order to confirm the diagnosis of DVT, ultrasound imaging is generally needed. PE may be suspected in case of elevated D-dimer in the bloodstream and altered arterial blood gas (ABG) test (low levels of arterial pO2). The electrocardiogram (ECG) may also show some alterations, but only with a ventilation/perfusion lung scintigraphy or a computed tomography (CT) angiography may the diagnosis be confirmed.
- Fat embolism syndrome: It is a potential complication of long-bone or pelvic fracture. This condition occurs when a piece of fat, deriving from the yellow bone marrow of the fracture site, enters the bloodstream and causes a blockage of blood flow in vessels of the lungs, brain, kidneys or skin. Symptoms may include petechial rash, shortness of breath, fever, acute kidney injury and altered level of consciousness.

28.8.2 Late Complications

It includes complications occurring after many weeks, months or years after the trauma:

- Post-traumatic arthritis: It is a secondary osteoarthritis frequent in patients who experienced a previous articular fracture. Over time, despite surgical reduction, articular discrepancies may cause an early form of arthrosis. Symptoms include swelling, synovial effusion and pain.
- Avascular necrosis: This condition results from the temporary or permanent loss of blood supply to the bone. When it occurs, the bone tissue dies and the bone collapses. It should be

investigated in patients with long-term pain and disability in the site of a previous trauma. If not treated, avascular necrosis evolves into secondary osteoarthrosis. The most common sites affected are the femoral head, talus, scaphoid and humeral head.

- Heterotopic ossification: It is the presence of bone in soft tissue, where bone normally does not exist. It is frequently seen next to a previous site of trauma.
- Algodystrophy: Also known as complex regional pain syndrome, it is a painful disease characterized by oedema, functional impairment, pain, redness, and sensory and vasomotor disturbances. It affects all of the tissues of a specific site: it justifies the variability of the clinical manifestations. The term Sudeck's disease generally refers to a form of algodystrophy of the lower extremities. Conservative and symptomatic treatment is the first choice.
- Osteomyelitis: It is an infection of the bone marrow that secondarily affects the bone. The most common form of the disease in adults is caused by injury exposing the bone to local microorganisms. In most cases, patients present fever, swelling, tenderness, redness and warmth in the area of the infection. The diagnosis is based on X-rays, blood tests, MRI and bone scans. A bone biopsy is generally needed to determine the germ. Most people with osteomyelitis are treated with long-term antibiotics, surgery or both.

28.9 Child Bone Fractures

Fractures occurring during growth have a unique pattern because bones are still growing. In children (persons younger than 18 years of age), the bone is more elastic, as it contains a greater amount of collagen fibres. Moreover, the periosteum is thicker, with a greater osteogenic potential. This results in a faster rate of consolidation and a higher rate of bone remodelling. Thus, even fractures not perfectly aligned or reduced can heal without sequelae in children. Another peculiarity of the growing bone is the presence of growth plates, which may be included in the fracture line.

Three types of fractures occur only in growing bones:

- *Greenstick fractures*: As mentioned before, they are flexion fractures that do not break the two cortical bones. In this case, because of the thick periosteum, the bone bends and cracks, instead of breaking completely into separate pieces.
- In some fractures of the metaphyseal-diaphyseal zone, the epiphysis is introduced into the metaphysis.
- *Growth plate lesions or physeal fractures*. They are total or partial traumatic detachment or compression of the growth plate (or physis). Salter and Harris classified these lesions into five types (Fig. 28.3):
  - Type 1: separation through the physis
  - Type 2: fracture enters in the plane of the physis and exits through the metaphysis
  - Type 3: fracture enters in the plane of the physis and exits through the epiphysis
  - Type 4: fracture crosses the physis, extending from the metaphysis to the epiphysis
  - Type 5: a crush injury resulting in a partial or total bone bridge

Grades 4 and 5 have a bad prognosis. To prevent the sequelae of a physeal fracture, it is recommended to make an early anatomical reduction. A physeal fracture can produce a total or partial closure of the growth plate, leading to shortening or deviation of the limb.

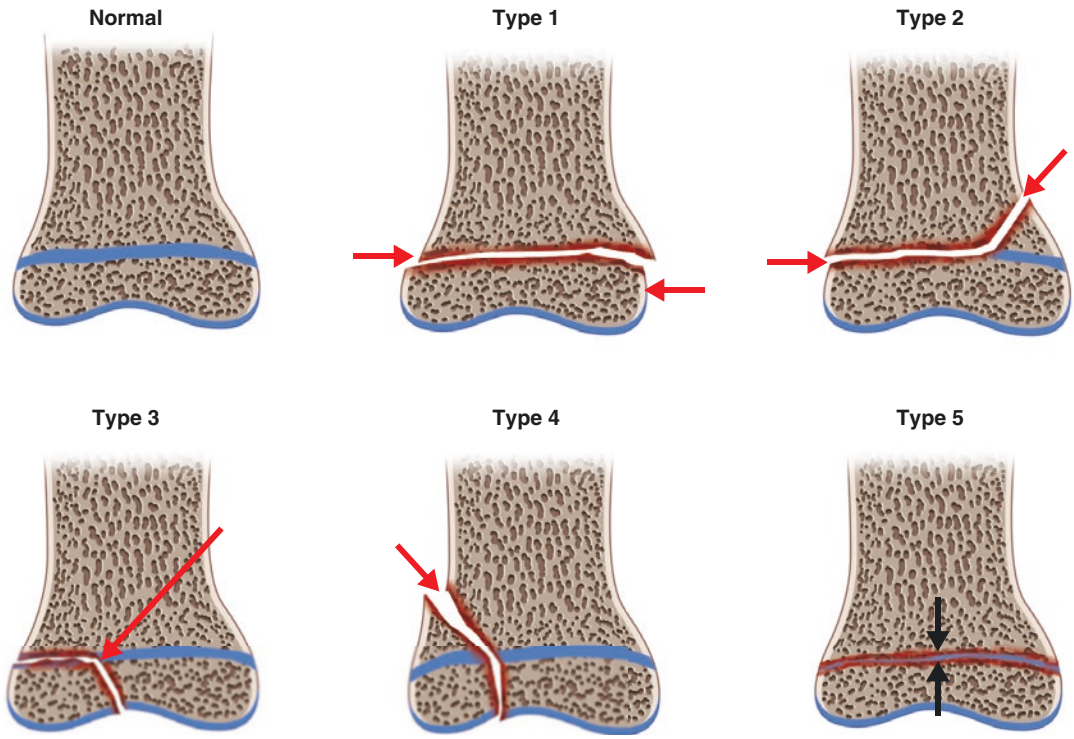

Fig. 28.3 Salter-Harris classification

Take-Home Message

- A bone fracture is the interruption of bone and cartilage continuity due to mechanical trauma.
- Fractures during growth have unique characteristics since the bone tissue is more elastic and the growth plate is open.
- The standard classifications of fractures could be anatomical and depend on each bone. However, comprehensive systems were proposed for a systematic classification of fractures with a common terminology.
- A rigid fixation osteosynthesis produces little granulation tissue, and the external callus is inadequate or non-existent since a high rigidity renders the presence of growth factors useless.
- There are several fracture complications, such as bone healing disorders and associated complications.

Summary

A bone fracture is defined as a medical condition in which a partial or complete interruption of the bone integrity occurs due to mechanical trauma. The specific features of a bone fracture depend on extrinsic and intrinsic factors. Extrinsic factors include the type, duration and direction of the traumatic force. Intrinsic factors are the bone quality, its deformability and resistance. However, features of fractures are different in patients of different ages: during childhood, bones are more elastic, becoming more rigid with ageing. The two most widely used fracture classification systems are the AO/ASIF for long bones and the Orthopaedic Trauma Association (OTA). The OTA classification, revised every 3 years, is based on the AO system, introducing the short bones, the pelvis and the spine. Bone healing is a dynamic process that can be divided into three sequential phases: inflammation, consolidation and remodelling. The natural process of healing a fracture starts when the trauma occurs and ends with the

formation of new bone in the site of fracture. Patients with fractures may develop specific complications. They can be classified into two groups: bone healing disorders and associated complications (localized or systemic).

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. The characteristics of a bone fracture depend on:
   (a) External and internal factors
   (b) Mechanism of injury
   (c) Patient's characteristic
   (d) Characteristics of the injured site
2. Pauwels' classification is used for:
   (a) Femoral neck fracture
   (b) Femoral diaphysis
   (c) Proximal humerus
   (d) Distal radius
3. Denis' classification is used for:
   (a) Spine
   (b) Femur
   (c) Lower limbs
   (d) Shoulder
4. Repair fracture process consists of:
   (a) Inflammation, repair phase, remodelling phase and tissue formation
   (b) Remodelling phase, inflammation, repair phase and tissue formation
   (c) Remodelling phase, tissue formation and inflammation
   (d) Inflammation, remodelling phase and tissue formation
5. Pseudoarthrosis could be divided into:
   (a) Hypertrophic, atrophic and normotrophic
   (b) Hypertrophic, viable and congenital
   (c) Congenital and acquired
   (d) Stable and unstable

Further Reading

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Fracture Treatment: Basic Concepts

29

Francisco Forriol and Alessandro Mazzola

Overview

Fracture healing involves a complex and sequential series of events to restore injured bone to pre-fracture conditions. The bone healing process is influenced by many factors, including mechanical stress, biochemical mediators, and neural and endocrine influences. In brief, any factor capable of altering the metabolism of osteogenic cells may influence bone healing.

29.1 Introduction

Prompt patient stabilization and immobilization of the fracture site decrease the risk of further injury and increase the chances of a successful fracture repair. Bone fracture treatment can be either conservative or surgical. The latter could also be divided into closed (percutaneous) or open surgery. Fixation devices are necessary to stabilize the fracture site. They could be classified into rigid or elastic, also called dynamic (Fig. 29.1).

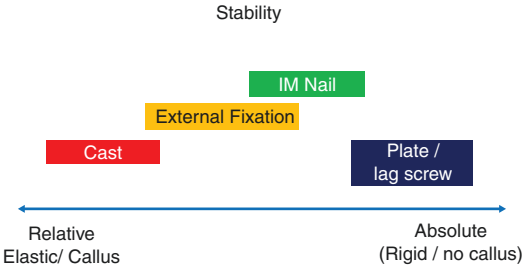

Fig. 29.1 Stability of the systems for fracture treatment

A rigid fixation aims to achieve an anatomical reduction of fragments without movement between bones. In contrast, elastic or dynamic systems stabilize the fracture but do not entirely restrict mobility, allowing controlled micromovements between fragments. How much motion is optimal and the best time for definitive fixation are still unclear. Rigid fixation minimizes the formation of the granulation tissue. Bone is formed directly between fragments, without an external callus: it realizes a primary consolidation. On the contrary, elastic systems induce the formation of an exuberant callus in order to restrict movements. The mobility allowed by these devices needs to be minimal, and rotational movements or shear stresses must be avoided. Excessive mobility of fragments leads to delayed healing or nonunion.

Vascularization should also be considered: periosteal and endosteal vascularization also contribute to fracture healing. Conservative treatment

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allows the formation of a hematoma: the rupture of vessels causes extravasation of blood between the broken fragments. Proinflammatory cytokines reach the fracture site through the blood and stimulate the repair process. In contrast, open fixation requires deperiostization and muscle detachment with consequent devascularization of the cortical bone. In case of intramedullary nailing, only endosteal vascularization is injured, whereas muscles and periosteal vascularization are respected. Fracture consolidation should respect five important conditions: mechanical stability, adequate vascularization, osteoinduction, osteogenesis, and osteoconduction.

29.2 Biomechanical Aspects of Fracture Treatment

In order to achieve a stable and valid fixation, some rules need to be respected. Both biological and mechanical factors must be considered: the energy of trauma, initial displacement, integrity of soft tissues, and comorbidities of the patient influence the healing process. Moreover, the fracture focus tends to displace because of muscle contractions. The ideal conditions for treating fractures are stable fixation, good vascularization, and absence of soft tissue alterations or infections.

29.3 Principles of Immobilization

Rigid fixation uses plates and interfragmentary lag screws to stabilize fracture fragments. Dynamic or elastic devices allow the transmission of forces through the focus of fracture (cast, intramedullary nail, or external fixator), permitting micro-movements.

Depending on the severity of fracture, elective or emergency treatment may be required. Treatment may also be temporary or definitive.

Each implant needs to protect the fracture focus, preventing mechanical stress on the injured site. This concept is termed “stress shielding” (or stress protection) and occurs when metal implants are used to repair fractures. These implants help to maintain contact between fragments. The stiffness of the implant results in bone loss as a consequence of decreased physiologic loading of the bone. The stress shielding needs to be reached during the early stages of the healing process. Subsequently, when the callus appears, mechanical loads provide a stimulus for bone formation. If excessive rigid fixation with plating occurs, mechanical stress cannot reach the fracture site and bone formation is hindered. In case of cast, splint, nail, or external fixation, a small proportion of forces can pass through the fracture site. In particular, pins of external fixation and intramedullary nails could deform with the load, but their elasticity protects them from breakage (elastic deformation).

29.4 Conservative Treatment

Conservative treatment includes bandages, casts, and splints. The choice between conservative and surgical treatment depends on the type of fracture, associated injuries, age, comorbidities of the patient, and his/her social circumstances. Conservative treatment with a plaster bandage aims at the formation of an exuberant periosteal and endosteal callus. Plasters are made from circular bandages of different materials, fiberglass or polyurethane compounds. The most frequently used is the cast of Paris, which hardens after it has been made wet. When a cast is placed after a fracture, it should be opened longitudinally and subsequently wrapped with a bandage to prevent excessive compression or compartmental syndrome. At the first signs of compression, the cast must promptly be opened. According to this, in the early stages, a temporary or permanent immobilization device formed by a cast surrounded by a bandage (splint) could be placed. In some cases (particularly for tibial fractures), the length of the cast should be reduced to improve the patient’s function. When the callus becomes radiographically visible, a functional splint is preferred. This technique guarantees movements to the joints next to the bone fracture (in case of tibial fractures, to the knee and ankle joints). Moreover, the load transmission through the fracture focus stimulates the bone formation process.

29.5 Surgical Treatment

29.5.1 Plates

Compression plates provide a stable primary osteosynthesis. They are placed once the fracture has been anatomically reduced, opening the fracture focus and separating and disinserting the muscles and the periosteum next to the fracture site. Conventional plating inhibits the formation of the periosteal callus, while it does not influence the endosteal callus. There are three different interfaces in a plate construct:

- Screw-bone interface
- Screw-plate interface
- Bone-plate interface

The number of screws depends on the fracture type and the size of the bone. A short plate or few screws may be mechanically insufficient. In contrast, a plate with many screws may lead to osteopenia. Moreover, when the plate is removed, holes in the bone increase the risk of refracture.

To avoid large incisions and massive periosteum removal, low-profile plates have been designed with locked polyaxial screws. These plates are inserted through a small incision, and screws are placed with a percutaneous technique.

29.5.1.1 Screw-Plate

The screw-plate-type dynamic hip screw (DHS) combines a screw of 13 mm in diameter with a 135–150° angled plate. This plate can be used for proximal femoral fractures.

29.5.2 Screws

Screws are generally adopted to stabilize fragments in combination with other fixation systems (nails or plates). Many screws make the system stable but, at the same time, more rigid (Fig. 29.2). A possible solution to the problem is to place one or more screws far from the fracture site or to use a lag screw (Fig. 29.3). The latter improves interfragmentary compression, but if used alone, it may not be sufficient.

29.5.3 Intramedullary Nails

The intramedullary nail fixes the fracture by occupying the medullary canal. It is inserted from an entry point far from the focus of fracture. It preserves the periosteum and muscular insertions. The external blood supply is maintained, but intramedullary vessels are injured. It results in an exuberant periosteal callus formation, with consequent inhibition of the endosteal callus.

Gerhard Küntscher was a German surgeon who inaugurated the intramedullary nailing of long-bone fractures: the Küntscher nail was the first type of elastic nail designed. It has a cloverleaf section with a longitudinal groove for a simplified insertion technique. Furthermore, the nail

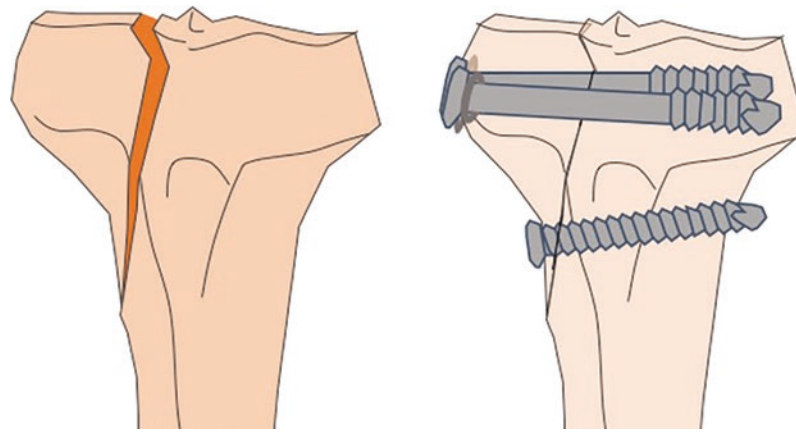

Fig. 29.2 An interfragmentary compressive screw should be placed perpendicular to the fracture line

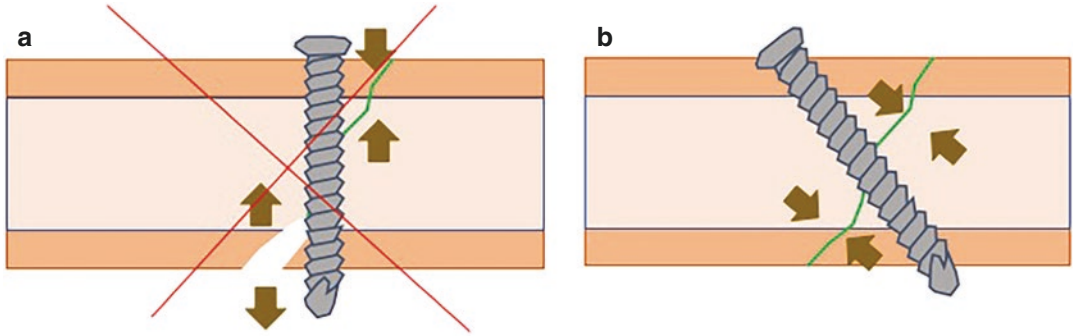

Fig. 29.3 To achieve adequate compression and good fracture repair, the screw must perpendicularly cross the fracture line, (**a**) incorrect, (**b**) correct

flexibility allows the transmission of forces through the focus of fracture, permitting micro-movements.

The anatomy of the medullary canal is not homogeneous; it alternates between wide and narrow areas. Intramedullary nails can be reamed or not. The intramedullary reaming results in an easier passage of the nail through the canal. However, the reaming injures the endosteal vascularization and forms detritus with high osteogenic capacity (increasing the risk of heterotopic calcification). Un-reamed nails are typically smaller in diameter and, consequently, they have less control of the fracture stability.

The interlocking nail provides good relative stability to the fractured bone: it is a Steinmann nail with holes, through which screws can be inserted and fixed to bone cortex. The interlocking nail, thanks to screw fixation, prevents implant migration. However, interlocking nails with proximal and distal transfixing screws are rigid constructs. Therefore, the passage of forces through the fracture line is not permitted. Removing proximal or distal screws from an interlocking nail changes the system from static to dynamic, permitting the transmission of stresses through the fracture focus. Static locking maintains bone length and prevents torsional stresses and fracture collapse. Therefore, its use is recommended in complex fractures.

Intramedullary nails could be inserted from proximal to distal (antegrade) or distal to proximal (retrograde). Intramedullary nails are indicated for diaphyseal fractures of the long bones (humerus, femur, tibia). The interlocking nail is helpful in proximal and distal metaphyseal fractures of the long bones and in complex or multifragmentary diaphyseal fractures.

29.5.3.1 Elastic Intramedullary Nailing

Elastic nails are smaller and more flexible than solid nails. Elastic nailing is recommended in children's fractures with open growth plate, using Kirschner wires or 2–4 mm diameter titanium nails (TEN) (Fig. 29.4). They are introduced far from the fracture focus, protecting the growth plate. Nails have a preset curved configuration, and they always have at least three contact points with the bone. They have many advantages: easy introduction, low infection rate, protection of the growth plate, and rapid consolidation.

29.5.3.2 Gamma Nail

The gamma nail is an intramedullary nailing system for treating proximal femur fractures and, in some cases, femoral shaft fractures. Gamma nails can have different lengths and diameters. The nail is crossed by a lag screw (angled at 45°) that passes through the neck of the femur from the outer cortex to reach the femoral head. It is a simple and stable system. It allows a fast discharge with an early weight-bearing. No reject of the implant is reported in the long-term follow-up.

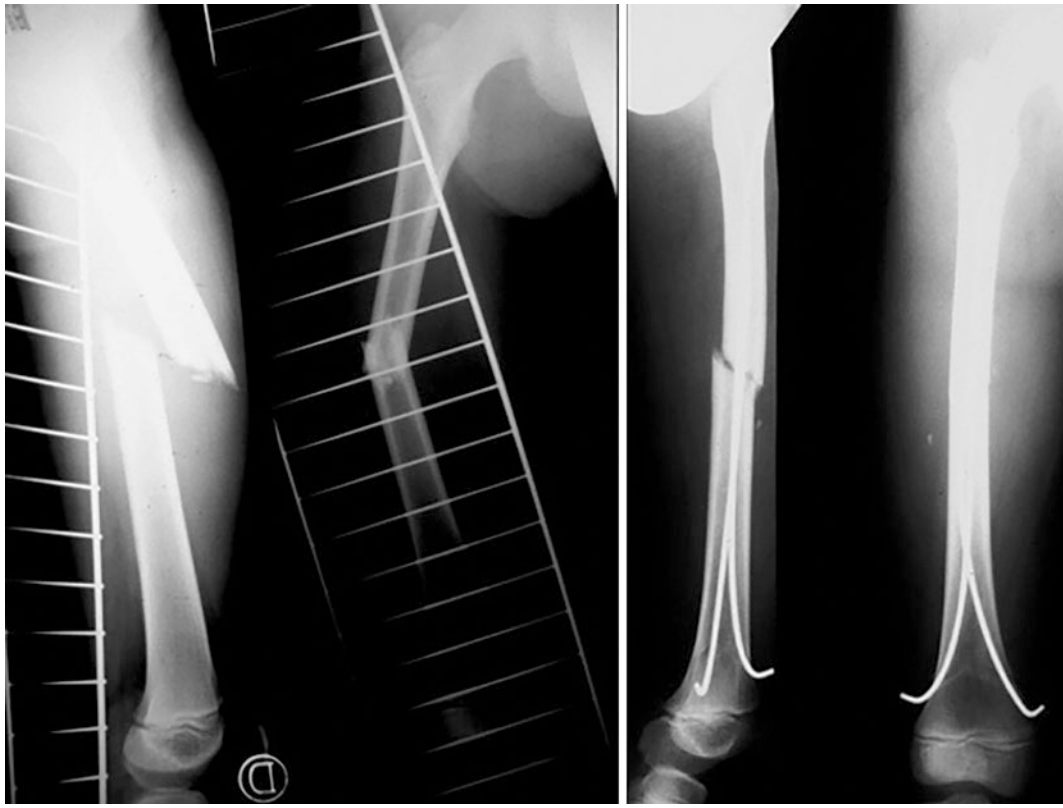

Fig. 29.4 Elastic nail in children's femur fracture

29.6 External Fixation

External fixation is a surgical treatment by which pins or wires are inserted into bone percutaneously and held together via an external scaffold. It respects the biology of the healing process, as it does not interfere with the focus of fracture, the soft tissues, and the periosteal and endosteal blood supply. It was initially described by Malgaigne in 1853. Nowadays, this method is predominantly used for fracture fixation in pediatric or adult patients who have open fractures with severe soft tissue and/or wound contamination. It is generally adopted as a temporary treatment, in order to provide provisional alignment and stability to the affected area, or as a definitive treatment in selected pelvic fractures, compound long-bone fractures, and periarticular fractures. There are different types of external fixators: some can be applied only after a proper reduction, whereas other types are used to manipulate and reduce fracture fragments (osteotaxis). Depending on the geometry of the fixator, pins can be transfixing (they completely pass through the limb) or not. An external fixator must respect the anatomy of the affected region: pins or wires are placed in safe areas without risking muscular or neurovascular injures. Custom-made configurations may be necessary in the treatment of complicated cases, using different frames, pins, and wires. External fixators are also divided into permanent (non-modifiable) and variable (allow mechanical changes, modulating the elasticity of the fixator) systems. According to this, fixators adapt to the type of fracture as they can be

arranged in neutralization, compression, or distraction. Neutralization is used to maintain bone length in comminuted fractures. A slight compression may be helpful in transverse fractures. In case of severe bone loss, a distraction is recommended.

In case of open fractures or other musculoskeletal emergencies, an external fixator can be used as a bridge treatment for the first days after trauma. After the patient is stabilized and soft tissue exposure is reduced, definitive osteosynthesis is generally performed.

More specifically, an external fixator is indicated in case of open fractures with soft tissue damage, infected pseudoarthrosis, consolidation delays, arthrodesis, unstable pelvic fractures, bone elongation, and fracture treatment when internal fixation is dangerous or sterile conditions are not guaranteed. The external fixation allows complementary treatments such as wound cleaning, dressing changes, skin or bone grafting, and irrigation, without altering the fracture reduction. Moreover, early mobilization of the patient is possible and, in some cases (pelvic fractures or multiple trauma injuries), it does not interfere with hygiene care.

External fixation may also be indicated in fractures of the upper and lower extremities in children. The surgical treatment is minimally invasive, and it can be carried out, in cases of extreme need, only with local anesthesia. Moreover, at the end of the treatment, implanted external devices and pins may also be removed without an operating room. Loosening and infection are the two most important complications. The infection rate with external fixation is very low; however, superficial infection of the pins or wires is common and can be controlled with daily cleaning. Chronic osteomyelitis and nerve and vessel injury are possible. Differences between internal and external fixation are reported in Fig. 29.5.

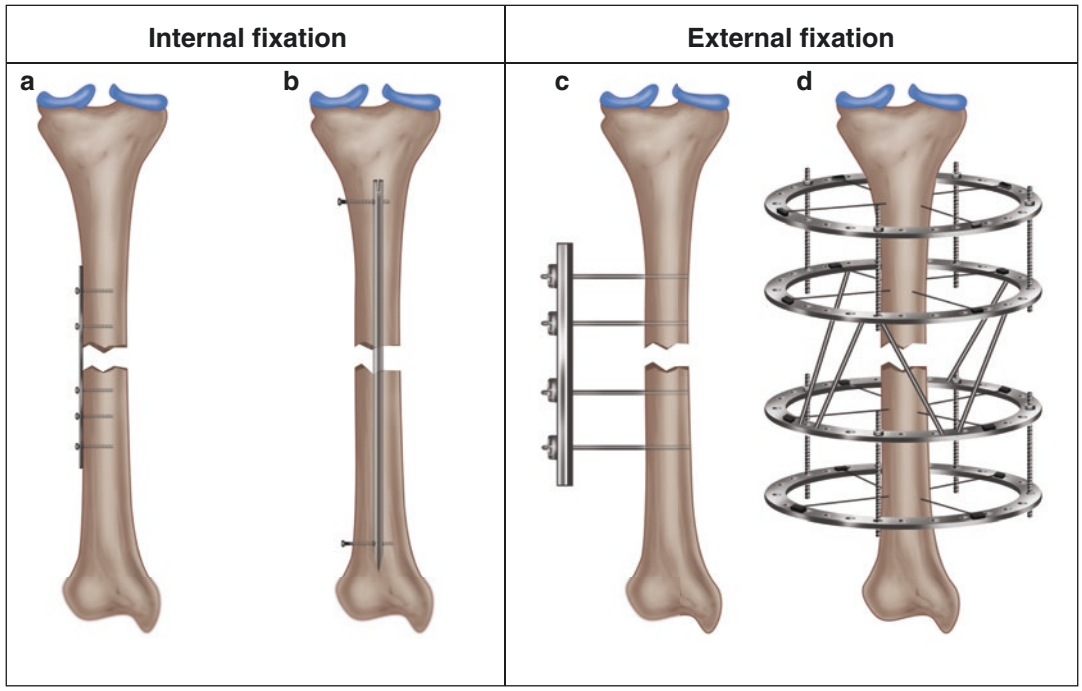

Fig. 29.5 Differences between internal and external fixation. (**a**) plate, (**b**) intramedullary nail, (**c**) unilateral external fixator, (**d**) circular external fixator

29.7 Osteoporotic Bone Fractures

Osteoporotic bone fractures (fragility fractures) are a result of osteoporosis, a condition in which the bones become more fragile due to bone deterioration or low bone mass. Osteoporotic fractures are produced by minimal trauma due to bone fragility. Bones commonly affected by these fractures include the spine, hip, distal radius, and proximal humerus. Although osteoporotic bone is structurally different, osteosynthesis techniques are nevertheless the same. The development of new implants for osteoporotic bone should improve the design of the devices and increase the fixation power of the screws. Indeed, metallic implants have a cutting effect on the osteoporotic bone, so it is possible to detect implant movements or breakage (named as "cut-out" or "pullout"). Cutout is the impaction of the screw with intra-articular penetration; screw pull-out corresponds to "en bloc" pullout of the screw from the bone at one or both ends of the plate.

Sometimes, fractures in the osteoporotic bone imply an irreversible crushing of the cancellous bone. In these cases, the augmentation with bone cement is necessary to fill cavities or to increase the anchoring of the implant.

29.8 Open Fractures: Principles of Treatment

Open fractures present more complications than closed fractures, with higher risks of infection, nonunion, and coverage defects. The treatment generally depends on the type of fracture and the degree of soft tissue injury. It is mandatory to clean wounds properly and remove necrotic tissues before fixation, avoiding any contact of the implants with the infected area. In some cases, it may be necessary to treat the soft tissue injury while trying to consolidate the bone fracture. In cases of severe bone loss (due to the trauma or necrotic tissue removal), osteosynthesis with an external fixator may be helpful, allowing to treat firstly the soft tissues and subsequently the bone. In open fractures, antibiotic management (with oral, systemic, or local administration) is mandatory. In specific antibiotic-loaded cement, the antibiotic can directly reach the infection site.

Take-Home Message

- Callus development depends on mechanical factors and biological factors, such as vascularization, blood supply, growth factors, oxygen tension, and tissue conditions.
- Treatment techniques can be divided into conservative or surgical methods.
- Rigid fixation minimizes the formation of the granulation tissue and external callus. It achieves a primary consolidation.
- The dynamic or elastic treatments allow the passage of stresses through the focus of fracture (cast, intramedullary nail, or external fixator) and permit micromovements between fracture fragments.
- Fracture treatment can be emergency, temporary, or definitive.
- Typical conservative treatments are bandages, casts, and splints, placed in a suitable position for the time necessary to achieve a stable consolidation.

Summary

Fracture healing is a physiological process that aims at a complete functional recovery without deformities. It implies an anatomical process of bone consolidation, rehabilitation of the muscle tone, and articular function. Callus development depends on mechanical factors and biological factors, such as vascularization, blood supply, growth factors, oxygen tension, and tissue conditions. Bone fracture treatment can be divided into conservative or surgical. The latter could also be divided into closed (percutaneous) or open surgery. Rigid fixation minimizes the formation of the granulation tissue and external callus. It achieves a primary consolidation: bone is formed

directly between fracture fragments, without the need for an external callus. In contrast, elastic systems induce the formation of an exuberant callus that aims to restrict mobility. There are several principles in the treatment of fractures to achieve stable fixation: the fracture focus has a displacement tendency due to muscle contractions. Compression must counteract these stresses and achieve a good contact between fragments. Rigid treatments (plates or lag screws for interfragmentary compression) do not allow the passage of stresses through the fracture focus, and there is no mobility between fracture fragments. Dynamic or elastic treatments allow the passage of stresses through the fracture focus (cast, intramedullary nail, or external fixator) and permit micro-movements between fragments. Fracture treatment can be emergency, temporary, or definitive. The typical conservative treatments are bandages, casts, and splints, placed in a suitable position, for the time necessary to achieve a stable consolidation.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. An exposed fracture of the tibial diaphysis should be fixed with:
   (a) External fixation
   (b) Casting
   (c) Plate and screws
   (d) Intramedullary nail
2. The main principles of exposed fractures are:
   (a) Debridement, washing and fracture stabilization
   (b) Skin suture and traction
   (c) Skin suture and antibiotics
   (d) Medication that keeps the skin opened and sterile
3. The most common treatment of diaphyseal femoral fracture is:
   (a) Intramedullary nailing fixation
   (b) Plate and screws
   (c) External fixation
   (d) Screw fixation
4. Intramedullary nailing is commonly adopted in:
   (a) Diaphyseal fractures
   (b) Metaphyseal fractures
   (c) Epiphyseal fractures
   (d) Meta-diaphyseal fractures
5. One of the most common casting complications of diaphyseal femoral fractures is:
   (a) Knee stiffness
   (b) Varus angulation of the fracture
   (c) Valgus angulation of the fracture
   (d) Nonunion

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Fracture Complications

30

Stephen Gibbs, Madison Colcord,
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Overview

The majority of fractures heal uneventfully. Complications are defined as deviations from the normal, expected posttreatment course, whereas sequelae are negative effects related to the injury itself or inherent to the treatment.

30.1 Introduction

The majority of fractures heal uneventfully. However, negative outcomes in the treatment of fractures can occur as the result of associated injuries, complications of treatment, or sequelae. Complications are defined as deviations from the normal, expected posttreatment course, whereas sequelae are negative effects related to the injury itself or inherent to the treatment. The designation of one particular negative outcome as an associated injury, complication, or a sequela depends on when and how it occurred. This chapter provides an overview of injuries associated with fractures, complications of fracture care, and fracture sequelae.

30.2 Arterial Injury

Arterial injuries can occur in association with fractures or as a complication of fracture surgery.

30.2.1 Epidemiology

The overall rate of arterial injury in association with fractures caused by blunt trauma is 1% (Coleman Am Surg 2016). The rate varies by anatomic location and injury mechanism. Some blunt trauma injuries have a strong association with arterial injury: knee dislocations have a popliteal artery injury rate of 16%.

30.2.2 Etiology/Pathogenesis

Arterial injury can be caused by penetrating injury, sharp fracture fragments, avulsion of the artery from tethering structures, vasospasm, or increased intracompartmental pressure. Arterial injury as a complication of fracture surgery usu-

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ally occurs by laceration from a sharp instrument, such as a knife or drill bit.

30.2.3 Classification

While the detailed classification of peripheral vascular injury is beyond the scope of this text, the orthopedic surgeon must determine whether the arterial injury has resulted in compromised perfusion to the extremity.

30.2.4 Diagnosis

Physical examination of the fractured extremity is the key to early diagnosis. The "hard signs" of arterial injury (external arterial bleeding, rapidly expanding hematoma, palpable thrill/audible bruit, and signs of distal ischemia) should be recognized. Distal peripheral pulses should be checked and compared side to side for any differences. The best noninvasive tests for arterial injury are the ankle-brachial index (ABI) for the lower extremity and the wrist-brachial index (WBI) for the upper extremity. In these tests, the systolic blood pressure distal to the injury (ankle or wrist) is divided by the proximal (brachial) systolic blood pressure to produce a ratio. An ABI less than 0.9 is indicative of an arterial injury. Invasive methods used for diagnosis of arterial injuries include angiogram and CT angiogram.

30.2.5 Treatment

A dysvascular limb is a surgical emergency requiring arterial repair or revascularization. Often, the orthopedic surgeon's role is to provide skeletal stability with external or internal fixation of the fracture at the time of vascular repair. Prophylactic fasciotomies of the extremity distal to the injury may be indicated to reduce the effects of reperfusion injury. Arterial injuries that do not compromise distal perfusion may be ligated for hemostasis, depending on the specific clinical scenario. Intraoperative arterial injury can be managed as indicated with either ligation or repair; if the complication is recognized postoperatively and perfusion of the limb is compromised, the patient must return to the operating room immediately for exploration.

30.3 Peripheral Nerve Injury

Peripheral nerve injuries are nerve injuries outside of the brain or spinal cord.

30.3.1 Epidemiology

The rate of associated peripheral nerve injury is influenced by fracture location, local anatomy, and mechanism of injury. The incidence of peripheral nerve injury in extremity trauma is between 1% and 3%. Some fracture types have well-known associations with peripheral nerve injury. Fractures of the humerus involving the distal one-third of the diaphysis are associated with radial nerve palsy; fractures of the sacral ala medial to the neural tunnels (Denis Zone III) have a 56.7% incidence of neurologic injury. As a complication of surgery, the rate of peripheral nerve injury varies by anatomic location and surgical approach. Postoperative ulnar nerve palsy following operative fixation of distal humerus fractures ranges from 0% to 21%. Iatrogenic sciatic nerve injury during acetabular fracture surgery occurs in 5.6% of cases.

30.3.2 Etiology/Pathogenesis

Peripheral nerve injury occurs by laceration, stretch, compression, or contusion.

30.3.3 Classification

Peripheral nerve injuries are classified as neuropraxia, axonotmesis, or neurotmesis. Neuropraxia, the mildest form of nerve injury, is characterized by focal segmental demyelination without disruption of axons. Axonotmesis, comparably more severe, is characterized by

axonal disruption with intact endoneurium. Neurotmesis, the most severe form of injury, is complete transection of a peripheral nerve.

30.3.4 Diagnosis

A detailed motor and sensory examination of the extremity will allow diagnosis of most clinically significant peripheral nerve injuries.

30.3.5 Treatment

In order to prevent surgical complications, forceful retraction of tissue near nerves should be minimized, and an adequate neurolysis of the nearby nerve should be performed when necessary to gain exposure to a fracture. An intraoperative peripheral nerve laceration should be repaired, if the surgeon is capable of doing so, or tagged for later repair. If a postoperative peripheral nerve palsy is assumed to be a stretch injury, the limb should be positioned to reduce stretch on the nerve, and any compressive bandages or splints should be loosened. Treatment of peripheral nerve injuries varies based on the specific clinical scenario. Neuropraxia injuries often have a good prognosis for recovery with observation alone, and neurotmesis injuries have a poor prognosis without nerve repair. Tendon transfers may be indicated to restore function in the case of certain severe peripheral nerve injuries.

30.4 Venous Thromboembolism

Venous thromboembolism (VTE) includes deep vein thrombosis (DVT) and pulmonary embolism (PE). Patients with fractures of the pelvis or extremities are at risk for VTE, which can result in significant morbidity and mortality.

30.4.1 Epidemiology

The reported incidence of VTE varies widely according to the anatomic site, the mode of detection, and the study sample size. Without VTE prophylaxis, the rate of DVT in a trauma patient population is cited to be as high as 80%. With prophylaxis, one randomized controlled study found an 11% rate of DVT and 1% rate of clinically significant PE.

30.4.2 Etiology/Pathogenesis

Virchow's triad of venous stasis, endothelial injury, and hypercoagulable state is thought to mediate the development of VTE.

30.4.3 Classification

There is no widely used orthopedic classification system for VTE. Thrombi can be categorized as superficial or deep and by location in the upper or lower extremities.

30.4.4 Diagnosis

Clinical signs and symptoms associated with DVT include unilateral limb swelling, redness, and tenderness. Pulmonary embolism can present with tachypnea, tachycardia, dyspnea, or hemoptysis. However, physical examination of a patient with DVT or PE can be completely normal and therefore cannot be used to rule out those diagnoses. When DVT is suspected, the diagnosis can be confirmed through duplex ultrasound or CT venography. CT pulmonary angiography or ventilation-perfusion scintigraphy is used to diagnose PE. The Orthopaedic Trauma Association, the American Academy of Orthopaedic Surgeons, and the American Academy of Chest Physicians guidelines recommend against the routine screening of asymptomatic patients.

30.4.5 Treatment

The practice of VTE prophylaxis is generally accepted for orthopedic trauma patients. The tim-

ing of prophylaxis initiation, duration of prophylaxis, and agent are debated. Mechanical prophylaxis, with compression stockings or intermittent pneumatic compression devices, is a low-risk adjunct to chemoprophylaxis. In patients contraindicated for chemoprophylaxis, inferior vena cava filters can be implanted to reduce the risk of PE. Therapeutic anticoagulation is indicated for confirmed DVT or PE.

30.5 Infection

Fracture-related infection (FRI) is infection occurring after fracture fixation.

30.5.1 Epidemiology

The overall rate of postoperative infection after fracture fixation is approximately 11%. The association between open fractures has long been appreciated, with infection seen in 1–2% of Gustilo-Anderson type I fractures and 15–60% of type III fractures.

30.5.2 Etiology/Pathogenesis

Risk factors for infection include open fracture, severe soft tissue injury, immunocompromised host, and tobacco use. FRI in closed fractures treated with internal fixation is thought to arise from bacterial inoculation at the time of surgery. Infection of open fractures can be related to bacterial contamination at the time of injury or surgical treatment. The US Centers for Disease Control estimates that half of the surgical site infections are preventable.

30.5.3 Classification

The FRI international consensus group proposed confirmatory (infection definitely present) and suggestive (infection possibly present) criteria for diagnosis of fracture-related infection. A distinct time frame that differentiates acute from chronic FRI is not available in the literature.

30.5.4 Diagnosis

Diagnosis of infection is based on clinical judgment combined with laboratory tests, imaging, and microbiological evidence. Confirmatory criteria for FRI include fistula, sinus tract, wound dehiscence, purulent drainage, presence of pus, or >5 polymorphonuclear neutrophils per high-power field (×400) in chronic cases. Suggestive criteria include clinical signs such as redness or fever, radiologic signs, new-onset joint effusion, elevated serum inflammatory markers, or persistent wound drainage. On surgical exploration, growth of a microorganism on two separate deep specimens or histopathological evidence of infection is confirmatory.

30.5.5 Treatment

Antibiotic prophylaxis at the time of surgery is indicated to reduce the risk of infection. In open fractures, administration of appropriate intravenous antibiotics within 60 min of injury is the most important intervention to reduce the risk of infection. Timely surgical debridement of open fractures is indicated. In the case of severe contamination or soft tissue injury, staged management of open fractures with external fixation may reduce the risk of infection. When infection is identified, the specific clinical scenario will direct treatment. Timing of symptom onset, progress of fracture healing, type of fixation construct, soft tissue condition, host physiology, and infectious organism will all influence treatment. When FRI is identified prior to fracture consolidation, debridement with antimicrobial therapy and implant retention (DAIR) may be reasonable to maintain fracture stability until bony union. FRI identified after fracture healing, or chronic infection, should be treated with debridement, implant removal, and antibiotic therapy.

30.6 Nonunion

Nonunion is failure of a fracture to heal. A pragmatic definition of a nonunion is a fracture in which the healing process is arrested and will not progress to union without further intervention.

30.6.1 Epidemiology

The overall rate of nonunion in a large study of 18 human bones was 4.9%. Closed fractures had a nonunion rate of 4.7% overall, and open fractures had a nonunion rate of 10.9%. Nonunion rate varied by fracture site, with scaphoid fractures and tibia fractures exhibiting the highest nonunion rates at 15.5% and 14%, respectively.

30.6.2 Pathogenesis

Nonunion can be caused by infection, impaired biology (poor vascularity), mechanical issues (such as instability, malalignment, or fracture gap), metabolic conditions, host factors, or a combination of these.

30.6.3 Classification

Nonunions are characterized by their radiographic appearance. Nonunions of all types show persistent fracture lines. Atrophic nonunions, associated with impaired local biology, show little to no callus formation at the fracture site. Hypertrophic nonunions, associated with mechanical instability, show abundant callus formation. Oligotrophic nonunions show intermediate features, with modest callus formation caused by disturbed local biology and mechanical instability.

30.6.4 Diagnosis

Nonunions may be painful with motion or weight-bearing of the affected extremity. On radiographs, nonunions show persistent fracture lines and arrest of progress toward healing. When evaluating a nonunion, it is important to recognize contributing factors such as infection, mechanical instability, malalignment, or systemic conditions. These will need to be addressed in the treatment strategy.

30.6.5 Treatment

Treatment varies by the nonunion bone, fracture location, type of nonunion, and other contributing problems, which need to be addressed to promote a successful outcome. For an atrophic nonunion, in which nonviable fracture edges and impaired local biology have resulted in failure to heal, a treatment strategy might involve placement of bone autograft. For a hypertrophic diaphyseal nonunion, in which mechanical instability has prevented healing, treatment might involve increasing stability at the nonunion site with more rigid internal fixation.

30.7 Fracture Sequelae

In the context of fracture treatment, sequelae are predictable “aftereffects” that are inherent to the injury. Sequelae of fractures include post-traumatic conditions such as stiffness, post-traumatic arthritis, osteonecrosis, and heterotopic ossification.

30.8 Stiffness

It is loss of joint range of motion. Stiffness occurs as the result of bony abnormality, soft tissue abnormality, or both. Examples of bony abnormalities include articular malreduction (intra-articular) and heterotopic ossification (extra-articular). Soft tissue abnormalities include capsular contracture (intra-articular) and ligamentous contracture (extra-articular). Prolonged immobilization is a risk factor for stiffness. Early motion can mitigate the risk of postoperative stiffness.

30.9 Post-traumatic Arthritis

It is a painful, debilitating condition characterized by loss of articular cartilage. Fractures involving the articular surface of weight-bearing joints are at increased risk of this sequela. The development of post-traumatic arthritis is related to abnormal wear of the cartilage due to articular incongruity, chondral damage, mechanical malalignment, or instability.

30.10 Osteonecrosis

Osteonecrosis or bone death occurs when vascular supply to a bone is disrupted by fracture or dislocation. Osteonecrosis is most commonly seen in areas with poor or tenuous vascularity such as the scaphoid, talus, or femoral head.

30.11 Heterotopic Ossification

Heterotopic ossification (HO) is the pathologic formation of bone in an abnormal location. The pathogenesis of HO is related to soft tissue damage at the time of injury or from surgical insult. HO ranges in severity from asymptomatic islands of bone to ankylosis. The most commonly affected areas are the hip and elbow.

Take-Home Message

- The majority of fractures heal uneventfully.
- Negative outcomes in the treatment of fractures can occur as the result of associated injuries, complications of treatment, or sequelae.
- Complications are defined as deviations from the normal, expected post-treatment course.
- Sequelae are negative effects related to the injury itself or inherent to the treatment.
- Patients with fractures of the pelvis or extremities are at risk for VTE, which can result in significant morbidity and mortality.
- The overall rate of postoperative infection after fracture fixation is approximately 11%.
- A pragmatic definition of a nonunion is a fracture in which the healing process is arrested and will not progress to union without further intervention.

Summary

The treatment of fractures can be complicated by the presence of associated injuries, iatrogenic surgical or nonsurgical complications, and sequelae inherent to the injuries themselves. Early recognition of common associated injuries and prevention of iatrogenic complications are necessary to ensure the best possible outcome.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. The diagnosis of an arterial injury after fracture consists of:
   (a) The "hard signs" of arterial injury (external arterial bleeding, rapidly expanding hematoma, palpable thrill/audible bruit, and signs of distal ischemia)
   (b) The "hard signs" of arterial injury (tachycardia, hypoglycemia)
   (c) Ultrasonography with echo-color Doppler
   (d) CT imaging
2. The most common nerve injury following operative fixation is the:
   (a) Ulnar nerve palsy
   (b) Radial nerve palsy
   (c) Sciatic nerve injury
   (d) Sciatic nerve palsy
3. Virchow's triad consists of:
   (a) Venous stasis, endothelial injury, and hypercoagulable state
   (b) Venous stasis, hypoglycemia, and tachycardia

(c) Tachycardia, venous stasis, and hyperglycemia
(d) DVT, uncontrolled bleeding, and tachycardia

4. Gustilo-Anderson classification is used for:
   (a) Open fractures
   (b) Closed fractures
   (c) Femoral fractures
   (d) Radial fractures
5. Nonunion could be classified into:
   (a) Hypertrophic, oligotrophic, and normotrophic
   (b) Stable and unstable
   (c) Static or progressive
   (d) Diffuse or localized

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The Polytraumatized Patient

31

Christopher D. Flanagan and Heather A. Vallier

Overview

This chapter focuses on the role of appropriate fracture management in the care of the polytraumatized patient. The trimodal temporal distribution of mortality following polytrauma is introduced; the role of appropriate musculoskeletal management in improving outcomes is explored. Classification systems utilized in polytrauma are discussed. The diagnosis and treatment evaluation of the polytraumatized patient are outlined. Treatment strategies are investigated.

31.1 Definition

The polytraumatized patient refers to any patient who has sustained injuries to multiple organ systems or, more specifically, in the case of musculoskeletal trauma, to multiple aspects of the same organ system. Strict, numerical definitions remain elusive, though quantifiable definitions do exist for standardized research reporting. Such patients require management by subspecialty providers to achieve optimal outcomes.

31.2 Epidemiology

The incidence and prevalence of polytrauma depend somewhat on the research classification used to define this entity. However, certain injury mechanisms and patient characteristics are associated with the polytraumatized patient. Most polytrauma patients sustain injury through blunt force mechanisms, often motor vehicle and motorcycle collisions. Males outnumber females in polytrauma reporting. While all age groups can sustain polytrauma, classically younger patients present secondary to high-energy mechanisms, while older patients present secondary to low-energy mechanisms, generating a bimodal age distribution. Associated factors also include potential drug or alcohol intoxication, as well as underlying mental health issues.

31.3 Etiology/Pathogenesis

Classically, mortality following polytrauma followed a trimodal temporal distribution. The first phase in this schema involved death within minutes at the scene of the accident. Non-survivable injuries, including aortic or major arterial rupture, or massive intracranial hemorrhage with

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herniation, accounted mechanistically for this first wave. Generally, this represents an area for which improvements in the delivery of polytrauma care will not ultimately change outcomes; however, systematic advances in safety measures, such as the widespread use of seatbelts and airbags, may reduce these deaths and allow patients to survive until hospital arrival. Survival to hospital presentation represents the second temporal category in this scheme. Mortality in this stage results from an inability to achieve resuscitation secondary to a lack of hemorrhage control. These deaths fall into the category of "possibly preventable." The prompt identification of injury, using clinical and radiographic tools, and the expeditious treatment of these injuries, may reduce morbidity or mortality. Improvements in trauma care delivery can influence mortality rates during this stage. The third temporal stage involves deaths days and weeks following injury. These deaths result from complications related to the initial trauma itself and also the care delivered. Infection, sepsis, blossoming pulmonary contusions, and deep vein thrombosis/pulmonary embolism (DVT/PE) are examples of mortality drivers during this third temporal stage.

Musculoskeletal injuries contribute substantially to hemorrhage and later complications following polytrauma; appropriate management of fractures can improve patient outcomes in the second and third temporal windows. Limb-threatening injuries, such as compartment syndrome, can have devastating systemic effects; in the setting of delayed management, massive myonecrosis produces a nidus for infection. Electrolyte abnormalities, especially hyperkalemia, related to rhabdomyolysis can result in severe cardiac and renal dysfunction. In addition, fractures themselves can also produce considerable blood loss; treating providers must recognize this fact. For example, a closed femoral shaft fracture can result in 1000–1500 mL of blood loss into the thigh; this represents approximately 20% of an average adult circulating blood volume. Clearly, bilateral femoral shaft fractures would have considerable hemodynamic consequences. Judicious application of tourniquets in the field or on arrival can dramatically reduce bleeding. Pelvic ring injuries can result in similar large-volume blood loss into the pelvis. A classic motto to remember in polytraumatized patients is the five locations into which a patient can have life-threatening hemorrhage: the chest, the abdomen, the pelvis, the thigh, and the floor.

Appropriately timed management of musculoskeletal injuries can also reduce morbidity and mortality from complication related to immobility; this represents strategies to improve outcomes in the third temporal window. Prompt management of open fractures can reduce rates of future infection, osteomyelitis, and sepsis. Provisional and definitive fracture fixation improves patient mobility, which can reduce the risk of DVT/PE. Improved mobility mitigates the development of decubitus ulcers, which can serve as a nidus for infection. Prolonged immobility can also result in overall deconditioning, a process which occurs rapidly in older patients and from which they may never fully recover.

31.4 Classifications

Several classification systems exist to convey the degree of injury severity in polytraumatized patients. The Abbreviated Injury Scale (AIS) represents one such system. This scale involves sectioning the body into nine different areas: head, face, neck, thorax, abdomen, spine, upper extremities, lower extremities, and external. The clinician assigns a numerical value of 0–6, corresponding to the severity of the injury, to each region (Table 31.1). A lower score corresponds to a more minor degree of injury; conversely, a

Table 31.1 Abbreviated injury scale

Score	Definition	Musculoskeletal example
0	No injury	None
1	Minor	Metatarsal fracture
2	Moderate	Radial shaft fracture
3	Serious	Femoral shaft fracture
4	Severe	Traumatic below-knee amputation
5	Critical	Anterior-posterior compression type 3 pelvic ring injury (open-book pelvis)
6	Maximum	Hemipelvis transection

score of 6 represents a non-survivable injury. The Injury Severity Score, an extension of the AIS, combines the individual areas of trauma into a single overall score. To calculate the ISS, the clinician identifies the three areas with the highest AIS. The square of each of these AIS is calculated and then summed, producing the ISS. For example, if a patient sustained a rib fracture (thorax—AIS = 1), a femoral shaft fracture (lower extremity—AIS = 3), and renal artery laceration (abdomen—AIS = 3), the patient would have an ISS of 19 ($1^2 + 3^2 + 3^2$). If any single injury sustained by the patient has an AIS = 6 (non-survivable), the patient automatically receives an ISS of 75. A positive correlation exists between ISS score and mortality rate. For this system, an ISS of 15 or greater defines a patient as "polytraumatized."

Polytrauma patients often present with hemodynamic instability; identifying the cause of this instability will allow the clinician to pursue appropriate management. Generally, four categories of shock exist: hemorrhagic/hypovolemic, neurogenic, cardiogenic, and septic. Cardiogenic and septic shock may occur in trauma patients, but these occur less frequently in the acute period than hemorrhagic and neurogenic shock. Four classes of hemorrhagic/hypovolemic shock exist (Table 31.2). Importantly, providers must understand that a patient may lose 15% of their circulating blood volume prior to exhibiting changes in hemodynamic parameters. Additionally, providers must recognize that a decrease in blood pressure represents a late response to hemorrhage; tachycardia results first, with hypotension present only after approximately a 30% decrease in circulating blood volume. Furthermore, this schematic should emphasize the importance of musculoskeletal injuries to hemodynamic stability; a patient with isolated, bilateral closed femoral shaft fractures has an anticipated blood loss of approximately 2000–3000 mL, representing potential presentation to the hospital in class IV hemorrhagic shock. Hemorrhage results in reduced oxygenation capacity, generating metabolic acidosis. Identification and correction of metabolic acidosis are critical elements of initial care.

Table 31.2 Classes of hemorrhagic shock

Class	Blood loss (assuming 5 L circulating volume)	Heart rate (bpm)	Blood pressure
I	<15% (750 mL)	Normal	Normal
II	15–30% (750–1500 mL)	>100	Normal
III	30–40% (1500–2000 mL)	>120	Decreased
IV	>40% (>2000 mL)	>140	Decreased

Neurogenic shock in the polytraumatized patient also deserves special consideration, as treatment for this type of hemodynamic instability differs from the volume resuscitation required for hemorrhagic shock. Polytrauma patients may sustain ligamentous injuries or fractures to the spinal column, which results in spinal cord injury. Neurogenic shock, which represents in a loss of systemic sympathetic tone, may result. Bradycardia and hypotension are noted. In addition to fluid resuscitation, these injuries require vasopressor support to prevent circulatory collapse. Importantly, neurogenic shock should not be confused with the term spinal shock, which represents a different entity; these terms are not interchangeable.

31.5 Diagnosis

The management of a polytraumatized patient should proceed in a consistent, standardized, and systematic fashion which includes a primary, secondary, and tertiary examination. The primary survey occurs at the initial patient presentation. A common mnemonic for the primary survey is ABCDE—airway, breathing, circulation, disability, and exposure. The clinician should first determine the patency of the airway: Are breath sounds present? Is the patient talking? Second, breathing is assessed: Is the patient breathing? What is the respiratory rate? Third, circulation is determined: What is the pulse? What is the blood pressure? Are the extremities cold or well perfused? Fourth represents disability: What obvious extremity injuries are apparent? Is the patient moving all limbs? Is the pelvic ring stable? Exposure fol-

lows promptly: the patient's clothes are removed, allowing for a full examination to identify injury. A secondary survey should take place only after a dedicated primary survey has occurred and any immediate life-threatening issues are addressed.

In the clinically stable patient, a secondary survey then proceeds; this survey should occur immediately following the completion of the primary survey and the acute stabilization of the patient. In this survey, the clinician examines the patient for extremity injuries. All major joints and long bones should undergo palpation and, when appropriate, range of motion testing. Specific attention should focus on (1) identifying pelvic ring injuries; (2) identifying spinal column injuries; (3) identifying open (e.g., exposed to the external environment) fractures/dislocations; (4) identifying major long-bone injuries; (5) confirming limb perfusion and identifying limb ischemia; and (6) identifying current or impending compartment syndrome. In severely displaced pelvic ring injuries, open fractures may occur through the rectum or vagina; a rectal or pelvic exam is mandatory if such fractures are suspected. A cervical spine injury should be assumed to be present in all patients until excluded by further testing. Multiple providers must logroll the patient to exam for spinal column compromise; the provider should check for step-offs and areas of bogginess, as these may denote areas of injury. A rectal exam is mandatory in the setting of a spine injury.

Additional diagnostic tests may occur during the secondary survey. Performing ankle-brachial index (ABI) testing can identify occult arterial injuries. To perform this exam, the clinician takes a blood pressure reading at the ankle and in the arm; ABI equals the systolic blood pressure at the ankle divided by the systolic blood pressure at the arm. Values less than 0.9 should raise concern for occult vascular injury, and appropriate arterial imaging should follow. An assessment for compartment syndrome should take place during the identification of fractures; the clinical signs of pain, paresthesia, pallor, paralysis, and pulseless are considered diagnostic. However, severe pain in the presence of a tense limb suggests compartment syndrome and need for emergent fasciotomy. In patients with altered mental status, direct compartment pressure measurements may be warranted. Compartment pressure within 30 mmHg of the diastolic blood pressure diagnoses compartment syndrome.

Diagnostic accuracy hinges on obtaining expedient medical imaging in the polytraumatized patient. In the trauma bay, the primary survey should include plain chest and pelvis radiographs; these provide immediate information for interventions. Ultrasound to perform a Focused Assessment with Sonography in Trauma (FAST) exam of the abdomen is sensitive in detecting intra-abdominal hemorrhage. However, advanced imaging in the form of CT scans remains the most common imaging modality during initial trauma evaluation. Generally, at a minimum, scans of the chest, abdomen, pelvis, and cervical spine are completed on arrival. Lower extremity CT angiography should also occur if ABI is <0.9. All extremities that the patient identified as painful during the secondary survey, or that have concern for deformity, require plain radiograph evaluation. In general, the joints proximal and distal to a fracture also require plain radiographic imaging.

The tertiary survey represents the final phase of the trauma evaluation. This may occur days (in some cases, weeks) following injury. Ideally, a patient should be awake and alert enough to participate in this exam. Once again, the clinician palpates all bones and joints, paying close attention to the hands and feet, as missed occult injuries in these areas can occur secondary to other major distracting injuries. The clinician ranges all major joints. Any area of pain should undergo further evaluation with plain radiographs to exclude the presence of a fracture or dislocation.

31.6 Treatment

Treatment for the polytraumatized patient should proceed according to the results of the initial trauma survey. The clinician should secure the airway. Ventilatory support should be initiated, as indicated. Administration of crystalloids and/or blood products should proceed according to the hemorrhagic shock classification; vasopressor support should occur in the setting of neurogenic

shock. Immediate operative exploratory surgery should proceed as indicated in the unstable patient. As hemodynamic stabilization proceeds, simultaneous fracture management should occur to assist these resuscitative efforts.

In the trauma bay, the musculoskeletal focus should start with the pelvis and spine. A cervical collar should be placed and not removed until a traumatic injury has been excluded. Unstable pelvic ring injuries that result in volumetric expansion of the pelvis (e.g., open-book pelvic ring injuries) should be immediately addressed. Circumferential sheet or binder application represents a simple, effective method for reducing intrapelvic volume. Application should be overlying the ilea and the greater trochanters, so that when the sheet or binder is tightened, the diameter of the pelvis will be decreased, promoting volumetric control. Temporizing external fixation and/or definitive treatment of pelvic ring injuries should occur promptly to permit removal, in order to avoid skin compromise.

Long-bone injuries should be addressed next, as these can contribute to circulatory collapse. Femoral shaft fractures may undergo placement of skeletal traction by a trained orthopedic provider. This traction provides longitudinal tension which results in perpendicular compression around the fracture, leading to pain relief and reduced bleeding. Other long-bone fractures should undergo splint stabilization, as this method also reduces bleeding from the fracture site. These interventions will aid resuscitation.

Musculoskeletal attention should then focus on identifying open fractures, as these require urgent operative intervention for debridement and stabilization. Prompt administration of antibiotics reduces infection from open fracture. Tetanus vaccination should also be updated. These fractures should undergo provisional reduction and immobilization in the trauma bay. All other fractures and dislocations should undergo closed reduction maneuvers and be immobilized appropriately with splints or braces. Reduction and splinting of displaced fractures promote soft-tissue rest and reduce complications related to both closed and operative management.

In general, orthopedic operative interventions proceed along one of the two pathways: damage control orthopedics (DCO) or early appropriate care (EAC). DCO occurs as part of the resuscitative process; the goal is to stabilize major fractures, often with external fixation, in order to assist in resuscitative efforts. EAC refers to the timing of definitive orthopedic care in the polytraumatized patient. In general, the EAC protocol recommends definitive fixation within 36 h of injury if the patient has been adequately resuscitated, as measured by improvement of metabolic acidosis. The following laboratory values should be met: lactate <4.0 mmol/L, pH ≥ 7.25, and base excess ≥−5.5 mmol/L, as outcomes are improved in this setting.

Take-Home Message

- Polytraumatized patients require a multidisciplinary approach to obtain optimal outcomes.
- Mortality following trauma follows a trimodal temporal distribution. Appropriate fracture management in the second and third waves can reduce mortality rates.
- Fractures may be associated with hemodynamic consequences related to blood loss; providers must remember that fracture stabilization supports hemodynamic resuscitation efforts.
- ISS and hemorrhagic shock class represent important metrics for the classification and stabilization of trauma patients.
- Neurogenic shock requires vasopressor support.
- Primary, secondary, and tertiary exams are mandatory in the polytraumatized patient.
- Suspected injuries must undergo appropriate advanced or plain radiographic imaging to ensure diagnostic accuracy.
- In general, fractures should undergo prompt closed reduction and manipulation. Open fractures require antibiotics and tetanus vaccination.
- Prompt definitive operative management of orthopedic injuries improves long-term patient outcomes after polytrauma.

Summary

The polytrauma patient requires a multidisciplinary approach. Appropriate, timely fracture management can serve as a critical adjunct to initial resuscitative efforts as well as improve long-term outcomes in a challenging patient population.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Do males or females have higher rates of polytrauma?
   (a) Males
   (b) Females
   (c) Males with BMI >30
   (d) No differences between sex
2. What are some causes of mortality in each of the three classic temporal stages of polytrauma?
   (a) First: aortic shearing or massive intracranial hemorrhage with herniation. Second: inability to control hemorrhage. Third: infection and sepsis
   (b) First: inability to control hemorrhage. Second: aortic shearing or massive intracranial hemorrhage with herniation. Third: infection and sepsis
   (c) First: aortic shearing or massive intracranial hemorrhage with herniation. Second: infection and sepsis. Third: inability to control hemorrhage
   (d) First: inability to control hemorrhage. Second: aortic shearing. Third: infection and sepsis
3. Approximately how much blood loss occurs into the thigh after an isolated closed femoral shaft fracture?
   (a) 1000–1500 mL
   (b) 1500–2000 mL
   (c) 500–1000 mL
   (d) >2000 mL
4. How much blood loss must occur before hypotension is seen in a young adult? To which stage of hemorrhagic shock does this correspond?
   (a) 30–40%; stage III
   (b) 10–20%; stage II
   (c) 30–50%; stage IV
   (d) 10–30%, stage I
5. What simple, fast measure can be used to decrease intrapelvic volume in the setting of unstable pelvic ring injuries?
   (a) Circumferential sheet or binder application
   (b) External fixation
   (c) Packing
   (d) Ischemic tourniquet to inferior limbs

Further Reading

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Part VIII

Spine

Cervical Spinal Pain

32

Brandon Yoshida, Mohamed Kamal Mesregah, Brandon Ortega, Jeffrey C. Wang, and Zorica Buser

Overview

This chapter first defines cervical spinal pain and reviews the prevalence, risk factors, and common concomitant pathologies upon presentation. Following sections will elaborate on the pathophysiology of cervical spinal pain, classify the different types of cervical spinal pain, and explain how mechanical and neuropathic cervical spinal pain compare and contrast with each other. Finally, the chapter discusses about pertinent findings in the history and physical that may support a diagnosis of cervical spinal pain as well as effective means to diagnose and treat.

32.1 Definition

Cervical spinal pain is a leading cause of disability worldwide and is commonly described as pain involving the neck and/or shoulder regions. Specifically, the International Association for the Study of Pain defines cervical spinal pain as pain perceived anywhere in the posterior region of the cervical spine between the superior nuchal line and the first thoracic spinous process; notably, pain located in the anterior cervical spine is commonly referred to as throat pain. Cervical spinal pain can be further subdivided based on region, with upper and lower cervical spinal pain being defined as pain above or below C4, respectively.

32.2 Epidemiology

Cervical spinal pain is a common musculoskeletal disorder with approximately half of all individuals experiencing at least one episode of clinically important neck pain over the course of their lifetime. It ranks as the fourth most common cause of disability in the United States behind back pain, depression, and other musculoskeletal disorders. Annual prevalence rates of neck pain range between 15% and 50%, and prevalence of neck pain typically peaks in middle age, although some studies cite that a high prevalence rate of neck pain is maintained from middle age and beyond.

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Cervical spinal pain may present alone or, more commonly, may present with several comorbidities such as depression, back pain, arthralgia, and headache. However, there is also a significant overlap between cervical spinal pain and other musculoskeletal and rheumatologic conditions as well as mental health disorders, sleep disorders, smoking, sedentary lifestyles, obesity, poor general health, and other psychosocial factors.

Risk factors for cervical spinal pain are primarily of traumatic origin or from participation in sports such as race car driving, football, wrestling, and ice hockey being the most strongly associated with the development of future neck pain. However, the most common risk factor for cervical spinal pain is whiplash, defined as forward acceleration of the head and neck while the trunk remains immobile. Besides traumatic events, other occupations including office workers, manual laborers, and healthcare workers also have a high incidence of neck pain. Low job satisfaction and perceived poor workplace environment are also commonly associated with neck pain.

Variables predicting worse prognosis of cervical spinal pain include the older age, female sex, radicular symptoms, and coexisting psychosocial pathology.

32.3 Etiology/Pathogenesis

The etiology of cervical spinal pain primarily originates from a local, identifiable pathologic cause of either musculoskeletal or nervous origin. In the event of traumatic injury such as whiplash, the shearing stress of a rapidly accelerating head and neck against a relatively immobile trunk violates the integrity of intervertebral disks and facet joints connecting adjacent vertebrae. In presentations of chronic cervical spinal pain, the origin and exact pathophysiologic mechanisms are obscure. Additionally, numerous studies have cited neck pain of nonorganic origin stemming from psychosocial factors. As a result, the etiology of cervical spinal pain may also be multifactorial and consistent with the biopsychosocial model, with psychosocial stress contributing to increased muscular tension and perceived pain and physical strain leading to microtrauma in connective and bony tissues.

32.4 Classifications

Numerous classifications of cervical spinal pain exist and can broadly be categorized by the pathophysiology, duration of symptoms, and severity of pain.

The most common classification of cervical spinal pain is based on one of the three pathophysiologic mechanisms of pain: mechanical, neuropathic, and secondary to another cause (e.g., referred pain). Mechanical pain is the most common type of neck pain and stems from microtrauma in the spine or its supporting structures, including the intervertebral disks, ligaments, musculature, fascia, and especially cervical facet joints (i.e., arthritis). Prior studies have demonstrated that mechanical pain secondary to cervical facet joint pain makes up 40–60% of all non-neuropathic cervical spinal pain. Mechanical cervical spinal pain is commonly described as a throbbing, dull ache in a non-dermatomal distribution.

Neuropathic pain is the second classification of cervical spinal pain according to pathophysiologic mechanism and makes up approximately 7% of cases. Neuropathic pain arises from disease or injury of the peripheral nervous system when cervical nerve roots are irritated, mechanically or chemically, from surrounding structures. Such irritation may result from spinal stenosis (e.g., cervical myelopathy) or a herniated disk or osteophyte compressing a nerve root. Neuropathic cervical spinal pain presents as a unilateral, shooting, sharp, electrical- or stabbing-like pain, commonly in a single- or multi-level dermatomal distribution depending on the number of cervical spinal roots affected (Table 32.1). Patients presenting with neuropathic pain typically experience higher levels of functional impairment and psychopathology compared to patients with mechanical pain.

In approximately 50% of cases, cervical spinal pain presents as a combination of both mechanical

Table 32.1 Differences between mechanical and neuropathic pain

	Mechanical pain	Neuropathic pain
Origin	The spine or its supporting structures (muscles, ligaments, and joints)	Nerve roots, or spinal cord
Potential causes	Degenerative disk disease, facet joint arthropathy, or muscle sprain	Nerve compression from a potential disk herniation, or osteophytes compressing the nerve roots causing radicular symptoms, or the spinal cord causing myelopathic symptoms
Character	Achy, dull, or throbbing	Shooting, burning, stabbing, tingling, or electrical-like
Localization	Usually localized, and it follows non-dermatomal distribution if referred	Radiates into one or both upper limbs following dermatomal distribution
Duration	Resolves with healing of damaged tissue but may be chronic	Mostly chronic
Intensity	Decreases with time	Increases with time
Hypersensitivity	Uncommon	Numbness, paresthesia, or dysesthesia
Motor deficits	Unusual	Present if motor nerve is affected
Aggravating factors	Exacerbations associated with activity	Unpredictable intense exacerbations

and neuropathic pain. This high proportion of patients is likely explained by the fact that neuropathic pain is commonly secondary to degenerative conditions such as cervical spondylosis, which predispose the patient to mechanical pain. Common examples of mixed mechanical and neuropathic pain include postlaminectomy (failed neck surgery) syndrome and intervertebral disk degeneration, where the neuropathic and mechanical pain components originate from the herniated nucleus pulposus compressing a cervical nerve root and disruption of the annulus fibrosus, respectively. Distinguishing between mechanical and neuropathic pain is clinically important, as the correct diagnosis directs clinical decision-making and defines which treatment options will be effective, from conservative to surgical modalities.

Cervical spinal pain can also be classified by the duration of symptoms. Like other disorders, neck pain can be subdivided into acute, subacute, and chronic states. According to the International Association for the Study of Pain (IASP) and the Neck Pain Task Force, acute, subacute, and chronic neck pain are defined to last less than 7 days, between 7 days and 3 months, and 3 months or more, respectively (note that terms used by the Neck Pain Task Force are transitory neck pain, short-duration neck pain, and chronic neck pain in lieu of acute, subacute, and chronic, respectively). This modality of classifying cervical spinal pain is the best predictor of prognosis, with cases presenting with shorter duration of symptoms being associated with better prognosis and resolving as quickly as 2 months in presentations of acute cervical spinal pain; however, it should be noted that while half of the patients presenting with acute cervical spinal pain have their symptoms resolve within 2 months, the other half of patients either continue to have low-grade symptoms or continue to seek treatment for over 1 year after presentation.

The final modality of classifying cervical spinal pain is based on the severity of symptoms. According to the Neck Pain Task Force, cervical spinal pain can be classified as one of the four different grades:

- Grade I: No signs of major pathology and no or little interference with daily activities
- Grade II: No signs of major pathology, but interference with daily activities
- Grade III: Neck pain with neurological signs or symptoms
- Grade IV: Neck pain with signs of major pathology (e.g., fractures, infections, neoplasm, spinal cord injuries, or systemic diseases)

Patients with grade III or IV cervical spinal pain should be referred for further medical management to seek out nerve decompression.

32.5 Diagnosis (Clinical and Imaging)

Diagnosing cervical spinal pain begins with a thorough patient history. Patients presenting with mechanical pain usually describe a throbbing, dull, aching pain in a non-dermatomal distribution that may be highly variable between different cases of mechanical cervical spinal pain. In cases that involve the atlantoaxial, atlantooccipital, or upper cervical facet joints or disks, patients may also describe pain radiating to the occiput. Generally, mechanical cervical spinal pain presents with early-morning stiffness and a low baseline level of pain that worsens with activity. Oftentimes, symptoms have an insidious onset secondary to progressive, nontraumatic strain on the intervertebral disks and cervical facet joints from repetitive, low-level stress.

Neuropathic pain, on the other hand, presents as a shooting, sharp, electrical- or stabbing-like pain in a unilateral dermatomal distribution with the C6 and C7 dermatomes being affected the most (which presents as pain radiating to the thumb or the second and third fingers, respectively). Neuropathic pain may also present with additional symptoms also common in patients with mechanical pain such as stiffness, but numbness, paresthesia, and/or dysesthesia are specific for neuropathic pain only, with sensory symptoms rarely presenting in cases of mechanical cervical spinal pain. Compared to mechanical pain that is often predictably worsened with activity, episodes of neuropathic pain are less predictable and are of higher intensity. Head turning or bending to the ipsilateral side as well as coughing, sneezing, or other Valsalva-like maneuvers that increase subarachnoid pressure may exacerbate the pain. The onset of isolated neuropathic pain is typically in an acute setting after a specific event, as herniated intervertebral disk is the most common etiology; in older patients, however, the onset may also be progressive since degenerative conditions may lead to both mechanical and neuropathic pain.

The role of the physical examination is to confirm findings in patients' history, provide additional diagnostic insight, and direct further workup management. Patients' general appearance should always be appreciated to help confirm patients' symptoms in the presence of associated facial expressions and behavioral indications. Malingering or factitious disorders should be suspected in cases where typical behaviors and expressions associated with pain are absent. Additionally, provocative maneuvers, while not always specific, are helpful in diagnostic workup. For example, the Spurling shoulder abduction and neck distraction tests have moderate sensitivity (approximately 50%) but high specificity (>80%) for cervical radiculopathy. The Hoffmann test also has moderate sensitivity (approximately 50%) but high specificity (approximately 80%) for cervical myelopathy. Range of motion is often limited in cases of mechanical neck pain, but both passive and active cervical range of motion only have moderate interrater reliability. Long-standing neuropathic neck pain may present with weakness. However, true neurologic weakness may be difficult to distinguish from pain-induced weakness; thus, testing for muscle strength and endurance also has only moderate interrater reliability and diagnostic utility.

Outside of the history and physical, numerous clinical assessment tools and questionnaires exist to help quantify patients' symptoms. The Neck Pain Task Force lists a number of red flags for providers to be aware of, including but not limited to pathologic fractures, infections, neoplasm, systemic inflammatory diseases, cervical myelopathy, and history of previous neck surgery (Table 32.2). Since neck pain can usually be alleviated with rest, severe, unrelenting pain should also be considered a red flag and may suggest a diagnosis of infection, neoplasm, or other primary neurologic conditions. With regard to self-reported pain scales, the visual analog scale (VAS) is frequently used for its simplicity and strong psychometric properties. While other pain scales exist such as the simple descriptive scale

Table 32.2 Neck pain red flags

Red flags	Possible conditions
Age <20 years	Congenital anomalies
Age >50 years	Metastases, vascular disease
Bowel/bladder dysfunction	Cervical myelopathy
Infectious symptoms (e.g., unexplained fever, history of intravenous drug use)	Vertebral osteomyelitis, spinal abscess, meningitis
Constitutional symptoms (e.g., fever, unexplained weight loss, history of malignancy)	Metastases Rheumatologic conditions
Significant trauma	Vertebral fractures or ligamentous disruption
History of rheumatoid arthritis	Atlantoaxial subluxation
Simultaneous chest pain, or shortness of breath	Myocardial ischemia
Tearing pain	Carotid/vertebral arterial dissection

(SDS) and numeric rating scale (NRS), the VAS scale is the most reliable and valid for measuring pain intensity and is therefore considered the gold standard. Semantic differential scales, like the McGill Pain Questionnaire (MPQ) or Short Form MPQ (SF-MPQ), also exist which utilize word lists and categories to help patients describe the quality of pain in three dimensions: affective, evaluative, and sensory. Unfortunately, these scales are less convenient and time consuming to complete and require a high patient literacy level.

Pain questionnaires provide self-reported information regarding the impact of neck pain, perceived functional ability, deficit, psychosomatic status, change of condition over time, and effectiveness of treatment intervention for both clinicians and patients. Examples of pain questionnaires include the Extended Aberdeen Spine Pain Scale (ASPS), the Bournemouth Questionnaire (BQ), and the Whiplash Disability Questionnaire (WDQ) amongst many others.

Other commonly utilized questionnaires include the Neck Disability Index (NDI), which measures pain, function, and disability, and the SF-36 Health Survey, which quantifies patients' physical and mental health statuses. These questionnaires are considered the gold standard for other questionnaires measuring similar criteria and are often used in research studies.

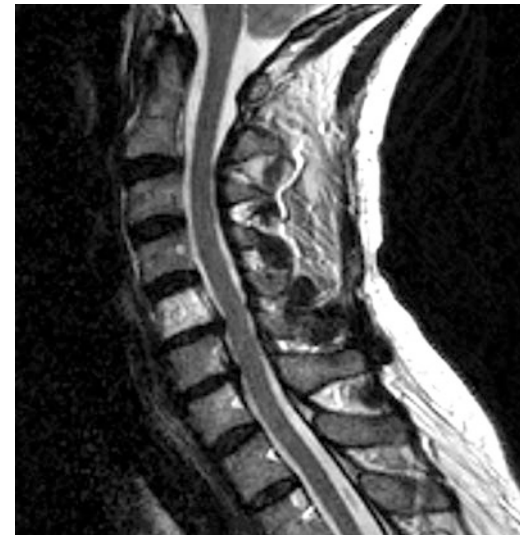

Fig. 32.1 T2-weighted sagittal magnetic resonance image of a 52-year-old female patient showing disk bulging at the levels of C5–C6 and C6–C7, with bone marrow edema of the C5 vertebra

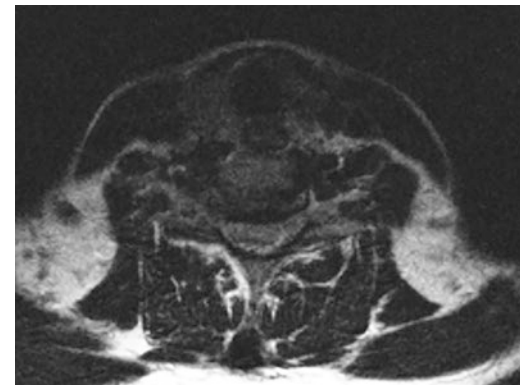

Fig. 32.2 T2-weighted axial magnetic resonance image through C6–C7 level of the same patient showing central spinal stenosis caused by the disk bulge

Regarding imaging modalities, magnetic resonance imaging (MRI) remains the most sensitive test for detecting soft-tissue abnormalities (Figs. 32.1, 32.2). However, MRI is also characterized with a rate of detecting abnormalities in otherwise asymptomatic patients as high as 60% and 80% in asymptomatic patients in their 40s and 60s, respectively. Therefore, MRI should only be used to rule out red flags in patients presenting with signs of serious neurologic deficits or for further diagnostic workup for procedural

interventions. In patients with persistent pain that is unresponsive to conservative management, MRI may be considered.

Plain radiographs also offer some clinical utility, though they are more useful in working up suspected structural abnormalities such as scoliosis, spondylolisthesis, and fractures. Electrodiagnostic testing, such as electromyography and nerve conduction studies, can be considered in patients who present with equivocal symptoms or imaging findings or in cases where peripheral neuropathy should be ruled out. Selective nerve root blocks are also a diagnostic tool that has been used to identify the involved nerve root, select patients for surgery, and improve surgical outcomes.

32.6 Treatment

Treatment options for patients with cervical spinal pain range from conservative to operative, but the majority of patients respond to conservative therapy alone. Specifically, conservative treatment may consist of strengthening exercises and cervical and scapular stretching, which has been proven to provide intermediate relief for mechanical neck pain due to endorphin secretion improving sleep and mood while reversing and/or preventing deconditioning. Alternative and integrative medicine options, such as acupuncture, spinal manipulation, massage, yoga, and meditation, have been demonstrated to be superior to no treatment, but evidence is weak.

Pharmacologic therapy consisting primarily of systemic nonsteroidal anti-inflammatory drugs (NSAIDs) and acetaminophen has been proven beneficial in patients with cervical spinal pain. Muscle relaxants, especially at intermediate or high doses between 15 mg and 30 mg/day, may also provide relief, although they are usually more beneficial for acute rather than chronic pain. Benzodiazepines, by providing a similar muscle relaxant property, can also be effective; however, due to their potential for abuse and lack of superiority over other muscle relaxants, benzodiazepines should only be used when other muscle relaxants have failed and in the presence of clearly defined goals, time frames, and surveillance.

Injections are an effective treatment modality, especially for neuropathic cervical spinal pain. Epidural corticosteroid injections with or without local anesthetic have been shown to have lasting benefits up to 1 year. However, studies have shown that a combination treatment of epidural corticosteroid injections plus conservative management such as adjuvants and physical therapy is superior to either treatment alone.

Treatment of mechanical cervical spinal pain with injections, however, has demonstrated mixed evidence. Trigger-point injections of local anesthetic or botulinum toxin, for example, provide relief in patients with mechanical neck pain secondary to myofascial involvement, but some studies report no difference in patient symptoms compared to saline or dry needle injections. However, the efficacy of corticosteroid and/or anesthetic injections is limited in patients with mechanical neck pain caused by the cervical facet joints. Instead, radiofrequency ablation of the medial branch (facet joint nerve) has proven to have a weakly positive effect in both pain relief and functional improvement.

When conservative treatment options have failed, surgery may be considered. In a randomized study comparing anterior decompression and fusion (ACDF), physical therapy, and hard collar immobilization for patients with cervical radiculopathy, the surgical group experienced significantly greater improvements in muscle strength and sensory loss and greater reductions in pain than the other treatment groups. More recently, cervical disk replacement or cervical disk arthroplasty (CDA) has been suggested as an alternative to ACDF. Compared to ACDF, CDA has demonstrated to have greater reductions in neck disability, greater patient satisfaction, and fewer complications and reoperation rates. Additionally, patients with good functional preoperative capacity, nonsmoking status, and patients of the male sex are associated with greater long-term treatment results and likelihood of success.

For disk herniations, patients with neck and/or arm radiculopathy also experience significant improvements in both pain and function after plasma disk decompression for up to 1 year after surgery. However, some studies have demon-

strated that while surgery may have superior outcomes compared to physical therapy at 1 year after surgery, the difference between groups by 2 years is no longer statistically significant.

Treatment of cervical spinal pain is continuing to advance. Emerging treatment modalities that have demonstrated promising evidence in chronic pain conditions include biological therapies, such as stem cell therapy, platelet-rich plasma, and nerve growth factor inhibitors; however, these therapies are yet to be studied in patients with cervical spinal pain. Though not yet studied in patients with neck pain, clinical trials have demonstrated that ketamine is beneficial in treating patients with neuropathic pain and pain secondary to central sensitization like fibromyalgia.

Take-Home Message

- Cervical spinal pain is a common musculoskeletal disorder and a leading cause of disability worldwide.
- Providers should be cognizant of common red flags that necessitate further diagnostic imaging and potentially operative management. Such red flags include infection, neoplasm, pathologic fractures, systemic inflammatory diseases, cervical myelopathy, history of previous neck surgery, or other primary neurologic conditions.
- Correctly diagnosing patients' cervical spinal pain is crucial, as it defines what treatment modalities will be most efficacious.
- Depending on the type of cervical spinal pain, numerous treatment options have proven effective, from conservative therapy such as physical therapy and NSAIDs to surgical treatments.

Summary

Cervical spinal pain is a common, leading cause of disability worldwide that affects approximately half of all adults at least once in their lifetime. The etiology of neck pain is multimodal, and while the majority of cases are secondary to acute trauma or chronic degenerative conditions, nonorganic etiologies including psychosocial factors contribute to the development of cervical spinal pain. Cervical spinal pain, amongst other classification definitions, is commonly categorized as either mechanical or neuropathic pain. Because treatment for mechanical and neuropathic cervical spinal pain differs, diagnosing the type of neck pain correctly is crucial. When treating cervical spinal pain, providers should be aware of the various modalities that help treat neck pain, both conservative and invasive, and which treatment options are effective or ineffective for the two types of neck pain.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. What is the annual prevalence of neck pain?
   (a) The reported rate in most studies ranges from 15% to 50%
   (b) The reported rate in most studies ranges from 5% to 15%
   (c) The reported rate in most studies ranges from 15% to 50%
   (d) The reported rate in most studies ranges from 5% to 15%
2. Is pain located in the anterior aspect of the neck considered cervical spinal pain?
   (a) No, cervical spinal pain is located in the posterior region of the cervical spine
   (b) Yes, cervical spinal pain could be anterior or posterior, depending on the cause
   (c) No, cervical spinal pain is located in the postero-lateral region of the cervical spine
   (d) Yes, cervical pain could be anterior, lateral or posterior, depending on the cause
3. Which occupations are reported to have a higher incidence of neck pain?
   (a) Manual workers, healthcare personnel, and office workers
   (b) Football players and employers
   (c) Spine surgeons
   (d) Computer programmers and dentists

4. What is the best predictor of the outcome of neck pain?
   (a) Pain duration. The shorter the duration, the better the prognosis
   (b) Type of pain. Anterior pain has a worst prognosis compared to posterior pain
   (c) Psychological factors. High levels of stress negatively impact the prognosis
   (d) Only depression could affect the outcomes
5. What is the most sensitive imaging modality for detecting soft-tissue abnormalities?
   (a) Magnetic resonance imaging (MRI)
   (b) Computed tomography (CT)
   (c) X-Rays
   (d) Ultrasounds

Further Reading

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Cervical Spondylotic Myelopathy

33

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Overview

Cervical spondylotic myelopathy (CSM) is a disorder with an increasing prevalence. It is a degenerative spine disease leading to compression of the spinal cord. Degenerative changes include bone spurs (osteophytes), disc bulges, and thickened ligaments. Cervical spondylosis is responsible for more than half of cervical myelopathy cases and is the most common cause of spinal cord dysfunction in patients over 55 years of age. CSM causes progressive disability and impairs the quality of life. A complete recovery is rare after it sets in.

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33.1 Definition

Cervical spondylotic myelopathy (CSM) is an impaired function of the spinal cord caused by degenerative changes in the cervical spine that results in compression of the spinal cord. Cervical myelopathy typically presents generally in the sixth decade of life with symptoms such as weakness, numbness, tingling, or, rarely, pain in the arms, hands, and legs. Delayed diagnosis is common due to the lack of pain and the wide array of bizarre symptoms observed in CSM.

33.2 Epidemiology

CSM mostly involves patients above 55 years old. The age of onset can vary depending on the degree of congenital spinal canal narrowing. Radiographic evidence of cervical spondylosis can be found in more than 50% of middle-aged people, but myelopathy occurs only in 5–10% of cases. CSM is the main cause of spasticity acquired in the aged population. Given the increased life expectancy, the incidence of CSM is likely to increase.

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33.3 Etiology/Pathogenesis

Three pathophysiologic factors contribute to the development of CSM:

1. Static mechanical (reduction of spinal canal diameter)
2. Dynamic mechanical (flexion and extension of the neck)
3. Spinal cord ischemia

The process that leads to spinal cord compression is a result of arthritis in the neck (also called spondylosis). The degenerative changes that occur in the spine are the result of the aging process that involves joints, intervertebral discs, ligaments, and connective tissue of the cervical vertebrae.

Cervical spondylosis is a slow and progressive phenomenon. Early disc degeneration typically precedes major bone changes by 10 years. Tears of the intervertebral disc are evident from the second decade of life and increase over time (Fig. 33.1). The resulting loss of disc height alters the mechanics of the facets, placing greater stress on the articular cartilage of the vertebrae and their respective end plates. Osteophytic spurs develop at the margins of end plates obstructing the transverse foramen rather than the central canal (Fig. 33.2). Osteophytes increase the weight-bearing surface of the end plates and, therefore, decrease the effective force being placed on them, but also cause a reduction in the joint range. In addition, with aging, the ligamentum flavum may stiffen and buckle into the spinal cord dorsally, further reducing the spinal canal (Fig. 33.3).

Flexion and extension of the neck may exacerbate compression. During flexion, the spinal cord

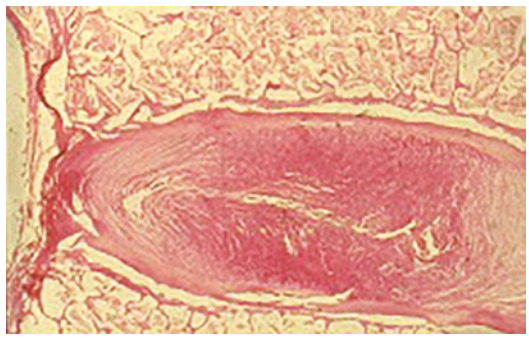

Fig. 33.1 Early disc degeneration consists of tears with different orientation

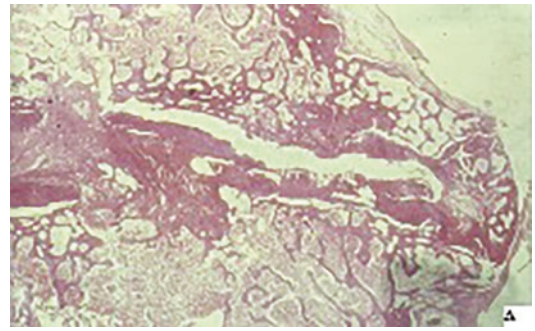

Fig. 33.2 Osteophytic spurs

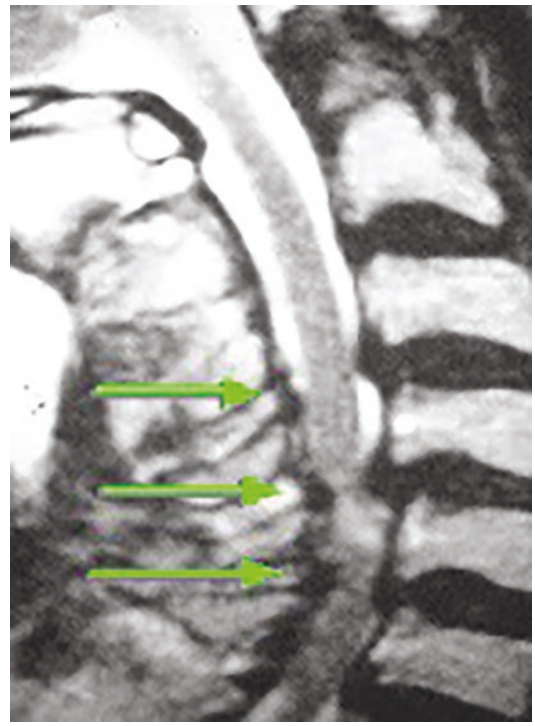

Fig. 33.3 The ligamentum flavum may stiffen and buckle into the spinal cord dorsally

elongates and sprawls over ventral osteophytes. During extension, the available space for the spinal cord is reduced by the inward buckling of the ligamentum flavum.

The role of spinal cord ischemia in the pathophysiology of CSM is not well understood. Histopathological evidences show an involvement of the gray matter, with minimal involvement of the white matter. This pattern corresponds to an ischemic insult rather than to a direct mechanical compression. Moreover, symptoms often extend cranially to the compressed level. Vascular impairment can originate from compression of the anterior medullary artery, segmental medullary artery in the foramen, or distortion of the sulcus arteria vertebralis.

Clinical presentation of cervical spondylosis can be characterized by neck pain, radiculopathy, myelopathy, or a combination of the three in relation to the compressed area.

33.4 Diagnosis (Clinical and Imaging)

The diagnosis of CSM is based on the patient's history and clinical examination. Imaging and electrophysiology should support and confirm the diagnosis. Focus on MRI or CT before considering the signs and symptoms of the patient is one of the main causes of diagnostic errors.

Regarding patient history, many patients are asymptomatic at first. Symptoms are believed to arise when the spinal cord has been reduced by at least 30%. Symptoms often develop insidiously, and periods of stability alternate with periods of worsening.

Clinically, the most characteristic symptoms of the CSM are the instability of gait, loss of fine motor control of the upper limbs, numbness in the hands, weakness of the hands and legs, neck pain and stiffness, and urinary emergency.

The vague nature of early myelopathic symptoms, such as weakness in the lower limbs, or walking impairment, is often responsible for the diagnostic delay.

The physical examination aims to identify the physical signs of disease. Neurological examination is crucial in the diagnostic process because several pathologies can have a similar presentation.

Physical examination should start from the head, with the evaluation of the cranial nerves and cervical range of motion, and should include the neuromuscular examination of the upper and lower limbs. It is also important to evaluate walking and balance, especially in the dark since visual information is less effective in replacing the proprioceptive information of the feet.

Typically, the following signs can be found: limitation of range of motion; spasticity, with increased reflexes below the compromised level; positive Babinski sign; absence of abdominal reflexes; reduction of proprioceptive and vibratory sensitivity; and abnormal walking. Attention should be paid to concomitant lumbar stenosis, which can be present in 15–33% of patients with cervical spondylotic myelopathy, resulting in lower limb hyporeflexia.

Ranges of motion are measured in flexion, extension, lateral bending, and rotation in flexion and neutral position. With aging, all ranges of motion are reduced except for rotation in flexion, as in this position, the main contribution comes from the atlantoaxial joint that is less commonly affected by degenerative changes. Thus, a significant reduction of rotation in flexion compared to neutral position suggests the spondylotic involvement of the atlantoaxial joint; conversely, similar ranges in flexion and in neutral position indicate spondylosis of the middle and lower cervical spine. Significant spondylosis of facet joints limits extension more than flexion and limits lateral bending in the direction of the most significant degenerative changes.

Radicular compression produces a dermatomal distribution of pain, reproducible with cervical extension, lateral banding, and axial compression (Spurling maneuver), and can be resolved by plac-

ing the ipsilateral hand on the head (shoulder abduction maneuver). Myelopathy produces an electric shock along the back to the extremities triggered by cervical flexion (Lhermitte's sign).

The examination of strength, sensitivity, and reflexes allows differentiating radicular problem from central and systemic problems. In addition to tactile and pain sensitivity, vibratory sensitivity can be strained to assess the function of the posterior columns and any overlapping peripheral neuropathy.

Additional diagnostic studies, such as radiographs, magnetic resonance imaging (MRI), computed tomography (CT), bone scan, electromyography (EMG), somatosensory evoked potentials (SEPs), and motor evoked potentials (MEPs), can help to provide further information useful for the management of the patient.

Patients with neurological disorders, over the age of 50, history of neck trauma, or failure of conservative treatment should undergo radiographic study. The cervical spine must be evaluated for alignment, congenital abnormalities, arthritis, fractures, subluxations, lytic lesions, fractures, and osteoporosis. The anteroposterior projection allows identifying cervical ribs and scoliotic deformities. It allows good visualization of the uncovertebral joints (joints of Luschka). The lateral projection highlights reduction of disc space, osteophytes, sagittal alignment, diameter of the medullary canal, and ossification of the posterior longitudinal ligament. Lateral flexion/extension views rule out translational instability of the cervical vertebral bodies, which often occurs at adjacent levels to the more spondylotic and hypomobile segments. The diameter of the medullary canal in dynamic films is usually smaller than in static conditions. Oblique views may be required to visualize foraminal narrowing.

Although degenerative changes in the cervical spine are evident in patients with myelopathy compared to those without myelopathy, the correlation of these abnormalities with clinical deficits is less clear. The percentage of imaging anomalies in the asymptomatic population ranges from 25% to 50% at 50 years and from 75% to 85% at 65 years.

Persistent or progressive neurological deficits or signs of myelopathy require further instrumental investigation. Magnetic resonance imaging (MRI) and computed tomography (CT) can identify soft tissues, such as herniated discs, hypertrophy, and protrusion of the yellow ligament, which can contribute to medullary or nerve root compression.

MRI is useful to detect signal changes due to myelomalacia and allows evaluating suspected spinal stenosis, congenital anomalies (i.e., Chiari malformations), syringomyelia, spinal cord neoplasm, multiple sclerosis, and disc degeneration.

The exam should be performed at least with two pulse sequences, a combination of T1-, T2-, and STIR-weighted sequences. T1-weighted images allow a contrast evaluation of bone marrow and extradural soft-tissue structures. T2-weighted images focalize on intramedullary pathology, providing a myelographic-like image of epidural impressions owing to degenerative changes. STIR-weighted images detect cord pathologies and highlight soft-tissue edema and infiltrative processes. Paramagnetic contrast media helps to diagnose intramedullary disease and, in the postoperative spine, to detect epidural inflammation and fibrosis. MR myelography can be useful for the evaluation of nerve root compression and spinal stenosis.

CT is complementary to MRI. It is able to rapidly assess specific details of osseous injuries, spinal stenosis, and osteophytes. It may aid in the determination of the anatomy associated with nerve root and spinal cord compression.

If the history, physical examination, and imaging are not well correlated, another pathology should be suspected, and other studies should be undertaken. Neurophysiological tests are useful

supplementary tests for the evaluation of cervical myelopathy.

Electrodiagnostic studies include needle EMG, electroneurography, and evoked potentials (EPs). They allow distinguishing a lesion in the periphery from a nerve root lesion, to differentiate normal conditions from a diffuse polyneuropathy, focal entrapment neuropathy, radiculopathy, myelopathy, myopathy, and disorder of the neuromuscular junction. They should be considered as an extension of the neurologic examination.

Needle EMG records the electrical activity of the muscle cell membrane. EMG can be useful in any suspected peripheral nerve injury, peripheral neuropathy, radiculopathy, localized entrapment, or disease of the motor unit potential.

Somatosensory evoked potentials (SSEPs) or motor evoked potentials (MEPs) provide a measure of the electrical conductivity of the spinal cord across the compressed segments. Such testing may also be performed as a baseline in anticipation of monitoring of the spinal cord during surgery itself with the same studies.

SEP studies are particularly valuable in case of suspected multiple sclerosis, Lyme disease, systemic lupus erythematosus, neurosyphilis, spinocerebellar degenerations, familial spastic paraplegia, and deficiency of vitamin E or B12, among other disorders.

The differential diagnosis includes any condition associated with neck pain, arm pain, motor-sensory-reflex changes, and signs of spinal cord dysfunction such as hypertrophic cervical dural meningitis, syringomyelia, amyotrophic lateral sclerosis, spinal muscle atrophy, extramedullary tumors, intramedullary tumors, multiple sclerosis (particularly, the spinal progressive type), nerve entrapment syndromes (carpal tunnel and thoracic outlet), spastic spinal paraparesis, neuralgic amyotrophy (Parsonage–Turner syndrome), funicular myelosis, paraneoplastic myelopathy, acute traumatic myelopathy (centro-medullary syndrome), Friedreich syndrome, posterior cervicosympathetic syndrome, supraspinal (central) pathology, cerebral tumors, cortical atrophy, and psychoneurosis (“the great imitator”).

33.5 Treatment

The choice of treatment should take into consideration patient’s symptoms, comorbidities, and expectations.

Conservative treatment is considered in the absence of clinical and instrumental deficit. The aim is to decrease pain and improve patient’s function and ability to perform daily activities.

Treatment consists of analgesic such as steroidal and/or nonsteroidal anti-inflammatory medications to reduce spinal cord and nerve root inflammation, or codeine and paracetamol. Additionally, low dose of antidepressants (amitriptyline) and antiepileptics can be used to manage pain intensity.

Muscle relaxants, peripheral or central (diazepines), can be used as treatment adjuvants, as contracture is often the main cause of acute neck pain in cervical spondylosis.

Temporary immobilization of the neck, using a semirigid Schanz collar with proximal occipito-mandibular support, can be considered for patients with spondylotic cervical pain without medullary or radicular compromise. It limits joint excursion and promotes muscle relaxation, relieving pain of contracture. In case of myeloradiculopathy, a collar can worse the pain as it prevents the patient from assuming an analgesic position suitable to relieve root compression.

Manipulations of the cervical spine (Hippocratic manipulations) must be reserved exclusively for early cervical spondylosis without involvement of spinal cord or roots. They can

cause serious neurological damage in case of improper and/or violent execution. For this reason, physiotherapeutic or osteopathic treatment of patients with spondylotic neck pain must be preceded by a complete orthopedic and radiological evaluation, which has excluded compression of the spinal cord. In the case of exclusive involvement of the vertebral joints, cervical traction can be beneficial, as it opens the disc space, widens the foramina, stretches the posterior longitudinal ligament, and extends contracted muscles.

The natural history of CSM is variable, and a precise prognosis cannot be defined. Once moderate signs and symptoms have developed, spontaneous improvement is unlikely to occur, and the patient will benefit from surgical treatment.

Surgical treatment is performed when symptoms fail to improve after 4–6 weeks of nonsurgical management, or progress in spite of nonsurgical treatment. The indication is rigorous and proceeded by a complete preoperative evaluation. The outcomes are influenced by the duration and worsening of symptoms and concomitant systemic metabolic (diabetes, dyslipidemia) or vascular (vasculopathy, hypertension) or organ pathologies (liver, kidney, heart). In severe spondylotic myelopathy, a chronic damage is established and removing the cause of stenosis can only limit the progression. Outcomes are better if surgery is performed when first neurological signs appear.

Surgery consists of spinal cord decompression and aims to directly remove the cause of compression and stenosis. The approach can be anterior or posterior, depending on the location of the stenosis elements.

Indications to posterior approach can be precisely defined, such as in case of spinal cord compression caused by posterior osteophytes, posterolateral (uncal) osteophytes, calcification of the posterior longitudinal ligament, hypertrophy and thickening of the ligamentum flavum, and malformation of the pedicles or laminae. In patients with a segmental compression (only one level), internal fixation is mandatory to eliminate any residual motion and avoids further medullary damage.

An anterior approach (Cloward instrumentation with or without multiple corpectomies) is preferred in case of anterior osteophytes protruding onto the anterior surface of the cord (Fig. 33.4). In such instances, it is inappropriate to limit surgery to a laminectomy because it only indirectly addresses the cause of compression, with relevant risks of failure or worsening. Instrumentation (plates and screws) can be used to provide immediate internal support for the cervical spine and to promote bone graft healing.

Multilevel stenosis (4–5 levels) can be managed with either an anterior or a posterior approach, aiming to decompress the spinal cord and restore the normal sagittal alignment. However, anterior decompression by

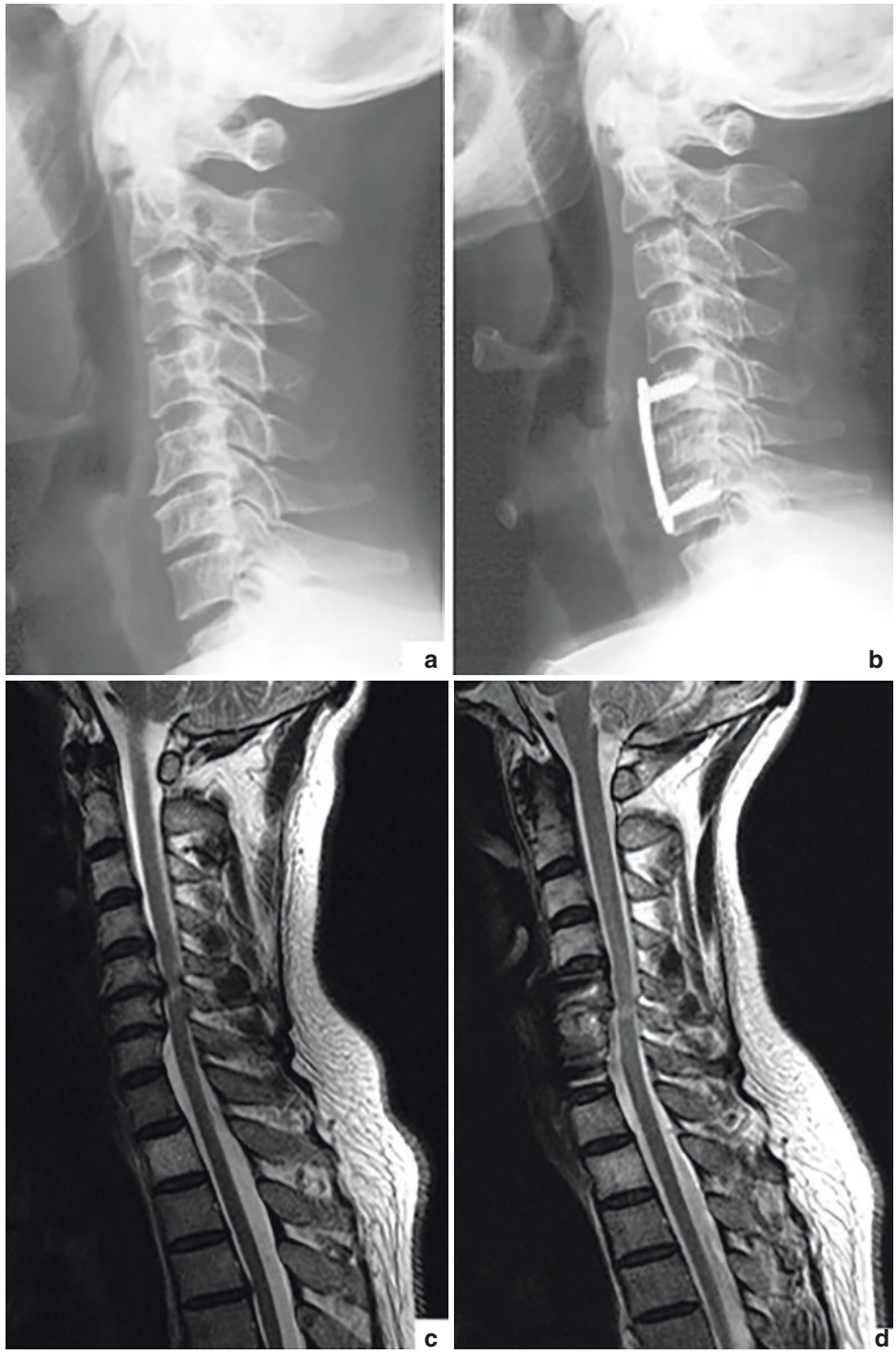

Fig. 33.4 Patient M, 47 years old with cervical spondylotic myelopathy, underwent anterior discectomy C5–C6 and C6–C7 and subtotal somatectomy C6, arthrodesis with transplant taken from the iliac crest, and stabilization with plates and screws. (**a**) Preoperative X-ray shows reduction of the disc space, osteophytes, and loss of cervical lordosis; (**b**) postoperative X-ray; (**c**) preoperative MRI shows anterior cord compression with myelopathy; (**d**) postoperative MRI shows the enlargement of vertebral canal after removal of the compression elements

multiple subtotal somatectomy can be too aggressive. It has been associated with a high incidence of cord injury, cerebrospinal fluid leakage, graft failure, pseudarthrosis, infection, and dysphagia. Posterior decompression by wide laminectomies (4–5 levels), associated with stabilization in lordosis of the operated segment, allows the circumferential decompression and the backshift of the spinal cord. The dorsal migration of the spinal cord clears ventral compression allowing for indirect decompression of the anterior spinal cord (Fig. 33.5). The fusion eliminates the movement of the operated segment (Fig. 33.6).

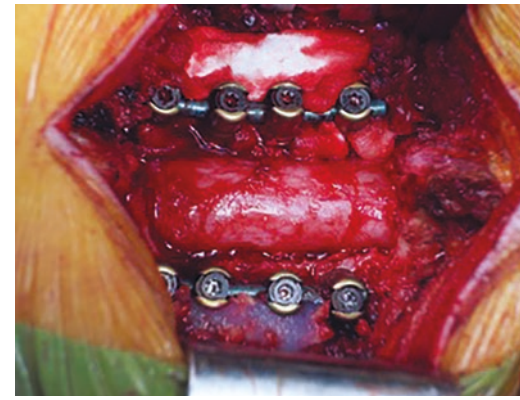

Fig. 33.5 The dorsal migration of the spinal cord clears ventral compression allowing for indirect decompression of the anterior spinal cord

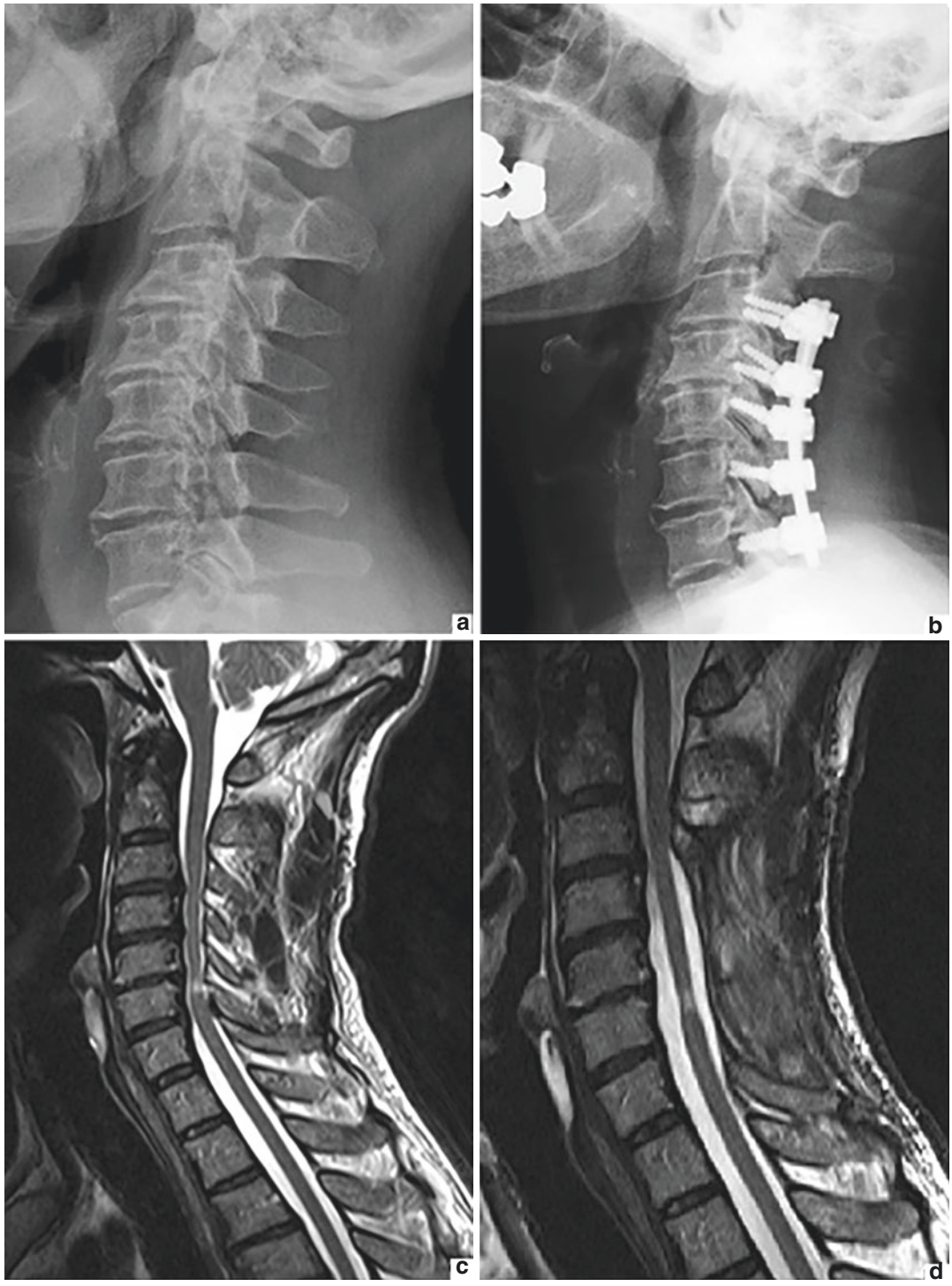

Fig. 33.6 Patient, 76 years old, F, with cervical spondylotic myelopathy, treated with laminectomy and C3–C7 stabilization with rods, screws, and autologous graft added with platelet gel. (**a**) Preoperative X-ray; (**b**) postoperative X-ray; (**c**) preoperative MRI; (**d**) postoperative MRI

Take-Home Message

- CSM is a degenerative spine disease leading to compression of the spinal cord.
- The process that leads to spinal cord compression is a result of arthritis in the neck (also called spondylosis).
- Cervical spondylosis is the most common cause of spinal cord dysfunction in patients over 55 years of age.
- The vague nature of early myelopathic symptoms, such as weakness in the lower limbs, or walking impairment, is often responsible for the diagnostic delay.
- Focus on MRI or CT before considering the signs and symptoms of the patient is one of the main causes of diagnostic errors.
- The percentage of imaging anomalies in the asymptomatic population ranges from 25% to 50% at 50 years and from 75% to 85% at 65 years.
- Electrodiagnostic studies allow distinguishing a lesion in the periphery from a nerve root lesion, to differentiate normal conditions from a diffuse polyneuropathy, focal entrapment neuropathy, radiculopathy, myelopathy, myopathy, and disorder of the neuromuscular junction.
- Surgical treatment is performed when symptoms fail to improve after 4–6 weeks of nonsurgical management, or progress in spite of nonsurgical treatment.
- Surgery consists of spinal cord decompression and aims to directly remove the cause of compression and stenosis.
- Indications to posterior approach can be precisely defined, such as in case of spinal cord compression caused by posterior osteophytes, posterolateral (uncal) osteophytes, calcification of the posterior longitudinal ligament, hypertrophy and thickening of the ligamentum flavum, and malformation of the pedicles or laminae.
- An anterior approach (Cloward instrumentation with or without multiple corpectomies) is preferred in case of anterior osteophytes protruding onto the anterior surface of the cord.
- For multilevel stenosis, posterior decompression by wide laminectomies, associated with stabilization in lordosis of the operated segment, allows the circumferential decompression and the backshift of the spinal cord.

Summary

Cervical spondylotic myelopathy (CSM) is an impaired function of the spinal cord caused by degenerative changes in the cervical spine that result in compression of the spinal cord. CSM is the main cause of spasticity acquired in the aged population.

The diagnosis of CSM is based on the patient's history and clinical examination. Imaging and electrophysiology should support and confirm the diagnosis.

The natural history of CSM is variable, and a precise prognosis cannot be defined. Once moderate signs and symptoms have developed, spontaneous improvement is unlikely to occur, and the patient will benefit from surgical treatment.

Surgery consists of spinal cord decompression and aims to directly remove the cause of compression and stenosis. The approach can be anterior or posterior, depending on the location of the stenosis elements. Multilevel stenosis can be managed by posterior decompression by wide laminectomies, associated with stabilization in lordosis of the operated segment that allows the backshift of the spinal cord for indirect decompression.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. When does CSM typically present and what are the most common symptoms?
   (a) CSM typically presents insidiously in the sixth decade of life with symptoms

such as weakness, numbness, tingling, or, rarely, pain in the arms, hands, and legs.

(b) CSM typically presents insidiously in the third decade of life with symptoms such as weakness, numbness, tingling, or, rarely, pain in the arms, hands, and legs.

(c) CSM typically presents insidiously in the fifth decade of life with symptoms such as weakness, numbness, tingling, or, rarely, pain in the arms, hands, and legs.

(d) CSM typically presents acutely in the fourth decade of life with symptoms such as stiffness, tremors, and muscle spasms in the limbs.

2. What are the pathophysiologic factors that contribute to the development of CSM?
   (a) Static mechanical (reduction of spinal canal diameter); dynamic mechanical (flexion and extension of the neck); spinal cord ischemia
   (b) Static mechanical (reduction of spinal canal diameter); infections; hereditary factors
   (c) Dynamic mechanical (excessive neck flexion and extension); spinal cord inflammation; hormonal imbalances
   (d) Genetic mutations; metabolic disorders; autoimmune reactions
3. What are electrodiagnostic studies used for?
   (a) Electrodiagnostic studies allow distinguishing a lesion in the periphery from a nerve root lesion, to differentiate normal conditions from a diffuse polyneuropathy, focal entrapment neuropathy, radiculopathy, myelopathy, myopathy, and disorder of the neuromuscular junction.
   (b) It is used to make diagnosis of myelopathy.
   (c) It is used to monitor the progression of CSM over time.
   (d) Electrodiagnostic studies help assess psychological factors related to CSM.
4. When is surgery indicated?
   (a) Surgical treatment is performed when symptoms fail to improve after 4–6 weeks of nonsurgical management, or progress in spite of nonsurgical treatment.
   (b) Surgical treatment is performed when symptoms fail to improve after 1–2 weeks of nonsurgical management, or progress in spite of nonsurgical treatment.
   (c) Surgical treatment is performed when symptoms remain unchanged after 3-4 weeks of nonsurgical management.
   (d) Surgical treatment is considered immediately upon diagnosis of CSM.
5. When is anterior approach used?
   (a) In case of anterior osteophytes protruding onto the anterior surface of the cord
   (b) In case of posterior compression of the spinal canal
   (c) In case of lateral displacement of the spinal cord due to disc herniation.
   (d) In case of thoracic spine involvement with CSM.

Further Reading

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Cervical Spine Tumours

34

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Overview

This chapter focuses on the multimodality imaging findings of some of the commonly encountered tumours, which involve the bony elements of the cervical spine. Neurogenic tumours and soft-tissue tumours adjacent to the spine are not discussed as they are beyond the scope of this chapter.

34.1 Introduction

Metastatic disease, myeloma and lymphoproliferative disorders commonly present in adults as multiple lesions, and on seeing multiple lesions, the diagnosis is most likely to be one of these three disorders. If the lesion is solitary, a primary bone tumour must also be considered part of the differential diagnosis. The World Health Organization (WHO) has classified primary tumours on the basis of their cell type (Table 34.1).

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Table 34.1 Classification of primary bone tumours of the spine modified from the WHO classification of bone and soft-tissue tumours, 2020

Tumour type	Benign	Malignant
Osteogenic	Enostosis (bone island) Osteoid osteoma Osteoblastoma	Osteosarcoma
Chondrogenic	Osteochondroma Chondroblastoma	Chondrosarcoma (primary or secondary)
Haematological/vascular	Haemangioma	Haemangioendothelioma Haemangiopericytoma Plasmacytoma/myeloma Lymphoma
Undifferentiated small round-cell tumours		Ewing's sarcoma
Osteoclastic giant cell-rich tumour	Aneurysmal bone cyst Giant-cell tumour locally aggressive type	Malignant giant cell tumour
Notochordal	Benign notochord rest	Chordoma

34.2 Imaging Techniques

34.2.1 Radiographs

Radiographs are frequently the first imaging modality obtained in the evaluation of a patient presenting with neck pain, following trauma or for the evaluation of arthritis; however, their usefulness when assessing for tumour infiltration is limited by the fact that tumour detection by radiographs requires 30–50% of bone destruction as a minimum to be detected. Approximately 40% of lesions will not be seen on radiographs resulting in a false-negative result.

34.2.2 Computed Tomography

Multi-detector computed tomography (CT) can accurately demonstrate the degree of cancellous and cortical bone destruction. CT can recognise a destructive bony lesion up to 6 months earlier than on radiographs.

34.2.3 Magnetic Resonance Imaging

Magnetic resonance imaging (MRI) is the only imaging modality that allows accurate assessment of the bone marrow. Its superior soft-tissue resolution also allows soft-tissue extension of the tumour to be identified and to determine if there is any resulting spinal cord or nerve root compression. Standard sequences include sagittal and axial T1- and T2-weighted spin echo sequences. On T1-weighted sequences, marrow infiltration by tumour shows up as a loss of the normal fatty marrow signal giving a low signal rather than the bright signal from the marrow. On T2-weighted sequences, tumours may be seen as hyperintense lesions if they have relatively higher water content. Fat-supressed sequences such as fat-saturated T2-weighted or a short tau inversion recovery (STIR) can increase the conspicuity of such lesions compared to the T2-weighted sequences.

34.3 Osteogenic Tumours

34.3.1 Enostosis

Enostosis (bone island) is a benign osteogenic tumour. It is a focus of dense cortical bone within cancellous bone. They can be identified on radiographs, but CT is the imaging modality of choice. CT demonstrates a densely sclerotic lesion which has a characteristic 'brush border', which is fine projections at the periphery of the lesion which merge with the adjacent bone. They can vary in

size, but when 2 cm or greater in size, they are termed giant bone islands. While they are fairly characteristic, the differential diagnosis includes a sclerotic metastasis usually from breast or prostate carcinoma. CT is especially useful as precise attenuation (density) measurements can be performed. Due to the densely sclerotic nature of these lesions, they are characteristically of low signal intensity on both T1- and T2-weighted sequences. They may show increased activity on bone scintigraphy in up to 10% of cases. Most lesions remain stable, but up to 31% of lesions will show an increase in size.

34.3.2 Osteoid Osteoma

Osteoid osteoma is a benign osteoid-forming tumour. They usually occur in children and young adults commonly between the second and third decades of life. They can occur anywhere in the body with 10% of tumours located in the spine, mostly in the posterior elements. They are most common the lumbar spine (60%) followed by the cervical spine (27%), then the thoracic spine (12%) and the sacrum (2%). They usually present with pain which is classically at night and is relieved with salicylates. It is also associated with a scoliosis and is the most common cause for a painful scoliosis in children. The tumour is referred to as the 'nidus', which has a variable amount of surrounding reactive sclerosis. By definition, the nidus of an osteoid osteoma is less than 1.5 cm. If the nidus exceeds this, then the tumour is classified as an osteoblastoma. The nidus can have a variable amount of mineralisation.

Radiographs may show the surrounding reactive sclerosis, but the complex cervical spine anatomy can make the nidus difficult to see. A scoliosis may be present, classically with the osteoid osteoma at the concavity of the curve. CT is the imaging investigation of choice, which shows the nidus and the reactive sclerosis and allows it to be differentiated from a stress fracture or pars defect (Fig. 34.1). On MRI, the nidus can have a variable appearance due to the degree of mineralisation within it. On T1-weighted images, it is usually intermediate to low signal intensity and usually intermediate to high signal intensity on fluid-sensitive sequences. The reactive zone of the tumour extends over a much larger area than

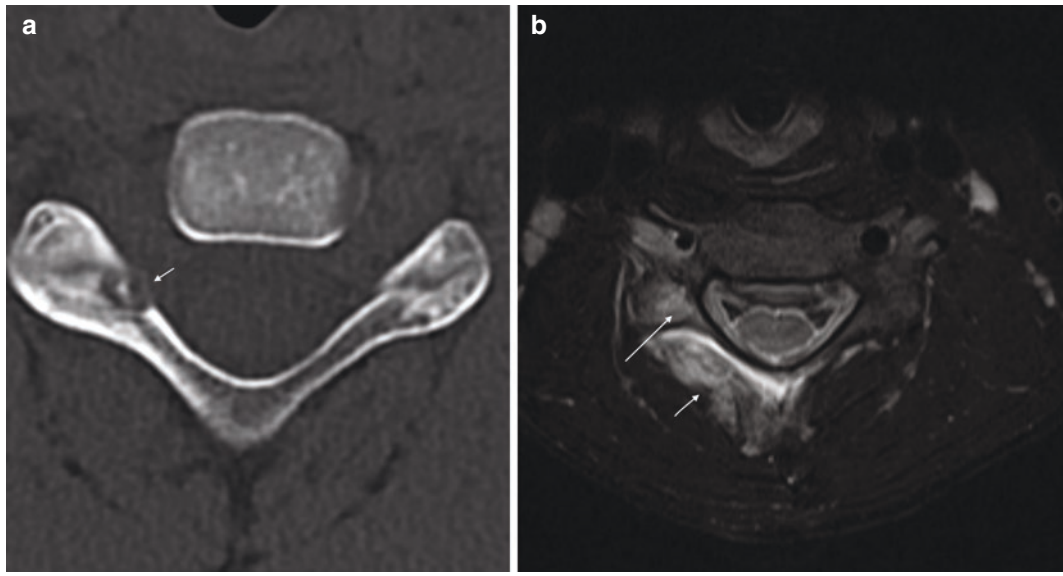

Fig. 34.1 Osteoid osteoma. (**a**) Axial CT image shows nidus of osteoid osteoma in the right lamina of C4 (white arrow). (**b**) Axial STIR image shows intraosseous oedema in the posterior elements of C4 (long white arrow) and inflammatory change in the paravertebral soft tissues (short white arrow)

the tumour and can extend into the paravertebral soft tissues and even adjacent vertebral levels. It is usually low signal on the T1-weighted images and high signal on the fluid-sensitive sequences (Fig. 34.1). Traditionally, the treatment of osteoid osteoma was surgical—by removing the nidus of the osteoid osteoma with or without stabilisation. Increasingly, percutaneous ablation of the nidus is being performed and has been shown to be safe in the cervical spine.

34.3.3 Osteoblastoma

Osteoblastoma is a benign osteoid-forming tumour. It is differentiated from osteoid osteoma macroscopically by its larger size (>1.5 cm). 40% of osteoblastomas occur in the spine. Most occur in the cervical spine (40%) followed by lumbar (25%), thoracic (20%) and sacrum (15%). They preferentially occur in the posterior elements; however, in the cervical spine, they can more frequently involve the vertebral body than other locations. There is a male-to-female ratio of 2:1, and they occur most frequently in the second to third decades. There are two subtypes, conventional osteoblastoma and a more aggressive type of osteoblastoma. The more aggressive variant is commonly larger with an increased area of bony lysis and can also extend into the epidural and paravertebral soft tissues. Radiographic appearances can be variable. They are commonly osteolytic with a variable amount of matrix mineralisation. A sclerotic or lytic appearance of the pedicle, expansion of the transverse process and an associated scoliosis are also features. CT is the imaging modality of choice. Lesions can look identical to an osteoid osteoma but with a larger nidus (Fig. 34.2); however, they can be osteolytic with a thinned sclerotic rim. There is a variable amount of mineralisation in the lesion. An expansile appearance, extension into the vertebral body and paravertebral/epidural soft-tissue extension are suggestive of the more aggressive subtype. MRI demonstrates an intermediate to low signal intensity on T1-weighted sequences and low to high signal intensity on fluid-sensitive sequences depending on the degree of mineralisation in the lesion. Peritumoral oedema, especially in the more aggressive subtype, can extend to the paravertebral soft tissue and can extend to adjacent vertebrae. MRI is the modality of choice for the demonstration of a soft-tissue mass. The more aggressive subtype (Fig. 34.3) can be confused with tumours such as osteosarcoma, Ewing's sarcoma and lymphoma. Treatment options include radical surgery, surgical curettage or percutaneous ablation.

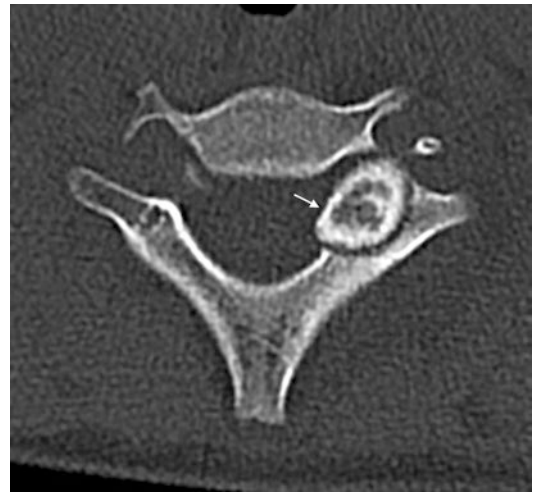

Fig. 34.2 Conventional osteoblastoma. (**a**) Axial CT shows mineralised nidus >2 cm in keeping with an osteoblastoma (white arrow)

34.3.4 Osteosarcoma

Osteosarcoma of the spine accounts for only 4% of all osteosarcomas. The cervical spine is the least common location of spinal osteosarcoma. In contrast to appendicular osteosarcoma which usually affects children and young adults, spinal osteosarcoma has a peak prevalence in the fourth decade. Secondary osteosarcoma arising in Paget's disease or following radiotherapy is observed in the elderly. Most cases involve the posterior elements but can extend into the body. Adjacent-level involvement is seen in 17%. Appearances on CT scanning can be variable depending on the degree of osteoid formation ranging from an ivory vertebra to a completely osteolytic appearance. There is usually a wide

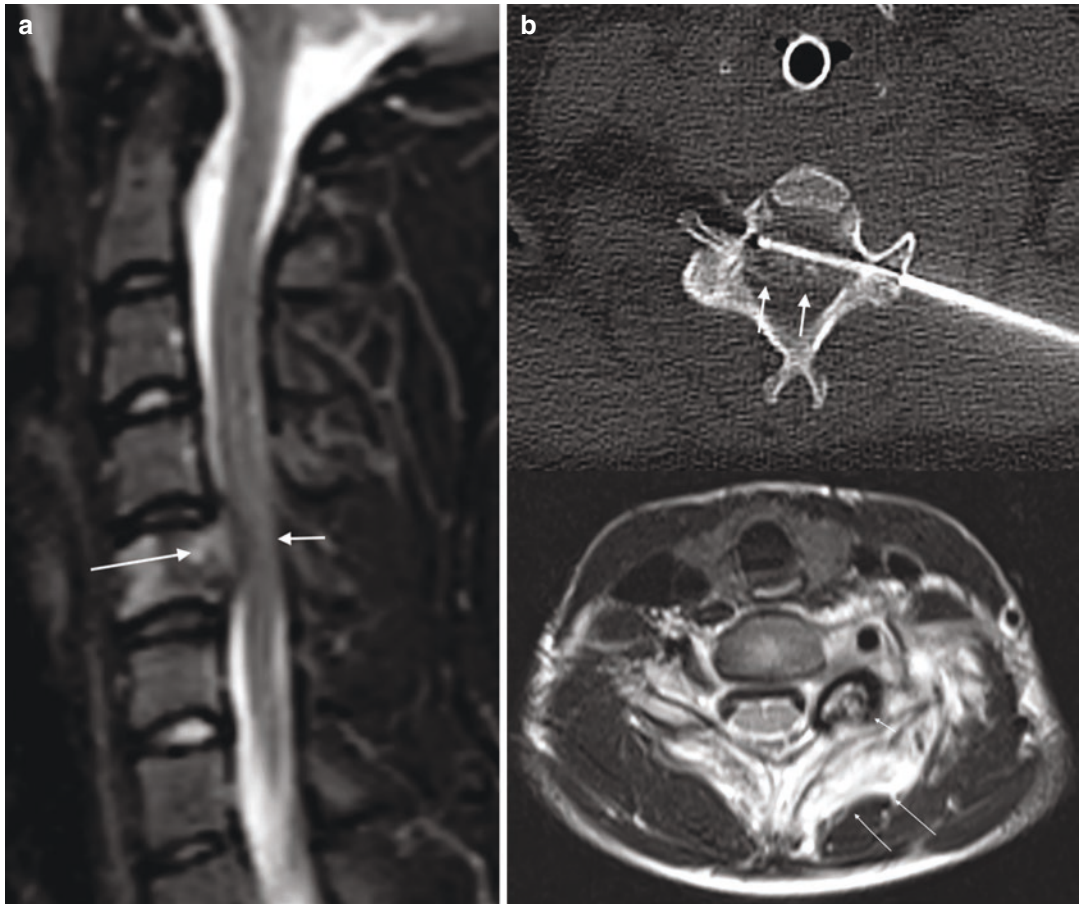

Fig. 34.3 Aggressive osteoblastoma. (**a**) Sagittal STIR MRI shows lesion in the body of C5 (long white arrow) with perilesional oedema and bowing of posterior cortex and cord compression (short white arrow), which is suggestive of an aggressive lesion. (**b**) Axial CT at the time of percutaneous radiofrequency ablation shows an expansile lytic lesion with a thin sclerotic rim (white arrows) and central cord narrowing. Note how this lesion looks more aggressive than the conventional type of osteoblastoma

zone of transition between the normal and abnormal bone and cortical destruction. The presence of a soft-tissue mass which contains immature osteoid is also commonly seen.

On MRI, there is an aggressive mass lesion involving the vertebrae. Signal characteristics depend on the degree of osteoid production of the tumour. Lesions which are densely ossified will be of low signal intensity on T1- and T2-weighted sequences. Lytic tumours are of low signal intensity on T1-weighted sequences and can be hyperintense on fluid-sensitive sequences. The telangiectatic subgroup of osteosarcoma is associated with fluid-fluid levels, but nodular areas of solid tumour mass should make the distinction between this and a primary aneurysmal bone cyst (ABC) relatively straightforward. MRI also allows the visualisation of cortical breakthrough, the demonstration of a soft-tissue mass and its effect on the underlying cord. The differential diagnosis includes a sclerotic metastasis, osteoblastoma, chondrosarcoma or Ewing's sarcoma.

Osteoblastoma like osteosarcoma is a rare subtype of low-grade osteosarcoma, which can resemble an osteoblastoma on imaging. They represent 1.1% of all osteosarcomas. These lesions have an overlap of radiological features of both osteosarcomas and osteoblastomas, although

there may be the presence of some aggressive features such as permeative borders on MRI and CT. The presence of an extraosseous soft-tissue mass is not usually a feature. Both osteoblastomas and osteoblastoma-like osteosarcomas will show a variable amount of osteoid formation. The presence of host bone permeation and the lack of 'maturation' towards the edge of the tumour are features seen in osteoblastoma-like osteosarcoma. The distinction is important as osteoblastoma-like osteosarcoma will require en bloc resection and chemotherapy.

34.4 Chondrogenic Tumours

34.4.1 Osteochondroma

Osteochondroma is the most commonly encountered bone tumour; however, spinal osteochondromas are relatively rare, representing 1–5% of sporadic osteochondromas and 1–9% in patients with hereditary multiple exostoses (HME). The cervical spine is the most common site for osteochondromas with a predilection for the C2 level. Most arise from the spinous or transverse process but can arise anywhere in the vertebrae. The peak age is between 10 and 30 years, and there is a male-to-female ratio of 3:1. The diagnosis can be difficult to make on radiographs due to the degree of bony overlap; however, one may see a sessile or pedunculated mass with corticomedullary continuity with the parent bone. If the cartilage cap is extensively mineralised, it may be visualised. CT is the imaging modality of choice for demonstrating corticomedullary continuity with the parent bone. CT will also demonstrate the presence of a cartilage cap, which will appear as a soft-tissue density at the periphery of the lesion and will clearly demonstrate any mineralisation within the cartilage cap (Fig. 34.4). MRI will also show corticomedullary continuity with

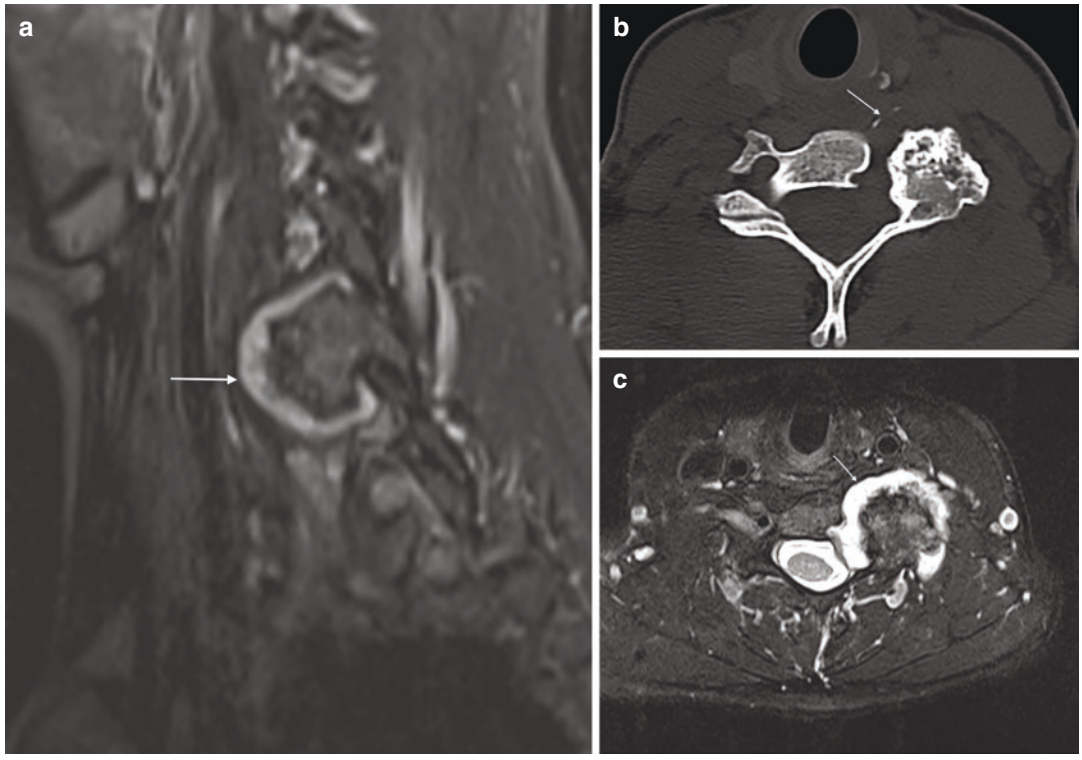

Fig. 34.4 Osteochondroma. (**a**) Axial CT showing osteochondromatous mass arising from the posterior elements of C5. Note the corticomedullary continuity and feint curvilinear calcification (white arrow) of the cartilage cap (not typically chondroid on CT). (**b, c**) Sagittal and axial STIR shows thin cartilage cap <2 cm in keeping with a benign osteochondroma

the parent bone, the cortex will be of low signal intensity on T1- and T2-weighted sequences and the medullary component will mirror the appearances of normal bone marrow on all sequences. The presence of matrix mineralisation within the cartilage cap will appear as focal low signal areas on both the T1- and T2-weighted sequences. MRI is the modality of choice for the evaluation of the cartilage cap. On T2-weighted sequences, the cartilage cap appears as a thin lobulated hyperintense mass (Fig. 34.4). The cartilage cap is commonly seen in children; it is less pronounced in adults as it tends to get thinner during growth. As in the appendicular skeleton, a cartilage cap thickness of >2 cm is concerning for secondary chondrosarcomatous transformation, and a biopsy followed by surgical excision will be required and is seen in <1% of sporadic cases and 3–5% of cases of HME.

34.4.2 Chondroblastoma

Chondroblastoma is a benign bone tumour most frequently seen in the epiphyses of the long bones in children and young adults. It is rare in the spine with only 1.4% of chondroblastomas occurring in the spine. Most occur in the thoracic spine but have been described in the cervical region. They can arise in the vertebral body or the posterior elements. The imaging findings are nonspecific; however, in most cases, they appear as an aggressive lesion with bone destruction with the presence of a soft-tissue mass. Interestingly, perilesional bone marrow oedema and secondary ABC formation are not features on MRI in contrast to chondroblastoma of the appendicular skeleton. Lesions can demonstrate a chondroid pattern of calcification, which can be easily seen on CT and will appear hypointense on both T1- and T2-weighted sequences. Due to the aggressive appearance on cross-sectional imaging, a biopsy is required to make the formal diagnosis.

34.4.3 Chondrosarcoma

Chondrosarcoma is a malignant tumour of cartilage. It is the third commonest primary malignant bone tumour involving the spine after myeloma and chordoma. The peak age of presentation is between 40 and 60 years. It is four times more common in males than females. They are less frequently seen in the cervical spine than the thoracic and lumbar spine. Most involve the posterior elements and the body together followed by the posterior elements alone and the body alone. Most spinal chondrosarcomas are primary; however, secondary chondrosarcomas can occur in a pre-existing osteochondroma. Secondary transformation should be suspected if there is pain or an increase in size of a pre-existing osteochondroma following skeletal maturation or an increased thickness of the cartilage cap >2 cm (Fig. 34.5).

Radiographs can demonstrate an aggressive osteolytic lesion with or without chondroid matrix mineralisation. CT better demonstrates the osteolytic lesion and the chondroid matrix calcification, which when present is typically of ‘ring and arc’ appearance but can be amorphous and extensive. The extraosseous soft-tissue mass when present is of lower attenuation than skeletal muscle due to the increased water content of car-

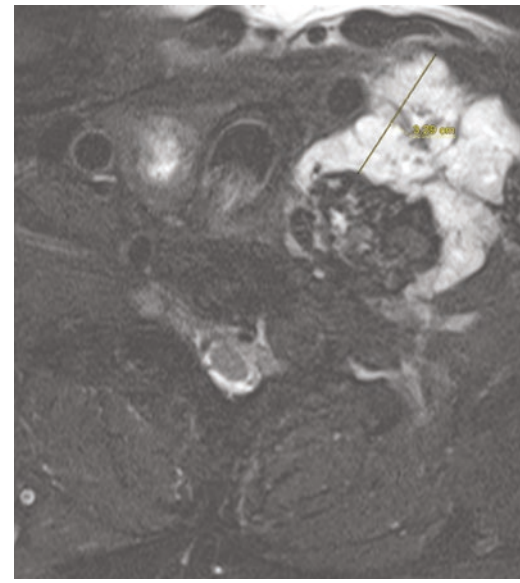

Fig. 34.5 Secondary chondrosarcoma. Axial STIR MRI shows osteochondromatous mass arising from the cervical spine. Note the increased thickness of the cartilage cap (>2 cm) in keeping with secondary chondrosarcoma from an osteochondroma

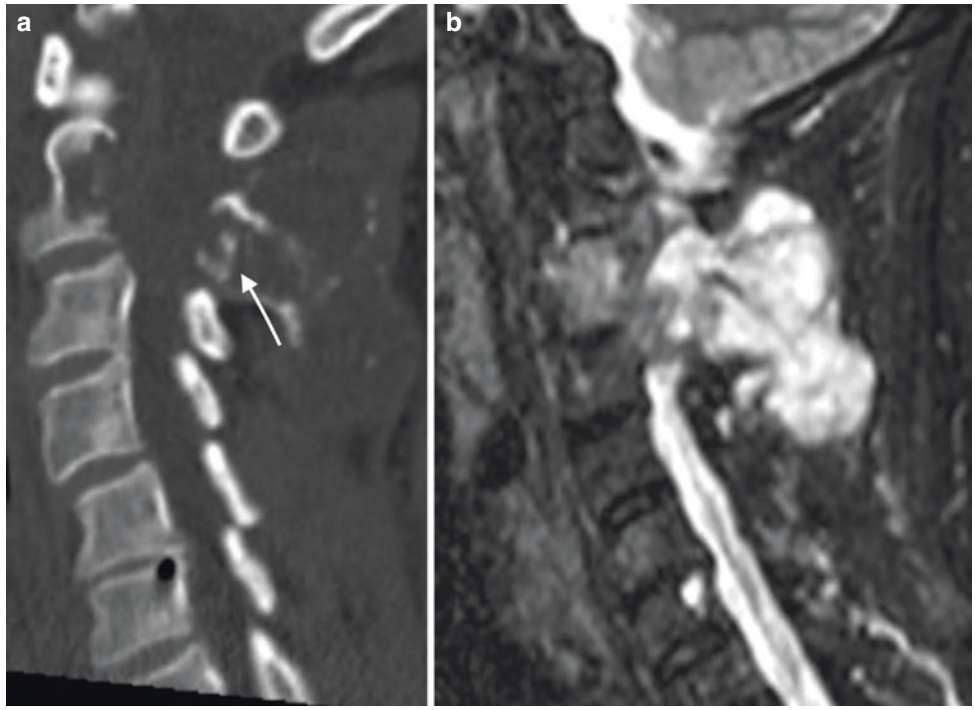

Fig. 34.6 Primary chondrosarcoma. (**a**) Sagittal CT shows destructive lesion in the body and posterior elements of C2 with areas of chondroid calcification (white arrow). (**b**) Sagittal STIR MRI shows lobulated high T2 signal lesion with low signal septa in keeping with a chondrosarcoma

tilage cells. The tumour has a lobulated outline and can invade adjacent vertebral bodies. MRI will show an osteolytic mass with or without a soft-tissue component. It has a uniformly hypointense signal intensity on T1-weighted sequences and is markedly hyperintense on T2-weighted sequences due to the water content of the cartilage cells (Fig. 34.6). Septa between individual cartilage lobules will be of low signal on T2-weighted sequences. When chondroid calcification is present, it will be hypointense on all MRI sequences. When there is extensive calcification within a lesion, it can be confused with osteoblastic lesions such as osteosarcoma, osteoblastoma or a sclerotic metastasis.

34.5 Small Round-Cell Tumours

34.5.1 Ewing's Sarcoma

Ewing's sarcoma is a small round-cell tumour of bone, cartilage or soft tissue (Fig. 34.7). Histologically, these tumours are characterised by small, round, undifferentiated cells. Because of the undifferentiated nature of these tumours, they can be difficult to diagnose without immunohistochemical techniques. Sometimes, there is a greater degree of neuroectodermal differentiation within the tumour, and historically, it has been called primitive neuroectodermal tumour (PNET). 3.5% of all cases of Ewing's sarcoma involve the spine. Most cases occur between the ages of 10 and 30 years with a slight male preponderance. Most cases in the spine involve the sacrum (55%), with the cervical spine the least commonly affected site (3% of cases). Greater than one segment involvement is seen in 8% of cases. On radiographs, most lesions are lytic, but lesions can also be sclerotic or mixed lytic and sclerotic. The sclerosis is due to a host bone response to the tumour rather than matrix production by the tumour. Other radiographic findings include vertebra plana.

CT demonstrates a permeative pattern of bone destruction with a wide zone of transition.

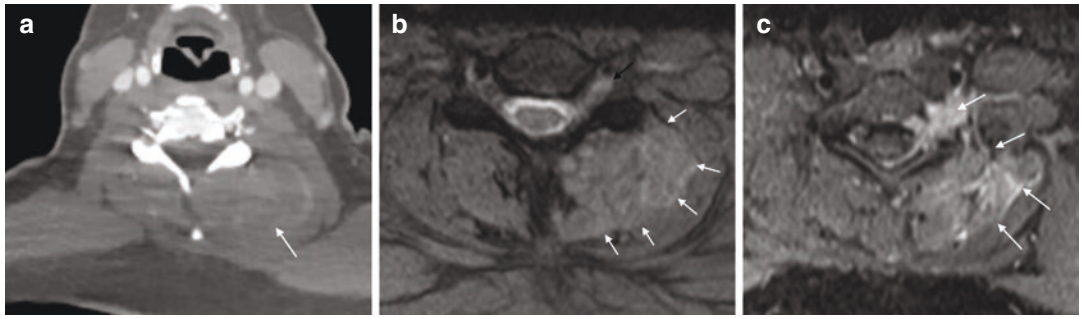

Fig. 34.7 Ewing's sarcoma. (**a**) Axial CT showing lytic lesion involving posterior elements without matrix mineralisation with a comparatively larger soft-tissue component (white arrow). (**b**) Axial STIR MRI shows larger soft-tissue component (white arrows) and some extension into the left neural foramen (black arrow). Note that large areas of bone destruction are not a feature. (**c**) Axial fat-saturated T1 post-contrast scan shows mild enhancement of the tumour (white arrows)

An associated soft-tissue mass is commonly seen; however, large areas of cortical destruction are not usually a feature; rather, the tumour penetrates through tiny perforations in the cortex. The soft-tissue component is often larger than the osseous component of the disease. Lesions are usually hypointense to isointense to skeletal muscle on T1-weighted imaging. Due to the abundance of red marrow in the spine, the tumour can be difficult to see on T1-weighted images. On T2-weighted sequences, tumours are usually hyperintense. MRI is the best modality to demonstrate the soft-tissue component of the disease, which is present usually without significant cortical destruction. The presence of central necrosis is commonly seen, which results in non-enhancing areas following gadolinium administration on T1-weighted sequences.

34.6 Haematopoietic Tumours

34.6.1 Haemangioma

Vertebral haemangioma is a commonly encountered benign lesion within the spine and is the commonest benign tumour of the spine in adults. Histologically, they consist of thin-walled channels lined with vascular endothelium within the medullary cavity. They also contain variable amounts of fatty tissue. They are commonly multiple (30%). Most are confined to the vertebral body but can extend into the posterior elements. Most are completely intraosseous often involving the entire vertebral body, but they can extend into the epidural soft tissues giving the impression of a more aggressive lesion; such lesions are coined aggressive haemangiomas. Lesions are typically asymptomatic and are usually discovered incidentally during CT and MRI examinations of the spine for other reasons. Peak incidence is usually in young adults, and there is a slight female predilection. They are commonest in the thoracic spine followed by the lumbar spine and least common in the cervical spine.

Lesions can be visualised on radiographs especially when large. They appear as a radiolucent lesion with coarsening of the trabecular giving a typical 'corduroy' appearance. The lesion is much better appreciated on CT. The characteristic appearance on axial CT is of 'polka dots', which represent the thickened trabeculae in axial section. On coronal and sagittal sections, the 'corduroy' appearance is seen. MRI appearances can be variable due to differences in the amount of fatty and vascular tissue within the haemangioma. Typical haemangiomas will show hyperin-

tense signal on both T1- and T2-weighted sequences. The thickened trabeculae will show up as linear areas of low signal on both T1- and T2-weighted images (equivalent of the polka dots on CT). When the vascular component in a haemangioma predominates, the signal can be isointense to hypointense on T1-weighted sequences and can be confused with a marrow infiltrative process such as a metastasis. In such situations, chemical shift imaging can be helpful in determining the presence of a marrow infiltrative process (e.g. malignant) from a non-bone marrow infiltrative process (e.g. haemangioma, red marrow hyperplasia). A >25% signal dropout correlates with a 100% sensitivity and 86% specificity for a benign process (Fig. 34.8). MRI will also best demonstrate the presence of an associated soft-tissue mass, which is seen in the more locally aggressive form of the disease, and its effect on the adjacent cord or nerve roots. The diagnosis of a haemangioma is fairly straightforward; however, the differential diagnosis of the more aggressive form includes a vascular metastasis as the struts of trabeculae can be confused with flow voids (vessels in a vascular lesion).

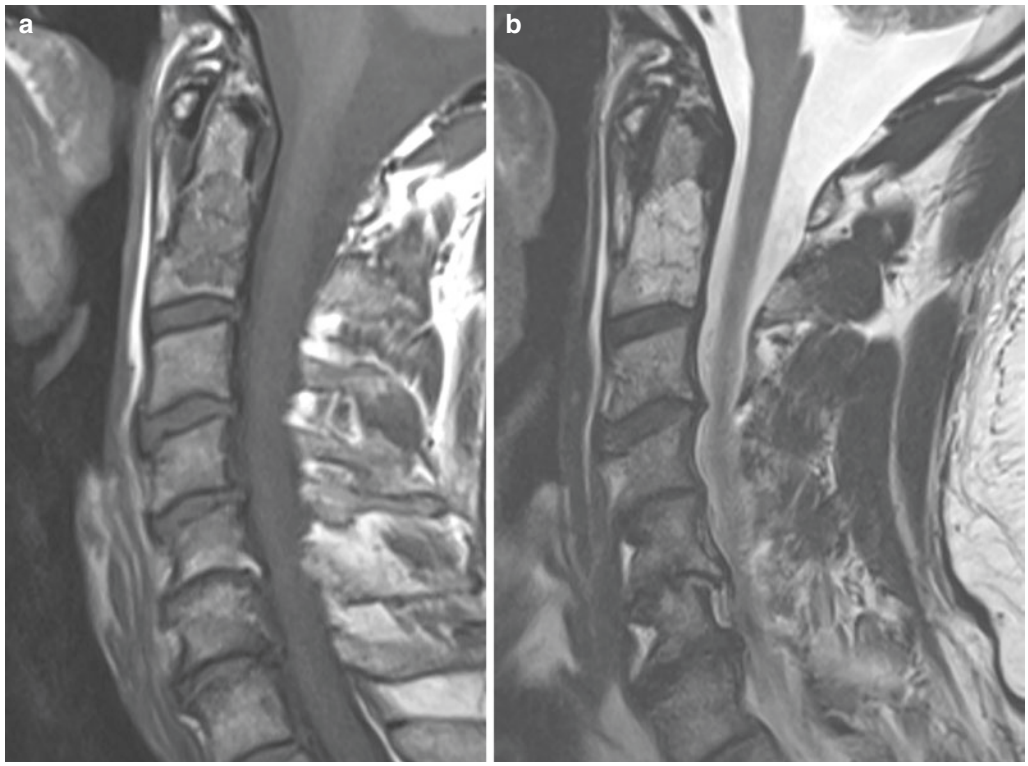

Fig. 34.8 Haemangioma. (**a**) Sagittal T1-weighted and (**b**) sagittal T2-weighted images show lesion in the body of C2. It is low to intermediate signal on T1 with a few focal areas of fat seen. (**c**) In-phase and out-of-phase imaging shows >20% signal dropout, suggesting that lesion contains a significant amount of fat and therefore no marrow infiltration. This has been stable on follow-up. (**d**) Sagittal CT shows coarsening of the trabeculae giving the classical 'corduroy' appearance. Myeloma

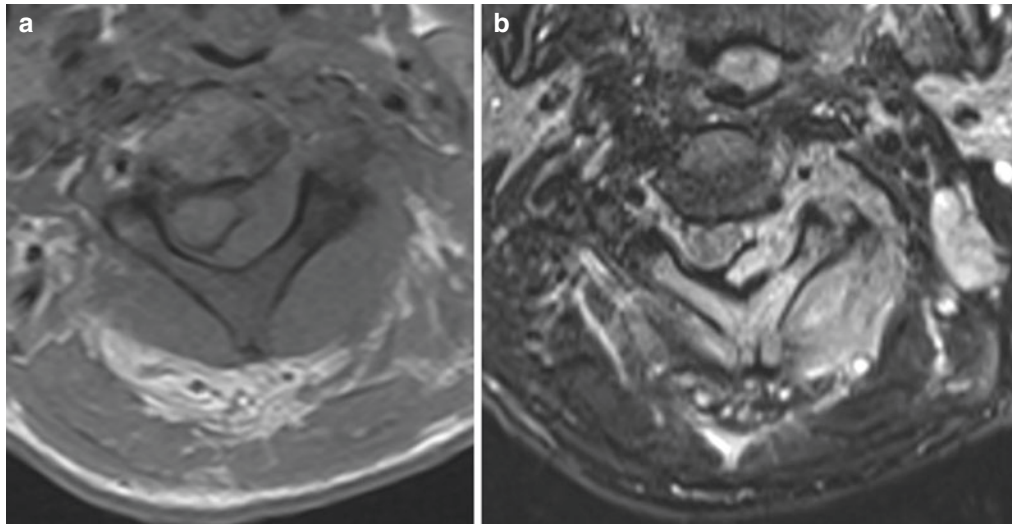

Fig. 34.9 Lymphoma. (**a**) Axial T1 and (**b**) axial T2-weighted images showing marrow infiltration in the posterior elements bilaterally with a left-sided epidural and paravertebral soft-tissue mass with cord compression. Not the relative preservation of the cortex of the bone, which is a typical feature of small round-cell tumours

34.6.2 Lymphoma

Lymphoma of the spine is usually due to the metastatic spread of primary lymphoma (Fig. 34.9). Lymphoma characteristics are described in Chap. 6.

34.6.3 Plasmacytoma

Plasmacytoma is a solitary plasma cell neoplasm of bone or soft tissue. It is considered to be an early form of multiple myeloma. The spine is the most common site of involvement. The thoracic spine is the commonest location, with the cervical spine being the least commonly affected location. It is usually located in the vertebral body and can extend into the posterior elements. Most patients are aged between 55 and 75 years of age.

Radiographs can be normal in the early stage. Lesions are commonly osteolytic, and vertebral body collapse is common. CT better demonstrates the bone destruction. Cortical thickening and vertical struts can form as a response to the osteolysis. Occasionally, lesions can be sclerotic, but this is uncommon. The degree of vertebral body collapse is easily assessed on sagittal reformatted CT images. On MRI, plasmacytoma is of low signal intensity on T1-weighted images and heterogenous hyperintense signal on T2-weighted sequences with varying degrees of vertebral compression. Infoldings of the cortex can be seen on the T1-weighted sequence giving a 'mini brain' appearance. MRI is the best modality to demonstrate the presence and extent of any soft-tissue extension, which is commonly seen. 1/3 of patients will have a second occult focus of disease elsewhere in the spine, so whole-spine imaging should be performed.

34.6.4 Multiple Myeloma

Multiple myeloma is a plasma cell neoplasm characterised by malignant proliferation of plasma cell in bone marrow (Fig. 34.10). Lymphoma characteristics are described in Chap. 6.

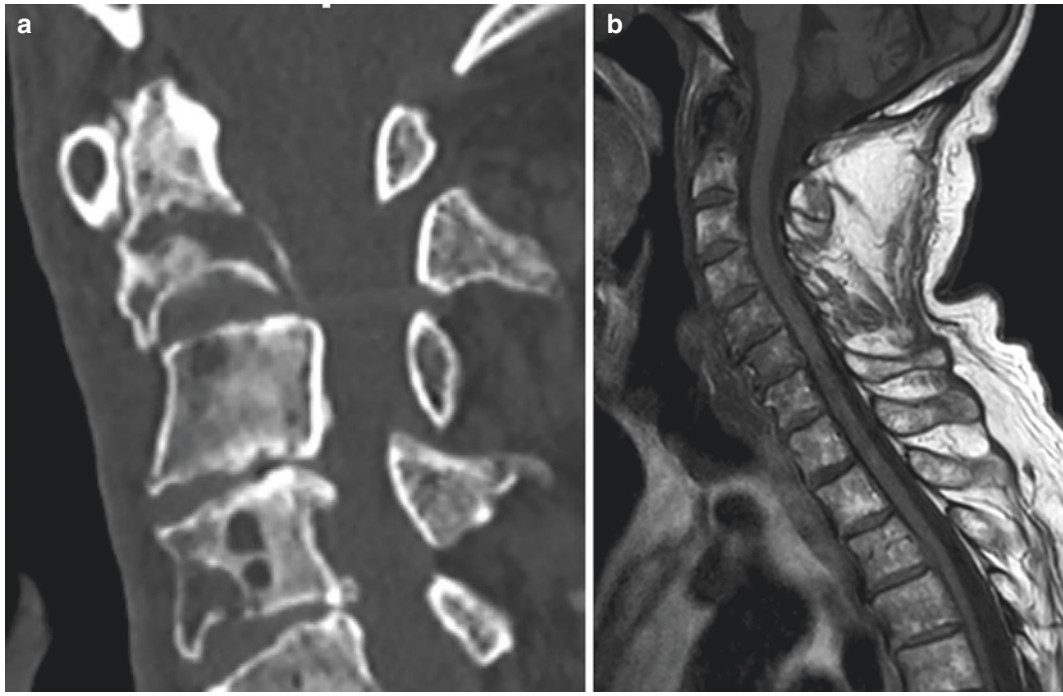

Fig. 34.10 Multiple myeloma. (**a**) Sagittal CT shows multiple lytic deposits in keeping with micronodular disease. (**b**) Sagittal T1-weighted image showing micronodular (salt and pepper) pattern of marrow infiltration

34.7 Osteoclastic Giant Cell-Rich Tumours

34.7.1 Giant-Cell Tumour

Giant-cell tumour of bone is a locally aggressive rarely metastasising tumour composed of stromal cells with macrophages and osteoclast-like giant cells. A small subset of this tumour is malignant. It typically affects the ends of the long bones but can affect the spine in 7% with most cases involving the sacrum. The cervical spine is the least common site of spinal involvement. There is a slight female preponderance and is mostly seen in the second to fourth decades. Most tumours involve the vertebral body but can extend into the posterior elements. A tumour involving the posterior elements alone is unlikely to be a giant-cell tumour of bone. Extraosseous involvement is commonly seen in up to 79% of cases. Tumours may extend through the disc to involve adjacent vertebrae. Pathological fracture may be identified.

Radiographs can show a lytic expansile lesion without matrix mineralisation. Cortical disruption may be observed. CT can better delineate the lesion, will clearly show any areas of cortical disruption and may show the presence of an extraosseous soft-tissue mass. MRI will show the lesion and any extension into the paravertebral and epidural soft tissues and its relationship to cord and nerve roots. Giant-cell tumours are usually of low to intermediate signal on both T1-weighted and fluid-sensitive sequences, which reflects the haemosiderin and collagen in the tumour (Fig. 34.11). Higher signal areas on

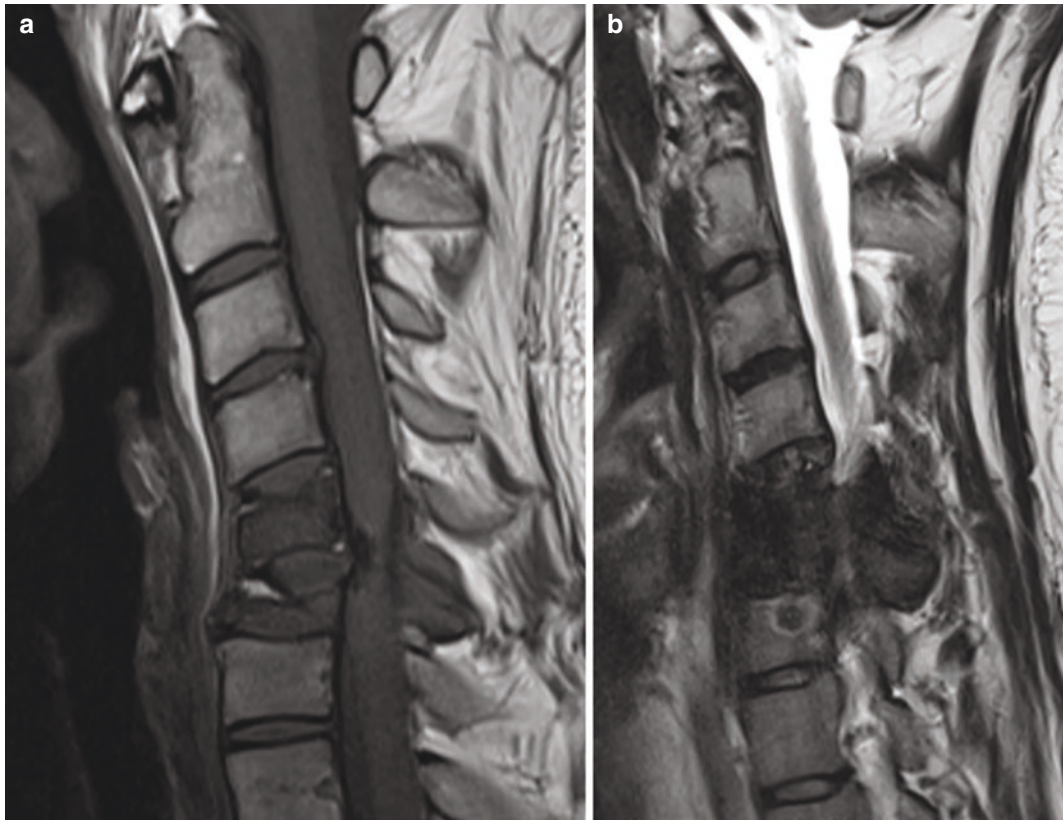

Fig. 34.11 Giant-cell tumour of bone. (**a**) Axial CT showing lytic lesion in the body of C5. (**b**) Sagittal T1- and (**b**) sagittal T2-weighted show lesion in C5 and C6, which is low signal on both T1- and T2-weighted sequences, which is characteristic of giant-cell tumour. Note the collapse of the C6 level

T1-weighted images and presence of fluid-fluid levels may be due to recent tumour haemorrhage and are a common finding.

34.7.2 Aneurysmal Bone Cyst (ABC)

Aneurysmal bone cyst is a benign bone tumour comprised of thin-walled blood-filled cystic cavities. Up to 30% of ABCs occur in the spine. Some studies have shown a predilection for the cervical and thoracic spine. ABCs are usually centred in the posterior elements commonly in the lamina with most extending into the vertebral body. These are much more common in those under 20 years of age, and there is a slight female predilection.

Radiographs can demonstrate a lytic expansile lesion with cortical thinning in the posterior elements extending into the body. On the AP radiograph, there may be loss of visualisation of the pedicle. Very occasionally, they can cause vertebra plana. CT can better delineate the abnormality. There should be a narrow zone of transition and a thinned cortical rim, which may be disrupted in places. Septations may be visible within the lesion. MRI demonstrates the expansile mass with multiple cystic cavities and the characteristic fluid-fluid levels on T2-weighted sequences due to bleeding within the cysts (Fig. 34.12). Secondary ABC formation can occur due to the presence of another lesion. Giant-cell tumour of bone, telangiectatic osteosarcoma and osteoblastoma are tumours, which commonly show sec-

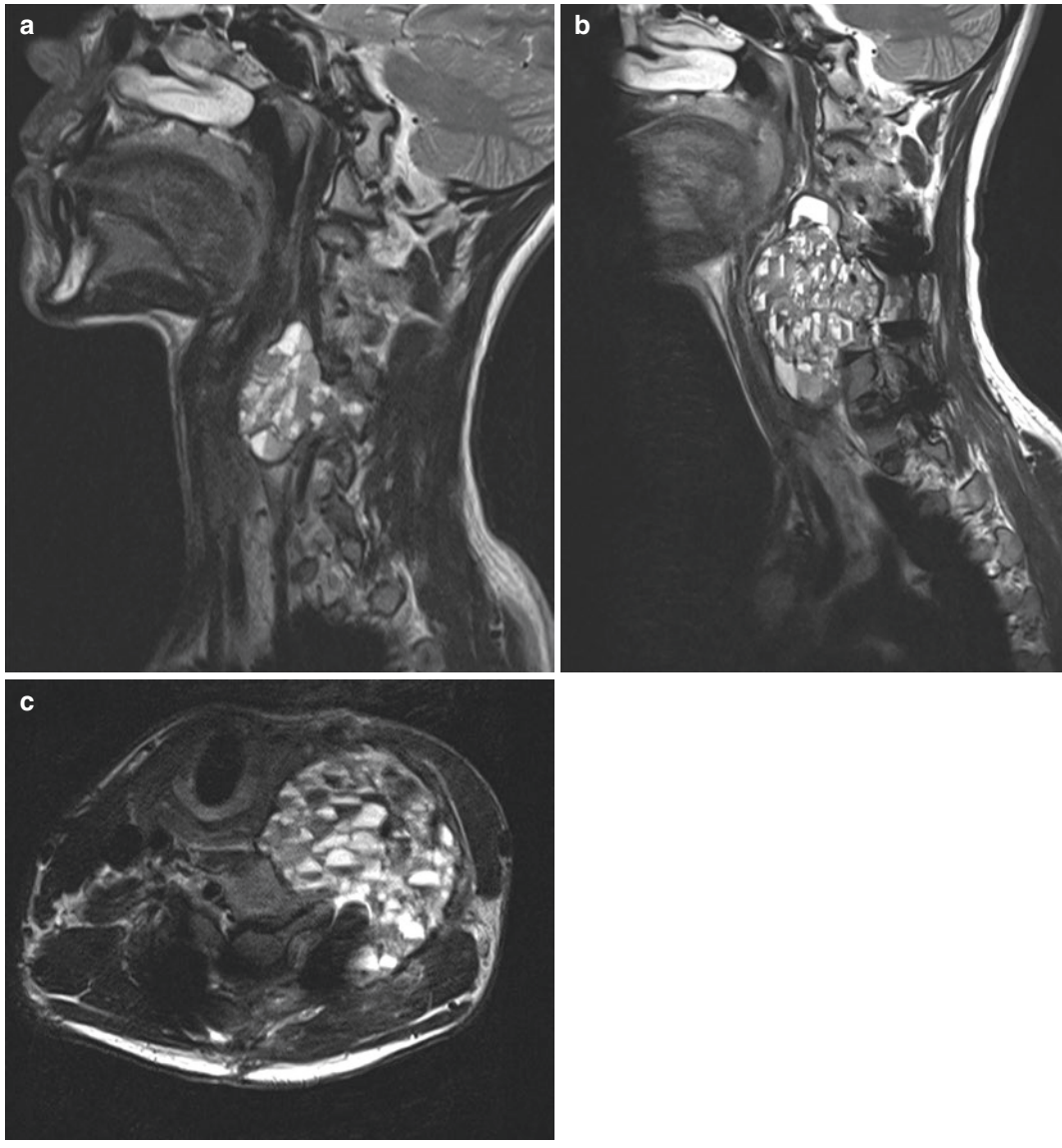

Fig. 34.12 Aneurysmal bone cyst. (**a**) Sagittal T2-weighted MRI shows lesion in the posterior elements and body of C5 with multiple fluid-fluid levels in keeping with an ABC. The patient underwent curettage and stabilisation. (**b**) Sagittal T2- and (**c**) axial T2-weighted images of the same patient 6 months after surgery showing marked increase in the size of ABC reflecting its locally aggressive nature

ondary ABC change. Vascular metastasis can also show fluid-fluid levels but is usually seen in an older age group. Contrast administration will show only peripheral or septal enhancement, but in most cases, the diagnosis will be made without the use of contrast. ABCs are locally aggressive and can grow rapidly; therefore, an urgent biopsy should be performed to confirm the diagnosis. This then allows for prompt definitive treatment in the form of sclerotherapy or surgical curettage ± stabilisation.

34.8 Notochordal Tumours

34.8.1 Chordoma

Chordoma is a slow-growing malignant tumour arising from notochord remnants in the axial skeleton. They are commonly seen in the clivus or the sacrococcygeal region. 15% occur in the spine. The cervical spine is the most common spinal location accounting for 20–50% of spinal lesions with C2 being the commonest location. Lesions involve the vertebral body primarily, and extension into the posterior elements is not a common feature. There is often a paravertebral/epidural soft-tissue mass, and tumours can also extend along nerve roots and can expand the neural foramen and therefore can be confused with neurogenic tumours. Due to the central position of the notochord remnants, tumours are usually centred over the midline. They can extend into the disc and can span more than one vertebral body. They can occur at any age but are most prevalent in the fifth to seventh decades. When they involve children and young adults, they are commonly located in the base of the skull and the upper cervical spine. There is a 2:1 male preponderance. Macroscopically, chordoma is a lobulated solid mass with a high myxoid content. Cystic degeneration and haemorrhage are also seen.

Radiographs may show a destructive lesion, which may involve more than one vertebral body; if there is a large soft-tissue mass, this may be visible. CT will better demonstrate the osseous abnormality and the soft-tissue component of tumour. The soft-tissue component can be comparatively larger than the bony component. Cystic and myxoid areas will have a hypodense appearance on CT. Amorphous calcification can be seen in up to 30%. On MRI, lesions are low or isointense on T1-weighted sequences. They are markedly hyperintense on fluid-sensitive sequences due to the high water content of the myxoid/gelatinous components of the lesion. Thin low signal septations may be seen. MRI is the best modality to appreciate the soft-tissue extension of the disease and its relationships to the cord and nerve roots. Any internal calcification will be of low signal on all MRI sequences.

34.9 Metastatic Disease

Metastatic disease is the commonest spinal tumour, and spinal metastases are the third commonest site for metastatic disease after lung and liver. An example of metastatic disease is reported in Fig. 34.13. Spinal metastasis characteristics are described in Chap. 26.

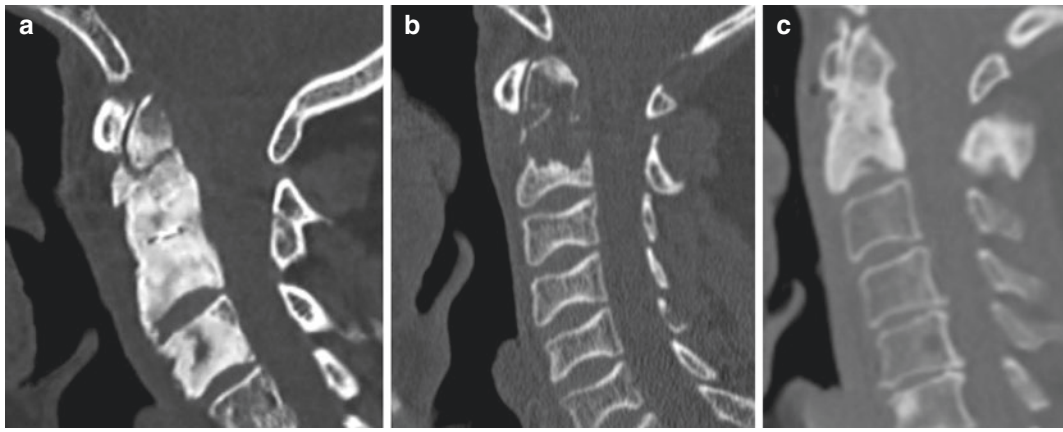

Fig. 34.13 Metastatic disease. (**a**) Sagittal CT showing multiple osteoblastic lesions in C2 and C3 with a pathological fracture in C2 in a patient with known prostate carcinoma. (**b**) Sagittal CT showing osteolytic metastasis in a patient with known breast carcinoma. (**c**) Same patient in (**b**) showing sclerosis in the metastatic deposit following treatment with systemic chemotherapy

Take-Home Message

- Metastatic disease, myeloma and lymphoproliferative disorders are commonly present in adults as multiple lesions, and on seeing multiple lesions, the diagnosis is most likely to be one of these three disorders. If the lesion is solitary, a primary bone tumour must also be considered part of the differential diagnosis.
- Enostosis (bone island) is a benign osteogenic tumour.
- Osteoid osteoma is a benign osteoid-forming tumour.
- Osteoblastoma is a benign osteoid-forming tumour.
- Osteochondroma is the most commonly encountered bone tumour; however, spinal osteochondromas are relatively rare, representing 1–5% of sporadic osteochondromas and 1–9% in patients with hereditary multiple exostoses (HME).
- Chondroblastoma is a benign bone tumour most frequently seen in the epiphyses of the long bones in children and young adults.
- Ewing's sarcoma is a small round-cell tumour of bone, cartilage or soft tissue.
- Vertebral haemangioma is a commonly encountered benign lesion within the spine and is the commonest benign tumour of the spine in adults.
- Lymphoma of the spine is usually due to the metastatic spread of primary lymphoma.
- Plasmacytoma is a solitary plasma cell neoplasm of bone or soft tissue.
- Giant-cell tumour of bone is a locally aggressive rarely metastasising tumour composed of stromal cells with macrophages and osteoclast-like giant cells.
- Aneurysmal bone cyst is a benign bone tumour comprised of thin-walled blood-filled cystic cavities.
- Chordoma is a slow-growing malignant tumour arising from notochord remnants in the axial skeleton.

Summary

Metastatic disease, myeloma and lymphoproliferative disorders are commonly present in adults as multiple lesions, and on seeing multiple lesions, the diagnosis is most likely to be one of these three disorders. If the lesion is solitary, a primary bone tumour must also be considered part of the differential diagnosis.

Enostosis (bone island) is a benign osteogenic tumour. It is a focus of dense cortical bone within cancellous bone. They can be identified on radiographs, but CT is the imaging modality of choice.

Osteoid osteoma is a benign osteoid-forming tumour. They usually occur in children and young adults commonly between the second and third decades of life. They can occur anywhere in the body with 10% of tumours located in the spine, mostly in the posterior elements.

Osteoblastoma is a benign osteoid-forming tumour.

They preferentially occur in the posterior elements; however, in the cervical spine, they can more frequently involve the vertebral body than other locations.

Osteosarcoma of the spine accounts for only 4% of all osteosarcomas. The cervical spine is the least common location of spinal osteosarcoma.

Osteochondroma is the most commonly encountered bone tumour; however, spinal osteochondromas are relatively rare, representing 1–5% of sporadic osteochondromas and 1–9% in patients with hereditary multiple exostoses (HME). The cervical spine is the most common site for osteochondromas with a predilection for the C2 level.

Chondroblastoma is a benign bone tumour most frequently seen in the epiphyses of the long bones in children and young adults. It is rare in the spine with only 1.4% of chondroblastomas occurring in the spine. It mostly occurs in the thoracic spine but has been described in the cervical region.

Chondrosarcoma is a malignant tumour of cartilage. It is the third commonest primary malignant bone tumour involving the spine after myeloma and chordoma. The peak age of presentation is between 40 and 60 years.

Ewing's sarcoma is a small round-cell tumour of bone, cartilage or soft tissue. Histologically, these tumours are characterised by small, round, undifferentiated cells. Because of the undifferentiated nature of these tumours, they can be difficult to diagnose without immunohistochemical techniques.

Vertebral haemangioma is a commonly encountered benign lesion within the spine and is the commonest benign tumour of the spine in adults. They are commonly multiple (30%). Most are confined to the vertebral body but can extend into the posterior elements.

Lymphoma of the spine is usually due to the metastatic spread of primary lymphoma.

Plasmacytoma is a solitary plasma cell neoplasm of bone or soft tissue. It is considered to be an early form of multiple myeloma. The spine is the most common site of involvement. The thoracic spine is the commonest location, with the cervical spine the least commonly affected location. It is usually located in the vertebral body and can extend into the posterior elements.

Multiple myeloma is a plasma cell neoplasm characterised by malignant proliferation of plasma cell in bone marrow.

Giant-cell tumour of bone is a locally aggressive rarely metastasising tumour composed of stromal cells with macrophages and osteoclast-like giant cells. A small subset of this tumour is malignant. It typically effects the ends of the long bones but can affect the spine in 7% with most cases involving the sacrum. The cervical spine is the least common site of spinal involvement.

Aneurysmal bone cyst is a benign bone tumour comprised of thin-walled blood-filled cystic cavities. Up to 30% of ABCs occur in the spine.

Chordoma is a slow-growing malignant tumour arising from notochord remnants in the axial skeleton. They are commonly seen in the clivus or the sacrococcygeal region. 15% occur in the spine.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. The imaging modality of choice for identify enostosis is:
   (a) CT scan
   (b) MRI
   (c) X-ray
   (d) Angiogram
2. Osteoid osteoma usually occurs:
   (a) Between 20 and 30 years
   (b) Between 40 and 60 years
   (c) Over 60 years
   (d) Under 20 years
3. Osteoblastoma is differentiated from osteoid osteoma:
   (a) By its larger size (>1.5 cm)
   (b) By its smaller size (<1.5 cm)
   (c) By the macroscopic aspect
   (d) By the microscopic aspect
4. Osteosarcoma of the spine:
   (a) Usually affects adults rather than children
   (b) Usually affects children rather than adults
   (c) Usually affects adolescent rather than children
   (d) Affects only people aged over 60 years old
5. A specific histological feature of the chondrosarcoma is:
   (a) The cartilage cap
   (b) The onion bulb aspect
   (c) The Codman triangle
   (d) The osteolytic lesion

Further Reading

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Management of Cervical Spine Tumours

35

Vincenzo Denaro, Umile Giuseppe Longo, Sergio De Salvatore, and Luca Denaro

Overview

There are no specific guidelines regarding cervical spine tumour surgical management; therefore, this surgery needs to be tailored to the patient. Instead, in case of neurological deficits, surgery represents the best option possible to improve the chance of recovery. Cervical spine surgery is burdened with a high rate of complications.

35.1 Introduction

Progress in technology, radiotherapy, surgical devices and technique has significantly improved the quality of cervical spine tumour management. It is possible to obtain satisfactory spine fusion with new bone substitute devices without using an allograft. Furthermore, it is possible to obtain an accurate diagnosis and information regarding the tumour extension and localization employing modern imaging techniques (volumetric spiral CT, magnetic resonance imaging (MRI) and angio-MRI). Combined management procedures provide a more favourable prognosis in some patients. Biopsy is necessary to establish the histological diagnosis. The biopsy can be performed with a percutaneous needle under radiographic or CT guiding. In lesions located in the anterior portion of C1–C2, and the anterosuperior portion of C3, a transoral approach could be used to perform the needle biopsy. In C3–C7 lesions, the anterolateral presternocleidomastoid approach is chosen. After the visualization and the protection of the neurovascular bundle (the vagus nerve, the common carotid artery and the internal jugular vein), the biopsy is performed. Biopsies of the posterior portion of the vertebrae could be addressed with a posterior approach. In cases that require urgent surgical decompression, the biopsy can be performed intraoperatively.

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35.2 Surgical Management

The surgical treatment is based on the type of tumour. From a technical point of view, there are no specific guidelines concerning cervical spine tumour surgical management. Therefore, the experience of the surgeon is crucial. Generally, cervical tumours require further stabilization after tumour excision to stabilize the operated segment. Bone grafting procedures, osteobiologic devices and spinal fusion instrumentation are adopted to reconstruct spinal stability. The surgery needs to be tailored to the patient; therefore, the surgical approach, the type of fixation and the postoperative management must be modified to the single case.

Chemotherapy, radiation therapy and hormone therapy could be administrable before or after surgery as adjuvant or neoadjuvant therapy. Some studies reported better outcomes in patients with spinal cord compression from metastatic cancer treated with surgery and radiotherapy than those treated by radiotherapy alone. Bladder continence, muscle strength and long-term survival rate were higher in the former group. Prophylactic embolization with angiography (24 h before surgery) could reduce the intra- and postoperative bleeding and facilitate the surgical excision.

Cervical spinal tumour surgery is challenging due to the continuous proximity to several vascular and neurological structures encountered during the procedure; therefore, a combined approach is required several times. Even in benign tumours, compression of the canal structures could be present, requiring complete excision and segmental stabilization (Figs. 35.1 and 35.2). The surgery, even in benign lesions, can guarantee good prognosis quoad vitam, but poor quoad valetudinem.

Cervical spinal tumour surgery may have two different aims: palliative treatment (neural decompression and stabilization) or curative treatment (radical tumour resection and stabilization). The former focuses on pain reduction, segmental stabilization and prevention of further damages. It is usually adopted in metastasis cases, particularly those with worst prognosis tumours (e.g. brain metastasis, lung tumour) or pathologic fractures. In cases of osteoblastic metastatic tumours, without neurological deficits, conservative therapy could be used. It includes bracing with orthosis, chemotherapy or radiation therapy.

Instead, surgery is mandatory for osteolytic lesions to prevent the progressive worsening of neurological status due to vertebral pathologic fractures. For this type of lesions, a wide tumour excision with stabilization and reconstruction of the bone loss is required. Various bone substitutes could be used, either synthetic or biologics. Allograft harvested from the iliac crest of the fibula represented the most common bone substitute used. Otherwise, with techno-

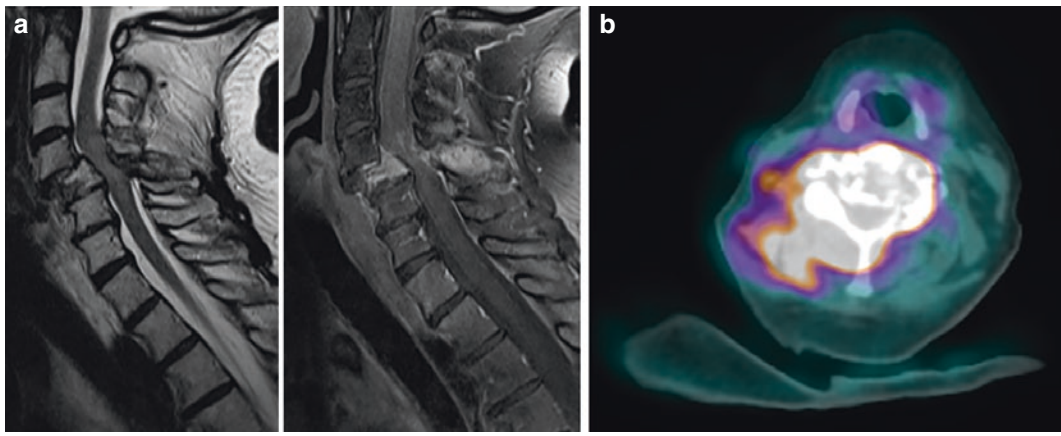

Fig. 35.1 MRI scans (**a**) show pathological fracture of the C5 vertebra for osteolytic lesion due to thyroid metastasis with cervical compressive myelopathy. PET-CT (**b**) shows FDG high focal uptake at the level of the C5 vertebra. The patient presented with complete tetraplegia and severe actinic dermatitis due to previous irradiation of the lesion

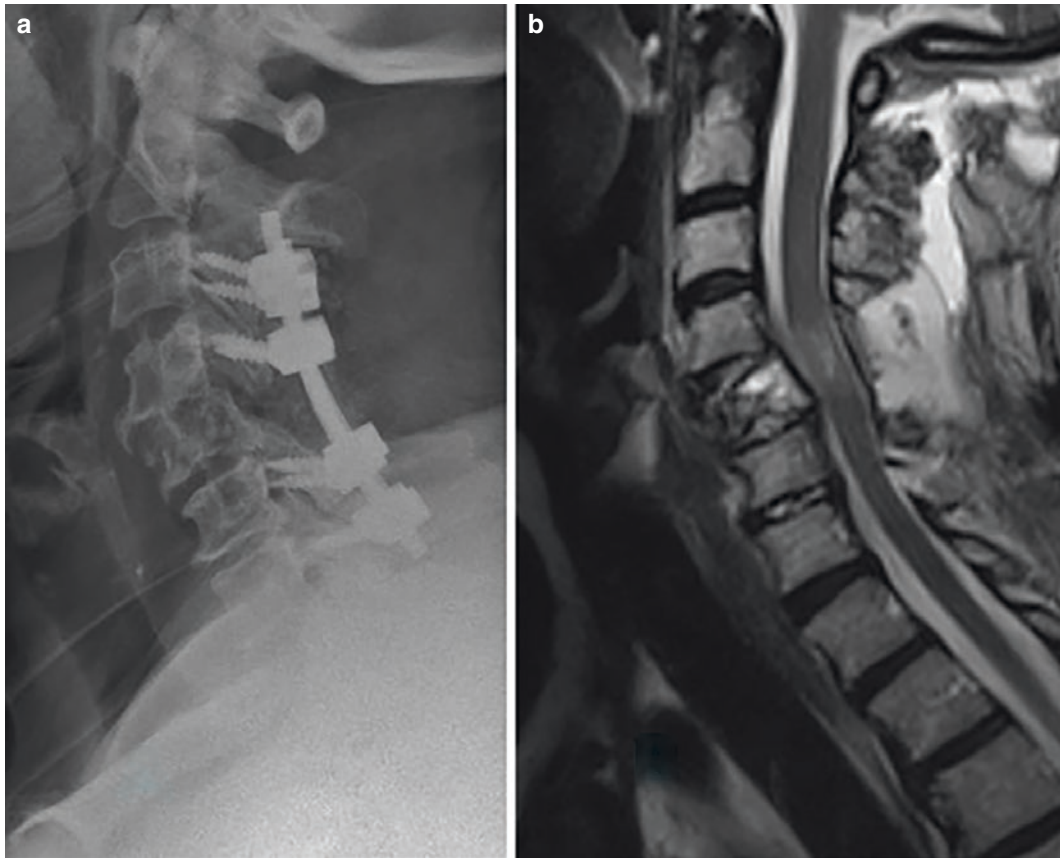

Fig. 35.2 Postoperative X-ray (**a**) and MRI (**b**) show posterior C5 decompression and lateral mass screw fixation at C3, C4, C6, C7. It has been performed an urgent posterior approach because the patient had been previously irradiated. The cervical decompression and fixation resulted in a neurological recovery first of the upper limbs and subsequently of the lower limbs

logical improvements, titanium or carbonium fibre mesh cage filled with homologous bone graft, acrylic cement or other osteobiologic augmentation devices represent a valid alternative nowadays. In case of combined approaches, anterior plating and screws with plates or longitudinal bar and lateral mass screws posteriorly can be used.

In case of poor prognosis, conservative treatment is the first choice; instead in patients with poor prognosis but good general status, it is possible to perform an "internal bracing" stabilization using metal wiring stabilized with acrylic cement. This surgical technique provides lowered operatory times with moderate perioperative morbidities.

35.3 Complications

Cervical spinal tumour surgery is burdened with a high rate of complications as vascular lesions, visceral structure injuries, neurological lesions, spinal cord canal lesions, bone graft and device complications, general complications and infections (Table 35.1). Furthermore, some risk factors could determine the success of the surgical procedures as prolonged chemotherapy, radiotherapy before surgery, sarcopenia and chronic use of steroids. The overall risk rate ranges from 19% to 25% including progressive neurological lesions, death (5.7%), intraoperative bleeding, implant failure (2.2%), wound complications (4%), dysphagia, deep vein thrombosis, cerebro-

Table 35.1 Summary of complications

Type of injury	Site of injury
Vascular lesions	• Carotid artery • Vertebral artery • Jugular vein
Visceral structures	• Pharynx • Oesophagus
Neurological lesions	• Vagus nerve • Phrenic nerve • Recurrent laryngeal nerves • Cervical plexus • Brachial plexus (mainly to the primary trunks near to the intervertebral foramina)
Spinal cord canal lesions	• Lesions to the spinal cord • Nerve roots • Dura mater
Implant failure	• Breakage • Mobilization • Migration
Bone graft complications	• Rupture • Collapse • Resorption
General complications	• Pulmonary • Urological • Cardiocirculatory • Deep vein thrombosis • Intra- and extradural hematomas
Infections	• Wound infections • Systemic infections

spinal fluid leakage, iatrogenic spinal injuries or compression, paravertebral hematoma, progressive renal failure and stress ulcer bleeding.

Take-Home Message

- Cervical spinal tumour surgery is challenging and therefore associated with high difficulty and complications.
- The tumour resection can require combined approaches.
- Vascular and neurological injuries are frequent.
- Preoperative identifications of potentially good prognosis patients are mandatory to select more aggressive surgical treatments.
- Preoperative radiotherapy, chemotherapy, sarcopenia and chronic use of steroids could increase the risk of complications.

Summary

- Advancement in technology, radiotherapy, surgical devices and technique improved the quality of cervical spine tumour management. Furthermore, it is possible to obtain a more accurate diagnosis and information regarding the tumour extension and localization of modern imaging techniques.
- There are no specific guidelines regarding cervical spine tumour surgical management; therefore, the experience of the surgeon is crucial. More often, cervical tumours require a further stabilization after tumour removal to stabilize the treated segment. The surgery needs to be tailored to the patient; therefore, the surgical approach, the type of fixation and the postoperative management need to be adapted to the single case.
- Cervical spinal tumour surgery is challenging due to the constant proximity to various vascular and neurological structures encountered during the procedure; therefore, a combined approach is required several times.
- Cervical spinal tumour surgery may have two different aims: palliative treatment or curative treatment. The former focuses on pain decrease, segmental stabilization and prevention of further injuries. It is usually adopted in case of osteoblastic metastases. Instead, surgery is mandatory for osteolytic lesions, and wide tumour excision, stabilization and bone reconstruction are required. In case of combined approaches, anterior plating and screws with plates or longitudinal bar and lateral mass screws posteriorly can be used.
- In case of poor prognosis, conservative treatment is the first choice; instead in patients with poor prognosis but good general status, it is possible to perform an "internal bracing" stabilization using metal wiring stabilized with acrylic cement. This surgical technique provides reduced operatory times with low perioperative morbidities.
- Cervical spinal tumour surgery is burdened with a high rate of complications as vascular lesions, visceral structure injuries, neurological lesions, spinal cord canal lesions, bone graft and device complications, general complications and infections.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. In case of lesion located:
   (a) In the anterior portion of C1–C2, and the anterosuperior portion of C3, a transoral approach could be used to perform the needle biopsy.
   (b) In the anterior portion of C2–C3, and the anterosuperior portion of C4, a transoral approach could be used to perform the needle biopsy.
   (c) In the anterior portion of C5–C6, and the anterosuperior portion of C7, a transoral approach could be used to perform the needle biopsy.
   (d) In the anterior portion of C1–C2, and the anterosuperior portion of C3, a posterior approach could be used to perform the needle biopsy.
2. In C3–C7 lesions that involve the bone of the vertebra:
   (a) The anterolateral presternocleidomastoid approach is adopted.
   (b) The posterior approach is adopted.
   (c) The combined anterior-posterior approach is adopted.
   (d) The transoral approach is adopted.
3. Combined posterior-anterior approach:
   (a) Is required in cases of anterior and posterior compression
   (b) Is required in each case of cervical spinal tumours
   (c) Is performed only with posterior laminotomy and anterior plating
   (d) Is performed with wide anterior decompression without bone grafting
4. Dysphagia:
   (a) Is a frequent complication in anterior cervical spine surgery
   (b) Is a frequent complication in posterior cervical spine surgery
   (c) Is not a common complication in cervical spine surgery
   (d) Occurs with posterior approach
5. Death:
   (a) Occurs in 5.7% of cases
   (b) Occurs in 1.2% of cases
   (c) Is not frequent in cervical spine surgery
   (d) Could occur only with posterior approach

Further Reading

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Cervical Spine Injuries

36

Julian Scherer and Georg Osterhoff

Overview

Cervical spine injuries, mostly due to high-energy trauma, can have a tremendous impact on each individual's ability and life. Severity can range from minor non-displaced fractures to osteo-ligamentous instability with concomitant spinal cord injury. Because of the unique anatomy, the cervical spine is divided into *upper cervical spine* (C0–C2) and *subaxial spine (C3–C7/T1)*. Prompt immobilization, rapid clinical and radiological assessment, and treatment are the key steps in dealing with these injuries. Definitive management is based on fracture pattern, stability, presence of neurological deficits, and other patient-related factors.

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36.1 Definition

Cervical spine injuries comprise injuries to the first to seventh cervical vertebrae or to the ligaments and discs connecting them.

36.2 Epidemiology

Between 19% and 51% of all spinal traumas account for cervical spine injuries. Approximately 2–3.5% of all trauma patients suffer from cervical spine injury. The incidence of spinal injury is doubled in unconscious trauma patients compared to alert trauma victims. Injury to the cervical spine occurs predominantly in the age of 20–45 years and 70–80 years. Injury to the spine occurs more frequently in male trauma patients (70%). Approximately 30% of cervical spine injuries affect the upper cervical spine, whereas 70% account for subaxial spinal injury. Occipital condyle fractures occur in 1–2% of severely injured patients and are mainly due to high-energy trauma. Fractures of C1 (atlas) occur in 3–13% of cervical spine trauma (Fig. 36.1), and in 40–44%, a combination of both C1 and C2 (axis) fractures is seen. Twenty-four percent of upper cervical spine fractures account for fractures of C2 with one-third showing odontoid fractures. A fracture of the odontoid process is the most common injury to the cervical spine in patients older than 80 years. The most frequently

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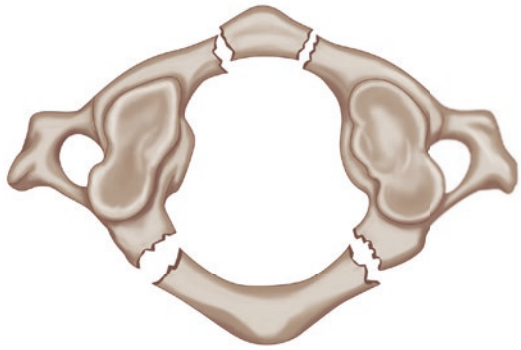


Fig. 36.1 Atlas fracture

seen fractures of the subaxial spine are fractures of C6 and C7, which account for 40% of all cervical spine injuries.

Localization (frequency)

Localization	Frequency
Occipital condyles	<3%
C1	70%
C2	30%
C3–C4	15%
C5	25%
C6	30%
C7	30%

36.3 Etiology/Pathogenesis

- Indirect force:
  - Axial compression
  - Hyperflexion/hyperextension
  - Translation
  - Rotation
- Direct force (stroke, blow) is rare.
- Typical mechanisms:
  - High-energy trauma (road accident, sport accident, industrial accident)
  - Fall in domestic setting
  - Low-energy trauma especially in geriatric patients (>65 years), typically hyperextension due to fall on head
- Predisposition:
  - Degenerative changes (osteochondrosis, disc herniation)
  - Ankylosing conditions (DISH, ankylosing spondylitis)
  - Tumor
  - Osteoporosis
  - Tendency to fall or gait disorder due to neurological conditions

36.4 Classifications

36.4.1 Upper Cervical Spine Fractures

Fractures of the **atlas (C1)** are traditionally classified according to the *Gehweiler classification*:

- Type I: Isolated anterior arch fracture
- Type II: Isolated posterior arch fracture
- Type III: Combined fracture of the anterior and posterior arch (*Jefferson fracture*):
  - Type IIIa: Transverse atlantic ligament intact, no or minimal horizontal displacement (stable fracture)
  - Type IIIb: Transverse atlantic ligament disrupted, horizontal displacement (unstable fracture)
- Type IV: Isolated fracture of the lateral mass
- Type V: Isolated fracture of the transverse process

In type 3b fractures according to the *Gehweiler classification,* it is helpful to classify the disruption of the transverse atlantic ligament according to the *Dickman classification*.

Fractures of the **axis (C2)** have to be divided into fractures of the odontoid, fractures of the axis ring resulting in spondylolisthesis, and atypical fractures of the axis body.

Fractures of the odontoid process are classified according to the *Anderson and D'Alonzo classification* accompanied by the *Eysel and Roosen classification* for subclassifying type II fractures*:*

- Type I: Isolated fracture of the odontoid tip or due to avulsion fracture of the alar ligament
- Type II: Fracture of the odontoid base:
  - Type A: Horizontal fracture
  - Type B: Fracture from anterosuperior to posteroinferior
  - Type C: Fracture from posterosuperior to anteroinferior

- Type III: U- or V-shaped fracture extending into the body of C2

Fractures of the axis ring ("hangman's fracture," traumatic spondylolisthesis) are typically classified according to the *Effendi classification*, which has been modified by *Levine and Edwards:*

- Type I: Isthmus fracture with displacement <2 mm, disc C2/C3 intact
- Type II: Isthmus fracture with displacement >2 mm, disc C2/C3 damaged, hyperkyphosis (Fig. 36.2)
- Type IIa: Displaced isthmus fracture with disc damage C2/C3, ruptured anterior longitudinal ligament, hyperlordosis type III: Displaced isthmus fracture with uni- or bilateral facet joint dislocation

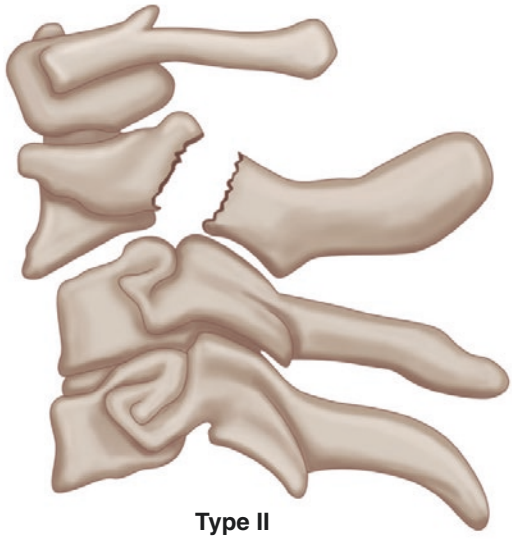


Fig. 36.2 Type II C2 fracture

36.4.2 Subaxial Spine Fractures

Because of high reliability and combination of morphological and clinical (neurological) findings, the *AOSpine classification for subaxial cervical spine injuries* is recommend for classification (Fig. 36.3). The classification consists of injury type (A/B/C), type of facet joint injury (F), neurological injury (N), and modifiers (M).

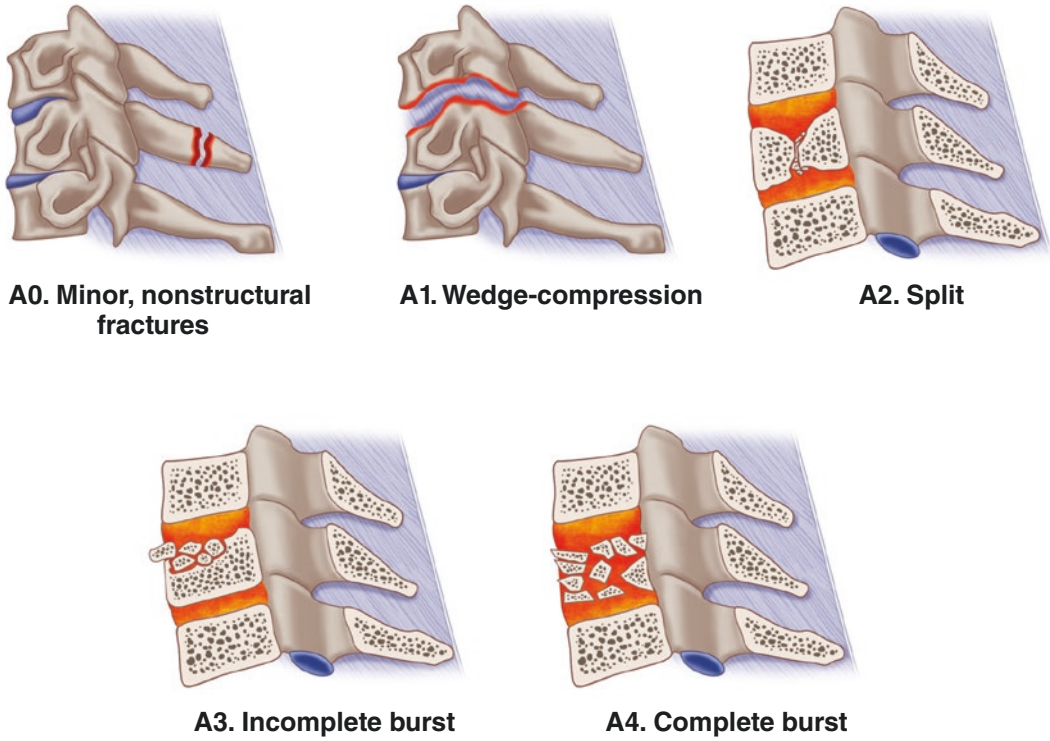
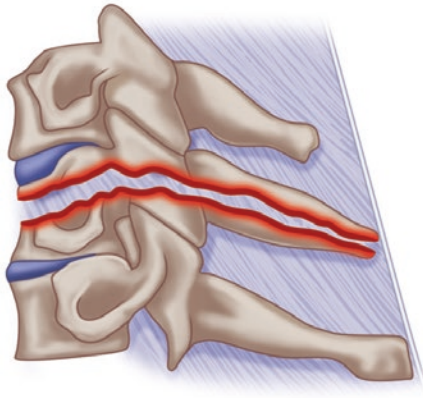
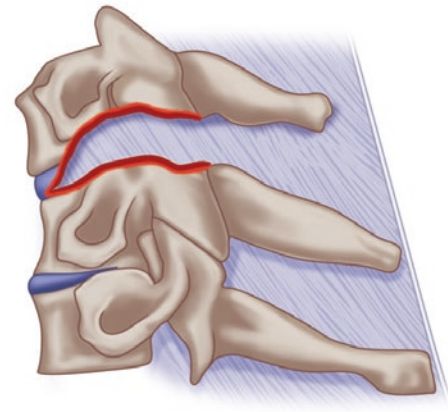


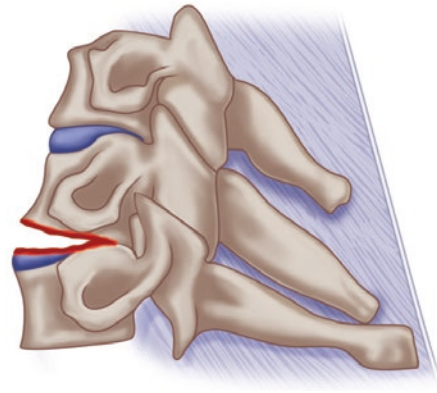
Fig. 36.3 The AO classification for subaxial spine fractures



B1. Posterior tension band injury (bony)

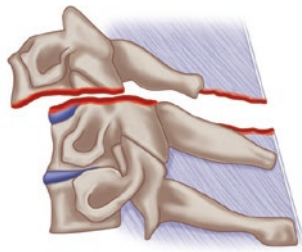


B2. Posterior tension band injury (bony capsuloligamentous, ligamentous)

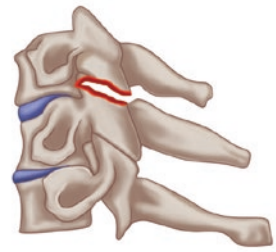


B2. Anterior tension band injury

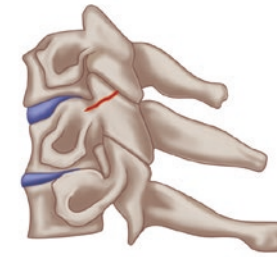
Fig. 36.3 (continued)



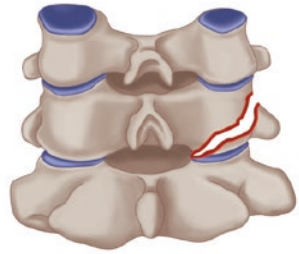
C. Translational injury in any axis-displacement or translational one vertebral body relative to another in any direction



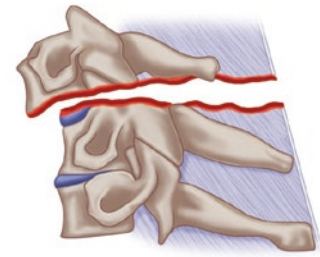
F1. Nondisplaced facet frature



F2. Facet frature with potential for instability



F3. Floating lateral mass



F4. Pathologic subluxation or perched/dislocated facet

Fig. 36.3 (continued)

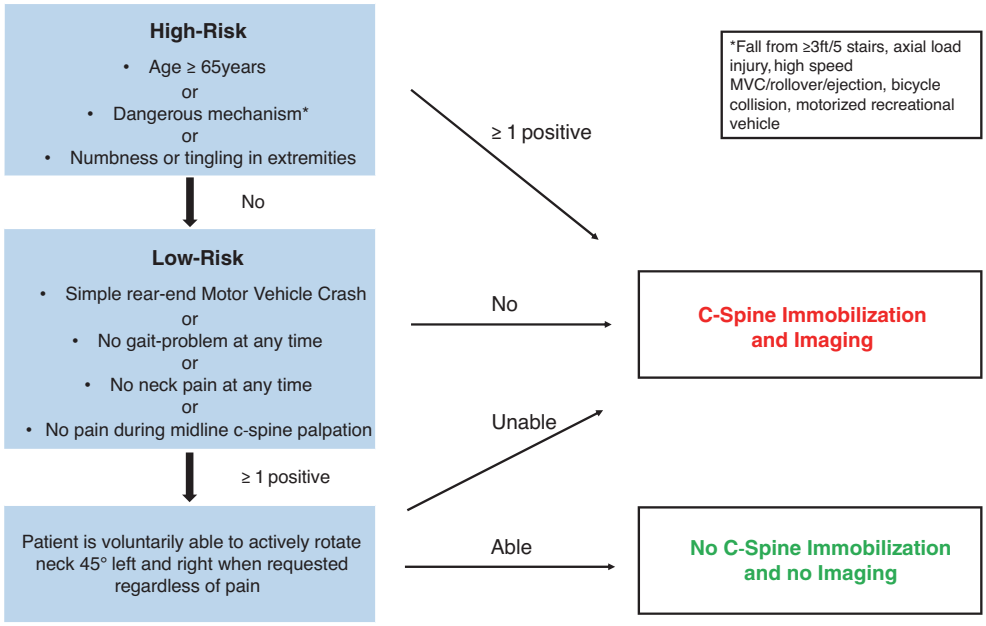
36.5 Diagnosis (Clinical and Imaging)

Cornerstone of diagnosis is the knowledge of the patients' history, analysis of trauma mechanism, clinical symptoms, and findings. Additional imaging may be required.

36.5.1 Clinical Examination

Clinical examination in alert patients is crucial and should always be performed prior to analgosedation. Examination must include a general trauma check (ATLS), tenderness to pressure along the cervical spine, and cervical muscle tension. In fully conscious patients (GCS 15), pain with rotation of the cervical spine can be evidential for spinal injury.

A full neurological status should be obtained including consciousness, cranial nerves, cerebral dysfunction, sensation of the occipital and cervical area, urinary and bowel function, as well as a full motor and sensory status. We recommend standardized assessment sheets as developed by the American Spinal Cord Injury Association. In alert patients, cervical spine injury can be excluded clinically with high reliability utilizing the NEXUS criteria or the figure for the Canadian C-Spine Rule and no imaging is required.



In severely injured patients (polytraumatized) with regularly reduced consciousness and concomitant injuries, a whole-body CT scan is highly recommended. Clinical examination should focus on obvious findings on the spine (bruises, steps, or offset of the spinal processes) as well as a trauma check according to ATLS.

36.5.2 Imaging

Generally, imaging is indicated in the following conditions:

- Polytraumatized patient
- Unconscious patient with suspected cervical spine injury
- Cervical spine symptoms after dangerous trauma mechanism
- Painful rotation of the cervical spine
- Clinical suspicion of structural injury to the cervical spine
- Sensory or motor deficits
- Preexisting ankylosing conditions

36.5.2.1 Radiography

Conventional radiography remains an imaging modality with ubiquitous availability, low radiation exposure, and low costs. However, the poor sensitivity and specificity of radiographs for detecting cervical spinal injuries must be considered, especially in the upper cervical spine. Radiographic imaging can be used for follow-up in known spinal conditions and as a screening tool.

36.5.2.2 Computed Tomography

Computed tomography (CT) is the gold standard in initial cervical spine injury imaging. Images with a maximum slice thickness of 1 mm should be obtained as well as at least 2-D reconstructions of the standard planes (coronal, sagittal). A 3-D surface reconstruction and axial plane reconstruction aligned to the superior end plate or parallel slicing in respect of the spatial planes of atlas (C1) and axis (C2), especially in upper cervical spine injuries, can be advantageous and should be performed whenever available. With CT imaging, injuries to the cervical spine can be detected with a sensitivity of 93.7–99% and a

specificity of up to 100%. Injuries to the ligaments or vertebral discs can be detected only indirectly and therefore are missed frequently.

36.5.2.3 Magnetic Resonance Imaging

MRI is the standard imaging tool for detection of injuries to the ligaments and discs as well as the spinal cord. Patients with unexplained neurological deficit after CT imaging must undergo immediate MR imaging. *Gehweiler* type 3 fractures of the atlas are without avulsion fracture in prior CT imaging, and fractures of the axis ring are with suspected instability of C2 and C3. In subaxial cervical spine injuries, prompt MR imaging is recommended in all facet injuries (F according to AOSpine) in order to rule out any discoligamentous instability.

In addition, MR imaging should be used if there is a suspected history of cancer in order to rule out metastatic disease or to differentiate acute bony injuries from subacute or old fractures, especially in patients with osteoporotic fractures.

36.5.2.4 Dynamic Radiographs

In neurologically stable patients with suspicion of segmental instability, dynamic radiographs should be obtained by an experienced surgeon in order to rule out instability of the cervical spine.

36.5.2.5 Angiography

Nowadays, CT angiography has replaced conventional angiography as the gold standard in posttraumatic cervical spine imaging. Patients with a cervical spine injury who are unconscious, especially after deceleration trauma, or who show clinical signs of a dissection of the vertebral artery, should receive angiographic imaging promptly. Angiography should be considered in all severely displaced fractures, fractures affecting the transverse foramen, and injuries to the facet joints (F3 and F4). In addition, angiographic imaging can be advantageous in preoperative planning and can detect vessel abnormalities such as a "high-riding" vertebral artery. MR angiography and CT angiography seem to be equal in terms of sensitivity, and modality decision should be made according to availability and in respect of the amount of radiation the patient may be exposed to.

36.6 Treatment

36.6.1 General Considerations

Spinal immobilization is crucial in patients with a suspected cervical spine injury. After admission, primary and secondary survey as well as treatment of concomitant injuries should be performed according to ATLS guidelines, including adequate analgesia according to the WHO ladder scheme. If bradycardia or arrhythmia in association with hypotension occurs, spinal shock must be considered and should be treated primarily by intravenous fluid resuscitation. If not successful, vasopressors and inotropes should be administered to maintain a mean arterial pressure (MAP) at 85–90 mmHg in order to prevent further damage to the spinal cord. Patients should receive drugs for the prevention of thrombosis (if cranial hemorrhage is absent). Primary reduction of displaced injuries should only be performed if direct external or internal fixation can be provided promptly after repositioning. For older patients, taking into account various perioperative risks based on their individual comorbidities, indication for surgery must be made carefully. If neurogenic deficits are present or imminent, prompt surgery is required. Apparent neurogenic symptoms (ASIA B-D) should be surgically treated (decompression) within up to 8 h. Unstable injuries (AO B and C) should be taken care of within 48 h. Stable fractures (AO A1 and A2) can be treated conservatively, and AO A3–A4 should be surgically managed in the interval. In patients with coagulopathy because of preexisting anticoagulative drug intake, normalization of hemostasis should be sought. In critical patients with unstable spine injury and neurogenic symptoms, surgery can be performed despite coagulopathy.

36.6.2 Upper Cervical Spine

1. Fractures of the atlas

   Nonoperative treatment is possible in *Gehweiler* type I, II, and V fractures according to the minimally displaced fractures of type IV and type IIIa with intact transverse ligament and usually consists of immobilization with a semirigid collar for 6–12 weeks.

   Operative treatment is recommended in types IIIb and IV with incongruence of the atlantoaxial or atlanto-occipital joints. Most cases with a type III injury can be addressed by posterior C1/2 fixation as described by Goel/Harms—either fusion or temporary instrumentation. Some displaced type III fractures can be addressed by direct osteosynthesis.

   Patients with a type IV injury should be managed with posterior fixation from C0 to C2.

   Whenever possible, fusion of the occipitocervical junction and C1/2 should be avoided due to limiting of spinal movement. Halo fixation is a valuable therapeutic alternative in atlas fractures that can be reduced and retained by this technique.

   As osteoarthritis of the atlanto-occipital and atlantoaxial joints develops slowly, halo fixation in displaced type III and IV fractures is not outweighed by its tremendous hazards in this patient population, however. Hence, these should be treated with a semirigid collar.
2. Odontoid fractures

   Nonoperative treatment with a semirigid collar for 6–12 weeks is possible in type I and non-displaced stable type II and type III fractures. Stability can be assessed by dynamic radiographic imaging. Close follow-ups are necessary in order to detect secondary instabilities.

   In young patients, unstable or displaced fractures of the odontoid process (type II) can be treated with direct anterior screw fixation. If anterior screw fixation is not possible (osteoporosis, large comminution zone, anatomic anomalies), atlantoaxial fusion is recommended in most of the cases. C1/C2 posterior fusion as described by Goel/Harms has shown to achieve the highest biomechanical stability, but other techniques as posterior or anterior transarticular screw fixation are valuable alternatives. Most unstable type III fractures will also primarily be treated with atlantoaxial fusion.
3. Fractures of the axis ring (hangman's fracture)

   Levine and Edwards type I and type II should be treated with a semirigid collar for 6–12 weeks. Extension position of the cervical spine is crucial for fracture compression due to ligamentotaxis by the preserved anterior ligaments.

   In selected cases, direct screw fixation as described by *Judet* can be performed in type II with intact disc C2/3. Type IIa and type 3 fractures which can be reduced closed are regularly treated with anterior plate fixation. In rare cases, where closed reduction is not sufficient, posterior spondylodesis in combination with screw fixation according to *Judet* can be necessary.

36.6.3 Subaxial Spine

In general, A0–A2 with less than 15° increase of the physiological kyphotic angle and A3 fractures with less than 15° increase of the kyphotic angle and little subjective discomfort should be treated conservatively. Gold standard in the treatment of these fractures is free range of motion without external immobilization and without extreme force to the spine. Physiotherapy and sufficient analgesia are required. Soft collars can be worn as a reminder and/or to reduce pain. Radiological follow-ups should be obtained fre-

quently. In mild SCIWORA symptoms, a Philadelphia collar is justifiable (see above).

In patients with an increase of the physiological kyphotic angle of more than 15°, A3–A4, B and C fractures, as well as compression of the spinal cord due to disc protrusion, epidural hemorrhage, or bone fragments, surgery is indicated. Analogous to the upper cervical spine, in displaced fractures or injuries, the disc must be judged radiologically or must be excised surgically prior to reduction in order to prevent further damage to the spinal cord. Depending on the fracture type, mono-, bi-, or multi-segmental fusion might be required. Principally, stabilization should be performed mono-segmentally using plates, or screw-rod systems. In comminuted fractures, bi- or multi-segmental stabilization might be required. Most commonly, the anterolateral approach according to *Smith-Robinson* is used. Advantage of this approach is the patient's supine positioning on the operation table, less blood loss, and safer anatomy which reduces soft-tissue damage. In the so-called "hooked dislocation," the dorsal approach might be required in order to achieve reduction and in conditions where decompression is necessary. Decompression of compressed spinal cord should be performed at the site of compression and additionally be stabilized.

36.6.4 Aftercare

Conservatively treated fractures should be left in external immobilization for 6–12 weeks. Active physiotherapeutic exercise should be started 12 weeks after initiation of treatment and after radiological proof of fracture consolidation.

In halo fixation treatment, frequent care and readjustment of the pins should be performed as well as dynamic radiographs after the end of the treatment to judge treatment success.

In operatively treated fractures, early physiotherapy-guided mobilization should be performed. External immobilization more than 2 weeks postoperatively is usually not necessary.

Take-Home Message

- Cervical spine fractures are common in polytraumatized patients and can be overseen.
- Immobilization is crucial in every patient with suspected cervical trauma or dangerous mechanism.
- CT imaging is the gold standard in the diagnostics of suspected cervical spine injury.
- Early decompression in patients with neurological deficits must be performed.
- Unstable fractures should be treated operatively as soon as possible.

Summary

Clinical management, diagnostics, and management of cervical spine injuries are demanding. Making use of defined protocols/algorithms can be advantageous for both the physician and the patient. Identifying the need for early immobilization and adequate imaging (CT/MRI) is crucial. Early decompression of patients with neurological deficit improves outcome significantly. Indication for surgery must be made carefully, taking both the patient's characteristics and injury characteristics into account.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. What are the typical mechanisms of cervical spine fractures?
   (a) High-energy trauma (road accident, sport accident, industrial accident)
   (b) Fall in domestic setting

(c) Low-energy trauma especially in geriatric patients (>65 years), typically hyperextension due to fall on head
(d) Low-energy trauma in young adults
(e) Strain due to sport

2. What are the high-risk factors for immobilization/imaging according to the Canadian C-spine rule?
   (a) Age over 65 years
   (b) Dangerous mechanism
   (c) Neurological deficit
   (d) Age below 30 years
   (e) Fall from more than 50 cm
   (f) Neck pain
3. What is the most common fracture of the cervical spine in patients aged more than 80 years?
   (a) Fracture of the odontoid process of the axis
   (b) Fracture of the atlas
   (c) Fracture of the body of the axis
   (d) Fracture of C7
   (e) Fracture of C3
4. According to the *AOSpine* classification, which fractures can be treated conservatively?
   (a) A0–A3 with less than 15° increase of the physiological kyphotic angle and no to moderate pain
   (b) A0 and A1 with less than 15° increase of the physiological kyphotic angle and no to moderate pain
   (c) A0–A2 with less than 15° increase of the physiological kyphotic angle and no to moderate pain
   (d) A0
   (e) B1–B2
5. What is a "hangman's fracture" and how is it classified?
   (a) Fracture of the axis ring, classification according to Effendi/Levine and Edwards
   (b) Fracture of the atlas, classification according to Gehweiler
   (c) Fracture of C2, classification according to AOSpine

Further Reading

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Yelamarthy PKK, et al. Management and prognosis of acute traumatic cervical central cord syndrome: systematic review and spinal cord society-spine trauma study group position statement. Eur Spine J. 2019;28(10):2390–407.

Longo UG, Denaro L, Campi S, Maffulli N, Denaro V. Upper cervical spine injuries: indications and limits of the conservative management in Halo vest. A systematic review of efficacy and safety. Injury. 2010;41(11):1127–35.

Low Back Pain

37

Mehmet Atif Erol Aksekili

Overview

Low back pain (LBP), which is not a disease but a symptom, can be caused by known and unknown abnormalities and many specific diseases.

37.1 Definition

Low back pain (LBP), which is not a disease but a symptom, can be caused by known and unknown abnormalities and many specific diseases. LBP can be defined as the pain seen between the lower costal border and the gluteal fold and often accompanied by sciatica.

37.2 Epidemiology

LBP is the number one reason for disability. Since it is the most common reason for abstaining from work and physical activities, it creates a huge medical and economic burden on the patient and society. It is the second most common reason for consulting a doctor after respiratory infections. Its general prevalence is about 18%.

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Its 1-month prevalence is 30%, while its lifetime prevalence is approximately 40%. Peak incidence is reached between the ages of 40 and 69 and starts to decrease after that age. The possibility of LBP's activity restriction increases with advanced age. It is 20% more common in women than men. The risk of recurrence within 1 year after an LBP attack is approximately 25%. In the socio-economically disadvantaged group, the possibility of persistent pain and affecting daily life activities is higher compared to socio-economically advantageous counterparts. Studies conducted in the last decade have revealed that the lifetime incidence of LBP in adolescence is approximately 40%. The incidence is higher for girls than boys (39% vs. 35.0%). Its prevalence increases with age. It is reported as 1% for 7 years old, 6% for 10 years old, and 18% for 15 years old.

37.3 Etiology/Pathogenesis

LBP is a complex and multifactorial symptom that develops as a result of the relationship between many risk factors. Risk factors in LBP formation can be listed as structural deficiencies of musculoskeletal tissues; excessive static and dynamic loading, emotional state, and environmental and behavioral factors; beliefs and expectations about what can happen with low back pain in the future and whether it can be con-

UG. Longo, V. Denaro (eds.), *Textbook of Musculoskeletal Disorders*,
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trolled; social system; and social approach to the patient after LBP develops. The individual and variable response to musculoskeletal injury is a determining factor for LBP development. To explain the complexity of LBP, instead of the purely biomedical approach, the biopsychosocial approach can be applied as a framework.

Sources of pain in LBP encompass three different regions as specific lumbosacral, radicular, and reflected pain. Most frequently, no identifiable underlying pathology can be found at a rate of 90%. The rate of LBP seen with identifiable pathologies such as traumatic or osteoporotic fracture, stenosis, and lumbar disc hernia is approximately 5%. The rate of LBP that directly affects spinal structures such as neoplasia, metastasis, and infection is 1%. Approximately 2% of LBP comes from visceral organs that we call reflected pain.

Studies on the genetic origin of LBP have focused on genes that provide the formation of cartilage and bone structures and are accompanied by morphological signals in MRI. A genetic relationship has been identified between intervertebral disc height loss and disc herniation. Only a small part of genetic influences are caused by genes that affect disc degeneration. Therefore, it suggests that genetic variants of pain perception, signal transduction, psychological processes, and immune genes contribute to the heritability rate of chronic back pain. Genetic variability in pain transmission pathways may contribute to individual sensitivity to pain.

The pathophysiological mechanism underlying LBP has not been fully revealed. Pain in LBP can be caused by ligaments, facet joints, spinal nerve roots, vertebral periosteum, annulus fibrosus, and vascular structures. The basic sensation of the lumbar spinal structures is provided by the sinuvertebral nerve. The sinuvertebral nerve makes sensory innervation of the posterior longitudinal ligament, posterior annulus fibrosus, epidural vessels, posterior vertebral periosteum, and dural nerve root sleeve. The posterolateral part of the discs is innervated by the adjacent ventral primary rami and gray rami communicators near their junction with the ventral primary rami. The lateral part of the discs is innervated by branches from the rami communicantes. The medial branch of the posterior primary ramus of the spinal nerve provides the innervation of the facet joint, the deep and superficial lumbar muscles, and the overlying skin and interspinous ligament. Repetitive movement patterns, strenuous activity, poor posture, and sports activities may cause microtrauma or stress on lumbar anatomical structures. Muscles and myotendinous junctions can be directly injured by a crushing force applied to the spine, and this will lead to lumbar strain. These forces can cause stress in the facet joint capsule, annular fissure, and/or ligament damage. Substance P, which causes lumbar pain syndrome as a result of these injuries, may provoke the neurohumoral cascade that will lead to the release of prostaglandins, leukotrienes, vasodilators. Pain affecting the patient is ultimately perceived by the central nervous system. In the central nervous system, LBP is shaped by all of the factors that make up the memory and perception of pain, such as genetically determined perception and conduction capacity, familial and social conditioning, work-related socioeconomic factors, and the individual's own experiences. All these factors should be considered when the patient comes with a complaint of LBP.

37.4 Classifications

LBP is divided into three categories according to the duration of symptoms. If the symptoms last less than 6 weeks, it can be defined as acute LBP; if it lasts for 6–12 weeks, it can be defined as subacute; and if it lasts longer than 12 weeks, it can be defined as chronic LBP.

LBP is divided into four main categories according to the diagnostic triage approach (Fig. 37.1). These categories are:

1. Nonspecific LBP
2. Radicular syndromes
3. Specific spinal disorders
4. Visceral disorders

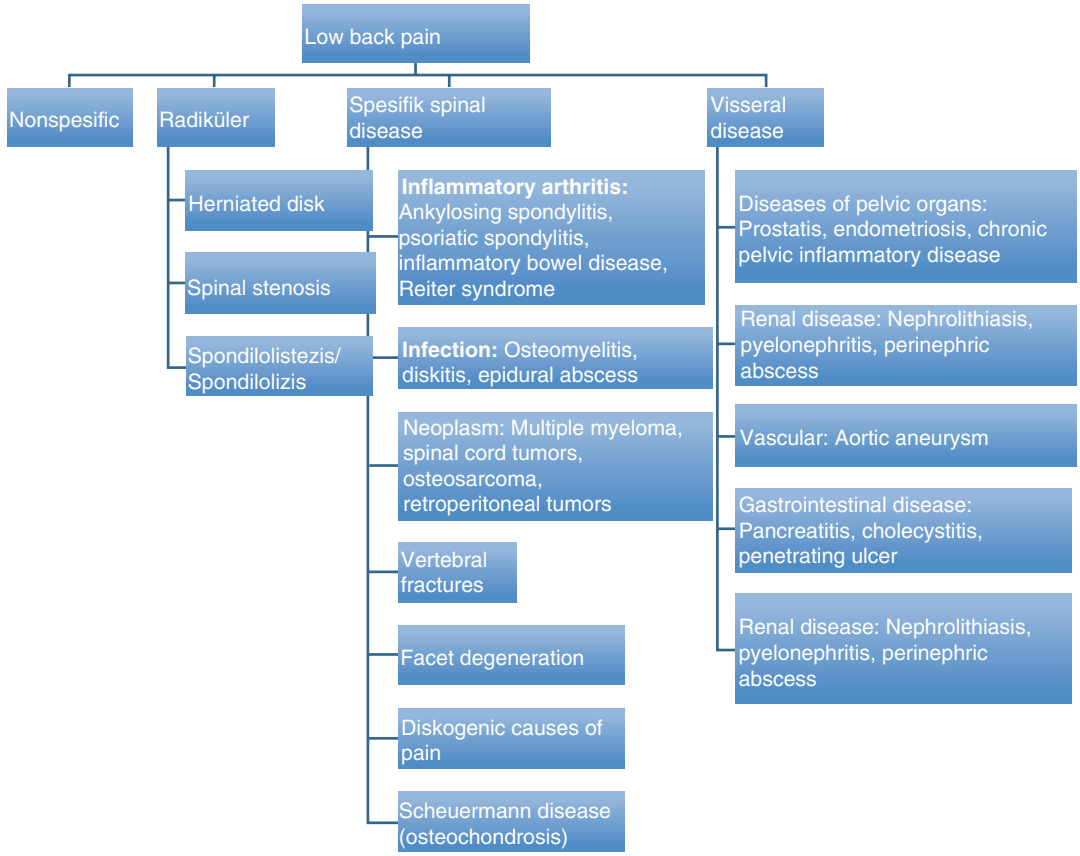

Fig. 37.1 Etiological classification of LBP

37.5 Diagnosis

37.5.1 Anamnesis

Since approximately 85% of the patients who apply for LBP in primary care are nonspecific LBP, it may not be possible to reveal the exact cause. Dividing the symptoms into three as acute, subacute, and chronic according to the duration of the symptoms helps us to determine the treatment. The location and spread of pain should be differentiated as axial or radicular. The level of pain should be determined by specific scales (visual analog scale or numerical rating scale score) as the current, worst, and average levels. The characteristics of pain should be questioned for burning, lancinating, aching, numbing, and electric shock sensations. Questioning factors that increase and improve pain, such as sitting, walking, climbing stairs, and reaching, help to clarify the differential diagnosis. Documentation of a previous history of similar symptoms reveals whether the nature of the symptoms is intermittent or not. Previous diagnostic studies and patient's response to previous treatments should be questioned in terms of subsequent treatments. The pain and functionality of the patient during work and daily life activities may affect the treatment. Besides, the anamnesis should include screening questions regarding structural symptoms that indicate a potentially progressive or unstable cause of pain such as cancer, infection, trauma, and neurological problems (red flags) (Table 37.1).

Table 37.1 Explanation of LBP according to the flag model

Red flags	*Indicates potential serious pathology*
1. Cancer-related signs	A. History of cancer B. Unexplained weight loss >10 kg within 6 months C. Age over 50 years or under 18 years old D. Failure to improve with therapy E. Pain persists for more than 4–6 weeks F. Night pain or pain at rest
2. Infection-related signs	A. Persistent fever B. History of intravenous drug abuse C. Severe pain D. Lumbar spine surgery within the last year E. Recent bacterial infection • Urinary tract infection or pyelonephritis • Cellulitis • Pneumonia • A wound in the spine region F. Immunocompromised states
3. Cauda equina-related signs	A. Urinary incontinence or retention B. Saddle anesthesia C. Anal sphincter tone decreased or fecal incontinence D. Bilateral lower extremity weakness or numbness E. Progressive neurologic deficit
4. Vertebral fracture-related signs	A. Prolonged use of corticosteroids B. Age greater than 70 years C. History of osteoporosis D. Mild trauma over the age of 50 years (or with osteoporosis) E. Recent significant trauma at any age
Yellow flags	*Include risk factors for chronicity and psychosocial issues*
	Expecting negative results from treatment, delayed return to work, anxiety, fear, negative thoughts about the recovery of pain, overreliance on passive treatments
Orange flags	*It includes psychiatric risk factors in the development of LBP*
	Depression, personality disorders
Blue flags	*Occupational problems*
	Believing that the job is too laborious and will likely result in further injury. Believing that the workplace supervisor and colleagues are not supportive
Black flags	*System or contextual obstacles*
	Heavy work with little opportunity to change tasks. Conflict with insurance personnel for the alleged injury

37.5.2 Physical Examination

Inspection: Inspection of the thoracolumbar spine gives information about posture and spine alignment (kyphosis, lordosis, scoliosis). Gait pattern gives information about etiologies; that is, antalgic gait indicates osteoarthritis, steppage gait indicates drop foot, duck-like gait indicates weak hip abductors, shear gait indicates spastic paralysis, and when the patient walks in an anterior trunk flexion posture, it indicates spinal stenosis. The skin should be evaluated in terms of scars, swelling, redness, trauma, or inflammation marks. While hair growth in the sacral area may indicate the underlying vertebral anomaly, dermatomal rashes indicate zoster infection.

Palpation: Although the tenderness localized in the spinous processes is nonspecific, it may indicate an abscess, an epidural tumor, or a vertebral compression fracture. Paraspinal tenderness may indicate facet syndrome. Copeman nodules can be palpated in the sacroiliac area.

Assessment of range of motion: Limitation of range of motion or development of pain with movement can guide us about the cause of pain. The normal range of motion is 90° forward flexion, 30° extension, 60° lateral rotation, and 25° lateral flexion. While the pain caused by extension or lateral rotation suggests facet arthropathy, pain caused by forward flexion suggests discogenic etiology.

Neurological examination: Lumbar and lower extremity motor and sensory examination gives information about the function of nerve roots and the level of pathology. Ankle dorsiflexion (L5), thumb extension (L5–S1), knee flexion (L5–S1), hip flexion (L2–L4), and knee extension (L2–L4) constitute the basic muscle strength examination. While the patellar tendon reflex shows the functional integrity of the L2, L3, and L4 roots, the Achilles tendon reflex shows the functional integrity of S1. Reflexes may not be obtained in people over 60 years old.

Straight leg raising test: While the patient is in the supine position, the examiner lifts the affected leg by holding the heel without bending the knee.

The occurrence or worsening of radicular symptoms between 30 and 70° indicates a positive test. The test suggests an L5–S1 lesion. Pain localized to the back of the thigh during the maneuver suggests hamstring tension.

Lasègue's test: It is a modification of the SLR test. When the pain occurs or worsens during the SLR test, the leg is lowered 5–10° and the ankle is brought to dorsiflexion. The occurrence of pain indicates a positive test.

Femoral stretching test: While the patient is in the prone position, the leg is slightly lifted and the knee is flexed. The development of pain in the anterior aspect of the thigh suggests L2–L4 radiculopathy.

Cross straight leg raising test: It is the occurrence or worsening of radicular symptoms in opposite leg while performing a straight leg raising test. It is more specific than SLR but not as sensitive.

Bragard sign: During the SLR test when pain is triggered, examiner brings the ankle in dorsiflexion position, resulting in worsening of symptoms.

Patrick test (FABER test): While the patient is in supine position, examiner lifts the affected leg by holding the heel, then the knee is brought to flexion position, the hip is externally rotated, and the ankle of affected leg is placed on the opposite knee. Pain in the groin during this maneuver suggests hip pathology, and pain in the hip and waist suggests sacroiliac joint pathology.

Nonphysiological back pain: Low back pain with no organic cause, often related to secondary gains or psychosocial problems. Waddell identified five signs to differentiate nonphysiological pain; the presence of three suggests nonphysiological back pain (Table 37.2).

Table 37.2 Waddell's nonorganic signs

Tenderness	It is not related to a specific skeletal or neuromuscular pathology; often superficial and/or nonanatomic
Simulation tests	These tests give the patient the impression of a specific examination, even though it has not been performed
Distraction tests	Routinely demonstrated positive physical sign is checked again while the patient is distracted; if the symptom disappears when the patient is distracted, an inorganic component may be present
Regional disturbances	Dysfunction involving a large area of body parts (e.g., sensory, motor) that cannot be explained anatomically; must be distinguished from multiple nerve root involvement
Overreaction	It may be in the form of disproportionate verbalization, facial expression, muscle tension and tremors, collapse, or sweating; the decision must be made carefully, and the supervisor's own emotional response must be minimized

37.5.3 Diagnostic Tests

Further diagnostic studies are not required in nonspecific LBP and radicular syndromes. Diagnostic studies are required when specific spinal pathology or visceral disorders are suspected at initial clinical evaluation. In diagnostic imaging methods, the first choice is two-way direct graphs. It gives information about diseases such as osteoporotic fractures, benign and malignant tumors, osteomyelitis, and spondyloarthritis. If there is a suspected bony lesion on direct radiography, we can have detailed information about the pathology with computerized tomography (CT). With CT, we can evaluate the disc, ligaments, adipose tissue, and nerve roots. CT provides valuable information in terms of surgical planning, especially in patients with trauma, spinal **deformity**, and stenosis. Since radiation exposure is high in CT, it should be used when it is the only option that will provide a diagnosis. Pathology can be better characterized by MRI when soft-tissue abnormalities, disc pathologies, myelopathy, infection, tumors, and stenosis are suspected. The diagnostic value of laboratory studies is low, except for patients with red flag findings. For patients with suspected malignancy and infection, complete blood cell count, C-reactive protein (CRP), and sedimentation rates are indicated.

37.5.4 Extrinsic Causes of LBP

Intrinsic and extrinsic factors could cause LBP. The former are due to traumatic, pathologic, or degenerative conditions of the lumbar spine. The latter are caused by visceral diseases that produce radiating pain to the spine. Notwithstanding degenerative disc disease is the most common cause of LBP, other conditions are elusive in diagnosis and resulting in a treatment failure. Degenerative disc disease could be present in patients of all ages, and in case of a clear-cut diagnosis, it is attributed as the primary cause of LBP. The clinician needs to focus the attention on other underlying pathologies to rule out extrinsic causes of LBP.

37.5.5 Vascular Disease

The most frequent vascular disease that produces LBP or symptoms resembling sciatica is the abdominal aortic aneurysm (Fig. 37.1).

This condition leads to progressive and irreversible dilatation of the aortic wall. It is usually provoked by atherosclerosis and determines the flattening of the muscular and elastic fibers of the aorta. The systolic pressure progressively increases the artery diameter due to the reduction of elasticity of the aortic wall. It could be asymptomatic in the early stage of the disease, even if a persistent deep-seated lumbar pain unrelated to physical activity could be present. One of the earliest symptoms of this situation is the LBP. In case of increased aortic diameter, the artery could irritate the periosteum and the intervertebral disc with every systole, causing LBP. In the late stages, the aortic wall could tear, causing extensive bleeding and death in 90% of cases. Moreover, in case of significant dilatation, intra-aortic thrombosis may occur, with consequent stroke due to embolization of atherosclerotic and thrombotic debris.

Another vascular disease that causes LBP is the insufficiency of the superior gluteal artery. This condition may produce a buttock pain with a claudicant character, aggravated by walking and relieved by standing still. A sciatic pattern of the pain may be present. However, bending, lifting, and other activities that involve spine movements do not exacerbate the pain. The intermittent claudication (with intermittent calf pain) caused by peripheral vascular deficiency could simulate sciatic pain. In this condition, the pattern is specific because walking increases pain while patients are relieved by standing still. These symptoms are similar to pure spinal stenosis claudication, but in this case, the pain is not relieved by standing still. In the case of vascular LBP, the pain is present during the day, and there is no muscular contracture.

37.5.6 Viscerogenic LBP

Viscerogenic back pain may be derived from disorders of the bowel, liver, kidneys, pelvic viscera, lesions of the lesser sac, and retroperitoneal tumors. Otherwise, LBP is usually accompanied by other symptoms in case of visceral disease. The most important difference with intrinsic causes of LBP is that the pain is not exacerbated by activity, and it is not relieved by rest. The Luschka nerve innervates the articular apophyses of L1 and L2. The pain is due to overloading of this joint. This pain reflects the topography of the roots L1–L2 (the lateral region of the lumbar spine, upper buttock, and groin area). For this reason, this pain is often attributed to appendicitis or abdominal conditions. One of the identifying signs of Luschka nerve irritation is the "pincé roulé sign." Picking up the skin (skin and subcutis of the gluteal or inguinal region) and moving it under the finger, the patient experiences significant pain.

37.5.7 Kidney Diseases

Ureteral stones and pyelonephritis could cause LBP on the affected side.

37.5.8 Liver Diseases

The hepatosplenomegaly in cirrhosis patients or toxic hepatopathies could increase the Glisson's capsule tension. Due to the innervation of this

capsule, a radiating continuous and persistent pain to the right flank could be referred by the patient. Moreover, cholelithiasis and cholecystitis could present the same pain pattern.

37.5.9 Intestinal Diseases

Transverse colon and colic flexure diseases (colitis, tumors, and malabsorption) could produce LBP.

37.5.10 Gynecological Diseases

Polycystic ovary syndrome (PCOS), endometriosis, retroverted uterus, and gynecological tumors could cause LBP. Uterus and ovaries are attached to the suspensory ligaments. In case of irritation of these structures, radiating pain to the spine could be present.

37.5.11 Psychogenic LBP

Patients that suffer from LBP or cervical pain nonresponding to conservative treatment and NSAIDs are frequent. The lumbar and the cervical spine are the most frequent contracture sites in case of psychological problems. Pure psychogenic back pain is rarely seen in clinical practice. Instead, diffuse and nonmetameric pain is more commonly observed in depressed patients. The clinician must learn to recognize the presence of an emotional breakdown, and it is essential not to forget that an underlying organic disease could be present.

37.6 Treatment

Since biopsychosocial factors contributing to LBP are prominent, the treatment should be viewed from the same perspective. With this approach, treatment should include both physical therapy and other dimensions (psychological, social, or occupational). Patients should be taught to manage their condition on their own. The patient should be informed about the natural course and prognosis of the disease, importance of staying active, and multimode self-care in intermittent symptoms. Unnecessary and complicated treatments should be avoided by applying a step-by-step treatment approach. Relieving pain and preserving function are the main goals of treatment. Acute LBP improves by 90% in the first 6 weeks.

In chronic LBP, self-management and physical therapy should be recommended before pharmacological treatment. Supervised exercise programs are effective in preventing LBP and chronic LBP.

Physical therapy and rehabilitation can also be used together with other methods to increase functionality and pain management. Stretching exercises are effective in reducing pain, and strengthening exercises are effective in increasing functionality. The effectiveness of modalities such as waist school, superficial cold or hot application, massage, and TENS has not been demonstrated by randomized controlled trials.

Pharmacologic treatment: NSAIDs are effective in short-term pain treatment in both acute and chronic pain. Acetaminophen up to 4 g per day has been found to be as effective as NSAIDs in acute pain, but its effectiveness is lower in chronic pain. It is recommended that NSAIDs should be used for the shortest duration—the lowest dose due to side effects. Muscle relaxants are also effective in acute low back pain. Opioid analgesics should be reserved for severe pain that cannot be controlled with the aforementioned agents. Tricyclic antidepressants have also been shown to be effective in chronic LBP. It is particularly effective in chronic LBP patients with fibromyalgia. Antiepileptics can also be used in the treatment of chronic LBP. Gabapentin has been shown to have analgesic efficacy in chronic LBP accompanied by radicular pain.

Psychosocial approaches are suitable for patients with acute pain who are candidates for chronicity with a yellow flag sign as well as patients with chronic pain. Behavioral cognitive therapy and biofeedback are examples of psychosocial approaches.

Acupuncture, one of the complementary medicine approaches, has been found to be effective

in reducing pain and increasing functionality from the treatment of chronic low back pain.

Minimally invasive approaches such as facet injection, medial branch nerve block, RF neurotomy, sacroiliac joint injections, and epidural steroid injections can be used in patients who do not respond to multidisciplinary conservative treatment methods.

37.6.1 Surgical Methods

Surgical treatment is not recommended for nonspecific LBP unless the underlying anatomical cause is revealed. Surgery is applied as the last-step treatment depending on the cause of the specific pathology. Surgery is indicated in some patients with red flag symptoms, such as cauda equina syndrome. In cauda equina syndrome, decompressive surgery is required within 48 h to preserve neurological functions.

When disc herniation is diagnosed, disc excision can be performed using methods such as microdiscectomy, percutaneous suction discectomy, and percutaneous laser discectomy. Spinal decompression and fusion surgery can be performed when there are symptoms of spinal instability and stenosis.

Take-Home Message

- It is the most common cause of avoiding physical activity and work life.
- It is also very common in the adolescent period.
- In the last decade, in LBP etiology, the biopsychosocial model which explains the etiology more comprehensively has been emphasized instead of the simple injury model.
- Nonspecific LBP is the most common type with a rate of 85–95%.
- Red flags are symptoms that indicate infection, fracture, malignancy, inflammatory disease, and neurological disease by causing LBP.
- If patients presenting with LBP have red flag findings, further investigation is required.
- Acute LBP resolves spontaneously in 80% in 2 weeks.
- The importance of keeping the patient active during the treatment process should be emphasized.
- Unnecessary treatments can be avoided by applying a step-by-step treatment approach.
- The treatment should be specified to the patient with a multidisciplinary approach by evaluating pharmacological treatment, physical therapy and rehabilitation, psychological treatment, and social aspects.

Summary

LBP is the most common cause of disability globally. The burden on the individual and society continues to increase. Therefore, it is a public health problem. In most patients, LBP is short-term, and specific nociceptive causes cannot be revealed. For this reason, it is called nonspecific. The persistence of underlying physical, psychological, and social factors increases the likelihood of chronicity. Imaging and laboratory examination may be required, based on history and physical examination, and red flag findings. NSAIDs and myorelaxants are recommended as first-line treatment in pharmacological treatment. If there is no response to treatment, a minimum dose of opioids may be prescribed. Physical therapy and rehabilitation treatment should be the other dimension of the treatment. During the treatment process, it is aimed to relieve the pain, prevent it from becoming chronic, and return to social life as soon as possible.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. A 30-year-old man feels pain in his lower back while lifting a heavy object at work 3 days ago. Because of this pain, he cannot go to work. He states that this pain is for the first time. On physical examination, there is tenderness in the lumbar paraspinal muscles with palpation. Neurological examination is natural. Which of the following should not be applied?
   (a) Range-of-motion exercises
   (b) Lumbar X-ray
   (c) NSAID
   (d) Reassurance
   (e) Suggesting to remain active in daily life
2. Which of the following is the probability that acute low back pain will improve within 6 weeks?
   (a) 50%
   (b) 60%
   (c) 70%
   (d) 80%
   (e) 90%
3. Which of the following nerves innervates the posterior annulus fibrosus?
   (a) Dorsal ramus ventral branch of spinal nerves
   (b) Dorsal ramus lateral branch of spinal nerves
   (c) Sinuvertebral nerve
   (d) Gray ramus communicans
   (e) Ventral ramus of spinal nerves
4. A 40-year-old male patient applies to your clinic with complaints of back pain and both leg pain for 3 days. Which of the following physical examination findings suggests that the complaint is associated with L5–S1 radiculopathy?
   (a) Patrick test
   (b) Gaenslen test
   (c) Straight leg lift test
   (d) Femoral stretching test
   (e) Hoffmann's test
5. Which of the following findings does not require a diagnostic workup in the patient who applied to your clinic with low back pain complaint?
   (a) Pain persists for more than 4–6 weeks
   (b) Immunocompromised states
   (c) Bilateral lower extremity weakness
   (d) Prolonged use of corticosteroids
   (e) Tenderness to light touch

Further Reading

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Lumbar Herniated Intervertebral Disc

38

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and Hiroyuki Yoshihara

Overview

Lumbar herniated intervertebral disc (LHID) is a common cause of low back pain and is a source of enormous healthcare expenditures all over the world. Common symptoms associated with LHID include low back pain, radiating leg pain, paresthesia in a dermatomal distribution, and weakness in a myotomal distribution according to the level affected. It is thought that the symptomatology associated with LHID is caused by both direct compression of lumbar nerve roots and local inflammatory changes that irritate lumbar nerve roots. Diagnosis of LHID begins with physical examination, but imaging modalities such as plain radiographs and magnetic resonance imaging (MRI) are often necessary to confirm the diagnosis. Various conservative modalities, including anti-inflammatory medications, physical therapy, and epidural steroid injections, are often effective in treating this pathology; however, surgery is indicated to remove the herniated disc and decompress neural elements when conservative treatment fails. Techniques in surgical treatment include conventional lumbar discectomy, microdiscectomy, and endoscopic discectomy.

38.1 Definition

While the clinical manifestations of lumbar radiculopathy were first described by both ancient Greeks and Egyptians, the distinct understanding of lumbar herniated intervertebral disc (LHID) can be attributed to Mixter and Barr, who were the first to identify the connection between intervertebral disc pathology and symptoms of lumbar radiculopathy. In the early twentieth century, space-occupying lesions in the lumbar spine were often surgically excised with symptomatic improvements; however, they were routinely misdiagnosed as enchondromas or osteochondritis dissecans. In 1932, Mixter and Barr performed the first surgery carrying a pre-

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operative diagnosis of "ruptured intervertebral disc" after noting that the symptoms of lumbar radiculopathy arose too acutely to be associated with tumor or mass. Since then, technological advancements of imaging studies such as computed tomography (CT) and magnetic resonance imaging (MRI), as well as improvements in surgical technique, have helped further understanding of this pathology.

Today, the combined task forces of the North American Spine Society (NASS), the American Society of Spine Radiology (ASSR), and the American Society of Neuroradiology (ASNR) define an LHID as a "localized or focal displacement of disc material beyond the limits of the intervertebral disc space." Using this definition, it is important to note that LHID may or may not cause clinical symptoms. In fact, spine literature has demonstrated high rates of lumbar disc pathology in asymptomatic individuals, including LHID rates of up to 36% in asymptomatic patients over 60 years of age. Moreover, it is possible to distinguish between degenerative disc disease/discopathy and discal herniation. In the former, the intervertebral disc progressively loses its height reducing the distance between end plates. This condition could not produce radicular symptoms, as it is present in the majority of the elderly population. Discal herniation instead affects young people in working age.

38.2 Epidemiology

The incidence of symptomatic LHID in the general population is about 5–20 cases per 1000 annually with a prevalence of around 1–3%. LHID more commonly affects men compared to women and usually afflicts adults in the fourth or fifth decade of life.

Disc herniations are much more common in the lumbar spine relative to the thoracic spine due to increased biomechanical forces caused by increased mobility of the lumbar spine (Fig. 38.1). A huge majority of LHIDs (roughly 95%) occur at the L4–L5 level or the L5–S1 level, largely due to the increased mobility of the spine at these levels.

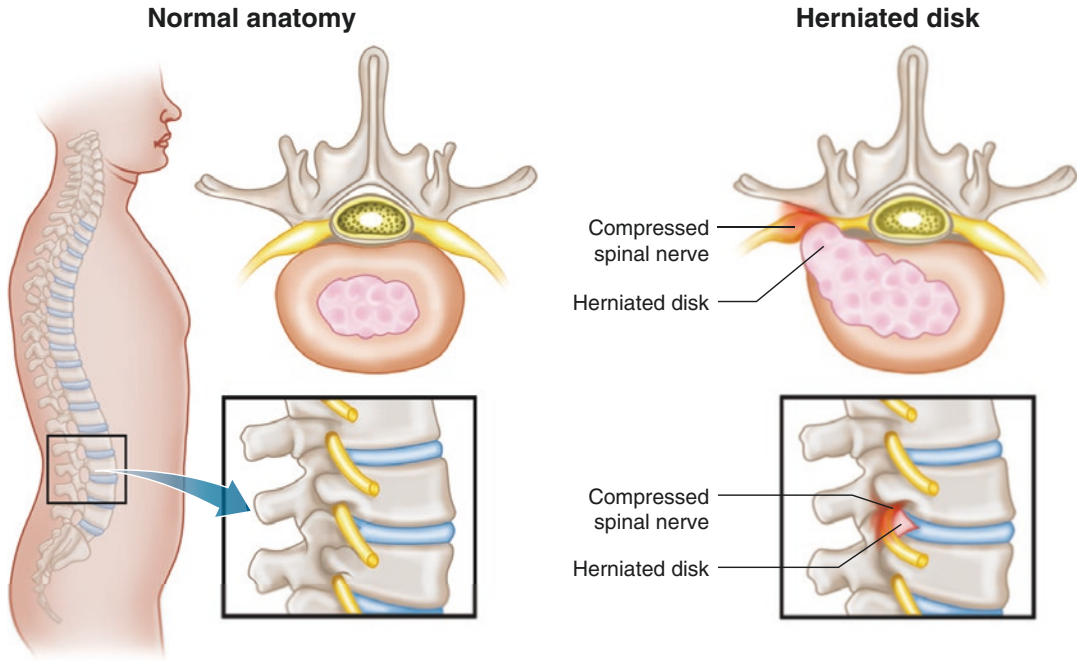

Fig. 38.1 Lumbar herniated intervertebral disc

According to the 2010 Global Burden of Disease Study, back pain is the number one cause of disability in the world and is responsible for over 100 billion dollars in healthcare expenditures every year in the United States alone. LHID is one of the most common causes of low back pain, accounting for as much as 40%, and is responsible for significant healthcare expenditures worldwide.

38.3 Pathogenesis

Intervertebral discs are composed of an inner nucleus pulposus, surrounded concentrically by the annulus fibrosis.

A herniation of the nucleus pulposus through the annulus fibrosis can occur from a variety of causes including degenerative changes, trauma, or congenital disorders and can cause a compression of neural elements triggering pathologic symptoms. The most common cause of LHID is degenerative changes in the lumbar spine, leading to degenerative disc disease and weakening of the annulus fibrosis.

Dehydration of the intervertebral disc has been shown to contribute to degenerative disc disease and increase the risk of disc herniation. Furthermore, it has been shown that degeneration of the intervertebral disc is associated with the formation of clefts within the annulus fibrosis as well as increased apoptosis of fibroblast-like cells. It has been postulated that these degenerative changes weaken the outer layer of the annulus, predisposing to disc herniation. The dehydration of the disc leads to a specific MRI pattern called "black disc" Several risk factors have been established for the development of LHID, including genetic factors, microtrauma, and medical comorbidities. It has been estimated that the origin of LHID is driven mostly by genetic factors. Literature has shown that allelic variants of collagen IX and XI proteins, that are abundantly present in both the nucleus pulposus and annulus fibrosis, show a much higher propensity for intervertebral disc disease, which is the most common cause of LHID. Other genes have been implicated in the increased likelihood of disc degeneration and LHID as well, including matrix metalloproteinases, apoptotic factors, growth factors, various cytokines, and even vitamin D.

In addition to genetic predisposition, microtrauma caused by excessive axial loading onto the intervertebral disc has also been implicated in advancing disc degeneration and LHID. Obesity has also been shown to be associated with accelerated intervertebral disc degeneration due to increased axial loads on the lumbar spine. It has been shown that obese patients are both more likely to have a disc herniation and more likely to have recurrent herniations.

LHID is believed to cause clinical symptoms due to a combination of mechanical compression of neural elements and local inflammation. Herniation of the nucleus pulposus through the annuls fibrosis can compress local nerve roots, which may lead to local ischemia and radicular pain. In addition, disc herniation can cause significant local inflammatory changes, which are designed to resorb the herniated tissue, but also cause inflammatory pain to local nerve roots as well. The intervertebral disc is composed of immune-privileged tissue. As such, extrusion of the nucleus pulposus into the epidural space causes a significant inflammatory reaction including increased vasodilation, vascular permeability, and recruitment of various inflammatory cytokines to the injury site.

Many inflammatory cytokines have been implicated in the symptoms of lumbar radiculopathy, but the exact mechanisms by which these proteins cause symptoms are not yet completely understood. Increased expression of TNF-alpha has also been shown to be directly related to symptoms of radiculopathy in several studies.

38.4 Classifications

The combined task forces of the NASS, ASSR, and ASNR have published consensus guidelines, which recommend describing herniations based on their anatomic location and also based on the morphology of the herniation.

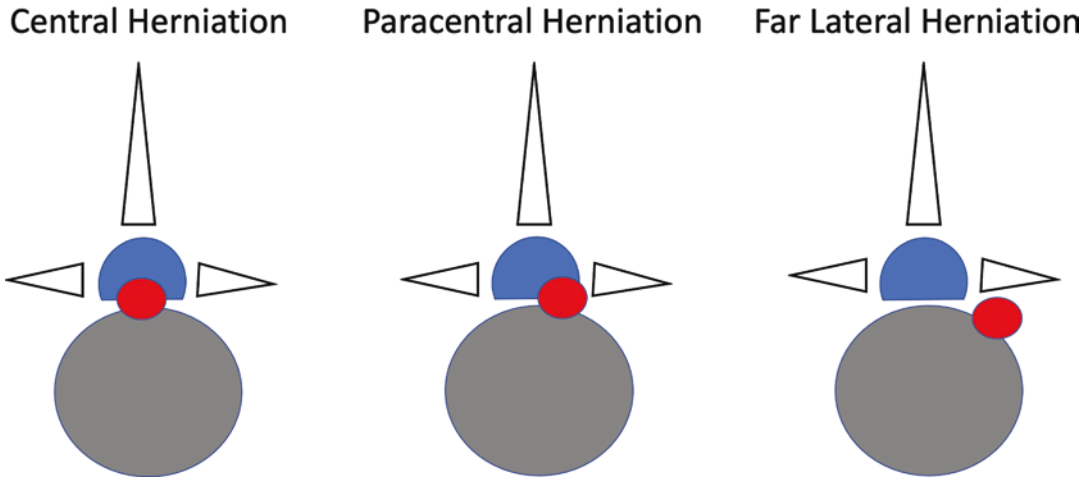

Fig. 38.2 Herniations may be described as either a central herniation, a paracentral (or posterolateral) herniation, or a far-lateral herniation

The anatomic location of the herniated disc carries clinical significance, as disc herniations cause different symptoms based on where they occur. Herniations may be described as either a central herniation, a paracentral (or posterolateral) herniation, or a far-lateral herniation (Fig. 38.2). In general, central disc herniations are typically associated with significant axial back pain but are less likely to cause radicular symptoms. However, large central herniations are capable of causing cauda equina syndrome, a devastating condition caused by compression of the spinal nerve roots distal to the spinal cord, leading to saddle anesthesia and severe autonomic dysfunction.

In contrast, paracentral disc herniations often cause radicular symptoms because of the proximity to the local nerve root. Paracentral herniations are the most common type of disc herniations due to the relative weakness of the posterior longitudinal ligament in this area. These herniations typically cause symptoms associated with the traversing nerve root, which passes the intervertebral disc in the canal.

Finally, a far-lateral disc herniation, while much less common than paracentral disc herniations, may cause much more severe pain because of direct compression of the dorsal root ganglion in this area. Due to the relatively lateral location of far-lateral disc herniations, these herniations typically affect the exiting nerve root in the far-lateral or foraminal area. As such, while a paracentral disc herniation at L4–L5 would affect the L5 nerve root, a far-lateral herniation would typically affect the L4 nerve root.

In addition to classifying LHID by their anatomic location, herniations can also be classified by the morphology. Herniations can be classified as either protrusions, extrusions, or sequestrations (Fig. 38.3). The smallest herniation is a protrusion, which is characterized by a herniation that is larger at the base than in the canal. In contrast, an extrusion is a herniation with a small base and a large herniation into the canal. Finally, a sequestration is defined by a disc herniation, which is not continuous with the intervertebral disc and is contained entirely within the canal. Classifying herniations by the anatomic location as well as by the morphology is important to understanding the clinical implications of this pathology and treatment plans.

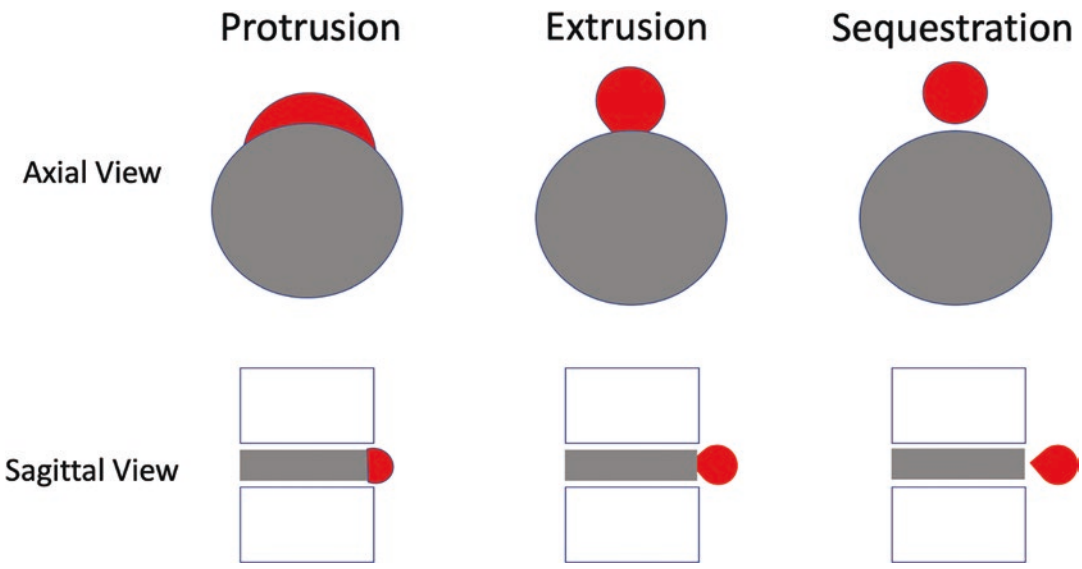

Fig. 38.3 Lumbar herniated intervertebral disc based on morphology

38.5 Diagnosis (Clinical and Imaging)

The clinical manifestations of LHID vary significantly based on the anatomic location and morphology of the herniation. The classic presentation of LHID is often associated with a memory of inciting event or injury. Oftentimes, patients may also describe pain with increased thoracic pressure, as when sneezing, straining, or coughing. In addition, sitting for long periods has also been associated with the development of intervertebral disc pathology and has been shown to raise pressure in the disc as well as significantly exacerbate symptoms.

Pain is often described as intense and may be associated with neurologic complaints, such as radiating pain or decreased sensation in dermatomal distribution. In severe cases, patients may also describe weakness in myotomal distribution depending on the affected nerve root, which depends on the level of the herniation and its anatomic location in the canal. A paracentral herniation typically affects the traversing nerve at the level of the disc herniation, while a far-lateral herniation affects the exiting nerve. Depending on the nerve affected, symptoms will manifest in different dermatomal or myotomal distributions.

For example, a disc herniation affecting the L4 nerve root may cause sensory symptoms over the knee and medial lower leg and weakness in extension of the knee as well as dorsiflexion of the ankle due to the weakness of quadriceps and tibialis anterior muscles. In contrast, a disc herniation impinging on the L5 nerve root may cause weakness in the extensor hallucis longus muscle as well as the gluteus medius, presenting weakness in dorsiflexion of the great toe and Trendelenburg gait due to weakness in hip abduction. L5 nerve sensory symptoms are typically over the lateral lower leg and dorsal foot. Finally, a herniation affecting the S1 nerve may cause weakness of the gastrocnemius muscle leading to weak ankle plantarflexion, while sensory symptoms may be affected over the posterior leg and plantar foot.

Some clinical signs could help in defining the position of the herniation. Scoliotic deviation of

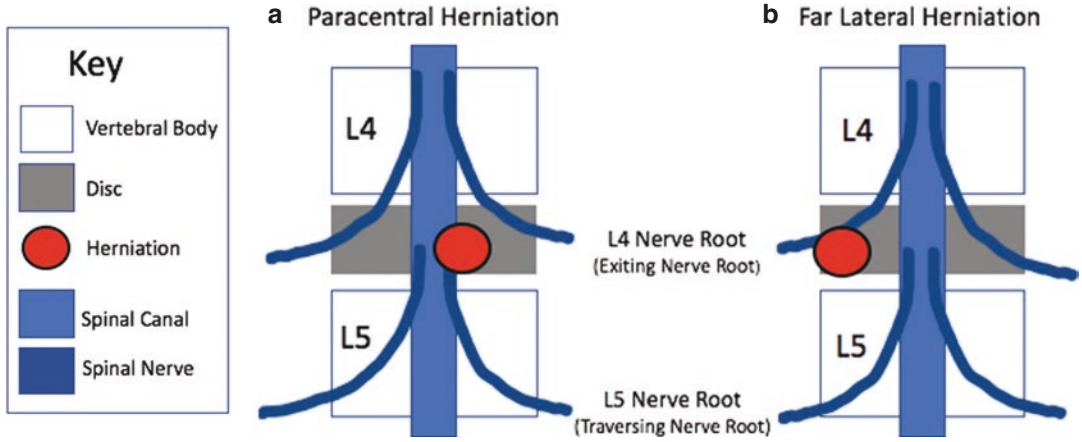

Fig. 38.4 (**a**) Disc herniation lateral to the nerve root. Theoretically, lateral flexion to the same side would increase the pain. (**b**) An axillary disc herniation. Theoretically, lateral flexion to the opposite side increases the pain

the trunk is frequent in patients with LHID. If the disc herniation is lateral to the nerve root, lateral flexion to the same side would increase the pain. In case of an axillary disc herniation, the lateral flexion to the opposite side increases the pain (Fig. 38.4).

In 2014, the NASS recommended that the gold standard for diagnosing LHID was the testing of muscle weakness, sensory symptoms, and supine straight leg raise test. The straight leg raise test has been shown to have excellent sensitivity but relatively low specificity in diagnosing lumbar radiculopathy.

While the diagnosis of lumbar radiculopathy can often be made with a thorough history and physical examination, diagnostic imaging is also important for diagnostic purposes as well as surgical planning. Radiographs should be obtained as basic imaging modality. AP, lateral, and flexion/extension radiographs may help to diagnose intervertebral collapse and associated bony pathologies such as osteophytes, spondylolisthesis, and instability. However, radiographs alone are insufficient to visualize the anatomic location and morphology of the disc herniation.

In addition to radiographs, MRI is the gold standard for diagnosing LHID. MRI has been shown to have excellent sensitivity and specificity in diagnosing disc herniation. MRI allows for direct visualization of the soft tissues in the lumbar spine, so the anatomic location and morphology of the disc herniation can be visualized. MRI is mandatory for surgical planning as it allows the surgeon to understand the correlation of clinical symptoms with the patient's anatomic pathology. However, while MRI is considered the gold standard in diagnosing lumbar disc pathology, it must be noted that such pathology found on MRI has been shown to be quite common even in asymptomatic patients. Therefore, practitioners treating lumbar disc pathology must rely on both clinical symptomatology and imaging studies to avoid misdiagnosis and overtreatment.

In cases where MRI may be unavailable or unsafe, as in the case of noncompatible implantations such as pacemakers, CT may be used to detect LHID as well. CT myelography has been recognized by the NASS as an acceptable imaging modality to diagnose LHID as an alternative to MRI. However, CT myelography may be associated with side effects such as headaches, cerebrospinal fluid (CSF) leakage, infection, and bleeding. MRI remains the gold standard in diagnosis and treatment planning of LHID. A thorough understanding of clinical symptomatology as well as imaging studies is necessary to treat patients with LHID and improve their symptoms.

38.6 Treatment

The first-line treatment for patients with LHID is conservative treatment. More than 90% of patients with LHID show significant improvement with conservative modalities. In fact, it has been shown that most patients with acute LHID will improve without any treatment at all. Conservative treatments most commonly include medications, activity modification such as rest and a gradual return to activity, physical therapy, and epidural steroid injections.

The mainstay of medical treatment for LHID is nonsteroidal anti-inflammatory drugs (NSAIDs), but several other medications are also used ubiquitously by general practitioners and spine surgeons alike, including muscle relaxants, TNF-alpha inhibitors, steroids, or short courses of opioids. Oral steroid tapers have been shown to provide symptomatic relief in the acute setting. Currently, the NASS recommendations suggest that there is insufficient evidence to recommend many medications commonly used in the treatment of radicular pain, including amitriptyline, gabapentin, agmatine sulfate, 5-hydroxytryptophan inhibitors, and glucocorticoids, and Grade B evidence to suggest no benefit of TNF-alpha inhibitors.

Physical therapy, including exercises focusing on core strengthening, extension exercises, and joint mobility, has been shown to improve symptomatology in patients with LHID. In addition, one landmark study using the Spine Patient Outcomes Research Trial (SPORT) trial found that formal physical therapy was associated with less low back pain at 1 year and also less opioid use in patients with LHID compared to those who did not undergo physical therapy.

Epidural steroid injections (ESI) can be a powerful treatment tool in patients with LHID.

Surgical treatment is indicated after conservative modalities have failed or when the patient has neurological deficit such as severe acute weakness of muscles and cauda equina syndrome. For patients with LHID who are nonresponsive to conservative management, surgery has been shown to improve symptoms faster than continued conservative care. In addition, the benefits of early and effective surgical treatment are becoming increasingly apparent in the literature. It has been suggested that long-term symptomatology is predictive of poor outcomes with nonsurgical management. As a result, some practitioners are in favor of immediate surgical management without a course of conservative treatment if symptoms have been present for over 6 months to a year.

Historically, it has been suggested that surgery provides faster symptom relief than conservative treatment but is associated with similar long-term outcomes. However, more recently, the long-term benefits of surgery have been made clearer. The SPORT trial, one of the most widely cited trials in spine surgery literature, has demonstrated the clinical superiority of surgical vs. nonsurgical treatment in the management of LHID for both short-term and long-term outcomes. Factors that have been shown to lead to better outcomes after surgery for LHID include significant leg pain, positive straight leg raise test, and weakness correlating MRI findings.

The first surgery for a diagnosis of LHID was performed in 1932, which was an L2–S1 laminectomy. Since that time, the incidence of surgical treatment for LHID has increased dramatically, and less invasive techniques have been developed over time. All surgical techniques for LHID throughout history have similar goals of removing the herniated disc and decompressing local neural elements to relieve symptoms.

Later, in the 1930s and 1940s, radical discectomy was common for the treatment of LHID with curettage of the nucleus pulposus and cartilaginous end plates in order to prevent reherniation. Due to high rates of postoperative instability associated with radical discectomy, adjunctive fusion was common. However, by the middle of the twentieth century, partial discectomy rather than radical discectomy became the standard and, as surgical technique and technology continued to improve rapidly, discectomy surgery has evolved in a way that would minimize soft-tissue dissection and damage to surrounding structures.

In 1938, Love described an extradural approach to the interverbal disc, where the ligamentum flavum was removed but bone loss was minimized. By utilizing the Love method, a standard nucleotomy could be performed without large amounts of bony dissection, which would decrease the risk of destabilizing the spine. The Love method quickly became the gold standard for lumbar discectomy before more minimally invasive techniques were described and led to a sharp increase in lumbar discectomy surgeries in the middle of the twentieth century. In Love's first case series, the Love nucleotomy was shown to have good outcomes in 100 patients. Furthermore, more recent literature has shown that Love's nucleotomy has excellent long-term results in terms of reherniation, patient satisfaction, and complication rates which are comparable to modern minimally invasive techniques.

In 1967, Yaşargil pioneered the use of an intraoperative microscope for lumbar discectomy surgery, which gained traction through the following decades. The use of microscopic surgery allowed for the first "minimally invasive" discectomy to be performed, which employed less invasive surgical dissection compared to the standard open nucleotomy described by Love. By the twenty-first century, the standard for surgical treatment of LHID was the microdiscectomy, which refers to the hemi-laminotomy, medial facetectomy, and partial discectomy (herniotomy). Microdiscectomy has been shown to have success rates as high as 90% in ideal patient population and is the most common surgical technique for LHID.

While midline incision approach is employed to treat paracentral disc herniations, this approach for far-lateral disc herniations requires extensive tissue dissection to access the disc herniation. In 1968, Wiltse et al. described a muscle-sparing approach to the lateral aspect of the vertebral column, which utilizes the plane between the multifidus and longissimus muscles. Though this approach was originally described for spinal fusion, it was adapted to allow for direct access to far-lateral disc herniations in order to minimize soft-tissue dissection and retraction while providing direct access to the far-lateral herniation.

More recent advancements in the surgical technique of LHID include the development of endoscopic techniques and approaches. While microdiscectomy is often considered the gold standard for the surgical treatment of LHID, endoscopic discectomy is less invasive in the soft-tissue dissection and manipulation of neural elements as well as dura compared with microdiscectomy. There are several endoscopic approaches to the intervertebral disc, including interlaminar, transforaminal, posterolateral, and transiliac, based on the anatomic location and morphology of the disc herniation. Therefore, both paracentral and far-lateral disc herniations can be treated by percutaneous approaches using endoscopy technique. Endoscopic discectomy has been associated with long-term outcomes similar to open discectomy and has been shown to be associated with less operative time and less blood loss due in part to their minimally invasive nature.

Despite the profound improvement in outcomes for lumbar discectomies over the years, these surgeries are not without the possibility of complications. One systematic review comparing the postoperative complication rates of open microdiscectomy, endoscopic discectomy, and percutaneous microdiscectomy found postoperative complication rates of 12.5%, 13.3%, and 10.8%, respectively. Common postoperative complications following lumbar discectomy include CSF leak, nerve root injury, and postoperative pain. However, while complications are indeed possible, significant improvements have been made in the way of lumbar discectomy, and the various surgical techniques used to treat LHID today are generally accepted to be safe, effective, and associated with excellent outcomes.

Take-Home Message

- Lumbar herniated intervertebral disc (LHID) is one of the most common spinal pathologies and is a major cause of low back pain and lumbar radiculopathy.
- Clinical symptoms of LHID are caused by a combination of mechanical compression of neural elements and local inflammation.
- LHID is diagnosed with physical examination as well as imaging studies such as radiographs and magnetic resonance imaging (MRI).
- The first-line treatment for patients with LHID is conservative treatment including medications, physical therapy, and epidural steroid injections. Surgical intervention is indicated when conservative modalities fail or when the patient has neurological deficit such as severe acute weakness of muscles and cauda equina syndrome.
- Surgical and nonsurgical treatment of LHID leads to excellent outcomes.

Summary

LHID is one of the most common spinal pathologies and is a major cause of low back pain and radiculopathy. Since the discovery of LHID in the 1930s, the scientific understanding of this ailment has increased. Disc herniations can cause direct compression of local neural elements and cause a local inflammatory response, which can lead to significant pain and disability. A thorough history and physical examination, in addition to routine diagnostic imaging studies with radiographs and MRI, can adequately diagnose disc herniation. In general, most patients with LHID will recover without the need for surgical treatment. Surgical intervention is indicated when conservative modalities fail or when the patient has neurological deficit such as severe acute weakness of muscles and cauda equina syndrome. Surgical techniques have become more and more minimally invasive and have been associated with excellent clinical outcomes.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. In a patient with MRI showing left intraforaminal L2–L3 herniation, which muscle will be affected by strength deficit?
   (a) Left quadriceps femoris
   (b) Right anterior tibial
   (c) Left anterior tibial
   (d) Extensor digitorum longus of the left big toe
2. Lasègue's sign is positive in cases of:
   (a) Back pain radiating to the posterolateral part of the leg
   (b) Arthrosis of the hip
   (c) Ankylosing spondylitis
   (d) Cervical disc herniation
3. Achilles hypo- or areflexia is frequently caused by:
   (a) L5–S1 disc herniations
   (b) Herniated discs L4–L5
   (c) L3–L4 disc herniations
   (d) Cervical disc herniations
4. All of these methods are part of the instrumental diagnosis of herniated discs with the exception of a:
   (a) Radiculography
   (b) CT
   (c) Arteriography
   (d) Magnetic resonance imaging
5. Which of these conditions is not the cause of lumbosciatica?
   (a) Herniated disc
   (b) Spondylolisthesis
   (c) Lumbar stenosis
   (d) Scheuermann's disease

Further Reading

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Spondylolysis and Spondylolisthesis

39

Jean-Marc Mac-Thiong and Hubert Labelle

Overview
This chapter reviews development spondylolisthesis, which is the most common type of spondylolisthesis observed in children, adolescents, and young adults. An overview of the fundamental concepts of sagittal balance is presented to better understand the biomechanics and pathogenesis involved with spondylolisthesis, and its relevance in the classification and management of spondylolisthesis in young patients.

39.1 Introduction

Spondylolysis involves a defect (or fracture) in the pars interarticularis, which is the area of the vertebral arch between the superior and inferior facets (Fig. 39.1). A spondylolisthesis is defined by the forward displacement of one vertebra on top of another vertebra. Most commonly, a spondylolysis will coexist with spondylolisthesis. On some occasions, spondylolisthesis will be associated with an elongated pars interarticularis following repetitive fracture and healing of the pars interarticularis. Spondyloptosis involves complete translation of one vertebra with respect to the caudal vertebra. This chapter focuses on spondylolysis and spondylolisthesis at the lumbosacral spine that occurs in children, adolescents, and young adults.

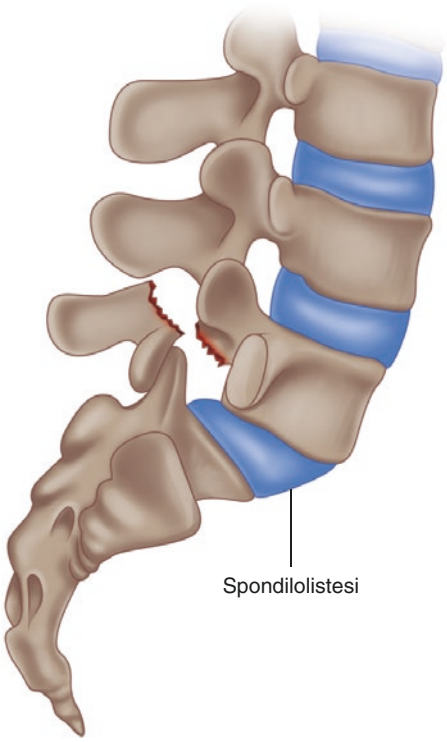

Fig. 39.1 Spondylolisthesis

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39.2 Epidemiology

In young patients, spondylolisthesis occurs predominantly at the lumbosacral junction. When occurring at L4–5 level, it is not uncommon to observe a transitional L5 vertebra involving variable degree of fusion between L5 and S1 vertebrae or sacralization of L5. In their prospective longitudinal follow-up of 500 first-grade children, Fredrickson et al. observed a prevalence of spondylolysis of 4.4% at 6 years old, 5.2% at 12 years old, and 6% in adulthood. By the age of 18 years, 74% of individuals with bilateral spondylolysis had developed a spondylolisthesis. Increased prevalence of spondylolysis and spondylolisthesis has been reported for certain ethnic groups such as Native American populations. Increased prevalence is also suggested for athletes participating in sports such as gymnastics and baseball.

39.3 Etiology/Pathogenesis

Spondylolysis and spondylolisthesis can be caused by different etiologies. However, in young individuals, the developmental type originally described by Marchetti and Bartolozzi is predominant. The ongoing development of the spine and pelvis will be altered locally (lumbosacral junction) and globally (sagittal alignment and balance) in a way that a spondylolisthesis can occur and/or progress (Fig. 39.2). The current authors have later clarified the concept of developmental spondylolisthesis by referring to a condition in which various degrees of predisposing and environmental factors will lead to a defect in the pars interarticularis.

This concept suggests a multifactorial etiology of developmental spondylolisthesis. It also underscores the biomechanical concepts involved with spondylolisthesis, beyond the presence of local trauma (or repetitive microtrauma) that may contribute to the development of spondylolysis. Spondylolysis is not observed in non-ambulant individuals, newborns, or quadrupeds. In the upright position, spinal loading is most prominent at the lumbosacral junction, while the L5–S1 facet joints support most of the shear forces and the L5–S1 disc supports most of the compression forces. Being a weak point of the vertebral arch, the pars interarticularis at L5 vertebra is subjected to high shear and compressive and tensile loads, particularly during repetitive flexion and hyperextension movements. With a spondylolysis at L5 vertebra, there is a dissociation

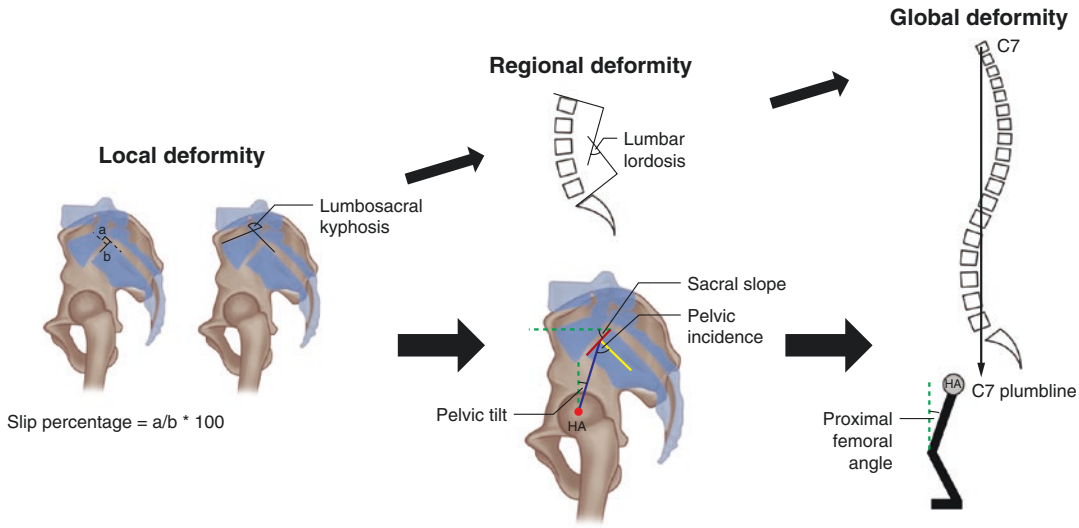

Fig. 39.2 Assessment of sagittal balance from parameters contributing to local, regional, and global deformity in spondylolisthesis. HA hip axis

between the anterior and posterior parts of the vertebra and loads will be transferred to the L5–S1 disc, thus predisposing to disc degeneration and dysplasia of bony elements (through impaired growth plate remodeling), and ultimately spondylolisthesis. Progression of the spondylolisthesis mainly occurs during growth, and the main risk factor for progression is the severity of slip at the initial presentation. Accordingly, the risk of progression is less likely when the slip percentage is less than 40%.

Altering the loading conditions and biomechanical environment at the lumbosacral junction will also impact the development and alignment of the entire spine and pelvis. A lumbosacral spondylolisthesis tends to induce forward displacement of the body's center of gravity (usually located over the femoral heads in order to minimize energy expenditure). To minimize energy expenditure and ensure proper location of the body's center of gravity, postural changes and compensation mechanisms through modifications in the sagittal balance of the spine, pelvis, and lower extremities are required, but may also further lead to abnormal biomechanical loads at the lumbosacral junction, thereby contributing to the development of spondylolisthesis.

In normal individuals, pelvic morphology modulates pelvic orientation and in turn regulates the alignment of the adjacent spine and lower extremities. In lumbosacral spondylolisthesis, an abnormal pelvic morphology combined with the local lumbosacral deformity will result in disturbed sagittal balance (incongruent alignment of spine, pelvis, and lower extremities). Using a postural model of sagittal balance, Mac-Thiong et al. have shown that a relatively normal posture was preserved in low-grade spondylolisthesis, while posture was abnormal in high-grade spondylolisthesis.

With progressive spondylolisthesis, lumbar lordosis increases to keep the center of gravity well positioned and the C7 plumb line behind the hip axis, thereby preserving a balanced posture. This first compensation mechanism involves an increase in lordosis in the lumbar spine and/or a decrease in kyphosis in the thoracic spine. When maximal lumbar lordosis (and minimal thoracic kyphosis) is achieved, progressive retroversion of the pelvis and verticalization of the sacrum occur in an attempt to maintain a balanced posture. Considering that each individual has a fixed pelvic incidence, this second compensation mechanism involves an increase in pelvic tilt and a reciprocal decrease in sacral slope. When these compensation mechanisms reach their limit, compensatory hip flexion (proximal femoral angle >10°) and/or spinal imbalance (C7 plumb line in front of hip axis) occur.

39.4 Classifications

Meyerding reported the first classification of lumbosacral spondylolisthesis based on the severity of spondylolisthesis, by assigning grade I, II, III, or IV to slips within 25%, 50%, 75%, or 100% of the dimension of the upper end plate of S1 vertebra, respectively. The Wiltse classification proposes five types of spondylolisthesis based on etiological and anatomical findings: dysplastic, isthmic, degenerative, traumatic, and pathologic. Although this classification can be useful to classify spondylolisthesis by defect type, it is limited by objective criteria to define (and differentiate) dysplastic and isthmic types, as young individuals will most often present concurrent dysplastic features and a defect in the pars interarticularis. Marchetti and Bartolozzi have proposed an etiology-based classification dividing spondylolisthesis into the developmental and acquired types. Developmental spondylolisthesis is categorized as low or high dysplastic based on the severity of bony dysplasia (lumbosacral kyphosis, trapezoid shape of L5 vertebra, sacral doming, etc.). However, the criteria for differentiating between low and high dysplastic types remain elusive. In addition, excluding stress fractures of the pars interarticularis from the developmental type is controversial considering that the great majority of patients classified as having a developmental spondylolisthesis will have a concomitant pars interarticularis defect.

More recently, Mac-Thiong and Labelle proposed a classification system from which originated the Spinal Deformity Study Group (SDSG)

Table 39.1 Spinal Deformity Study Group classification of spondylolisthesis

Slip severity	Pelvic balance	Spinal balance	Type
Low grade Slip <50%	PI <45°	–	1
	PI = 45–60°	–	2
	PI >60°	–	3
High grade Slip ≥50%	Balanced pelvis	–	4
	Unbalanced pelvis	Balanced spine	5A
			5B (LSK <80° or PFA >10°)
		Unbalanced spine	6

PI Pelvic incidence; *LSK* Lumbosacral kyphosis; *PFA* Proximal femoral angle

classification of developmental spondylolisthesis (Table 39.1). This classification system integrates important aspects related to the pathogenesis and outcome of spondylolisthesis, particularly the slip severity and the spino-pelvic balance.

The SDSG classification has been designed primarily to guide surgical treatment by arranging the different types in ascending order of severity such that the complexity of surgery will increase as the severity of spondylolisthesis increases. Likewise, the SDSG classification builds on the premise that a complex surgery is not necessary for all patients with spondylolisthesis. To classify a patient, slip severity and sagittal balance need to be assessed from lateral standing radiographs of the spine and pelvis.

39.4.1 Slip Severity

As a first step, slip severity is measured to determine whether it is low grade (<50% or Meyerding grades 1–2) or high grade (≥50% or Meyerding grades 3–4).

39.4.2 Pelvic Balance

In the presence of a low-grade slip, three types of spondylolisthesis are defined based on pelvic incidence: type 1, pelvic incidence <45°; type 2, pelvic incidence between 45 and 60°; and type 3, pelvic incidence >60°. With a high-grade slip, patients are first subdivided into two groups based on their pelvic tilt and sacral slope. The pelvic tilt and sacral slope are then reported in the nomogram provided by Hresko et al. to determine whether the pelvic is balanced (type 4) or unbalanced (types 5A, 5B, or 6).

39.4.3 Spinal Balance

For high-grade spondylolisthesis with unbalanced pelvis, spinal balance needs to be assessed based on the position of the C7 plumbline with respect to the hip axis. The spine is balanced when the C7 plumb line falls over behind the hip axis (type 5A or 5B) and unbalanced when the C7 plumb line falls in front of the hip axis (type 6). Patients with an unbalanced pelvis but balanced spine are further subdivided into type 5A or 5B based on the measurement of lumbosacral kyphosis and proximal femoral angle. Type 5B refers to a patient having a lumbosacral kyphosis <80° or a proximal femoral angle >10°.

39.5 Diagnosis (Clinical and Imaging)

Many individuals with spondylolysis and low-grade spondylolisthesis will be asymptomatic as they typically have a normal gait and posture. Low-back pain is the most common complaint in young patients presenting with developmental spondylolisthesis. Neurological symptoms are most common with high-grade spondylolisthesis, and L5 radiculopathy due to foraminal stenosis is typically observed. A cauda equina syndrome due to central stenosis is rarely observed in patients with a spondylolisthesis without spondylolysis (or with elongated pars interarticularis).

As the slip progresses, patients may have an increased lumbar lordosis with prominent abdomen, heart-shape buttocks, and a shortened-appearing trunk. Patients may flex their hips and knees in an effort to compensate for the forward displacement of the trunk. A scoliotic deformity may be observed and may be secondary to nerve root compression/tension or muscle spasm (anta-

lgic scoliosis), asymmetric slippage in the frontal plane (olisthetic scoliosis), or concomitant idiopathic scoliosis.

Symptomatic patients usually have variable degree of restricted and painful lumbar range of motion, particularly in extension. An increased popliteal angle with tight hamstrings may be due to knee flexion or nerve root tension/compression. The straight leg raise test is usually positive in the presence of nerve root tension/compression. A careful neurologic examination is required to detect signs of nerve root tension/compression or cauda equina syndrome.

Plain standing lateral and posteroanterior radiographs of the lumbosacral spine are obtained when suspecting spondylolysis and/or spondylolisthesis. When there is no evidence of interarticular pars defect on the lateral radiograph, oblique views are ordered when there is still a high suspicion of spondylolysis without spondylolisthesis. The posteroanterior radiograph is used to rule out the presence of scoliosis. In the presence of spondylolisthesis, long cassette standing radiographs including the spine and pelvis are recommended for classification purposes, treatment decision-making, and follow-up.

In children with back pain and no radiographic findings consistent with spondylolysis or spondylolisthesis, a CT scan is useful for identifying a pars interarticularis defect. A CT scan is also relevant for preoperative planning to assess the bony architecture. While it can be used to diagnose a spondylolysis, MRI is particularly useful to assess the cauda equina and nerve roots in the presence of neurological impairment. It is also sensitive for detecting early pars interarticularis stress reaction without overt spondylolysis. Preoperatively, MRI can be used to evaluate the status of adjacent L4–5 and S1–2 intervertebral discs.

39.6 Treatment

39.6.1 Nonsurgical Treatment

Asymptomatic individuals with low-grade spondylolisthesis and no sign of dysplasia require no activity restriction or treatment. Nonsurgical treatment is more likely to be successful in low-grade spondylolisthesis. Nonsurgical treatment of acute symptoms primarily relies on rest, physical therapy, and mild analgesia. Although a successful clinical outcome is not necessarily linked to successful healing of the pars defect, a trial of bracing can be attempted when an acute or stress fracture without spondylolisthesis is suspected. Brace treatment in neutral lordosis can also be attempted to alleviate pain in symptomatic patients with chronic spondylolysis and/or low-grade spondylolisthesis. Overall, a successful outcome will be achieved in about 85% of symptomatic patients with spondylolisthesis. Less than 10% of children and adolescents with low-grade spondylolisthesis will require surgery. With a low-grade spondylolisthesis of grade 1 or less, radiographic surveillance for slip progression every 6 months is recommended until the growth spurt has passed (Risser 1). In the presence of significant dysplasia and/or a slip severity >40%, follow-up until skeletal maturity is advised.

Nonsurgical treatment is more likely to fail in patients with high-grade spondylolisthesis. However, nonsurgical treatment and observation can be safe and successful in selected patients with slip <75% in the absence of neurological symptoms, if they are followed closely until skeletal maturity.

39.6.2 Surgical Treatment

The most common indication for surgical treatment of spondylolisthesis is to prevent the risk of further slip towards spondyloptosis. Accordingly, it is generally assumed that any patient with documented slip progression of 30% or more, immature patients with a slip greater than 50%, and mature patients with a slip greater than 75% are candidates for surgery. The presence of a neurological impairment from compression of nerve roots or the cauda equina is another indication for surgery. Persisting functional impairment or pain despite appropriate nonsurgical treatment, and progressive postural deformity or gait abnormality, is a relative indication for surgery.

Table 39.2 Proposed surgical algorithm for young patients with developmental spondylolisthesis based on SDSG classification

Global balance	SDSG type	Proposed surgical strategy
Well balanced	1	L5–S1 fusion
	2	L5–S1 fusion (consider pars repair if indicated)
	3	L5–S1 fusion (consider high-grade algorithm if ≥40% slip and significant dysplasia)
Relatively well balanced	4	In situ instrumented fusion vs. partial reduction of slip with 360° fusion
	5A	
Unbalanced	5B	Partial reduction with 360° fusion
	6	

Restoring or preserving adequate sagittal balance, achieving solid fusion, and decompressing neural elements are the main goals of surgery for developmental spondylolisthesis. It is expected that surgery will improve the quality of life in about 85% of patients. Table 39.2 proposes a surgical algorithm for young patients with developmental spondylolisthesis based on the SDSG classification.

Posterior L5–S1 instrumented fusion—without formal reduction—is the gold standard for low-grade spondylolisthesis, considering that these patients generally exhibit adequate sagittal global balance. Pars repair can be considered in low-grade patients with normal pelvic incidence (type 2). However, it should be avoided due to increased risk of failure when the pars interarticularis is subjected to high compression loading from a "nutcracker effect" in the presence of a low pelvic incidence (type 1) or to high tension loading in the presence of a high pelvic incidence (type 3). Type 3 patients with a slip between 40% and 50% and significant dysplasia (particularly sacral doming and/or trapezoidal L5 vertebra) represent a subset of patients that can be referred to as a "low/high-border" subtype for whom the risk of progression and outcomes are similar to those observed in high-grade spondylolisthesis. For this "low/high-border" subtype, treatment principles should follow the treatment principles for high-grade spondylolisthesis, based on the assessment of pelvic balance (from sacral slope and pelvic tilt), spinal balance, as well as lumbosacral angle and proximal femoral angle.

The most important consideration for high-grade spondylolisthesis is whether formal surgical reduction should be performed. Reduction of high-grade spondylolisthesis is suggested primarily to provide an enhanced biomechanical environment for fusion and optimal sagittal balance. Formal surgical reduction may not be required for types 4 and 5A spondylolisthesis considering that sagittal balance is relatively well preserved, since postural compensation mechanisms from the pelvis are not exceeded. If in situ fusion (postural reduction from patient positioning on the operative table without formal surgical maneuvers to reduce the spondylolisthesis) is performed, instrumentation should extend cephalad to L4 and distal to the iliac to decrease the risk of pseudarthrosis and instrumentation failure due to high tension loading on the graft and instrumentation in the presence of residual L5–S1 deformity. Otherwise, partial reduction to a low-grade slip with anterior column support can be performed in types 4 and 5A spondylolisthesis to maximize the likelihood to preserving a balanced pelvis. It is also believed that achieving an L5 incidence (similar to pelvic incidence but using the upper end plate of L5 rather than the upper end plate of S1 as a landmark) less than 65° through reduction will result in improved pelvic balance and quality of life after surgery. With formal reduction, fusion can be limited to L5 proximally, provided that the L4–5 disc is lordotic. Otherwise, fusion should be extended proximally to L4 to minimize the risk of supra-adjacent kyphosis and/or spondylolisthesis that may require revision surgery. In the presence of an open S1–S2 disc, instrumentation should extend to the iliac when performing reduction to minimize the risk of S1–S2 failure. In skeletally mature patients, particular consideration should be given to extending the instrumentation proximally to L4 and distally to the pelvis if forceful maneuvers have been required to reduce the spondylolisthesis, in order to reduce the risk of instrumentation failure. Types 5B and 6 spondylolistheses are best treated with partial reduction to a low-grade slip and L5 incidence less than 65° to maximize the likelihood of achieving normal sagittal balance, and with anterior column support to promote

solid fusion, reduce the risk of instrumentation failure, and facilitate correction of the local L5–S1 deformity. As for types 4 and 5A spondylolisthesis, the instrumented levels should be selected according to the status of the intervertebral discs adjacent to the spondylolisthesis and skeletal maturity.

Take-Home Message

- Spondylolysis involves a defect (or fracture) in the pars interarticularis, which is the area of the vertebral arch between the superior and inferior facets.
- A spondylolisthesis is defined by the forward displacement of one vertebra on top of another vertebra.
- Meyerding reported the first classification of lumbosacral spondylolisthesis based on the severity of spondylolisthesis, by assigning grade I, II, III, or IV to slips within 25%, 50%, 75%, or 100% of the dimension of the upper end plate of S1 vertebra, respectively.
- Low-back pain is the most common complaint in young patients presenting with developmental spondylolisthesis.
- The most common indication for surgical treatment of spondylolisthesis is to prevent the risk of further slip towards spondyloptosis.

Summary

Spondylolysis involves a defect (or fracture) in the pars interarticularis, which is the area of the vertebral arch between the superior and inferior facets. A spondylolisthesis is defined by the forward displacement of one vertebra on top of another vertebra. Most commonly, a spondylolysis will coexist with spondylolisthesis. In young patients, spondylolisthesis occurs predominantly at the lumbosacral junction. When occurring at L4–5 level, it is not uncommon to observe a transitional L5 vertebra involving variable degree of fusion between L5 and S1 vertebrae or sacralization of L5. Spondylolysis and spondylolisthesis can be caused by different etiologies. Meyerding reported the first classification of lumbosacral spondylolisthesis based on the severity of spondylolisthesis, by assigning grade I, II, III, or IV to slips within 25%, 50%, 75%, or 100% of the dimension of the upper end plate of S1 vertebra, respectively. The Wiltse classification proposes five types of spondylolisthesis based on etiological and anatomical findings. Many individuals with spondylolysis and low-grade spondylolisthesis will be asymptomatic as they typically have a normal gait and posture. Low-back pain is the most common complaint in young patients presenting with developmental spondylolisthesis. Neurological symptoms are most common with high-grade spondylolisthesis. Asymptomatic individuals with low-grade spondylolisthesis and no sign of dysplasia require no activity restriction or treatment. The most common indication for surgical treatment of spondylolisthesis is to prevent the risk of further slip towards spondyloptosis. The presence of a neurological impairment from compression of nerve roots or the cauda equina is another indication for surgery.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. A 16-year-old boy who practices sports complains for about 2 years intermittent low-back pain and is affected by bilateral spondylolysis. No neurological abnormalities. He has prospects of becoming a professional soccer player. You would suggest:
   (a) Not to play any kind of sport for 1 year
   (b) Bracing during sports activity
   (c) Foraminotomy
   (d) Posterior laminectomy
2. Spondylolysis is more frequent:
   (a) In pregnancy
   (b) In weightlifting athletes
   (c) In menopausal women
   (d) In children with rickets
3. Spondyloptosis is:
   (a) An extreme degree of spondylolisthesis
   (b) A tuberculous affection of the vertebrae

(c) A nonspecific infection of the intervertebral disc
(d) A radiculopathy

4. Spondylolisthesis can be due to:
   (a) Congenital abnormalities
   (b) Functional overload
   (c) Degenerative diseases
   (d) A + B + C
5. Spondylolysis is defined as:
   (a) A compression fracture of the vertebral body
   (b) The interruption of the vertebral isthmus
   (c) The interruption of the vertebral pedicles
   (d) The interruption of the vertebral laminae

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Lumbar Spinal Stenosis

40

Pilar Serra-Añó and Sara Mollà-Casanova

Overview
This chapter includes information about the most common types of spinal canal stenosis, its prevalence, and etiology. Further, the available tools and procedures for imaging and clinical diagnosis are defined. Finally, the description of the most usual therapeutic approaches for preventing and reverting the progression of the stenosis is addressed.

40.1 Definition

Lumbar spinal canal stenosis (SCS) is represented by an alteration in the relationship between the container and the content of the vertebral canal (Fig. 40.1). The French anatomist Antoine Portal (1742–1832) reported that the abnormal curvatures of the vertebral column could result in a more or less pronounced narrowing of the vertebral canal at the level of one or more vertebrae. The term "stenosis" derives from the Attic adjective "stenos" for the Ionic "steinos," meaning "being narrow." The Attif suffix -"osis" was added to indicate a pathologic condition.

In this pathology, the content is normal, while the container presents a reduced dimension. From a morphological and anatomical point of view, the vertebral canal is composed of bony ligamentous structures in which the bony parts are the posterior wall of the vertebra, the pedicles, the laminae, and the posterior articular apophyses. The ligamentous structures are the intervertebral disc, the posterior longitudinal ligament, and the ligamentum flavum. Stenosis of the lumbar spine is an anatomical alteration. In these patients, the vertebral canal presents from birth a smaller bony ligament component than normal. The studies conducted by Larsen and colleagues reported that a possible narrowing of the canal could be due to the reduced mid-sagittal diameter, indicating an isolated growth disturbance of the vertebral arch. Moreover, thickening of the laminae and an acute interlaminar angle could contribute to the narrowing. According to Verbiest and colleagues, it was possible to distinguish between *absolute stenosis* (in which signs and symptoms could appear also in the absence of additional compressive agents) and *relative stenosis*. In the latter condition, the narrowness was such that only in the absence of slightly bulging intervertebral discs, hard ridges, or infolding of the ligamentum fla-

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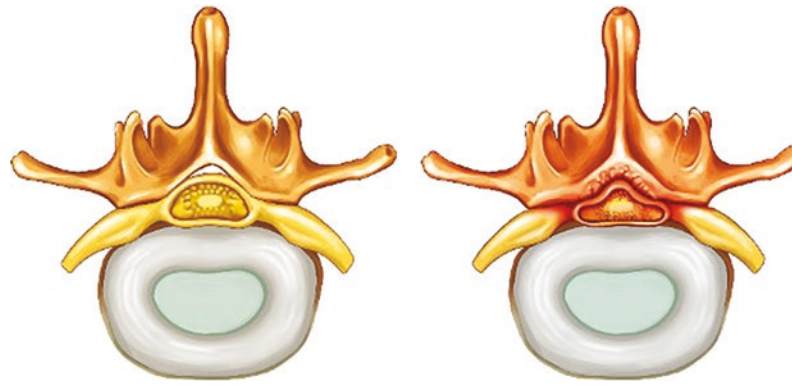

Fig. 40.1 Spinal canal stenosis. Figure shows a horizontal view of a normal lumbar vertebra (left) and a lumbar vertebra with spinal canal stenosis (right)

vum the patients remain asymptomatic. Moreover, in the absolute stenosis, there is no reserve capability of the extradural or intradural space to compensate for additional space-occupying agents. In relative stenosis, this reserve capacity is very small. However, relative stenosis is no pathologic condition. A narrowing that reduces the spinal canal to less than 10 mm in diameter is considered absolute stenosis, while a diameter between 10 and 12 mm is considered relative stenosis. It may persist for decades without showing symptoms. However, compression throughout the canal can eventually produce multiple symptoms of varying degrees of severity. This situation constitutes an unstable balance in which any pathology, either morphological (disc protrusion, osteophytosis due to arthrosis) or dynamic (instability with alteration of the posterior longitudinal ligament) that modifies the morphology of the container, can change the relationship between container and content. If this alteration occurs, there may be involvement of the myeloradicular structures. In this situation, in which in a normally sized spinal canal there would be no disease, in the case of stenosis, a clinical picture of pain, paresthesia, and claudication is triggered. Therefore, all those cases in which compression is secondary to other pathologies (fractures, tumors, severe instability, and iatrogenic injuries) should be excluded from the causes of lumbar stenosis.

40.2 Epidemiology

Clinical symptoms are not always correlated with the SCS diagnostic imaging findings. For this reason, epidemiologic information is limited. Kalichman's group reported a prevalence of 22.5% and 7.3% in relative and absolute acquired SCS, respectively, and 4.7% and 2.6% in relative and absolute congenital SCS, respectively. This prevalence increased at the lumbar spine in older people (>60 years), affecting both sexes equally.

40.3 Etiology and Pathogenesis

The pathogenic mechanism through LSS and nerve root compression resulting in the typical clinical pattern has not been elucidated. In the case of LSS, the central area of the spinal canal and the neural foramina are reduced, exerting high pressure on the venules surrounding the nerve roots (Batson venous plexus). The compression could lead to venous engorgement with consequent ischemic nerve injury. The ischemic theory of LSS and claudication intermittent may explain the typical reversibility of symptoms when the patients flex their spine forward. Another cause that could explain the claudicatio intermittens is that the increased lordosis during standing and walking produces additional compression of the

caudal nerve roots through plication of the dural theca and folding inward the vertebral canal of the ligamentum flavum.

SCS becomes apparent with the onset of insidious symptoms. The most common disease pattern consists of a simple radiculopathy with an acute painful crisis and functional limitation of proximal muscles. At the lumbar level, symptomatology increases with gait and decreases with rest.

In terms of etiology, SCS is commonly associated with acquired or congenital degenerative pathology that produces spinal canal narrowing, as described in the next section. In addition, other risk factors such as obesity and previous family history can affect this condition.

40.4 Classification

Congenital stenosis can be congenital (due to anatomical conditions or other pathologies: achondroplasia, Morquio syndrome, spinal dysraphism, or spondyloepiphyseal dysplasia), acquired, or iatrogenic. Acquired stenosis can be the result of an injury, trauma, neoplasm, or surgery. Nevertheless, the most common type is related to degenerative changes that affect the vertebral bodies (osteoarthritis, spondylosis, etc.), intervertebral discs (dorsal hernia or protrusion), yellow ligaments, and/or posterior longitudinal ligament. However, many cases of stenosis in clinical practice result from the aggravation of the congenital stenosis (mixed form).

Several classifications have been described in the literature, but the most commonly adopted are the etiological classification (Table 40.1) and the radiographic classification (Fig. 40.2).

Table 40.1 Etiological classification of lumbar spinal stenosis

Type of LSS	Age of onset
Congenital—developmental (short pedicles) • Idiopathic • Achondroplastic, Morquio syndrome, spinal dysraphism, or spondyloepiphyseal dysplasia	20–40 years
Acquired • Degenerative – Central canal (disc degeneration, facet osteoarthritis, ligament flavum hypertrophy) – Peripheral canal (sciatica-like presentation) Lateral recesses – Spondylolisthesis • Combination of congenital and degenerative	60–90 years
Iatrogenic • Post-laminectomy (LSS typically occurs at the adjacent level) • Post-fusion	–
Spondylolytic (associated with spondylolisthesis)	20 years
Post-traumatic	
Others • Corticosteroid excess – Iatrogenic – Cushing's syndrome • Paget's disease • Acromegaly	–

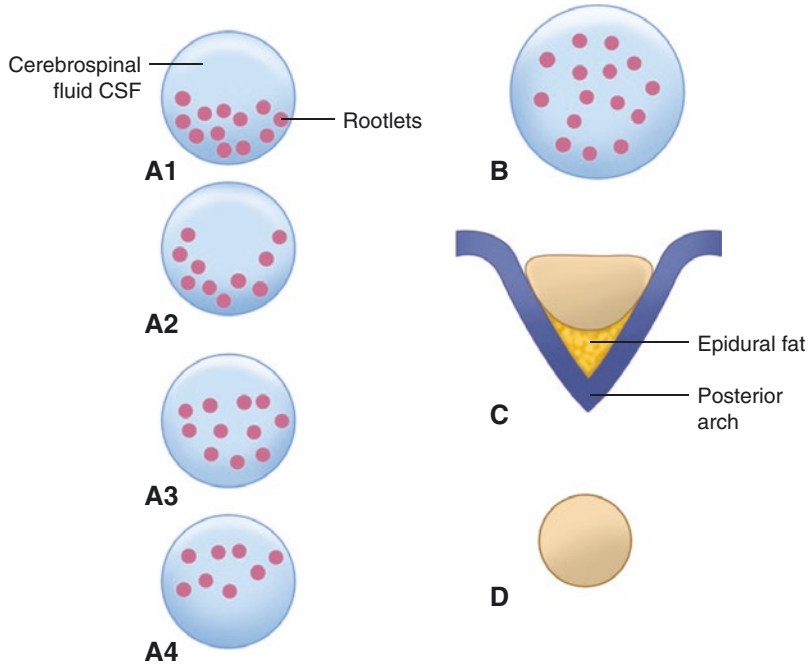

Fig. 40.2 Radiographic classification of lumbar spinal stenosis by Schizas et al. A1: the rootlets lie dorsally and occupy less than half of the dural sac area. A2: the rootlets lie dorsally, in contact with the dura but in a horseshoe configuration. A3: the rootlets lie dorsally and occupy more than half of the dural sac area. A4: the rootlets lie centrally and occupy the majority of the dural sac area. Grade B: the rootlets occupy the whole of the dural sac, but they can still be individualized. Some cerebrospinal fluid (CSF) is still present giving a grainy appearance to the sac. Grade C stenosis: no rootlets can be recognized, the dural sac demonstrating a homogeneous gray signal with no CSF signal visible. There is epidural fat present posteriorly. Grade D stenosis: no recognizable rootlets and no epidural fat posteriorly

40.5 Diagnosis (Clinical and Imaging)

An early and accurate diagnosis is important because there is never spontaneous regression, and treatment only prevents further progression of symptoms. However, there is no standard protocol for diagnosis, and it is the result of a combination of data from clinical history, physical assessment, and imaging assessment.

40.6 Physical Assessment

In the case of cervical impairment, exploration should include perceived pain, radicular symptoms, limb weakness, ataxia, and gait disorders. When the lumbar region is involved, most common symptoms are neurogenic claudication, myeloradiculopathy, sensory impairments, motor weakness, and pathologic reflexes. In this case, clinical testing such as lumbar extension loading can be useful. Further, physical assessment should evaluate how the related symptomatology may impact the patient's quality of life. The Romberg maneuver may reveal the involvement of the posterior column fibers. Otherwise, this sign could be differentiated with cervical or cerebellar injuries due to neurological or vascular conditions. In 80% of patients with LSS, a sensory or motor deficit could be observed, usually, assuming a bilateral polyarticular pattern. Although motor injury is mild, normally, it causes functional limitation.

Moreover, functional evaluation is important in these patients. Hip osteoarthritis, trochanteric

bursitis, and vascular claudication are to be considered as a differential diagnosis. Otherwise, the pain in hip osteoarthritis is referred to the groin, and it is exacerbated with internal rotation of the joint. Patients with trochanteric bursitis reported that they could not lie on the affected side, and palpation over the greater trochanter increases the pain. Vascular claudication is not influenced by lumbar movements (flexion or extension), and it is exacerbated by physical activity (walking).

A number of tests and scales with good psychometric properties have been developed to assess disability impact, such as the Modified Oswestry Disability Index, Modified Swiss Spinal Stenosis Scale, and Patient Specific Functional Scale. However, since neurogenic claudication is the most relevant symptom in lumbar SCS, gait tests and also gait analysis are important to assess functional capacity.

40.7 Imaging Examination

Overall, imaging examination includes the assessment of the foraminal narrowing and surrounding epidural fat.

The North American Spine Society guidelines recommend lateral X-ray as the most appropriate noninvasive method for detecting degenerative lumbar processes. By using this procedure, clinicians can evaluate bony abnormalities, listhesis and instability, translation and angulation, interpedicular distance, and transverse diameter. In addition, myelography (real-time X-ray imaging or fluoroscopy using contrast material) allows viewing the spinal cord and canal in detail. Nevertheless, to assess bone, intervertebral disc, and ligament condition, computed tomography (CT) provides a most accurate measurement. This method also identifies subluxation of the interapophyseal facet joints and allows measurement of spinal canal diameter (incipient narrowing is established with a sagittal diameter of between 10 mm and 13 mm, while greater narrowing suggests absolute root canal stenosis).

In addition, magnetic resonance imaging (MRI), as compared to CT, is able to more easily perform the analysis of the compression on the spinal cord, its extension, the effect on the nervous structures, and the possible suffering of structures. Therefore, MRI allows accurate assessment of acquired and congenital SCS because enhanced soft-tissue evaluation is possible. However, MRI can sometimes be inconclusive, and thus MRI or TC myelography is useful. Recently, MR myelography (MRM) has been developed as a new noninvasive technique to accurately evaluate spinal stenosis. MRM requires neither puncture nor contrast medium and causes no side effects.

40.8 Electromyography

This exam is not routinely warranted. It could be useful in patients with other neuropathies (e.g., diabetes) to determine which condition is responsible for symptoms.

Take-Home Message

- A common form of SCS in clinical practice is the mixed type (i.e., where congenital stenosis is aggravated by degeneration). Therefore, clinicians should take into account such possible degeneration when managing people with congenital stenosis.
- SCS affects the quality of life because of the derived symptoms. Therefore, a complete assessment should include the evaluation of daily tasks whose limitation can impact the quality of life. Knowing the potential limitations, health personnel should be capable of managing them.

Summary

SCS prevalence is expected to increase in the future, because of the ageing of the population. However, it has not yet been sufficiently studied. Available evidence suggests that an accurate diagnosis is important, not only based on causes (injury mechanisms), but also based on consequences (associated symptomatology).

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Which type of spinal canal stenosis is more common in clinical practice?
   (a) Mixed stenosis
   (b) Anterior stenosis
   (c) Foraminal stenosis
   (d) Posterior stenosis
2. What are the most common symptoms in lumbar impairment?
   (a) Neurogenic claudication, myeloradiculopathy, sensory disturbances, motor weakness, and pathologic reflexes
   (b) Vascular claudication and gait disturbance
   (c) Gait disturbance and motor weakness
   (d) Areflexia and pain
3. What clinical tests and scales are used to assess the impact on disability?
   (a) Modified Oswestry Disability Index, Modified Swiss Spinal Stenosis Scale, and Patient Specific Functional Scale
   (b) Visual analogue pain scale and numerical rating pain scale
   (c) KOOS, Japanese Orthopedic Association Score, and Modified Oswestry Disability Index
   (d) Satisfaction rating scale 36 and visual analogue pain scale
4. What type of diagnostic imaging provides a more accurate assessment of soft-tissue condition?
   (a) Magnetic resonance imaging
   (b) Computed tomography
   (c) X-ray
   (d) Computed tomography enhanced for soft tissue

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Lumbar Spinal Stenosis Treatment

41

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Sergio De Salvatore, and Luca Denaro

Overview

Lumbar spinal stenosis describes a broad spectrum of anatomical, clinical, and radiographic findings caused by a narrowing of the spinal canal and/or lateral recesses, leading to compression of the spinal cord and/or nerve roots.

41.1 Introduction

Lumbar spinal stenosis (LSS) describes a broad spectrum of anatomical, clinical, and radiographic findings (Fig. 41.1). The clinical manifestation is claudication, lower back and extremity pain, and reduced functionality. The use of a corset is indicated. Pain drug management of LSS includes acetaminophen, nonsteroidal anti-inflammatory drugs (NSAIDs) in case of acetaminophen non-responsiveness, and mild narcotics as the last line. The surgery aims to decompress the spinal canal and neural foramina, reducing the pressure of the nerve roots. The most commonly performed technique is decompression through laminectomy and partial facetectomy. Lumbar arthrodesis with fusion procedures is controversial, and the surgical indication needs to be tailored to the patient. Surgical treatment usually reduces symptoms, but a recurrence rate of 20% is observed after 7–10 years.

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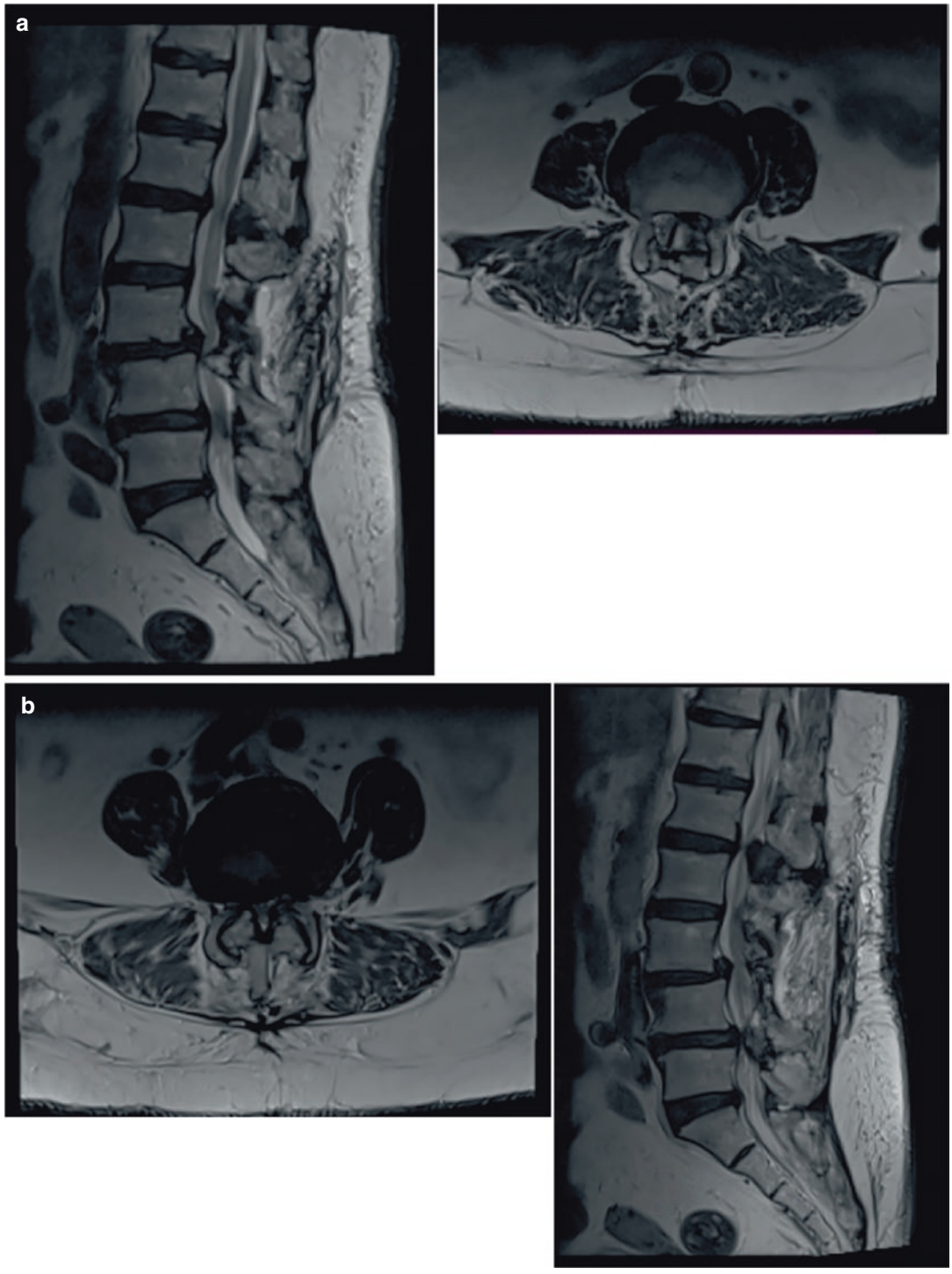

Fig. 41.1 MRI scans show severe stenosis of lumbar spine at L3-L4 (**a**) and L4-L5 (**b**) levels

41.2 Conservative Treatment

Anatomical anomalies of the spine (decrease or increase of lumbar lordosis) can alter the relationship between the containment and contents of the spinal canal. In some cases, restoration of normal lumbar lordosis can ensure optimal quality of life without neurological deficits. Therefore, in the absence of neurological symptoms (motor and sensory) or claudication, conservative treatment (postural gymnastics, strengthening of the abdominal and paravertebral muscles) may be indicated. However, surgical treatment will be necessary in cases in which the element that triggered the imbalance between the containment and the content of the spinal canal causes motor and sensory symptoms.

Global postural reeducation is characterized by a series of exercises to restore the physiological curves of the spine, particularly the lumbar lordosis. In lumbar stenosis in the early stages, the lumbar lordosis is inverted or significantly reduced. In these cases, exercises aimed at lengthening the iliopsoas muscle and strengthening the paravertebral muscles are helpful. By strengthening these muscles, including breathing exercises, and combining them with rebalancing exercises between the spine's posterior and anterior muscle chains, satisfactory results can be achieved. The combination of postural exercises, pharmacological treatment, and a corset is useful in resolving symptoms in patients with initial lumbar stenosis. However, this period of well-being is short-lived if the trigger is a herniated disc or an osteophyte from arthrosis or the loss of height of an intervertebral disc (with consequent shortening of the yellow ligament protruding into the vertebral canal).

However, there is a lack of high-quality studies on the conservative management of LSS.

The use of a corset is indicated. The assumption that the corset reduces the strength of the abdominal and paravertebral muscles is inaccurate. The corset has two functions: the first is to limit the excursions of the abdomen so that the patient feels sustained in the acute phase. The second is that the muscles of the anterior part of the abdomen and the posterior part (agonists and antagonists of the rachis) work in a craniocaudal sense. Therefore, the anterior and posterior muscles continue to work and do not develop atrophy. When the pain symptoms resolve, the corset can be removed.

Paracetamol is helpful in the early stages, but more importantly, a muscle relaxant should be used. It is beneficial to use centrally acting muscle relaxants (e.g., tricyclics such as Laroxyl) as they lower blood pressure and are better tolerated by patients than peripheral muscle relaxants. In addition, neuropathic pain modulators such as pregabalin may also play a role in treating chronic pain.

41.2.1 Epidural Corticosteroid Injections

In acute symptomatic patients with LSS, epidural corticosteroid injections could be adopted in the early stages or case of emergency. The injections (with or without anesthetics) could help reduce inflammation at the interface between nerve roots and compressing tissues, but their use is controversial. No high-quality randomized control trials proved the efficacy of this technique. However, many studies reported that epidural corticosteroid injections decrease leg pain for weeks to months.

41.3 Surgical Treatment

Claudicatio intermittens is a specific symptom. When the patients start to walk, they have no symptoms, but as they continue to walk, the pain (lower back and lower limbs) develops, and the patients are forced to stop. After a short period (5–15 min), the pain decreases, and the patient can resume walking. LSS is also called "shop window" disease, as patients stop and look at shop windows while waiting for the pain to recede. However, when the time interval between the onset of symptoms and walking reduces the walking autonomy to 100–150 m, the patient's quality of life is too limited, and surgical treatment is necessary. An additional indication for surgical treatment is the presence of radicular

pain (either metameric or diffuse). In these patients, stenosis of the lateral recesses and a reduction in the diameter of the spinal canal can cause compression of the contents of the spinal canal.

In case of a confirmed diagnosis of LSS with claudicatio intermittens and the presence of motor deficits in the lower limbs (with metameric distribution), surgical treatment must be carried out to avoid irreversible damage. In particular, in the case of compression of the L5 root, there is an inability to restore the complete functionality of the root. The two criteria for establishing a diagnosis of claudicatio intermittent include neural disturbances that impede the continuation of walking and that the walking can be resumed after 10–20 min of rest. A third criterion could be added: if the patients reported permanent neural disturbances, claudicatio is only accepted if it is caused by disturbances other than the permanent ones. Therefore, surgical treatment is mandatory in case of persistent symptoms after conservative treatments, in case of motor deficiency (also in the early stages), in case of sciatica, or in patients with severe claudicatio intermittens.

The surgery aims to decompress the spinal canal and neural foramina, reducing the pressure of the nerve roots (Fig. 41.2).

The most common procedure is laminectomy with partial excision of the lumbar facets to avoid destabilization of the vertebral segment. However, nowadays, total laminectomy is often replaced by a partial laminectomy called "recalibrage." Recalibrage consists of removing the hypertrophic yellow ligaments that protrude into the spinal canal, an economic hemilaminectomy of both the upper part of the lower lamina and the lower part of the upper lamina. The hemilaminectomy is performed longitudinally to maintain the relationships between the facet joints and release the foramina from which the nerve roots emerge. With this technique, it is also possible to reduce the thickness of hypertrophic laminae by removing the lower or ventral part of the laminae. These procedures have been called "recalibrage" because they aim to recalibrate the vertebral canal without the necessity of extensive operations. In addition, the less invasive nature of this technique allows to avoid or reduce postsurgical fibrosis, which is often the cause of chronic residual symptoms.

Lumbar arthrodesis with fusion is debated, and the surgical indication needs to be tailored to the patient.

In the absence of a herniated disc, it is not necessary to combine spinal fixation with recalibrage. In these cases, the intervertebral disc is often degenerated; therefore, it could not be removed. Furthermore, in the case of an uninjured intervertebral disc and longitudinal ligament, the elements of stability are preserved; therefore, spinal fixation is not required. Conversely, spinal fixation is indicated when the intervertebral disc is damaged, or elements of segmental instability are present (easily diagnosed by clinical examination and dynamic X-rays).

Performing a spinal fixation in the absence of signs of instability is therefore an overtreatment.

Spondylolisthesis represents a specific indication for spinal fusion, as some studies reported better results in patients treated by arthrodesis. However, there is no linear correlation between better results in spinal fusion and improvements in outcomes. Some authors reported that over 80% of patients who underwent spinal decompression improve after surgery. Otherwise, 7–10 years later, one-third of the patients experienced low back pain, reporting a reoperation rate of 10–23%. Patients with the worst functional and pain scores before surgery are the ones that reached better results.

Complications of decompression surgery include deep infections (1%) and recurrence (20%).

In highly narrow stenosis, particular care must be taken due to the high risk of dural tears. In these patients, the yellow ligament is hypertrophic and rigid and protrudes into the spinal canal. Over time, this degenerated ligament produces adhesions on the dural sac; therefore, there is a risk of dural tears when the yellow ligament is removed. A further complication is an insufficient decompression.

Removing the laminae and spinous processes without opening the conjugation foramen (without removing a large part of the inferior facet joint) can cause unstable compression and subsequent recurrences.

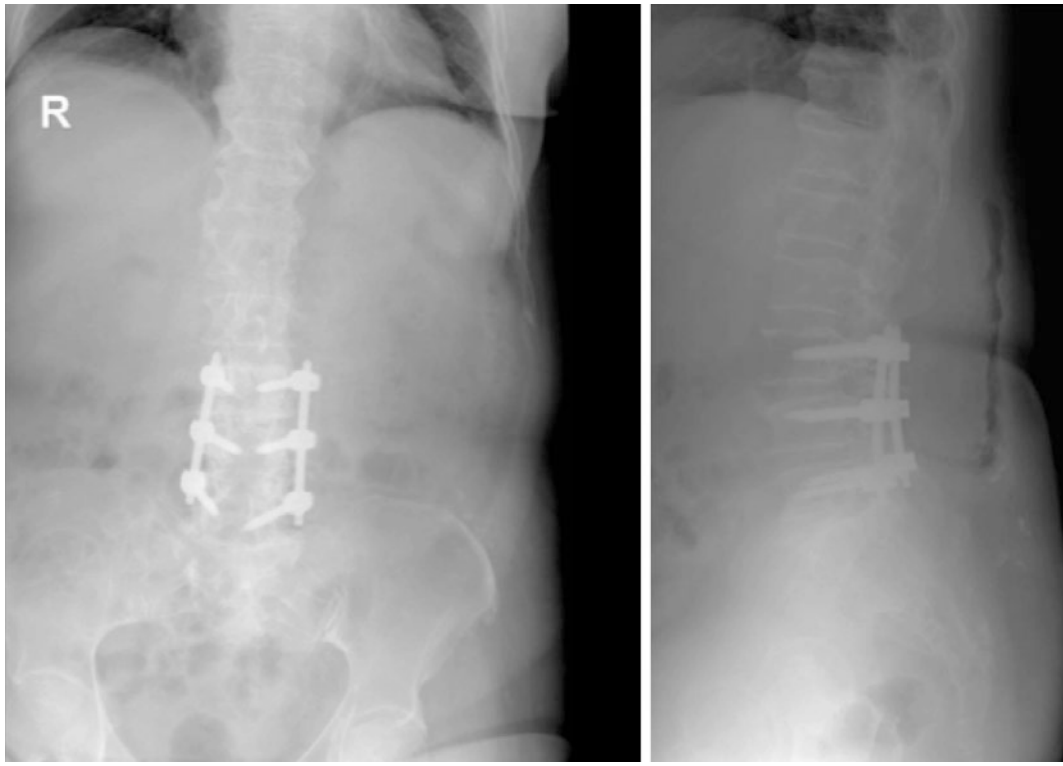

Fig. 41.2 Postoperative X-rays of posterior lumbar decompression at L3-L4 and L4-L5 levels and bilateral L3-L5 pedicle screw fixation

Finally, with the recalibrage, it is possible to suture the paravertebral muscles over the spinous and interspinous ligaments, increasing stability and providing a faster recovery than in patients treated with laminectomy.

Take-Home Message

- Lumbar spinal stenosis (LSS) can be managed with conservative treatments such as postural exercises, strengthening muscles, and the use of a corset.
- Global postural reeducation exercises can help restore the normal curves of the spine and improve early-stage lumbar stenosis.
- Further high-quality studies are needed to explore the effectiveness of conservative treatments for LSS.
- Surgical treatment becomes necessary when conservative measures fail or when there are neurological symptoms, motor deficits, or severe claudication.
- Surgical procedures aim to decompress the spinal canal and neural foramina, and the most common technique is decompression through laminectomy and partial facetectomy.

Summary

- Lumbar spinal stenosis (LSS) describes a broad spectrum of anatomical, clinical, and radiographic findings.
- LSS could often cause neurogenic claudication.
- The surgery aims to decompress the spinal canal and neural foramina, reducing the pressure of the nerve roots.
- The most commonly performed technique is decompression through laminectomy and partial facetectomy.
- Surgical management usually reduces symptoms, but a recurrence rate of 20% is reported after 7–10 years.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Surgical treatment of asymptomatic LSS:
   (a) It is not recommended as the majority of symptomatic patients treated conservatively do not worsen during the years.
   (b) It is recommended as the majority of symptomatic patients treated conservatively worsen during the years.
   (c) It is performed only in patients with a long life expectancy.
   (d) It is the gold standard to prevent progressive lumbar spine degeneration.
2. Pain drug management includes:
   (a) Acetaminophen, nonsteroidal anti-inflammatory drugs (NSAIDs) in case of acetaminophen non-responsiveness, and mild narcotics as the last line
   (b) Nonsteroidal anti-inflammatory drugs (NSAIDs), acetaminophen in case of NSAID non-responsiveness, and mild narcotics as the last line
   (c) Acetaminophen and mild narcotics in case of acetaminophen non-responsiveness
   (d) Nonsteroidal anti-inflammatory drugs (NSAIDs) as the gold standard
3. The aim of LSS surgery:
   (a) Is to decompress the spinal canal and the neural foramina
   (b) Is to reduce pain
   (c) Is to improve function
   (d) Is to prevent further spinal canal compression
4. Recalibrage:
   (a) Is a less invasive approach used to decompress the spinal canal in case of stable segments
   (b) Is a less invasive approach used to decompress the spinal canal in case of unstable segments
   (c) Requires a complete laminectomy
   (d) Needs to be performed only in the thoracic segment
5. With the recalibrage:
   (a) The inferior part of the upper lamina and the superior part of the inferior lamina are partially removed
   (b) The inferior part of the upper lamina and the superior part of the inferior lamina are completely removed
   (c) The superior part of the upper lamina and the inferior part of the inferior lamina are partially removed
   (d) The superior part of the upper lamina and the inferior part of the inferior lamina are completely removed

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Thoracolumbar Fractures

42

Moomal Rose Haris, Rajesh Botchu, and Harun Gupta

Overview

Thoracolumbar spinal injuries are commonly encountered, especially in the trauma setting. It is important for the radiologist to be aware of the main injury mechanisms and associated fracture patterns and to be able to convey meaningful information to the spinal team.

42.1 Introduction

Radiographs, CT and MRI are the most commonly used imaging modalities in the setting of spinal trauma. Though each modality has its own benefits and limitations, in combination, they often allow a more informed decision to be made regarding the injury sustained and patient management.

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42.2 Radiographs

Standard radiographic assessment of the thoracic and lumbar spine should include both anteroposterior (AP) and lateral projections. When forming an opinion regarding spinal radiographs, it is important that spinal alignment, presence of rotation, kyphosis, vertebral body height and any widening of the inter-pedicular or inter-spinous distance have been evaluated.

The upper thoracic spine is notoriously difficult to accurately assess on radiographs due to the overlying soft tissue and bones (shoulders, thoracic cage). This is important to realise as one cannot definitively exclude pathology in this area on radiograph alone—particularly in the trauma setting—and further imaging with CT is often indicated. Despite this, radiographs remain the recommended first-line investigation in patients with suspected T1–L3 injury with no neurological deficit.

Thoracolumbar alignment is assessed on both AP and lateral projections. There should be an equal distance between the spinous processes; additionally on the AP projection, the spinous process should be central with a symmetrical appearance of the pedicles and vertebral body. This appearance on the AP projection is colloquially known as an "owl's face" with the two pedicles representing the eyes and the spinous process representing the nose. If one of the pedicles is

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involved in metastatic process, a "winking owl" can be observed (Fig. 42.1).

The thoracolumbar transition zone and lumbar spine are relatively easy to demonstrate and assess on radiographs due to the even quantity of overlying soft tissue, which is useful in the trauma setting. Vertebral compression fractures in patients with osteoporosis can occur with relatively little trauma or force, and these are often detectable on radiograph. However, radiographs have limitations in ageing the injury, especially in the absence of prior relevant imaging. Generally, acute fractures are said to have sharp cortical margins and lucent fracture lines, whilst older fractures are thought to have smooth cortical margins with sclerosis. MRI is the superior imaging modality if ageing of a fracture is needed.

Thoracic soft-tissue swelling due to a paravertebral collection such as a haematoma in the trauma setting is best demonstrated on the AP projection, where there is focal widening of the mediastinum. In the lumbar spine, soft-tissue swelling is less discernible. Clues for this may include psoas muscle asymmetry on the AP projection.

Schmorl's nodes and limbus vertebrae are similar entities, which are commonly encountered pathology on thoracolumbar spinal imaging often on radiographs. Both these entities predominantly develop during adolescence. Schmorl's nodes are central protrusions of intervertebral disc material through the end plate of the vertebral body. These often occur spontaneously during adolescence but can also be seen in association with Scheuermann's disease, osteoporosis and other metabolic conditions. On radiographs, these appear as central end-plate lucencies often involving both superior and inferior end plates at a given level. These are generally asymptomatic but can occasionally be symptomatic due to irritation of the displaced intervertebral disc material contacting the vertebral bone marrow. Even if symptomatic, these are often managed conservatively. Limbus vertebrae most commonly occur

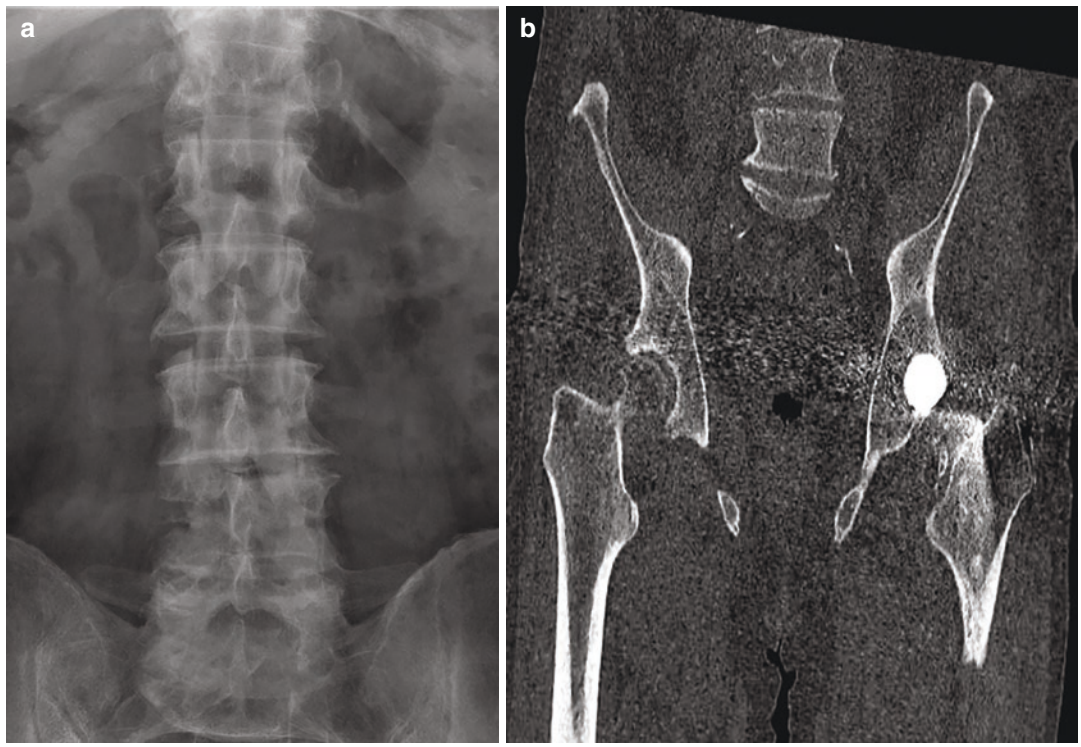

Fig. 42.1 (**a**) AP lumbar radiograph shows absence of the right L4 pedicle. (**b**) Coronal CT reformat of the pelvis of the same patient demonstrates lytic destructive abnormality of the right L4 pedicle

within the lower lumbar spine, specifically at the L4 level. They reflect anterior intervertebral disc protrusions with associated lifting of the ring apophysis. The typical location and well-corticated anterosuperior bone fragment should lead the clinician to recognise this as a limbus vertebra as opposed to a fracture.

In the trauma setting, it can be difficult and time consuming to obtain high-quality spinal radiographs in immobilised patients. Also, approximately 15% of spinal fractures occur at more than one level and are non-contiguous. It would therefore be wise to image the whole spine in the trauma setting to avoid a delayed diagnosis of a second fracture of which the current incidence is 23.1–83.3%.

Spinal radiographs also play a role in the follow up of stable burst fractures and post-surgical spines.

42.3 Computed Tomography (CT)

Dedicated soft tissue and bone window imaging through the thoracolumbar spine should be performed as part of the standard poly-trauma protocol. CT can be performed with relative ease and speed in the trauma setting. Various multi-planar reformatting (MPR) software exist to allow coronal and sagittal reformats of the spine. This allows easier identification and demonstration of bone fracture pattern, posterior process involvement and also fracture-dislocation injuries.

Multi-planar reformats should include the sternum as this forms part of the thoracic osseous ring. Sternal integrity is important with regard to the stability of a thoracic spinal fracture. CT is far superior in the detection of thoracic spinal fractures when compared to radiographs. Also due to the inherent rigid nature of the thoracic spine, osseous fractures are more common as compared to the mobile thoracolumbar transition zone where purely ligamentous injury can occur without any associated bone fracture (Fig. 42.2).

CT allows better assessment with regard to bone fracture pattern and dislocation injuries. MRI is superior to CT in the assessment of ligamentous injury. However, it is important to be aware of clues of ligamentous injury on CT. When assessing the posterior ligamentous complex on CT, features such as inter-spinous and facet joint widening, spinous process fracture and vertebral subluxation or dislocation should be determined. This in the least should prompt the radiologist to recommend an MRI to allow a more definitive assessment.

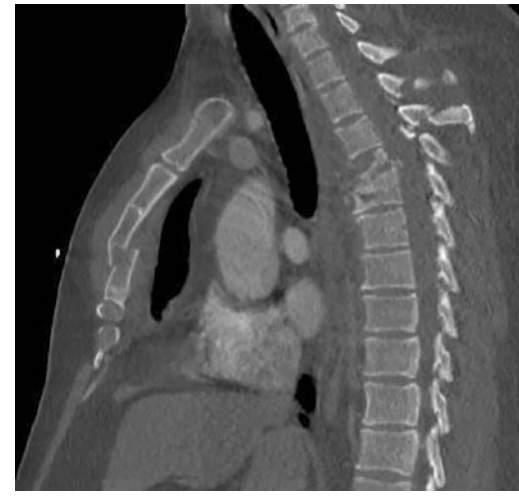

Fig. 42.2 Sagittal CT through the thorax demonstrates thoracic fracture dislocation with associated sternal fracture (disruption of the thoracic osseous ring)

The main concern with regard to radiographs and CT is radiation dose, especially in the paediatric population. A benefit-risk discussion should take place regarding the need for CT, but realistically in the trauma setting, the risk of significant and life-threatening injury or significant disability often overrides radiation risk.

42.4 Magnetic Resonance Imaging (MRI)

MRI is the gold standard in the evaluation of soft-tissue and ligamentous spinal injury. MRI is also useful in assessing spinal cord trauma, post-traumatic intramedullary abnormalities and traumatic disc pathology (Fig. 42.3).

MRI is helpful in ageing vertebral compression fractures. In the acute or subacute fracture stage, a larger area of geographical signal change is seen compared to in the chronic stage where a smaller

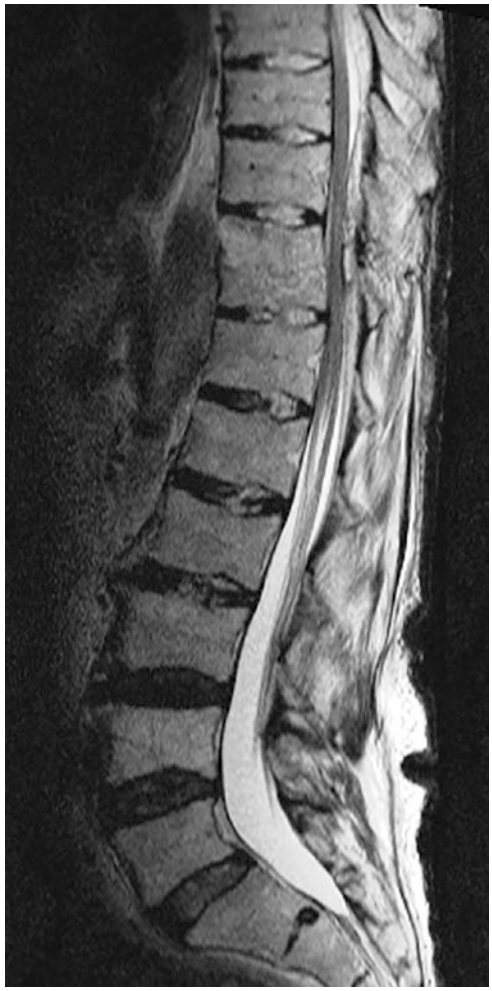

Fig. 42.3 Sagittal T2-weighted MR sequence of the lower thoracic and lumbar spine demonstrates T11 vertebral body fracture with posterior epidural haematoma at the T11 level with craniocaudal extension from lower T9 to upper L1 level

area of geographic signal change is observed with some restoration of normal fatty marrow. MRI can also help differentiate between benign and metastatic related fractures, especially with modified imaging sequences such as chemical shift, dynamic contrast-enhanced and fast spin-echo diffusion-weighted imaging. The presence of known background malignancy with osseous metastatic involvement and bulging of the posterior vertebral cortices are features which are more often associated with metastatic tumour infiltration.

The use of MRI, however, is restricted especially if there are contraindications, for example, but not limited to patients who have non-compatible cardiac pacemakers or metallic heart valves. MRI is also more time consuming and expensive when compared to CT and requires the patient to remain still for a prolonged period of time, which can be difficult if the patient is in acute pain.

42.4.1 Classification Systems

The Denis, AO and the Thoracolumbar Injury Classification and Severity Score (TLICS) systems are covered within this chapter. These are the most commonly utilised classification systems. Other classifications do exist; however, it is beyond the scope of this chapter to cover each and every spinal classification system. The primary aim of these classification systems is to give the radiologist a tool to aid in communicating important information regarding spinal injury to the surgeon. For the surgeon, it provides a guide to what management will be needed whether this is conservative or operative.

42.4.1.1 Denis Classification

Francis Denis developed a three-column theory of the spine in 1983. The three-column theory divides the spine into the following:

- Anterior column, which comprises the anterior longitudinal ligament (ALL) and the anterior two-thirds of the vertebral body.
- Middle column, which comprises the posterior longitudinal ligament (PLL) and the posterior one-third of the vertebral body including the posterior vertebral wall.
- Posterior column, which comprises all the structures posterior to the PLL. This includes the posterior osseous neural arch and the posterior ligamentous complex (PLC), which includes ligamentum flavum, inter-spinous and supraspinous ligaments and the facet capsule.

The basic principle of the Denis classification is that any injury which involves two out of three columns is unstable. This is now considered to be an oversimplification as one of the commonest two column thoracolumbar fractures is the burst fracture. However, many burst fractures are clinically stable and managed conservatively. Conversely minor end-plate fractures on a background of osteoporosis can progress to avascular necrosis, delayed vertebral collapse and deformity (Kummel disease). Sole assessment of the anatomical disruption of the spine is only one of the factors in determining the management of the patient, and it is important that the radiologist is able to identify the extent of bone and potential ligamentous disruption. Another issue with the Denis classification which was highlighted by the group forming the AO guidelines in the 1990s is that the middle column is a virtual column rather than an anatomic entity and therefore felt to be unfit to be used as a denominator of an injury type.

42.4.1.2 AO Classification

The AO classification is a comprehensive yet complex system. Due to this, its utility is generally reserved by specialist musculoskeletal radiologists and spinal surgeons.

The AO classification broadly consists of three separate components, of which only the first and third can be assessed, at least in part through imaging. The components include:

1. Morphology of the fracture
2. Neurological status of the patient
3. Presence of ligamentous injuries or co-morbid conditions (such as but not limited to osteoporosis and inflammatory spondylo-arthropathy)

The classification then subdivides each component into further subunits. For example, morphology of the fracture is divided into compression, distraction or dislocation injuries. These again are subdivided depending on the type of compression, distraction or dislocation injury sustained.

The second and third components are similarly subdivided. It is recommended that if this classification is used the radiologist should be descriptive as opposed to stating this is, for example, an A4 fracture, as this may not be understood by the referring clinician and may introduce an unnecessary clinical delay in patient management.

42.4.1.3 Thoracolumbar Injury Classification and Severity Score (TLICS)

The TLICS system was developed by the Spine Trauma Group in response to the fact that prior classification systems provided limited prognostic value and do not suggest onward treatment pathways. The TLICS system, similar to the AO system, is also divided into three components. The three components are also subdivided, but to a lesser extent than in the AO classification. A numerical score is then calculated for each of the above components; the higher the number, the more severe the injury needing urgent attention/management. The total score helps with decision-making with regard to conservative versus operative management. Generally, scores of 4 or greater often need operative management. If more than one fracture morphology exists, the single injury with the highest score is used. If multiple levels of injury are involved, each injury is assessed independently. The TLICS system highlights the importance of evaluation of the PLC integrity, done through MRI. It is essential for the radiologist to have knowledge of PLC anatomy and recognising whether it has been compromised (Fig. 42.4).

42.4.2 Fracture Mechanisms and Patterns

42.4.2.1 Compression Spinal Injuries

Compression injuries are secondary to axial loading, which refers to force directed through the top of the head and through the spine. These can occur in isolation or with a flexion component at the time of injury. An example of such an injury includes falling from a height and falling from a height with anterior bending, respectively. These types of injuries tend to result in vertebral com-

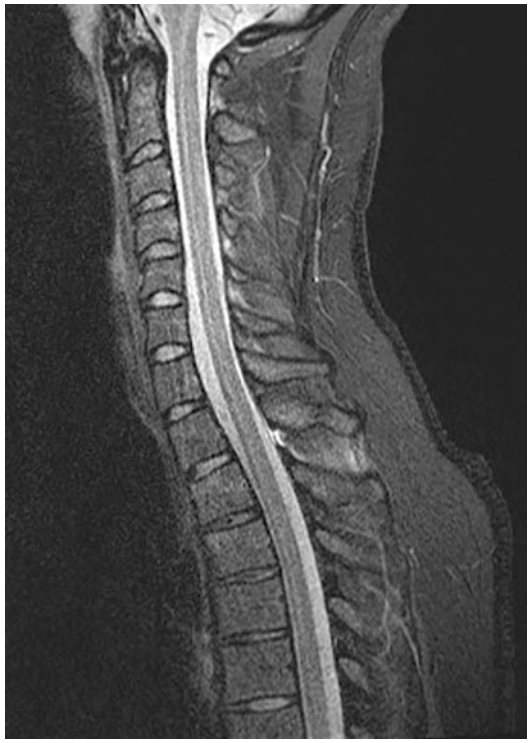

Fig. 42.4 Sagittal STIR MR sequence through the cervical and upper thoracic spine. Subtle bone marrow oedema at the T2 and T3 superior end plates with anterior wedging of the T2 vertebral body. Further oedema evident at the T1/2 level posteriorly secondary to PLC disruption (ligamentum flavum and supraspinous ligament disruption)

pression and burst fractures. The cervical and thoracolumbar transition zones are the most commonly involved in such injuries, though with flexion, the thoracic spine may also become involved.

Compression fractures are isolated vertebral body injuries resulting in shortening of the anterior column. Isolated end plate, wedge-type fractures and vertebral body collapse in osteoporosis are all examples of compression fractures. There can be multi-level involvement in 5–20% of cases. If there is 50% or greater loss of anterior vertebral body height on radiograph or CT, the radiologist should consider posterior ligament instability and perform an MRI for further evaluation. If this is ignored, it could result in progressive vertebral body collapse, posterior ligamentous involvement and delayed spinal instability (Fig. 42.5).

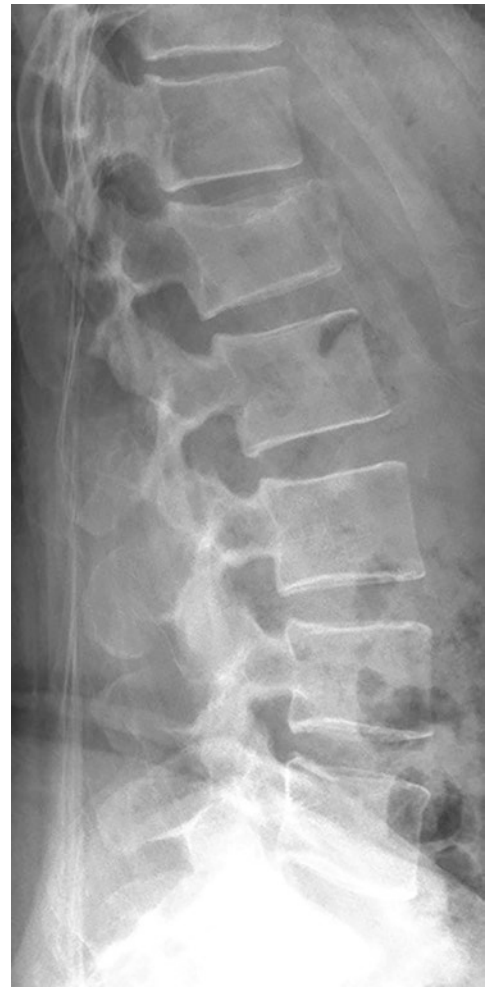

Fig. 42.5 Lateral lumbar radiograph. Mild superior endplate fracture of the L1 vertebral body with subsequent mild compression of the anterior vertebral body

Burst fractures are the most severe type of fracture resulting from axial compressive load with possible flexion injury. These fractures involve the posterior vertebral wall and can be incomplete, complete or burst split pattern. Often, there is a postero-superior fracture component which is dependent on size and retropulsion that can cause posterior longitudinal ligamentous disruption and spinal canal stenosis with injury. Typically, there is a loss of vertebral body height with possible increase in the inter-pedicular distance depending on the severity of the fracture. Approximately 25% of burst fractures are misdiagnosed as compression fractures if radiographs

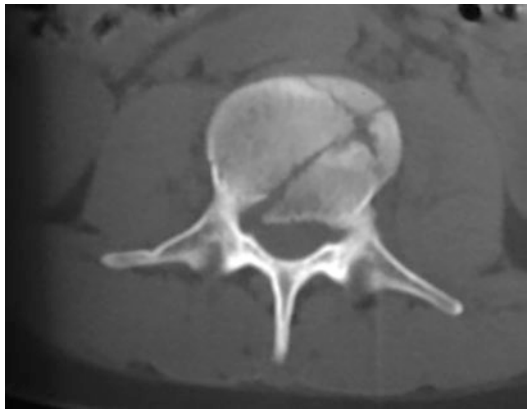

Fig. 42.6 Axial CT showing a burst fracture of the L2 vertebra

alone are evaluated. CT is therefore the preferred modality in the assessment of burst fractures with regard to assessing the degree and number of fracture fragment displacement. MRI may also be needed if there is associated neurology (Fig. 42.6).

42.4.2.2 Translation-Rotation Spinal Injuries

Translation-rotation injuries are often but not exclusively seen in the setting of road traffic accidents, where there is anterior bending and twisting at the waist level (seat-belt injury).

These injuries are defined at imaging as horizontal displacement or rotation of one vertebral body with respect to another. In most cases, there is failure of the posterior column to resist distraction with subsequent disruption of the PLC and also posterior column fracture. A variable degree of compression of the anterior column may also be evident. Posterior column disruption is significant in the classification of spinal injuries, as it indicates a high degree of severity of the injury with associated high incidence of neurological deficits. These injuries also often result in internal traumatic thoracic and abdominopelvic injury.

Seat-belt or Chance fractures are examples of injuries sustained post-translation-rotation injury. These fractures most commonly occur at the thoracolumbar transition zone. Chance fractures can occur at a single level with the fracture line extending from the vertebra into the pedicles and spinous process. This fracture pattern is termed a pure osseous Chance fracture. The PLC is not grossly involved. In other cases, there may also be extension of the fracture line through the pars inter-articularis to involve the PLC at the adjacent level. It is important to comment upon pars inter-articularis or adjacent vertebral level extension as this will affect surgical fixation. Widening of the inter-spinous distance should also be assessed. In these cases, where there is both osseous and ligamentous involvement, it is termed combined osseous and ligamentous Chance fracture. Conversely, if there is PLC disruption without osseous injury, this is termed a soft-tissue Chance injury—these can pose a diagnostic conundrum on radiographs and CT. Potential clues to a soft-tissue Chance injury include kyphotic spinal deformity. Kyphosis can occur with simple anterior compression fractures, but the degree of kyphotic abnormality is often proportionate to the degree of bone compression. In soft-tissue Chance fractures, the kyphotic deformity is exaggerated though the demonstration of this on radiographs and CT can be difficult due to patient positioning. However, the lack of kyphotic deformity cannot exclude a Chance injury. The neurological status of the patient is therefore of utmost importance in these cases. If there is any concern, MRI should be performed to allow optimal demonstration of PLC disruption. Chance fractures are often managed surgically; there is a better prognosis in pure osseous Chance fractures due to no ligamentous disruption compared to combined or soft-tissue Chance injuries (Fig. 42.7).

42.4.2.3 Fracture-Dislocation Spinal Injuries

Fracture-dislocation spinal injuries occur secondary to a combination of flexion and rotation such as a shearing mechanism where the upper part of the body moves in a different direction to the lower part of the body. These injuries are characterised by the rotation of two vertebrae against each other, with complete ligamentous disruption and abnormality in the sagittal and coronal planes (often both). These are high-force

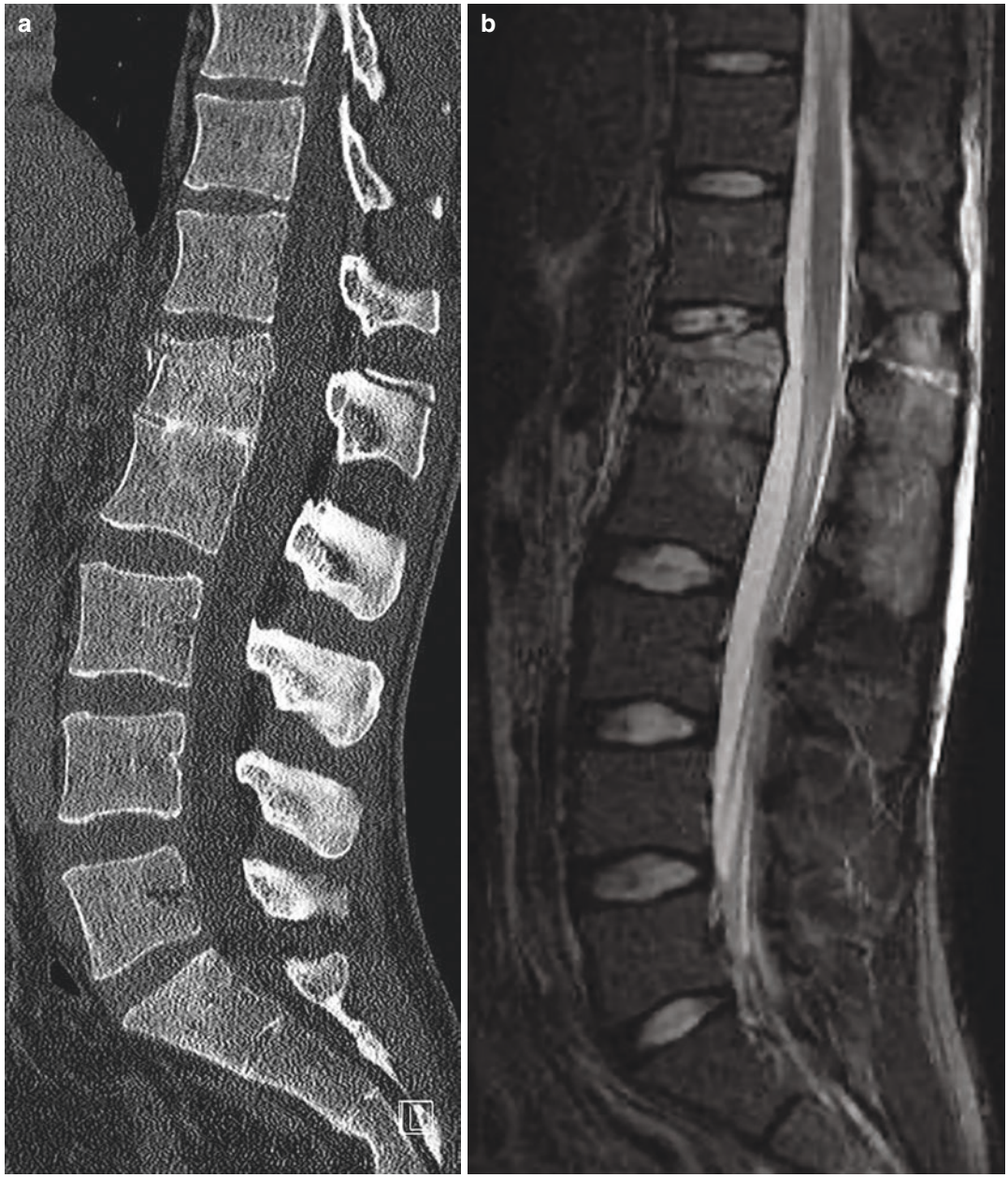

Fig. 42.7 (**a**) Sagittal CT through the lumbosacral spine. Chance fracture at the L1 level. Incidental fusion of L1/2 vertebra. (**b**) Sagittal T2-weighted MR sequence through the lumbosacral spine in the same patient. Re-demonstration of the Chance fracture at the L1 level. No epidural haematoma or conus abnormality

injuries and are innately unstable with a high incidence of severe neurological injury including cord transection. These types of injuries should not be mistaken for the commonly encountered spondylolisthesis seen at the L4–L5 level (often secondary to degeneration) and the L5–S1 level (either secondary to degeneration or bilateral pars inter-articularis defects). An urgent MRI should always be performed in patients with fracture-dislocation spinal injuries to assess neurological and ligamentous injury prior to intervention (Fig. 42.8).

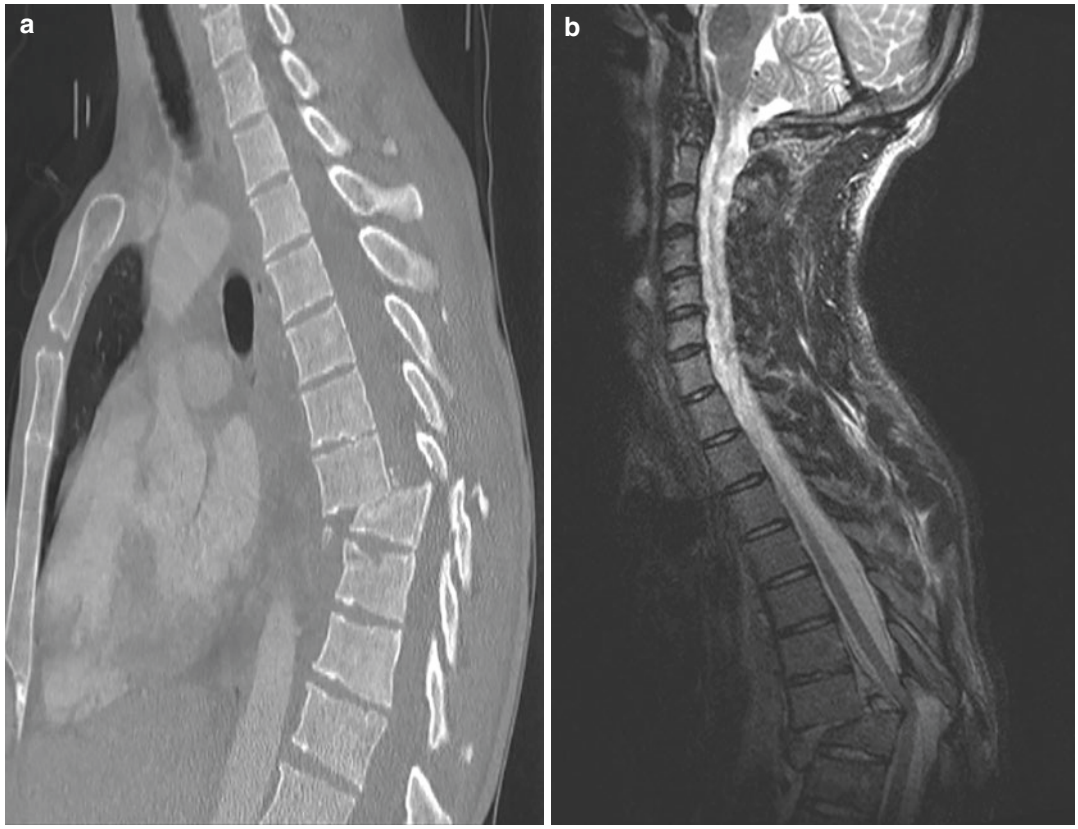

Fig. 42.8 (**a**) Sagittal CT through the thoracic spine demonstrates a fracture dislocation at the T8 level. (**b**) Sagittal STIR MRI through the cervicothoracic spine in the same patient shows the thoracic cord compression injury

42.4.3 Miscellaneous and Other Commonly Encountered Thoracolumbar Spinal Injuries

42.4.3.1 Lateral Compressive or Lateral Flexion Spinal Injury

These can occur when one falls on their side from a height or is struck from the side, respectively. These mechanisms of injury can result in asymmetric burst-type fractures with possible focal scoliosis.

42.4.3.2 Extension Injury

These can occur from a significant blow to the back of the spine/body. This mechanism of injury can result in anterior soft-tissue oedema with intervertebral disc widening and annular disruption as well as focal lordosis. These are uncommon fractures of the thoracolumbar spine.

42.4.3.3 Transverse Process Fractures

These can be seen in the context of trauma and can occur at several levels, but also be seen in isolation. In themselves, they are stable fractures and are often the result of avulsive stress from the psoas muscle or possibly related to paraspinal muscle contraction. If there is a transverse process fracture at the L5 level with concomitant ipsilateral sacral fracture, there may be associated nerve root (L5) and ligamentous injury. If there is any concern, MRI will be indicated.

42.4.3.4 Pars Inter-articularis Injury

Stress fractures of the pars inter-articularis tend to occur in adolescence and early adulthood. Established fractures of the pars are known as spondylolysis. These fractures are commonly seen in gymnasts and other athletes such as cricketers and football and rugby players who exert

significant flexion, extension and rotation of the lumbar spine. Patients tend to present with lower back pain, which is worse in extension, and there may be reduced straight leg raising ability and tightness in the hamstring muscle compartment. Imaging allows identification of such fractures and grading. Pars stress fractures most commonly occur at the L4 and L5 levels and can be unilateral or bilateral. CT is the gold standard for the assessment of the osseous posterior neural arch and so pars defects are readily identified. However, CT is limited in its assessment of ageing and grading such injuries. Isotope bone scans with SPECT-CT component and MRI are better placed in the assessment of acute or subacute pars fractures. Isotope bone scans with SPECT-CT do expose the patient to a large radiation dose, which must be taken into consideration in the younger age group. Figure 42.9 reports some examples.

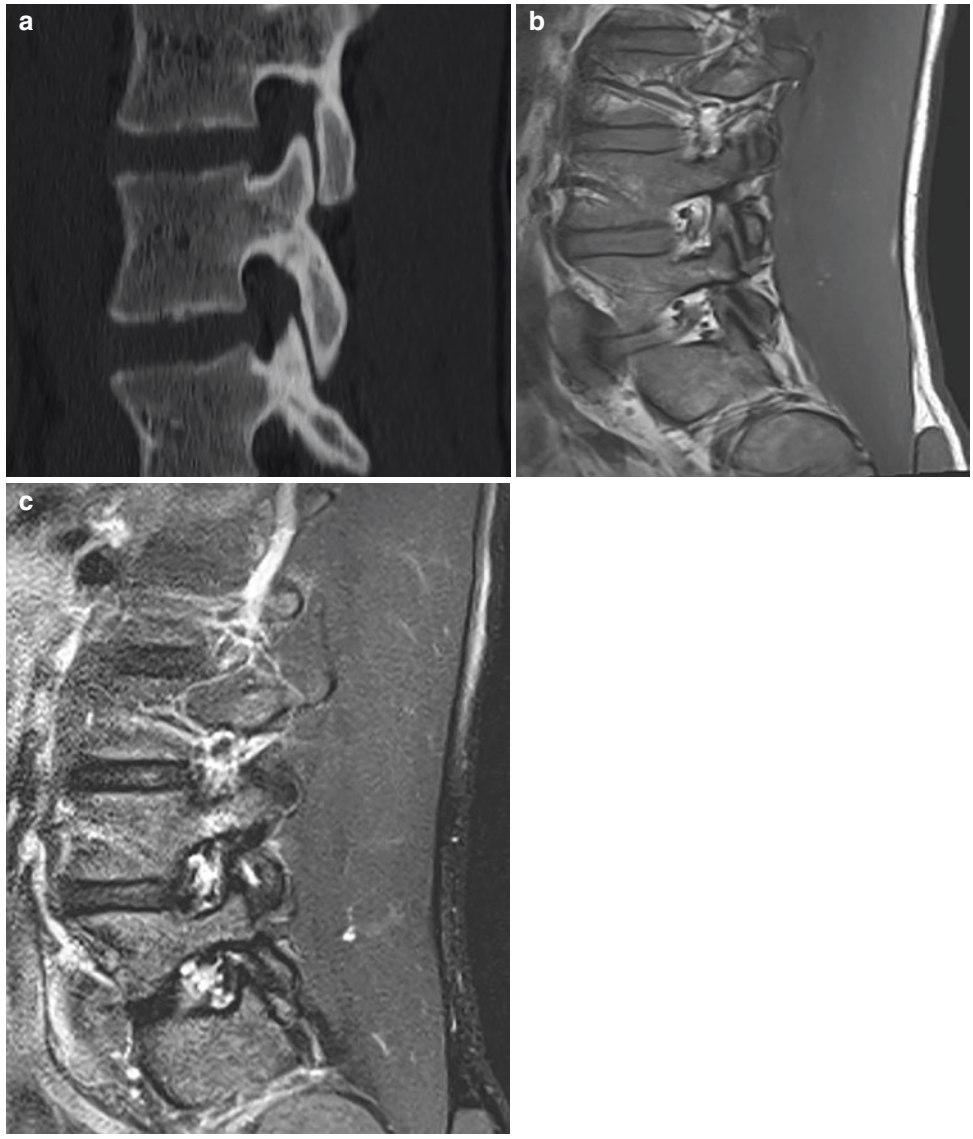

Fig. 42.9 (**a**) Coned in sagittal CT of the L4–L5 level. Lucent line at the L4 pars inter-articularis in keeping with an incomplete pars fracture. (**b**) Coned in sagittal T1-weighted MRI of the lumbosacral spine. Linear hypointensity at the L5 pars inter-articularis in keeping with pars fracture. (**c**) Coned-in sagittal STIR MRI of the lumbosacral spine. Bone marrow oedema at the L4 pars inter-articularis in keeping with pars stress change

42.4.4 Rigid Spine

Rigid spine can occur in patients who have ankylosing spondylitis (AS), diffuse idiopathic skeletal hyperostosis (DISH) and a surgically fused spine. The biomechanical stability in a patient with a rigid spine is altered. The lack of motion in the fused spine renders it with the biomechanical properties of a long bone. So, when a fracture occurs through the rigid/fused spine, it always occurs through the full width of the spine. Such trauma can occur with relatively minor impact injuries. The most common injury mechanism in such patients is hyperextension and often results in an unstable spinal injury (Fig. 42.10).

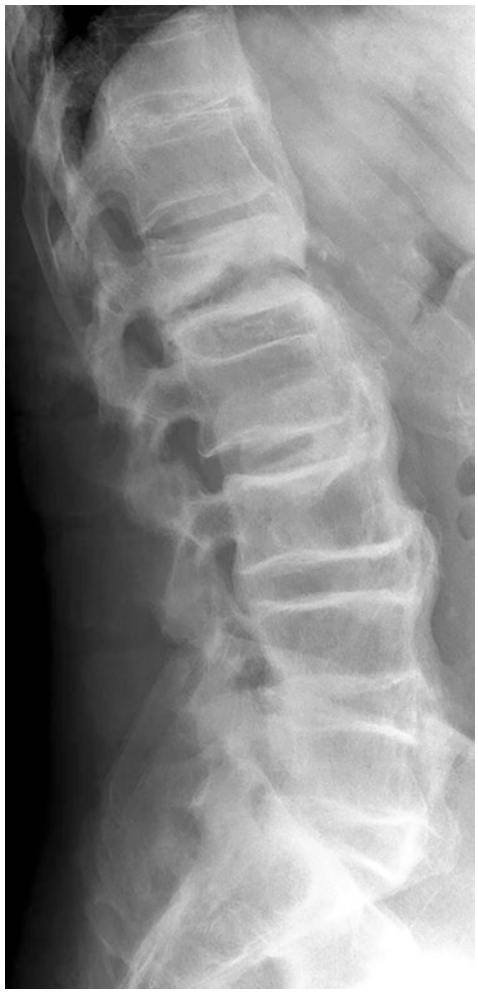

Fig. 42.10 Lateral radiograph of the lumbar spine in a patient with DISH. There is a transverse fracture through the entire width of the L1 level

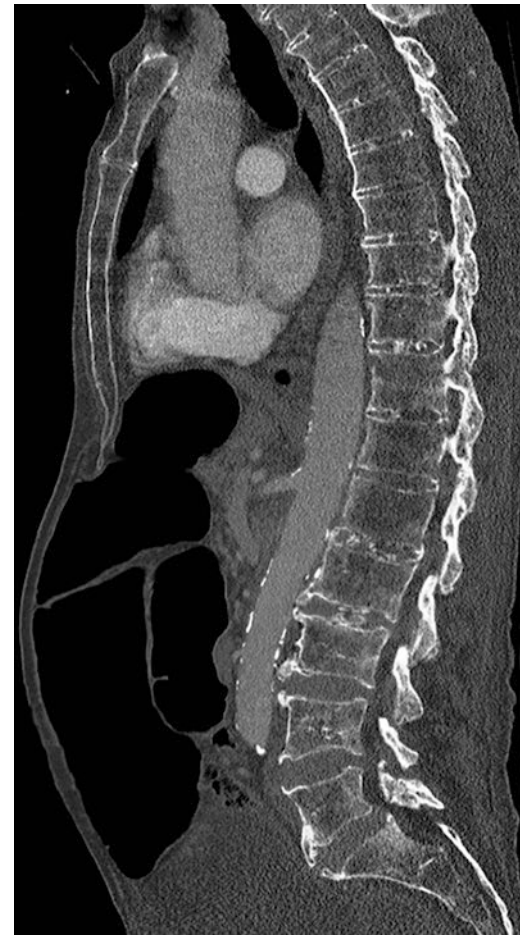

Fig. 42.11 Sagittal CT through the thoracolumbar spine. Full-width transverse fracture at the T11 level in a patient with DISH

CT is often considered first line in patients who are known to have a rigid spine and present with new back pain or suspected spinal trauma. Fractures through the fused spine often run in the transverse plane. It may be difficult to delineate the fracture on CT, if the fracture is un-displaced and the bones are osteoporotic. The presence of gas (vacuum phenomenon) within the fused spine is an indication of motion and hence fracture. There should be a low threshold in performing MRI in patients with a rigid spine due to concerns over occult fracture, epidural haematoma or spinal cord injury (Fig. 42.11).

Take-Home Message

- It is important for the radiologist to be aware of the main injury mechanisms and associated fracture patterns and to be able to convey meaningful information to the spinal team.
- AO classification system is the most common adopted.
- A multidisciplinary approach is often needed to ensure optimal patient management.

Summary

Thoracolumbar spinal injuries are commonly encountered, especially in the trauma setting. It is important for the radiologist to be aware of the main injury mechanisms and associated fracture patterns and to be able to convey meaningful information to the spinal team. This can be through descriptive reports or known classification systems, but this should be made clear on the report so there is no confusion. The incidence of multi-level fractures in the setting of spinal trauma should also be considered when protocolling requests. A multidisciplinary approach is often needed to ensure optimal patient management, and it is often better to have direct conversations with the surgeons especially in complex spinal trauma cases as they are also likely to have other traumatic internal injuries, which may affect prognosis or take priority.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. The diagnosis of a thoracolumbar fracture requires:
   (a) CT, MRI and X-ray
   (b) CT
   (c) MRI
   (d) X-ray
2. The Denis classification system divides the spine into:
   (a) Three columns: anterior, middle and posterior
   (b) Three columns: anterior, middle-posterior and posterior
   (c) Three columns: anterior, median and lateral
   (d) Two columns: anterior and posterior
3. The AO classification system consists of separate components:
   (a) Morphology of the fracture, neurological status of the patient and presence of ligamentous injuries or co-morbid conditions
   (b) Morphology of the fracture, presence of complications and age of patients
   (c) Age, BMI and comorbidities
   (d) Type of fracture and injury mechanism
4. The Chance fracture is reported in AO classification as:
   (a) Type B1
   (b) Type A1
   (c) Type A2
   (d) Type C
5. In case of rigid spine, the most common mechanism of vertebral fracture is:
   (a) Hyperextension
   (b) Compression
   (c) Distraction
   (d) Distraction and compression

Further Reading

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Part IX

Pelvis, Hip and Femur

Pelvic Fractures

43

Omer Kays Unal

Overview

Pelvis injuries are composed of a wide range of injuries, from low-energy osteoporotic fractures to high-energy disruption of the pelvic ring. Early mortality rate due to pelvic fracture is usually associated with hemorrhage or brain damage. Late mortality is usually due to severe sepsis and multiple-organ failure. Appropriate treatment strategy depends on the patient status and is extremely important in minimizing the mortality rate in such injuries. In addition, it is necessary to master the complex anatomy of this region for appropriate intervention. The main concept of treatment is stabilizing the patient's hemodynamic state. For this purpose, temporary fixation of the pelvis with some devices is widely used, and excessive bleeding from the venous plexus is rapidly prevented with this method. Open reduction and internal fixation are permanent treatments for pelvic fractures and should be performed when patients are hemodynamically suitable for this large-scale operation to avoid a second hit.

43.1 Definition

Before mentioning the pelvis fracture, understanding of the pelvis anatomy is crucial for correct intervention to the patient. The pelvic ring is composed of two paired innominate bones each is made up of three seperate bones (ilium, ischium, and pubis) which are connected with sacrum posteriorly and symphysis pubis anteriorly. Besides, ligamentous complexes (sacroiliac complex, sacrotuberous, sacrospinous, iliolumbar, and lumbosacral ligaments) contribute to the integrity of pelvic ring. Pelvic fracture is disruption of these bony and ligamentous structures, which surround and protect some vital organs such as external and internal iliac arteries which are derived from common iliac artery, complex venous plexuses, lumbar and sacral neural structures, and some parts of gastrointestinal and genitourinary systems.

43.2 Epidemiology/Etiology

Pelvic fracture's incidence is about 37 per 100,000 persons per year. Road traffic accidents are most common causes, and falls from height, crush injuries, etc. are other high-energy traumas which may cause life-threatening pelvic fractures. In the younger age group, it usually occurs as a result of high-energy injuries (falling from height, car accident, etc.), while in the geriatric

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group, mostly simple traumas can result as a pelvic osteoporotic fracture.

43.3 Pathogenesis

Stability of the pelvic ring depends on bone and ligament anatomy and requires high-energy traumatic events to cause fracture. Injury mechanism may differ according to the age for pelvic fractures. The direction and intensity of the force the pelvis is subjected to determine the type of fracture. Lateral compression forces that are commonly caused by side impact in motor vehicle accidents and falls force internal rotation in the pelvis. This usually results in anterior pubic ramus fracture and compression on the anterior part of the sacrum. If the anterior compression occurs directly through the symphysis pubis, pubic body overlap may be seen. Head-on motor vehicle accidents, falls, and crush injuries may cause anterior to posterior directed forces on the pelvis and create an external rotation moment on the hemipelvis, which leads to an "open book"-type injury. Disruption of the symphysis pubis is common with this injury, and rarely a pubic rami fracture may occur. If external rotation continues, the anterior sacroiliac ligament may be torn, but vertical stability can still be achieved with posterior ligaments. Vertical forces which are caused by falling from height create shearing force on pelvis. As a result of vertical forces acting on the pelvis, rupture occurs posteriorly in the SI ligament and anteriorly in the symphysis pubis ligament. Deterioration of the anterior and posterior support leads to vertical instability in the pelvis. In combined injury, there are multiple injuries in the pelvis as a result of the force applied from many aspects, and especially the loss of the posterior structures' support causes instability in the pelvis.

43.4 Classification

Pelvic fractures can be classified according to pelvic stability or direction of the trauma. Tile classification is based on pelvic stability and mostly used for preoperative planning guidance; besides, Young-Burgess classification is based on trauma mechanism and used for determining morbidity and mortality.

43.5 Diagnosis

The clinical assessment of the highly traumatized patient with suspected pelvic injury should be gentle but prompt. Pain is the main symptom, so after examination for abrasion and ecchymosis around the hip, manual compression at the iliac crests should be performed to assess abnormal movement and pain at the pelvic girdle. Because of the risk of rebleeding in self-limited hemorrhage, it is more appropriate to perform a clinical examination by the same physician at once. Additionally, asymmetry between the lower extremities is another sign of pelvic fractures.

Routine pelvic radiography after clinical examination in patients with high-energy trauma is recommended by the American College of Surgeons Advanced Trauma Life Support (ATLS) guideline. Direct X-ray of pelvis is extremely important in the detection of possible pelvic fractures and especially in terms of minimizing the mortality risk due to bleeding. Major anatomic landmarks and integrity of pelvis are checked at the pelvic X-ray (Fig. 43.1). If an

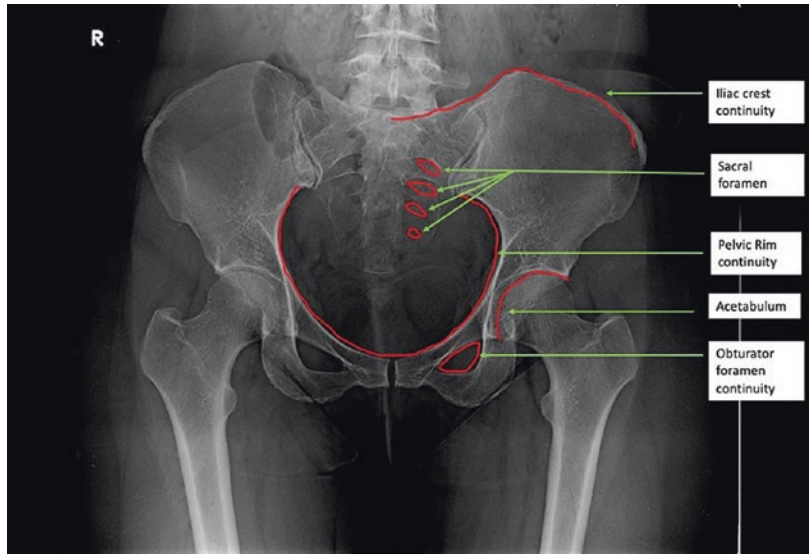

Fig. 43.1 Bony landmarks and anatomical lines should be checked at the AP pelvis X-ray

abnormal finding is detected on direct radiography, a detailed examination with computed tomography is required after the patient is stabilized hemodynamically.

43.6 Treatment

43.6.1 Acute Management

The most important issue in the treatment of a trauma patient is to ensure regaining of the vital functions. Especially for pelvic injuries, hemodynamic instability is rapid and life-threatening situation. Patients with pelvic fracture are at risk of major bleeding of presacral and paravesical venous plexus. Therefore, stabilization of the pelvis is crucial for controlling the hemorrhage. For this purpose, many devices (Sam pelvic sling, pelvic binder, T-pod, etc.) are developed and used in prehospital management or in the ER services. However, stabilization of the pelvis with a bedsheet, which is a simple method, is one of the most common methods even when a special device cannot be reached (Fig. 43.2). External fixators are another stabilizing method for insta-

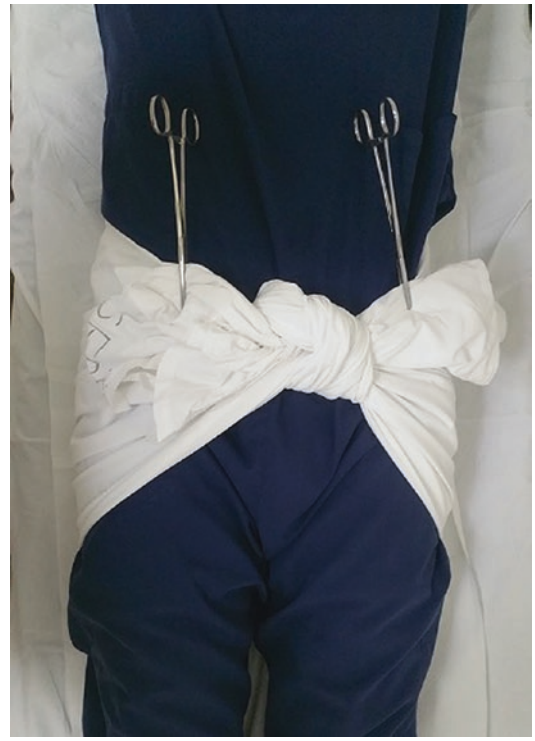

Fig. 43.2 Pelvic stabilization with bedsheet which covers both trochanter major and is knotted tightly at the level of symphysis pubis

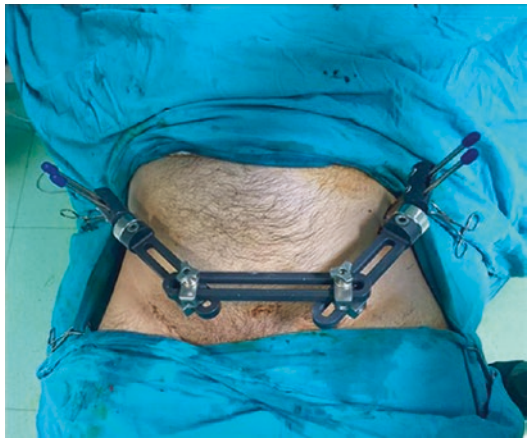

Fig. 43.3 External fixation of the pelvic fracture

ble pelvis fractures. These invasive methods can be done with rod–pin frame, or C-clamp. External fixator provides more stable pelvic fixation, but application requires expertise and special devices (Fig. 43.3).

43.6.2 Definitive Treatment

In orthopedic practice, the concept of damage-controlled orthopedics is an accepted treatment method, especially for polytraumatic patients. Delaying of definitive treatment until control of physiology is a priority in this modality. Permanent fixation of the pelvic fracture is planned according to the patient's hemodynamic status. When the patient is in a suitable condition for the operation, pelvic fractures are fixed with the appropriate surgical approach (Pfannenstiel approach, Stoppa extension, ilioinguinal approach, paramedian approach) and usually with plate-screw fixation (Fig. 43.4).

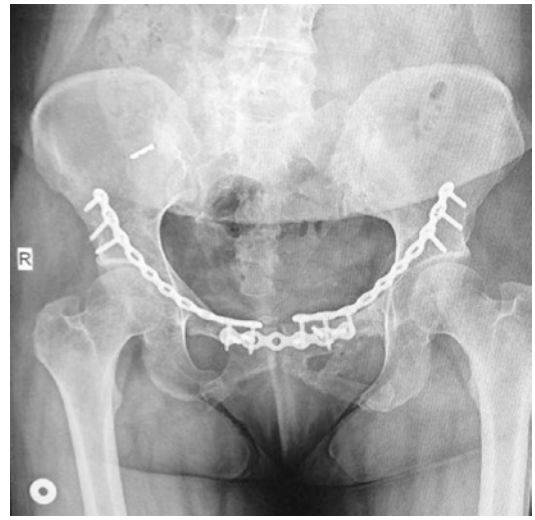

Fig. 43.4 Definitive fixation of the pelvic fracture with open reduction and plate-screw fixation

Take-Home Message

- Pelvic fracture is an orthopedic emergency and life-threatening injury.
- Hemorrhage is the most important reason of mortality.
- Immediate stabilization of the pelvis is essential and lifesaving.
- Definitive fixation should be delayed until hemodynamic stability.

Summary

Pelvis fractures are rare pathologies and usually result from high-energy trauma. It is extremely important that the patient should be evaluated and intervened by an experienced team in the process from the diagnosis at the time of the inci-

dent to the permanent treatment. Morbidity and mortality can be minimized with correct diagnosis and appropriate interventions.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. What is the main cause of mortality in pelvic fractures?
   (a) Major bleeding of presacral and paravesical venous plexus
   (b) Pulmonary embolism
   (c) DVT
   (d) Multi-organ failure
2. What are the main signs and symptoms of a pelvic fracture in a highly traumatized patient?
   (a) Ecchymosis in the pelvic area, pain on palpation, asymmetry in the lower extremities
   (b) Ecchymosis in the pelvic area, tachycardia, and diffused pain
   (c) Shock and diffused ecchymosis
   (d) Pain on palpation, symmetry in the lower extremities
3. What should be the first intervention for a patient with a pelvic fracture?
   (a) Stabilization of pelvis (sheet, pelvis stabilization devices, external fixator, etc.)
   (b) Fracture stabilization with plating
   (c) Fracture stabilization with open reduction and internal fixation (ORIF)
   (d) Pain management
4. Which organ or organ systems can be injured by the pelvis fracture?
   (a) Some parts of the genitourinary and gastrointestinal systems
   (b) Respiratory System
   (c) Liver
   (d) Spleen
5. When is the optimum time for definitive treatment of a pelvic fracture?
   (a) After hemodynamic stabilization of the patient
   (b) After trauma
   (c) After ORIF
   (d) Immediately

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Hip and Groin Pain

44

Madison Walker, Mahmoud Almasri,
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Overview

Hip and groin pain is a disabling condition, whose frequent causes are osteoarthritis, avascular necrosis, osteitis pubis, sports hernia, femoroacetabular impingement, bursitis, labral tears, hip dislocations, fractures, and snapping hip syndromes. The diagnosis is often difficult because of the complex anatomy of the area.

44.1 Introduction

Hip and groin pain is a disabling condition, whose frequent causes are osteoarthritis, avascular necrosis, osteitis pubis, sports hernia, femoroacetabular impingement, bursitis, labral tears, hip dislocations, fractures, and snapping hip syndromes. However, the diagnosis is often difficult because of the complex anatomy of the area. The aim of this chapter is to illustrate some of the most common causes of hip and groin pain, which will be discussed in detail in other chapters.

44.1.1 Osteoarthritis

Osteoarthritis (OA) is the most common form of arthritis and is a disease process of the articular cartilage and surrounding capsular tissue and synovium. It often affects weight-bearing joints in those of older age and results in significant pain and reduction in quality of life for patients. It has been estimated that hip and knee OA ranks as the 11th highest contributor to disability worldwide. Global prevalence estimates of radiographic confirmed OA are approximately 1–10%, dependent on the location, which makes it slightly less common than symptomatic knee OA. Rates of hip OA are higher in older individuals, and in women compared to men. Despite the lower prevalence, it is nonetheless important to

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address patient concerns and aim to improve pain and function. In fact, osteoarthritis carries an economic burden via three different mechanisms—direct costs, indirect costs, and costs associated with quality of life. This has been estimated to result in over $100 million per year of direct medical costs alone. Patients often present with hip pain localized to the groin and stiffness in the morning or after inactivity lasting less than approximately 30 min. Moreover, patients describe difficulties with activities such as walking or bending, and this disorder often leads to decreased range of motion.

Various classification systems exist for grading the extent of hip OA. These classifications are generally based on four characteristics: joint space narrowing, osteophytes, subchondral sclerosis, and subchondral cysts. As an example, hip OA may be graded according to the Tönnis classification as follows: Grade 0, no signs of osteoarthritis; Grade 1, mild joint space narrowing with sclerosis of the femoral head and acetabulum, and slight osteophyte formation; Grade 2, moderate joint space narrowing with small subchondral cysts of the femoral head and acetabulum, and moderate loss of femoral head sphericity; and Grade 3, severe joint space narrowing, large subchondral cysts, and severe femoral head deformity. However, it is important to note that symptoms do not always correlate directly with radiographic findings.

Osteoarthritis may exist as either primary OA of idiopathic origin or secondary OA in response to a disorder of the hip. This chapter reviews various causes of hip OA including avascular necrosis, developmental dysplasia of the hip, femoroacetabular impingement syndrome, and post-traumatic arthritis.

44.2 Avascular Necrosis of the Hip

Avascular necrosis (AVN) or osteonecrosis refers to the death of bone that results in disruption and collapse of the architecture of the bone. This in turn leads to progressive joint pain and loss of function of the joint. The ultimate driver towards bone death is due to a compromise in blood supply to the bone, regardless of the etiology. It can be further classified as traumatic or nontraumatic.

In traumatic AVN, there is a disruption to the vascular supply to the bone. In the hip, blood supply to the femoral head arises from the medial circumflex artery, which ascends anteromedially towards the femoral head. Various fractures, such as femoral neck or those of the femoral head, impair blood supply and can result in AVN. The causes of nontraumatic AVN are vast. The most common etiologies are corticosteroid usage and alcoholism, but other causes include sick cell disease, caisson disease, irradiation, idiopathic disease, and infection. Typically, these causes of AVN result in intravascular coagulation in the microvasculature. Arterial occlusion results in increased circulatory pressure and resistance in the bone, impairing the perfusion of the femoral head.

In either case, cell death occurs in the femoral head, and increased RANK-L results in osteoclast activation and bone resorption. This eventually results in microfractures throughout the bone, increasing pressure on the trabecular bone. The earliest radiologic sign visible is the crescent sign, representing the separation of the necrotic cancellous bone from the subchondral plate. Additionally, due to the reduced perfusion of the necrotic area, osteoblast recruitment is limited to the surrounding bone and forms a sclerotic band that prevents further penetration of the necrotic areas by growing blood vessels. Without blood supply to provide repair to the femoral head, the area ultimately fractures further and causes collapse of the femoral head. Flattening of the femoral head and degeneration of the joint result in abnormal hip mechanics and eventual osteoarthritis.

44.2.1 Treatment

Treatment for AVN is typically dependent on the stage of collapse. In the early, pre-collapse stage, some evidence has supported the use of bisphosphonates to prevent femoral head collapse; however, this has remained largely controversial.

Another nonsurgical option includes extracorporeal shock wave therapy, which has shown benefits in recent studies, particularly in improving Harris Hip Scores. While the exact mechanism of action is unknown, extracorporeal shock wave therapy is thought to induce neovascularization via increased expression of angiogenic growth factors and promotion of cell proliferation and osteogenesis.

Core decompression has also been utilized in the pre-collapse stage, where the goal is to reduce the hypertension in the femoral head. Core decompression with cell therapy, such as bone marrow aspiration and concentration, has shown promising results that may be superior to core decompression alone in early stages but inefficient in later stages. Other more invasive surgical options include femoral osteotomies, which transfer the weight-bearing area of the femoral head to an area of normal articular cartilage, or free fibula transfer that acts as a strut to prevent femoral head collapse. Finally, in the event of a visible crescent sign on X-ray or more advanced femoral collapse and arthritis, total hip surfacing or total hip arthroplasty (THA) is indicated. While older data suggested poor outcomes in terms of survivorship of THA following AVN, recent 10-year studies utilizing newer technologies and techniques have had favorable results, indicating that AVN no longer predicts poor outcomes following THA.

44.3 Developmental Dysplasia of the Hip

Developmental dysplasia of the hip (DDH) is a developmental hip disorder of acetabular dysplasia and a spectrum of hip instability, resulting in hip subluxation and gradual hip dislocation. Acetabular dysplasia indicates abnormal morphology of the acetabulum, but with maintained concentric articulation between the femur and acetabulum. Loss of this concentric contact between the articular surfaces indicates hip subluxation, while a dislocation refers to a complete absence of contact between the femoral head and the acetabulum.

Proper development of the hip relies on the concentric positioning of the femoral head in the acetabulum. Errors in this pathway are thought to arise from a number of factors, including female sex, intrauterine environment (oligohydramnios, breech presentation), family history, and maternal or fetal laxity. Anterosuperior acetabular deficiency, lateralization of the center of the rotation, and decreased contact area produce abnormal forces on the articular cartilage and lead to accelerated cartilage degeneration and possible development of early osteoarthritis.

Ideally, all children should be screened for DDH in infancy. Especially, those with an abnormal physical exam and presence of risk factors are advised to undergo ultrasound evaluation. The newborn screening examination includes the Barlow maneuver, which attempts to dislocate the femoral head via hip adduction and posterior force. Second, the Ortolani maneuver tries to relocate a dislocated femoral head with hip abduction and anterior force. However, these maneuvers are often only accurate for the first few months of life. Afterwards, asymmetry or limitations in abduction of the hips are the main sign and should be further evaluated. As well, limb-length discrepancies (Galeazzi sign) may indicate a dislocated hip. Infants less than 4–6 months of age should be screened using ultrasound imaging, evaluating alpha and beta angles, and screened for treatment according to their Graf classification. Those older than this should be screened using X-ray imaging. Various radiographic parameters exist to evaluate DDH, including Shenton's line and center-edge angle or acetabular index.

44.3.1 Treatment

The majority of DDH cases with dysplasia but no instability diagnosed within the first 6 weeks of life improve regardless of treatment. However, for infants older than 6 weeks of age and with ultrasound-diagnosed stable dysplasia, treatment with a Pavlik harness should be considered. Those that do not resolve should be treated with appropriate interventions. Higher age at presen-

tation is associated with worse outcomes following treatment as the ability of the acetabulum to remodel decreases after 3–4 years of age. For dislocated or subluxated hips, reduction of the femoral head is encouraged along with the usage a Pavlik harness to maintain the hip in flexion and 30–60° of abduction. This promotes stable, concentric positioning of the femoral head within the acetabulum and encourages acetabular remodeling. However, failure may occur and result in the development of late hip dysplasia. In cases of failure, or with children above 6–8 months of age, the hips should be reduced with either open or closed techniques and placed in a spica cast for immobilization.

In cases of persistent hip dysplasia despite immobilization techniques, surgical treatment in the form of femoral and/or acetabular osteotomies is needed to correct the hip dysplasia. The idea behind femoral osteotomies is to reorient the femoral head within the acetabulum, while acetabular osteotomies improve coverage of the femoral head. Such osteotomies include, but are not limited to, Dega, Shelf, or periacetabular osteotomies. Later in adolescence and adulthood, hip arthroscopy may be used to treat hip pain from labral tears or persistent hip dysplasia. However, the ability of hip arthroscopy to act as a hip preservation technique is limited to those with mild degrees of dysplasia, and in general outcomes have been reported as inferior compared to those reported in femoroacetabular impingement. Higher degrees of radiographic dysplasia with subjective hip instability may benefit from reorienting periacetabular osteotomies.

Patients with DDH often require total hip arthroplasty (THA) at younger ages than those with non-dysplastic hips. Following the development of osteoarthritis in those with DDH, approximately one in three patients require THA in 10 years' time and two in three patients in 20 years' time. However, this may be more technically challenging and may require specialized components, bone grafting, or osteotomies due to alterations in the anatomy of the acetabulum and proximal femur. As well, higher complication rates have been reported for those undergoing THA due to DDH versus primary OA. Complications include aseptic loosening, dislocation, delayed union of osteotomies, and heterotrophic ossification. Despite this, postoperative function has been comparable to individuals receiving THA for primary OA.

44.4 Femoroacetabular Impingement

FAI is a common hip pain-generating condition in young patients, often presenting with progressive onset of anterior groin pain worsened by activity. FAI syndrome encompasses three morphologic variations—cam-type, pincer-type, or mixed-type FAI. Cam type ("pistol grip") is believed to result from stress at the proximal femoral physis, which leads to cupping of the epiphysis during growth and thus asphericity of the femoral head. This bony deformity of the femur abuts against the acetabulum and may result in damage to the chondrolabral junction. In contrast, pincer type refers to acetabular overcoverage ("deep socket"), resulting in contact of the femoral neck and the acetabular labrum. This is thought to cause subluxation of the femoral head and a contra-coup injury to the posteroinferior capsular-labral junction.

The exact mechanism of how FAI results in osteoarthritis (OA) has not been completely described. There is a strong link between cam-type and OA and inconclusive data supporting a role of pincer-type FAI and OA. In fact, it is thought that pincer-type FAI almost exclusively damages the labrum. For cam-type FAI, repetitive abnormal contact between the femur and acetabulum is thought to result in labral and cartilage injuries, specifically cartilage delamination from the subchondral bone. As well, some reports have suggested a migration of the femoral head into damaged areas, primarily the anterolateral acetabulum. Such a migration alters normal joint contact area and joint forces, which may increase shear forces and lead to further damage of articular cartilage. These processes of cartilage damage can cause release of cartilage extracellular matrix degradation factors, including matrix metalloproteinases, that may trigger joint inflammation,

promoting catabolic processes in the hip and impairing joint lubrication, further increasing friction and wear in the joint and leading to the development of OA.

44.4.1 Treatment

The goal of treatment in FAI is to reduce pain and restore normal anatomy and mechanics of the hip joint. In terms of hip pain, different conservative options are available, including structured pelvic rehabilitation through physiotherapy (PT), nonsteroidal anti-inflammatory (NSAIDs), and activity modifications. PT targets muscular weakness, including core strengthening, and abnormal movement patterns associated with FAI.

Recent randomized control trials have suggested the superiority of surgical management over PT for hip-related quality-of-life outcomes in FAI. The goal of operative treatment for cam-type FAI aims to recreate sphericity of the femoral head to increase the head-neck offset of the femur, improving range of motion before impingement occurs. Traditionally, surgical treatment was completed through surgical dislocation of the hip. Advances in technology have allowed for these surgeries to be completed arthroscopically with similar results and improved return to play in athletes. During arthroscopy, osteochondroplasty can be completed under fluoroscopic guidance and improves outcomes compared to joint lavage alone. Suggested management of labral tears favors repair over labral excision due to better outcomes.

Rates of OA secondary to FAI have been reported as higher compared to healthy controls, with an increased risk of eventual total hip arthroplasty. However, controversial results exist regarding the salvage ability of arthroscopic procedures in preventing THA. Regardless, clinical outcomes for those undergoing THA after surgical treatment of FAI are believed to be similar to outcomes for primary THA.

44.5 Post-traumatic Osteoarthritis

Post-traumatic osteoarthritis (PTOA) of the hip typically presents in younger adults as a result of high-energy mechanisms such as motor vehicle accidents and sports injuries. Intra-articular fractures, as well as ligamentous and chondral injuries, are thought to contribute to the development of PTOA. The timing of symptomatic onset of PTOA varies greatly between patients. It may develop in as early as less than a year or may even develop 10–20 years following trauma.

The precise mechanisms of PTOA have been debated. Following acute injury, cartilage injury is thought to arise from three overlapping stages: 1) initial tissue damage, 2) acute inflammation and cell death, and 3) a chronic inflammatory phase. Joint instability and abnormal loading lead to stimulation of mechanoreceptors that stimulates catabolic cell pathways, causing production of matrix metalloproteinases (MMPs), and breakdown of the cartilage extracellular matrix. Further, damage to the articular cartilage results in the activation of various cellular responses, leading to alterations in chondrocyte metabolism and eventually cell death in nearby regions, preventing the maintenance of the articular cartilage. It is this combination of physiologic alterations in cellular processes and abnormal joint loading mechanics that leads to progressive pathophysiologic destruction and the eventual development of PTOA.

Fractures of the acetabulum have been associated with PTOA in particular. The risks of developing PTOA following acetabular fracture have been related to the type of fracture, poor fracture reduction (>2 mm step-off), and intra-articular fragments. Posterior wall fractures and posterior wall fracture-dislocation have been reported to have the highest rate of PTOA, ranging from 25% to 40%. In relation to posterior hip dislocation, femoral head fractures may occur in conjunction with posterior wall acetabular fractures and are associated with PTOA and poor outcomes. As well, femoral neck fractures may result in PTOA

through either insufficient reductions, malunion, or compromise of the vascular supply to the femoral head due to disruptions in the medial femoral circumflex artery, as discussed under the section of avascular necrosis.

44.5.1 Treatment

First and foremost, the goal should be the prevention of development of PTOA. This includes adequate steps towards proper fracture fixation to maintain joint congruity and prevent joint instability, such as open reduction and internal fixation of acetabular fractures, and either cannulated screws or hemiarthroplasty in terms of femoral neck fractures. Otherwise, treatment of PTOA should be aimed at reducing symptoms and preventing impairments on quality of life. This may include anti-inflammatory drugs, rehabilitation, and physical activity and lifestyle modifications. Patient education promoting realistic expectations and therapy aimed at optimization of muscle strength and control are important approaches to delay or prevent PTOA. Following conservative management, surgery may be indicated as a next step.

Often, treatment for PTOA involves THA. Rates of THA following open reduction and fixation of acetabular fractures range from 8% to 26%, often occurring approximately 2 years after the initial trauma. Compared to primary THA, these settings are associated with higher numbers of complications including heterotrophic ossification, infection, and dislocation. While THA is effective in reducing pain and improving function, long-term patient-reported outcomes have been reported as inferior to those for primary THA. This includes a lower 10-year survivorship for those who underwent THA due to acetabular fracture compared to primary THA (70% vs. 90%). Results of THA are dependent on a number of factors, including the status of the soft tissues, retained hardware, and bony deformities.

Take-Home Message

- Hip and groin pain is a disabling condition, whose frequent causes are osteoarthritis, avascular necrosis, osteitis pubis, sports hernia, femoroacetabular impingement, bursitis, labral tears, hip dislocations, fractures, and snapping hip syndromes.
- Patients with hip osteoarthritis typically present with pain in the groin region, stiffness, and limitations in range of motion.
- Radiographs typically demonstrate advanced joint space narrowing, subchondral sclerosis, and osteophyte formation.
- While hip osteoarthritis is often of idiopathic causes, secondary causes of hip osteoarthritis can be identified.

Summary

Hip and groin pain is a disabling condition, whose frequent causes are osteoarthritis, avascular necrosis, osteitis pubis, sports hernia, femoroacetabular impingement, bursitis, labral tears, hip dislocations, fractures, and snapping hip syndromes.

Hip osteoarthritis is a debilitating condition that results in hip pain, decreased function, and diminishing quality of life. Patients typically present with pain in the groin region, stiffness, and limitations in range of motion. Radiographs typically demonstrate advanced joint space narrowing, subchondral sclerosis, and osteophyte formation. While hip osteoarthritis is often of idiopathic causes, secondary causes of hip osteoarthritis can be identified. These causes include avascular necrosis, post-traumatic arthritis, developmental dysplasia, and femoroacetabular impingement. It has been estimated that hip and knee OA ranks as the 11th highest contributor to disability worldwide. Global prevalence estimates of radiographic confirmed OA are approxi-

mately 1–10%, depending on the location, which makes it slightly less common than symptomatic knee OA. Rates of hip OA are higher in older individuals, and in women compared to men. Despite the lower prevalence, it is nonetheless important to address patient concerns and aim to improve pain and function. In fact, osteoarthritis carries an economic burden via three different mechanisms—direct costs, indirect costs, and costs associated with quality of life.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. In grade 3 of Tönnis classification, it is possible to show:
   (a) Severe joint space narrowing, large subchondral cysts, and severe femoral head deformity
   (b) No signs of osteoarthritis
   (c) Moderate joint space narrowing with small subchondral cysts of the femoral head and acetabulum, and moderate loss of femoral head sphericity
   (d) Mild joint space narrowing with sclerosis of the femoral head and acetabulum, and slight osteophyte formation
2. In the early stage of AVN, the correct treatment involves:
   (a) Bisphosphonates to prevent femoral head collapse, and other nonsurgical options
   (b) Immediate surgery
   (c) Core decompression
   (d) Hip arthroscopy
3. The Ortolani maneuver is useful for:
   (a) DDH diagnosis
   (b) AVN diagnosis
   (c) Hip osteoarthritis diagnosis
   (d) FAI diagnosis
4. The surgical treatment preferred in FAI is:
   (a) Hip arthroscopy
   (b) Hip replacement
   (c) Hip resurfacing
   (d) Core decompression
5. In posterior hip dislocation:
   (a) There is a high risk of femoral head fracture and posterior wall acetabular fracture.
   (b) There is a high risk of femoral head fracture and anterior wall acetabular fracture.
   (c) There is a high risk of femoral neck fracture and posterior wall acetabular fracture.
   (d) There is a high risk of femoral neck fracture and anterior wall acetabular fracture.

Further Reading

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Hip Osteoarthritis

45

Biagio Zampogna and Rocco Papalia

Overview

Osteoarthritis represents the most common musculoskeletal chronic degenerative pathology. Several joints are mainly affected by OA, especially the hip, knee, hands, feet, shoulders, and spine. The hip joint is classified as enarthrosis (ball-and-socket type), which sustains an exceptionally high weight-bearing load. Hip articular cartilage underwent progressive wear due to static and dynamic stress during daily and sport activity.

45.1 Definition

Hip OA is defined as a degenerative process that involves the acetabular and femoral head articular surface. Hip OA results in pain, function loss, and worsening of quality of life for the patient.

45.2 Epidemiology

The hip is one of the most common joints affected by the degenerative process. According to principal surveys, hip OA's prevalence in the general population ranges from 4% to 10%. According to the National Healthcare Outcomes Program (PNE—2020 Edition), more than 100,000 total hip arthroplasties (THAs) per year are performed in Italy. A study published in 2014 highlighted that the number of implants would increase by 174% by 2030.

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45.3 Etiology/Pathogenesis

OA is the result of a different biomechanical and biochemical process involving the articular environment. Hip OA recognizes two etiology features: genetic and environmental.

According to genetic theory, it was demonstrated that patients suffering from hip OA compared to non-affected presented some specific genetic loci.

Environmental features are carried out from a biomechanical analysis of the hip joint. Patients with lubrication defects are predisposed to develop the disease because of increased friction and wear, like obese patients and manual workers. Joint congruency is mandatory for regular cartilage wear. Any anatomical defect, congenital or adolescence related, could affect congruency, especially in weight-bearing enarthrosis like the hip. Hip congruency could be affected by conditions like Perthes disease, avascular necrosis, prolonged steroid use, and pelvic trauma.

Another environmental factor is represented by the contact surface area that projects load force from the pelvis to the lower limb. In a few clinical patterns, like developmental dysplasia of the hip (DDH), the contact surface area is reduced with increased joint surface stress level.

45.4 Classifications

Hip OA is typically classified into primary and secondary diseases. Primary OA is age related, classically developed in the elderly population without any predisposing factor. Secondary OA is more often spread in younger patients suffering from one or more known predisposing factors that have altered the articular cartilage or the subchondral bone of the affected joints.

Primary hip OA, also defined as idiopathic OA, could be recognized as one of the following starter events: genetic factors, age-related chondrocyte degeneration, mechanical factors such as cartilage wear and tear (repetitive trauma, high-impact sports, abnormal joint alignment), and biochemical factors. Several features are recognized based on secondary OA: trauma, congenital anatomical anomalies, and inflammatory, neuropathic, hematologic, and metabolic joint disease (Table 45.1).

Table 45.1 Secondary hip OA etiology

Secondary hip OA etiology	
Femoroacetabular impingement (FAI)	Perthes disease
Developmental dysplasia of the hip (DDH)	Septic arthritis
Trauma (pelvic and proximal femur fractures)	Paget's disease
Hip avascular necrosis (AVN)	Hemophilia with recurrent hemarthrosis
Juvenile rheumatoid arthritis	Previous prolonged steroid intake
Ankylosing spondylitis	Cushing's disease
Slipped upper femoral epiphysis (SCFE)	*HFE* hemochromatosis (HH)

45.5 Diagnosis (Clinical and Imaging)

The patient's medical history and symptom characteristics should be primarily investigated. Several pathologies must be considered in differential diagnoses with hip OA: lumbar spine disorders, knee OA, trochanteric bursitis, psoas bursitis, myalgias, and pelvic inflammatory pathology. The main symptom is pain, and frequency, location, type, and irradiation have to be checked. Hip OA pain is classically located in the groin region and is often irradiated to the thigh. Patient inspection starts with walking analysis that shows antalgic gait due to pain during weight-bearing of the affected hip. Another typical gait pattern is Trendelenburg gait caused by the abductor's muscle insufficiency during gait.

The inspection phase is completed by pelvic tilt assessment, scoliosis presence, pelvis muscle tropism (gluteus and quadriceps), skin status (scars, swelling, and color), and knee joint evaluation.

Another hip physical exam step is palpation, although the hip joint position is more profound than the knee, ankle, or shoulder. Bony landmarks like great trochanter and ischial tuberosity and soft tissue like peritrochanteric muscles, hamstring, and adductor muscle groups are routinely checked. Then, lower limb discrepancy, with a possible compensation, needs to be investigated and evaluated through Coleman blocks. Hip range of motion (ROM) in OA's presence is severely reduced compared to normal values, especially for internal rotation. Both hips' active and passive ROM (flexion, extension, abduction, adduction, and internal and external rotation) need to be assessed. Normal hip joint ranges of motion are 120° for flexion, 45° for extension, 40° for abduction, 25° for adduction, and 45° for internal and external rotation. Then, a flexion deformity and abductor muscle deficiency could be evaluated by Thomas and Trendelenburg tests. Finally, lumbar spine and knee physical evaluation should equally be performed.

Hip OA diagnosis needs to be also confirmed by imaging techniques, while lab tests are instead helpful for differential diagnosis. Although MRI and CT scan provide several information, a plain pelvic X-ray with both hips' axial projection represents the widespread choice.

Pelvic X-rays account for four principal imaging features of hip OA: loss of joint space, osteophyte presence, subchondral sclerosis, and subchondral cysts (Fig. 45.1). Femoral head shape modification could be frequently located at the superolateral portion, especially in avascular necrosis.

Cartilage tissue presence guarantees joint space represented by radiographic radiolucency, while disease progression leads to contact between the acetabulum and femoral head. Osteophytes are composed of metaplastic tissue located at the acetabular joint rim and/or femoral neck. Structural weak and reactive areas are radiographically recognizable as subchondral cysts and subchondral sclerosis in advanced hip OA stages. Several radiographic classifications were described for hip OA. The Kellgren-Lawrence classification system described in 1957 is the most common for hip and knee OA, and it ranges from 0 (no OA) to 4 (severe OA). The system evaluates the following parameters: joint space narrowing, osteophyte presence, sclerosis, cyst formation, and bone deformity. Tönnis classification system is a qualitative tool to assess the hip OA stage on plain radiographs. It consists of a four-grade scale ranging from stage 0 (no OA) to stage 3. Arthroscopic assessment of coxofemoral cartilage damage can be performed through Outerbridge classification. CT scan (Fig. 45.2) is mainly requested for bone tissue evaluation in the presence of anatomical modification during preoperative planning for total hip arthroplasty (THA). MRI, on the contrary, is helpful to assess cartilage defect, acetabular labrum lesions, adductor and abductor muscle status, tendon pathology, and femoral or acetabular bone bruise or necrosis condition.

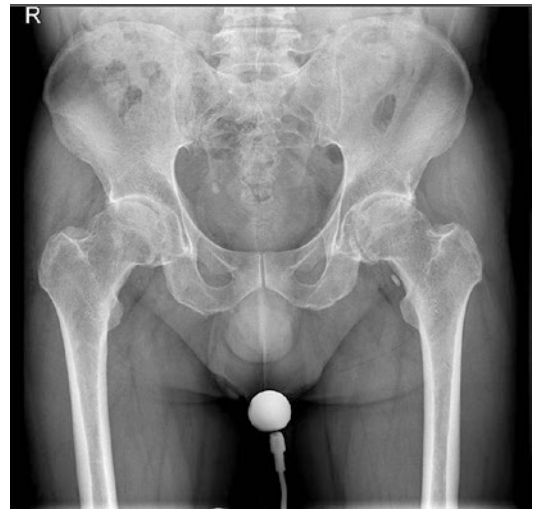

Fig. 45.1 X-ray of hip OA

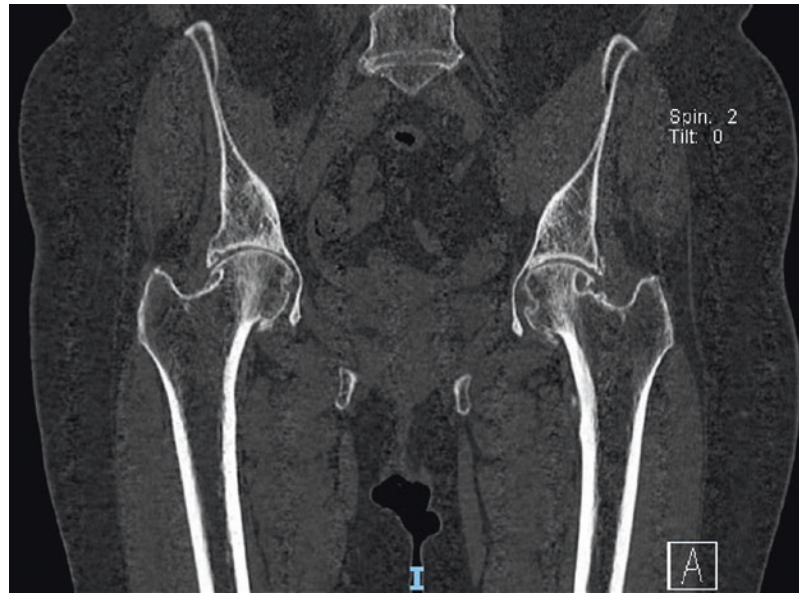

Fig. 45.2 Hip OA CT scan

45.6 Treatment

THA was defined as the operation of the century because of optimal results and very encouraging follow-up, and it is indicated for patients with end-stage OA. Several conservative treatments and joint-preserving arthroscopic or open surgical procedures have been described for early-stage hip OA.

Patient education (e.g., diet, weight loss) and individual risk factor management represent the first hip approach.

Pharmacologic strategy is crucial in the conservative management of hip OA. Analgesic therapy with paracetamol, NSAIDs, selective COX-2 inhibitors, and opioids should be tailored according to the patient's comorbidity and type and intensity of pain. Paracetamol is widely used in the early stages, and it is relatively free from adverse events. Across NSAIDs, selective COX-2 inhibitors are the most used, especially in patients with gastric ulceration history. Severe osteoarthritic pain, uncontrolled with NSAIDs, is frequently treated with opioid drugs, although they are not free from risks (dependence, constipation, and poor treatment compliance). In the last 10 years, a great interest has been in oral cartilage supplements, such as chondroitin sulfate and glucosamine.

Physical therapies, exercise, and adequate sports activity play a crucial role as nonsurgical solutions for degenerative hip disease. The role of core and periarticular muscles is fundamental in hip joint correct biomechanics. Several guidelines recommend regular physical activity in patients affected by moderate hip OA to control symptoms. Land-based or aquatic exercise (strengthening, aerobic, and flexibility exercises) and low-impact sports demonstrated effectiveness (reducing pain and improving function) as a conservative treatment for hip OA. As a consequence of physical activity, a body weight reduction could be useful in obese patients. Moreover, in the advanced OA stage, walking aids are often used with the aim of weight unloading.

Injective therapy is currently a developed and satisfying conservative approach. The aim of hip injective therapy is pain reduction and joint function reduction. Several injection substances are currently available: local anesthetic, corticosteroid, hyaluronic acid (HA), platelet-rich plasma (PRP), and mesenchymal stem cells (MSCs). The hip joint position often requires fluoroscopic or ultrasound assistance to ensure a real-time verification of intracapsular drug diffusion. Local anesthetics and corticosteroid injections also recover a diagnostic function in the differential

diagnosis of groin pain. HA recovers a visco-supplementative (synovial fluid properties' restoration) and visco-inductive role (endogenous production of HA induction). Several studies described the use of hip intra-articular PRP and MSC compared with HA with promising results.

The aim of hip joint-preserving surgery is to avoid conversion to THA. Several arthroscopic and open surgical procedures have been described: microfractures, simple debridement, autologous chondrocyte implantation (ACI), arthroscopic matrix-assisted autologous chondrocyte implantation (MACI), open ACI or MACI, and open mosaicplasty. These techniques aim to treat cartilage lesions often associated with the labral repair or anatomical anomalies' corrections as femoroacetabular impingement (FAI).

Hip OA conservative management treatment ensures a good quality of life but cannot slow down disease progression. Unfortunately, in a patient affected by severe end-stage hip OA, there is a need for joint replacement treatment. The modern satisfying THA results are mainly due to materials and surgical techniques across the last 40 years (Fig. 45.3). Several surgical approaches exist to perform hip replacement from direct anterior to posterior, aiming for tissue-sparing joint exposure. The implant is fixed to bone through two main fixation methods, with and without cement. The choice is performed according to bone quality, patient age, and intraoperative evaluation. Different THA-bearing surface materials are commonly implanted: ceramic or metal for femoral head and polyethylene, ceramic, and metal for the acetabulum. Modern material improvements ensure a higher implant duration nowadays.

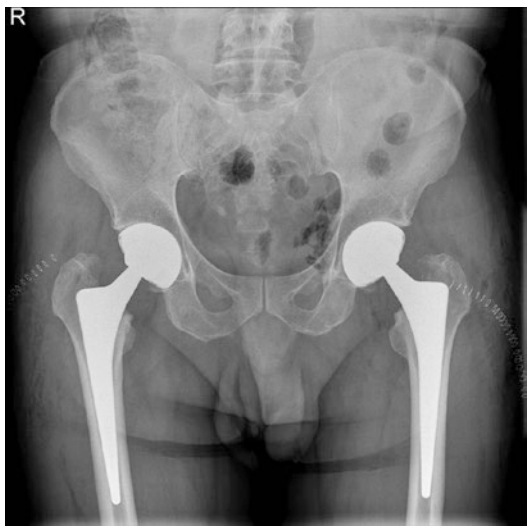

Fig. 45.3 X-ray of THA

Take-Home Message

Hip OA is a very disabling pathology that significantly affects the quality of life. Diagnosis needs to be confirmed by accurate clinical and radiological examination. Nowadays, several conservative or surgical strategies are available according to disease stage.

Summary

Hip OA is defined as a degenerative process that involves the acetabular and femoral head articular surface. Hip OA results in pain, function loss, and worsening of quality of life for the patient. According to principal surveys, hip OA's prevalence in the general population ranges from 4% to 10%.

A study published in 2014 highlighted that the number of implants would increase by 174% by 2030.

OA is the result of a different biomechanical and biochemical process involving the articular environment. Hip OA recognizes two etiology features: genetic and environmental. Hip OA is typically classified into primary and secondary diseases. Primary OA is age related, classically developed in the elderly population without any predisposing factor. Secondary OA is more often spread in younger patients suffering from one or more known predisposing factors that have altered the articular cartilage or the subchondral bone of the affected joints. The patient's medical history and symptom characteristics should be primarily investigated. Several pathologies must be considered in dif-

ferential diagnosis with hip OA: lumbar spine disorders, knee OA, trochanteric bursitis, psoas bursitis, myalgias, and pelvic inflammatory pathology. The main symptom is pain, and frequency, location, type, and irradiation have to be checked. Hip OA diagnosis needs to be also confirmed by imaging techniques, while lab tests are instead helpful for differential diagnosis. Although MRI and CT scan provide several information, a plain pelvic X-ray with both hips' axial projection represents the widespread choice. The Kellgren-Lawrence classification system described in 1957 is the most common for hip and knee OA, and it ranges from 0 (no OA) to 4 (severe OA). THA was defined as the operation of the century because of optimal results and very encouraging follow-up, and it is indicated for patients with end-stage OA. Several conservative treatments and joint-preserving arthroscopic or open surgical procedures have been described for early-stage hip OA. Patient education (e.g., diet, weight loss) and individual risk factor management represent the first hip approach, while surgery is reserved for late stages.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. What is the common cause of secondary hip OA?
   (a) Perthes disease
   (b) AVN
   (c) Both
   (d) None
2. Hip OA pain is often irradiated to:
   (a) Gluteus
   (b) Thigh
   (c) Lumbar spine
   (d) Abdomen
3. Trendelenburg test refers to:
   (a) Rotator cuff tear
   (b) Spine disease
   (c) Gluteus muscle insufficiency
   (d) Knee disorders
4. What is the best imaging exam to detect a hip AVN disease?
   (a) X-ray
   (b) CT scan
   (c) MRI
   (d) Ultrasound scan
5. Absolute contraindication in performing THA is:
   (a) Aged patient
   (b) Young patient
   (c) Presence of active infection
   (d) Severe pain

Further Reading

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Osteonecrosis

46

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and Umut Perçem Orhan Söylemez

Overview
Common causes and patients under the risk of ONFH have been well defined in the literature. Although total hip arthroplasty (THA) surgery is inevitable in advanced stages, it can be delayed by increasing the awareness among the patients that are at risk. This chapter provides a brief but thorough review of the current state of the art in femoral head osteonecrosis epidemiology, etiology/pathogenesis, diagnosis, and treatment.

46.1 Definition

Osteonecrosis (ON) (also referred to as avascular necrosis, aseptic necrosis, bone infarction, and spontaneous osteonecrosis) occurs in bones, particularly with poor blood supply, including femoral head, scaphoid bone, talus, etc. The femoral head is the most common site for ON. Osteonecrosis of the femoral head (ONFH) can cause serious mortality by disrupting the blood supply of the femoral head, and results in local necrosis, especially in the weight-bearing area of the head. This condition usually affects adult patients in the third to fifth decades of life. ONFH in children is called Legg-Calve-Perthes disease (LCPD) and differentiates from adult form both in diagnosis and treatment. The adult form is more common in men before the fourth decade and more common in women after the sixth decade. Etiologies, diagnosis, clinical manifestation, and prognosis of both forms have been well documented in the literature. Although several direct and/or associated risk factors have been reported to cause this condition, the exact mechanism of the disease remains obscure. Trauma-associated risk factors like femoral neck fractures, fracture-dislocation of the hip, sickle cell disease, and caisson disease directly disrupt extraosseous arterial circulation or venous drainage after the trauma. Nontraumatic risk factors including corticosteroid administration and alcohol abuse may cause arterial occlusion or venous compression after increased intraosseous tension due to metaphyseal edema. The result is almost always the same: the ischemia. The extent and location of the ischemia determine the stage of the disease. Early stages are better treated with minimally invasive surgical and physical therapy modalities. However, more complicated surgery is required in the advanced stages of the disease. Although sev-

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eral new nonsurgical treatment modalities have been recommended, ONFH is still a common reason for hip osteoarthritis, which is a severely disabling condition. Surgical interventions including joint-preserving procedures and total hip arthroplasty (THA) remain as major treatment modalities in adults. LCPD in children is a different entity regarding treatment as the bone has enormous healing and remodeling capacity at this age. Surgical interventions, including pelvic and/or femoral osteotomies and head-reshaping procedures with safe dislocations, aim to preserve the bone and obtain a relatively congruent joint. However, if the hip congruity cannot be achieved, osteoarthritis develops depending on the Stulberg stage between the third and sixth decades of life, and THA remains as the only treatment for these patients.

46.2 Epidemiology

Twenty to 30,000 people have been reported to be diagnosed with ON every year in the United States. Of all ONFH patients, 5–12% have been reported to have undergone a total hip arthroplasty. In the UK, the prevalence of the ONFH has been estimated to be around 1.4–3.0 per 1,000,000 patients, and it is the third most common indication for THA in people under 50.4 years old. The incidence rate of ONFH has been estimated to be around 1.91 per 100,000 in Japan. Prevalence of cases had been reported to increase from 20.53 per 100,000 to 37.96 per 100,000 between 2002 and 2006 in Korea. The prevalence of ONFH has also been reported to be 0.725% among the Chinese population.

Although men were found to be more prone to ONFH, man-to-woman ratio depends on the age and individual concomitant diseases. It is more common in men before the 45th decade and more common in women after the sixth decade of life. The male dominance of the disease has been referred to as a higher rate of alcohol consumption and smoking. Side dominance for adults has not yet been reported.

46.3 Etiology/Pathogenesis

The histopathology and pathomechanics of the ON have been well documented in the literature, and a consensus has been reached. However, there is an ongoing debate on risk factors and etiologies. The blood suppliers of the femoral head are the basicervical extracapsular articular ring, ascending branch of the medial femoral circumflex artery, smaller secondary contributions from inferior and superior gluteal arteries, and artery of the ligamentum teres. Disruption of these arteries as well as venous drainage may end with ONFH. Several factors may interrupt the circulation of the femoral head. These factors can be either extravascular or intravascular. Extravascular factors are trauma-related risk factors including femoral neck fracture, dislocation or fracture-dislocation, sickle cell disease, hemoglobinopathies, caisson disease (dysbarism), Gaucher disease, and radiation. Intravascular factors are referred to as nontraumatic risk factors as well. In these cases, embolic matters such as clots, lipids, immune complexes, or sickle cells occlude the terminal arterioles in the subchondral bone of the femoral head. Osteonecrosis is characterized by a typical pattern of cell death and a complex process of bone resorption and formation. After the necrosis of the hematopoietic cells and adipocytes, an interstitial bone marrow edema occurs. Osteocyte necrosis is reflected in empty osteocyte lacunae. Reactive hyperemia and capillary revascularization occur to a degree in the periphery of the necrotic zone, and with the entry of the new blood vessels, a repair process begins consisting of both bone resorption and production that incompletely replaces dead bone with living one. In the subchondral trabeculae, bone resorption exceeds formation and results in the net removal of the bone, loss of structural integrity of trabeculae, subchondral fracture, and joint incongruity. Lucencies on X-rays reflect bone resorption, while areas of sclerosis are comprised of both living reparative bone and dead trabeculae.

The causes of ONFH are reported as follows: nontraumatic risk factors including corticoste-

roid use, alcohol abuse, hospitalization, any cancer, osteoporosis, connective tissue disease, osteoarthritis within the past 5 years, high blood levels of triglycerides, cholesterol, male sex, urban residence, family history of osteonecrosis of the femoral head, heavy smoking, glucocorticoid intake, overweightness, obesity, systemic lupus erythematosus, Cushing's disease, hypersecretion of cortisol, chronic renal failure/hemodialysis, pancreatitis, pregnancy, organ transplantation, intravascular coagulation, thrombophlebitis, hyperuricemia/gout, and HIV. ONFH patients with no known history of the above causes are called idiopathic cases.

Of all these conditions, corticosteroid administration and alcohol abuse have been determined as the most common two major risk factors for nontraumatic ONFH. The most common cause of ONFH is corticosteroid use.

46.4 Classifications

Several classification systems depending on radiological findings and physical examination have been developed to classify the disease. Regarding these systems, Ficat and Arlet classification (FC) and Steinberg University of Pennsylvania classifications are the most used ones to anticipate the prognosis and aid the treatment. FC classification had been developed before the introduction of MRI. Thus, this classification has been adopted several times, and the most used version is the four-stage version. FC system classifies radiographic findings depending on the existence of sclerotic or cystic lesions, crescent sign, subchondral collapse, flattening of the femoral head, and osteoarthritis (Fig. 46.1). However, Steinberg classification employs X-rays, MRI images, and bone scintigraphy eval-

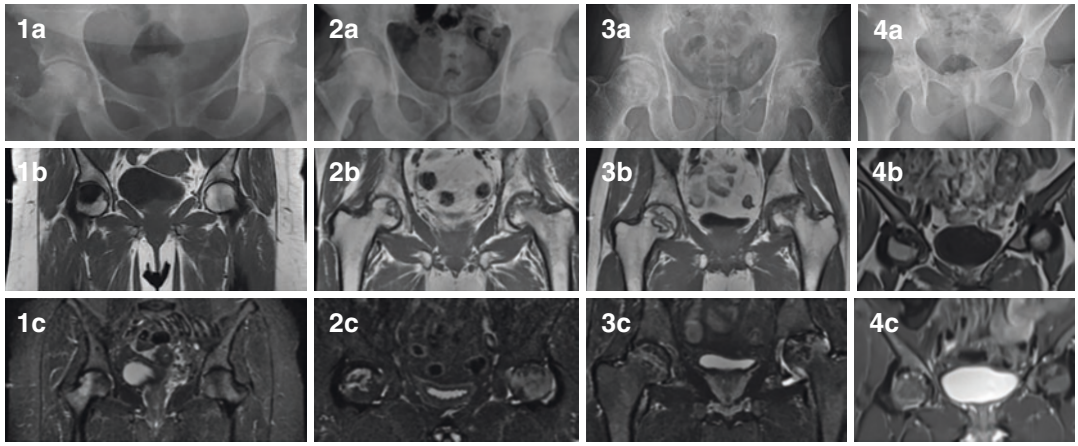

Fig. 46.1 *Ficat stage 1*: (**1a**) Mild sclerosis without crescent sign in the right femoral head, coxofemoral joint is preserved. (**1b**) Coronal T1-weighted image shows marked hypointensity due to edema in the right femoral head. (**1c**) Coronal T2-weighted image shows hyperintensity in the femoral neck due to bone marrow edema. *Ficat stage 2a*: (**2a**) Bilateral sclerosis in the femoral head without any collapse and subchondral fracture. (**2b**) Bilateral crescent-shaped areas, low-intensity serpentine band surrounding the normal tissue in coronal T1W sequence. (**2c**) Double-line sign in T2 sequence with fat saturation in the right femoral head. *Ficat stage 2b in right hip and Ficat stage 3 in left hip of the same patient*: (**3a**) Subchondral lucency with a rim of sclerosis in the medial aspect of the right femoral head and a curvilinear lucent line in the lateral part of the right femoral head; as for the left side, femoral collapse and subchondral fracture are seen but joint space is normal. (**3b**) Coronal T1W image, bilateral crescent-shaped areas. (**3c**) Coronal T2-weighted image shows bilateral bone marrow edema around the necrotic areas and effusion in the left joint, the serpiginous subchondral area across the upper portion of the left femoral head. *Ficat stage 4*: (**4a**) Subchondral cysts and osteoarthritic changes in both femoral head and acetabulum with narrowing of the joint on the right side. (**4b**) Coronal T1W image marked hypointensity in the femoral head. (**4c**) Coronal T2W sequences showing subcortical cysts in the femoral head and acetabulum, narrowing in the hip joint without bone marrow edema and joint effusion

uating the extent of the necrosis also with the factors that existed in Ficat and Arlet classification (Table 46.1). However, both systems have been criticized for their low inter- and intra-observer reliability. Therefore, the Association Research Circulation Osseous (ARCO) classification, which was adapted from Ficat and Steinberg classification, was developed. The ARCO had incorporated the radiographic findings with the location and extent of the lesion. However, later studies also found this system to be unreliable to predict the progress of the ONFH.

The modified Kerboul method is used to estimate the extent of necrosis. The sum of the angles in the mid-coronal and midsagittal MRI images is used to predict the collapse of the

Table 46.1 Classification systems

Ficat classification [21]			Steinberg classification [22]		ARCO classification [25]	
Stage	Radiographic signs	Symptoms	**Stage**	Criteria	**Stage**	Radiographic findings
0	Inconspicuous/ normal findings	–	**0**	Normal radiograph, bone scan, and MRI	**0**	Positive: histology negative/normal: radiograph/CT/MRI/ scintigraphy
I	Inconspicuous findings or minor changes (slight patchy osteoporosis, blurring of trabecular pattern, subtle loss of clarity)	+	**I**	Normal radiograph. Abnormal bone scan and/or MRI A: Mild (<15% of the femoral head is affected) B: Moderate (15–30% of the femoral head is affected) C: Severe (>30% of the femoral head is affected)	**I**	Positive: MRI and/or bone scintigraphy negative/normal: radiograph/CT *Subclass; localization of lesion, amount of involvement, length of crescent, and amount of depression*
II	Sclerotic or cystic lesions	+	**II**	Cystic and sclerotic changes in femoral head A: Mild (<15% of the femoral head is affected) B: Moderate (15–30% of the femoral head is affected) C: Severe (>30% of the femoral head is affected)	**II**	Radiograph: sclerotic, cystic, or osteoporotic changes of the femoral head *Subclass; localization of lesion, amount of involvement, length of crescent, and amount of depression*
IIA	Diffuse/focal radiological changes (osteoporosis, sclerosis, cysts)	+	**III**	Subchondral collapse without flattening (crescent sign) A: Mild (<15% of the articular surface) B: Moderate (15–30% of the articular surface) C: Severe (>30% of the articular surface)	**III**	Radiograph: subchondral fracture (“crescent sign”) *Subclass; localization of lesion, amount of involvement, length of crescent, and amount of depression*
IIB	Subchondral fracture (crescent sign^), segmental flattening of the femoral head (out-of-round appearance^)	+	**IV**	Flattening of the femoral head A: Mild (<15% of surface and <2 mm of depression) B: Moderate (15–30% of surface and 2–4 mm of depression) C: Severe (>30% of surface and >4 mm of depression)	**IV**	Radiograph: flattening of femoral head *Subclass; localization of lesion, amount of involvement, length of crescent, and amount of depression*
III	Broken contour of the femoral head, bone sequestrum, normal joint space	++	**V**	Joint narrowing or acetabular changes A: Mild B: Moderate C: Severe	**V**	Radiograph: flattening of the femoral head and osteoarthritic changes: decreased joint space and acetabular changes

Table 46.1 (continued)

Ficat classification [21]			Steinberg classification [22]		ARCO classification [25]	
Stage	Radiographic signs	Symptoms	**Stage**	Criteria	**Stage**	Radiographic findings
IV	Flattened contour of the femoral head, decreased joint space, collapse of the femoral head, acetabular osteoarthritic changes	+++	**VI**	Advanced degenerative changes	**VI**	Complete joint destruction

femoral head. Angles below 190° are predictive for low collapse risk, while angles above 240° are predictive for high risk. Independent from the size, the location of the lesion also becomes an important prognostic factor as lesions in a weight-bearing area can quickly progress into collapse. From this point of view, the Japanese Investigation Committee Classification (JICC) has been developed. According to this classification, lesions that occupy the medial one-third or less of the weight-bearing area have low collapse risk. However, lesions that occupy more than the medial two-thirds of the weight-bearing portion of the femoral head have a high risk for collapse. Also, femur head depression more than 2 mm is an indicator of poor prognosis, and the presence of bone marrow edema is largely related to the increase in symptoms and is a finding indicating that the structural damage is progressing into the collapse.

The Ficat classification is the earliest, yet still the most commonly used system. Although the latest classification systems like JICC systems have been reported to be promising and reliable, more evidence is required for more comprehensive use of the new systems.

46.5 Diagnosis (Clinical and Imaging)

Like all progressive musculoskeletal disorders, an early diagnosis of the ONFH may be beneficial for improving the success of the treatment outcome. Thus, general practitioners, musculoskeletal physiotherapists, and orthopedic surgeons must be aware of the predisposing risk factors for ONFH. Most of the ONFH patients refer to a general physician with nonspecific hip pain. A thorough history must be obtained to reveal the risk factors. The most common complaints are pain in the groin, buttock, thigh, or knee. Pain or limitation of internal rotation in a flexed hip may be indicative of ONFH. Excessive limitation of internal rotation shows a further collapse of the head. If physical examination reveals the limitation of ROM of the hip, an AP and frog-leg X-ray for hip and pelvis must be obtained. In the early stages of the disease, X-rays may not reveal any pathology (Fig. 46.1a). If any sign for ONFH is seen on X-rays, the patient must be referred to an orthopedic surgeon for further evaluation. Initial radiological findings of ONFH on X-ray are osteopenia, patchy sclerosis, and rim calcification. The major finding is the crescent sign that is indicative of subchondral fracture, the collapse of the femoral head, and the progression of the stage of the disease. In patients with normal X-ray findings, if pain persists for more than 6 weeks despite rest and NSAID use, the patient must be referred to an orthopedic surgeon. Also, patients under the aforementioned risk of ONFH must be referred to an orthopedic surgeon without any delay of more than 6 weeks. Further evaluation includes magnetic resonance imaging (MRI), computerized tomography (CT), technetium bone scans, and PET scans. MRI has a 95% sensitivity and specificity for detecting the ON. Major findings are as follows: a local area of a high-intensity signal on T2-weighted images and a low-intensity signal on T1-weighted images (Fig. 46.1 (1, 2, 3 and 4b, c)). CT scans are used for surgical planning rather than diagnosis. Although technetium bone scans and PET scans

have been reported to be sensitive for diagnoses, they are not used for initial diagnosis. They are rather used for screening the patients with risk factors, which may present ON in more than one bone (sickle cell anemia).

ONFH must be differentiated from osteoarthritis, transient osteoporosis of the hip, and subchondral insufficiency fractures of the femoral head. Imaging findings of these disorders resemble those of the early stages of ONFH. Transient osteoporosis of the hip is a syndrome of transient demineralization that causes acute hip pain and has MRI findings including bone marrow edema. In contrast to ONFH, transient osteoporosis of the hip is generally a self-limited condition that responds to symptomatic treatment and resting. In transient osteoporosis, there is a diffuse bone marrow edema extending from the femoral head to the intertrochanteric region. Osteoarthritis manifests with early joint space narrowing and subchondral cysts. Subchondral insufficiency fractures of the femoral head can be differentiated with MRI and histopathological examination. Flattening of the head and insufficiency fracture lines can radiologically be seen. The condition progresses very quickly to an advanced stage of osteoarthritis. The Japanese Investigation Committee Diagnostic Criteria for ONFH have also been reported to be a useful tool in the differential diagnosis.

46.6 Treatment

The prognosis of the disease depends on the extent of the osteonecrotic lesion, amount of depression, location of the lesion, and presence of bone marrow edema in the proximal femur, so effective management of the condition requires a thorough evaluation of these factors. The main goal of the treatment is the preservation of the congruity of the hip joint by preventing the collapse of the femoral head. This can be achieved with early diagnosis and appropriate treatment at the early stages of the disease. Thus, education of the patients who have risk factors will facilitate the early diagnosis and prevent rapid progression of the collapse of the femoral head. Treatment consists of nonsurgical and surgical modalities.

46.6.1 Nonsurgical Treatment

During the natural course of the disease, the collapse of the femoral head seems inevitable in most cases. Thus, the indication for nonsurgical modalities remains to be limited. Nonsurgical treatment modalities are applied to patients without collapse and with a small lesion at a non-weight-bearing area. These modalities consist of anticoagulants, statins, hyperbaric oxygen, bisphosphonates, vasodilators, biophysical modalities, growth factors, antioxidants, and hormones. Anticoagulant agents may be useful for early detected ONFH patients who have blood dyscrasia. Stains have also been reported to decrease the level of adipose tissue and osteonecrosis in animal models. This finding indicates that statins can be useful for the treatment of ONFH associated with corticosteroid use. However, treatment with these agents is still at the investigational level and more evidence is needed to prove their effectiveness.

46.6.2 Surgical Treatment

Surgical treatment modalities consist of head-preserving procedures and arthroplasty. Head-preserving procedures are used for Ficat stages I, II, and III in selected young patients. The major and most effective head-preserving modality is the core decompression alone or with vascular fibular autograft, nonvascular graft, tantalum rods, mesenchymal stem cells, and several other biological adjuvant agents. Trapdoor and lightbulb techniques have been defined for patients in Ficat stage I and pre-collapse stage II (Fig. 46.1 (1a–c, 2a–c)). With the trapdoor technique, curettage and grafting of the necrotic area are performed after safe dislocation of the hip. In the lightbulb technique, curettage and grafting are performed through a femoral neck window. The efficacy of these techniques after the collapse of the articular cartilage remains to be limited

despite promising results. Head rotation osteotomies can be used for Ficat stage 3 in young patients who have necrotic angles less than 200° (<15% of the femoral head is affected). The success of the procedure depends on the preoperative extent of the ON and the amount of the rotation. Reported results are contradictory, and the technique is demanding. Arthrodesis of the hip at a young age and the revision to TKA in older age are other bone-preserving options, particularly for laborers. However, due to difficulties and a high rate of complications after conversion to THA, arthrodesis in these patients is a rarely performed procedure.

THA and resurfacing arthroplasty are performed for patients with end-stage disease (Fig. 46.1 (3a–c, 4a–c)). Resurfacing arthroplasty is used to preserve the femoral head bone stock in young patients. However, the popularity of this procedure has decreased due to associated complications, early failure rates, and high rates of revision to THA. The rate of revision arthroplasties after THA for ONFH is higher when compared to primary osteoarthritis; however, better functional outcomes with long survival of THA have been reported with advanced techniques and new implants.

Take-Home Message

- Education of the high-risk patients may facilitate the early diagnosis and prevent the femoral head from a rapid progression of the collapse.
- The main goal of the treatment includes preserving the congruity of the hip joint and preventing the collapse of the femoral head and progression of the stage of the disease.
- MRI is the best tool for diagnosis in the early stages of the disease.
- Osteoarthritis, transient osteoporosis of the hip, and subchondral stress fractures of the femoral head must be differentiated from ONFH.
- The size, extent, depression, and location of the lesion must be considered for appropriate management of the disease.

Summary

ONFH remains to be a major reason for morbidity and THA in young patients. A thorough history of the patient must be obtained to detect the probable risk factors. Education of the high-risk patient may facilitate the early diagnosis and prevent femoral head collapse by limiting excessive weight-bearing or providing an early intervention chance with head-preserving procedures. MRI must be suggested for patients with persistent hip pain that lasts for 6 weeks as X-rays may not show the signs during the first 6 weeks. CT can be used to detect the subchondral fracture. Despite being the oldest one, Ficat classification system continues to be the major system in staging and aiding the treatment. Nonsurgical treatment modalities remain as ineffective because we currently have a superficial understanding of the real pathophysiology of the disease. Physiological modalities can be used to prevent the contracture of the muscles around the hip but do not affect the prevention of the progression of the femoral head collapse. Further evidence is needed to understand the proper use of the nonsurgical modalities in treatment. Although there is no consensus on a single surgical treatment algorithm, Zalavaras and Lieberman recommended to perform a head-preserving procedure for symptomatic osteonecrosis with small lesions in pre-collapse hip and large lesions in the pre-collapse hip of a young patient (Ficat I and II A), THA for large lesions in the pre-collapse hip of an older patient, and end-stage osteoarthritis (Ficat III, IV).

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. What are the most common two reasons for nontraumatic ONFH?
   (a) Corticosteroid use and alcohol abuse
   (b) Diabetes type 2
   (c) Obesity and alcohol
   (d) Mechanical stress and BMI
2. Which sign indicates the subchondral fracture and in which stage it can be seen?
   (a) Crescent sign can be seen in Ficat stage IIB

(b) Radiant sign can be seen in Ficat stage IA
(c) Crescent sign can be seen in Ficat stage IA
(d) Radiant sign can be seen in Ficat stage IIIC

3. Which radiological findings are associated with a poor prognosis?
   (a) B + C + D
   (b) Bone marrow edema
   (c) Lesions occupying more than the medial two-thirds of the weight-bearing portion of the femur
   (d) Head depression more than 2 mm
4. What is the best treatment option for Ficat stage 1 and 2 patients?
   (a) Core decompression
   (b) Hip replacement
   (c) Hip resurfacing
   (d) Arthroscopy
5. What is the indication for rotational osteotomy of the femoral head?
   (a) Young patient with a combined necrotic angle of less than 200° (<15% of the femoral head is affected)
   (b) Young patient with a combined necrotic angle of less than 100° (>15% of the femoral head is affected)
   (c) Old-aged patients with an angle of more than 200°
   (d) Old-aged patients with an angle of less than 200°

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47 Femoroacetabular Impingement and Acetabular Labral Tears, Tendinopathies, Snapping Hip, and Bursitis

Jacob G. Calcei and Marc R. Safran

Overview

The hip is a true ball-and-socket joint with the head of the femur articulating with the acetabulum of the pelvis. The depth of the acetabulum is increased by the acetabular labrum, and the surrounding musculature provides the dynamic mobility and additional stability of the hip joint. The complex anatomy of the hip is such that humans can walk upright, sit in a chair, and drop into a deep squat. The hip joint can withstand significant forces of up to six times body weight with everyday activities such as running and jumping. Patients often subject their hip joints to high forces, large ranges of motion, and compromising positions, particularly dancers and athletes as they run, jump, cut, pivot, and kick.

Hip pain can be significantly limiting to patients, serving as a common cause of lost quality-adjusted life years. There are a number of pathologies that can occur in and around the hip joint, and proper treatment starts with an accurate diagnosis, which is achieved through a thorough knowledge and understanding of the anatomy of the hip joint and surrounding structures. However, making the correct diagnosis about the hip can be challenging due to the difficulty of examining this deep structure, as well as the broad differential diagnosis of hip pain resulting from many surrounding structures and pain that may radiate to the hip joint (Table 47.1). The initial delineation begins with determining an intra-articular versus extra-articular cause of the patient's hip pain. In this chapter, we discuss the definition, epidemiology, pathogenesis, classifications, diagnosis, and treatment for each of the following pathologies of the hip joint:

- Intra-articular: femoroacetabular impingement and acetabular labral tears
- Extra-articular: tendinopathies, snapping hip, and bursitis

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UG. Longo, V. Denaro (eds.), *Textbook of Musculoskeletal Disorders*,
https://doi.org/10.1007/978-3-031-20987-1_47

Table 47.1 Common sources of hip pain in athletes

Intra-articular	Referred
• Femoroacetabular impingement • Labral tear • Chondral injury • Ligamentum teres tear • Loose body • Hip dysplasia • Hip microinstability • Hip dislocation	• Lumbar spine pathology • Radiculopathy • Pars injuries • Facet arthropathy • Disc degeneration • Disc herniation • Abdomen—gastrointestinal • Pelvis—genitourinary • Abdominal muscle strains • Hernia (inguinal)
Extra-articular	
• Iliopsoas strain and tendinopathy • Adductor strain and tendinopathy • Greater trochanteric bursitis • Gluteus medius strain and tendinopathy • External snapping hip syndrome • Internal snapping hip syndrome • Hamstring strain and tendinopathy • Piriformis syndrome • Core muscle injury/athletic pubalgia/sports hernia • Osteitis pubis • Sacroiliac joint dysfunction	

47.1 Femoroacetabular Impingement and Acetabular Labral Tears

47.1.1 Definition

FAI is a condition of the hip where aberrant anatomy of the acetabulum and femoral head-neck junction causes pathologic contact between the femoral neck and acetabular rim leading to irregular contact stresses and eventual labral damage and chondral injury. The two main forms of FAI are cam- and pincer-type deformities. Acetabular labral tears are injuries to the fibrocartilaginous seal/bumper along the outside of the hemispherical acetabulum.

47.1.2 Epidemiology

FAI and acetabular labral tears are a common finding in asymptomatic individuals in the general population. There has been an increased focus on and understanding of non-arthritic hip pathologies; thus, there has been an increase in diagnosis and treatment of FAI and acetabular labral tears in recent years. While isolated types of FAI are uncommon, there is a tendency for cam impingement to be seen more commonly in young, athletic males, while pincer impingement is more commonly seen in middle-aged, active females. However, combined types of FAI (both cam and pincer) are more frequently seen than either in isolation. Acetabular labral tears occur in the anterosuperior portion of the acetabulum as this is the most common impingement site—anterolateral femoral neck cam deformity impinging on an anterosuperior pincer deformity.

47.1.3 Etiology/Pathogenesis

A cam deformity is a bony prominence, commonly at the anterolateral femoral head-neck junction that creates an asphericity of the femoral head, causing a pathologic contact between the femur and the edge of the acetabulum. Pincer deformity is a bony overgrowth on the acetabular side creating an over-coverage of the femoral head and causing impaction with the femoral neck during extremes of motion. These deformities can be isolated, or some combination of the two pathologies leads to impingement pain and damage to the labrum, chondrolabral junction, and adjacent cartilage, with the end result being hip osteoarthritis.

While acetabular labral tears may be an end result of FAI, they can also occur in patients with dysplastic hips, hip instability, and traumatic dislocations and frequently are seen in asymptomatic individuals.

47.1.4 Classifications

There are three types of FAI pathology as mentioned above: cam, pincer, and combined. Cam and pincer deformities are identified and quantified using imaging studies, which are described below. An alpha angle of greater than 55° on radiographs and/or MRI is considered abnormal and is classified as a cam deformity. The presence of acetabular over-coverage of the femoral head on plain X-ray as seen in the crossover sign (Fig. 47.1a) and quantified by a lateral center-edge angle greater than 40° is consistent with the presence of a pincer deformity. Inherent in the name, combined FAI is some combination of cam and pincer deformities (Fig. 47.1b).

47.1.5 Diagnosis

As with any pathology around the hip, an accurate diagnosis of FAI and symptomatic labral tear can be achieved through a combination of a detailed history, thorough physical examination (Table 47.2), and select imaging. Pain within the hip joint typically manifests as anterior groin pain along the inguinal crease. However, referred pain to the thigh or even down to the knee can be a finding in patients with intra-articular hip pathology. Patients with intra-articular pain from FAI or labral tear will often form what is called the C-sign with their hand when asked to demonstrate the location of their pain, noting that the pain is deep, between the thumb and index finger forming a "C." The presence of mechanical symptoms, such as locking, clicking, or catching in the hip, is concerning for intra-articular pathology, specifically a labral tear or chondral flap. Additionally, patients with FAI and labral pathology may complain of pain with deep flexion and/or internal rotation of the hip.

During the history taking, the examiner should take note of how the patient is sitting. Patients with FAI will avoid sitting in low chairs, such as a low couch, and may not like sitting straight up in a chair to decrease hip flexion and impingement. Patients with FAI or labral tear will have pain at extremes of motion, which may manifest as a decrease in ROM as the patient avoids painful positions. Specifically, a loss of flexion and/or internal rotation is commonly seen in FAI as these are the positions of impingement. The

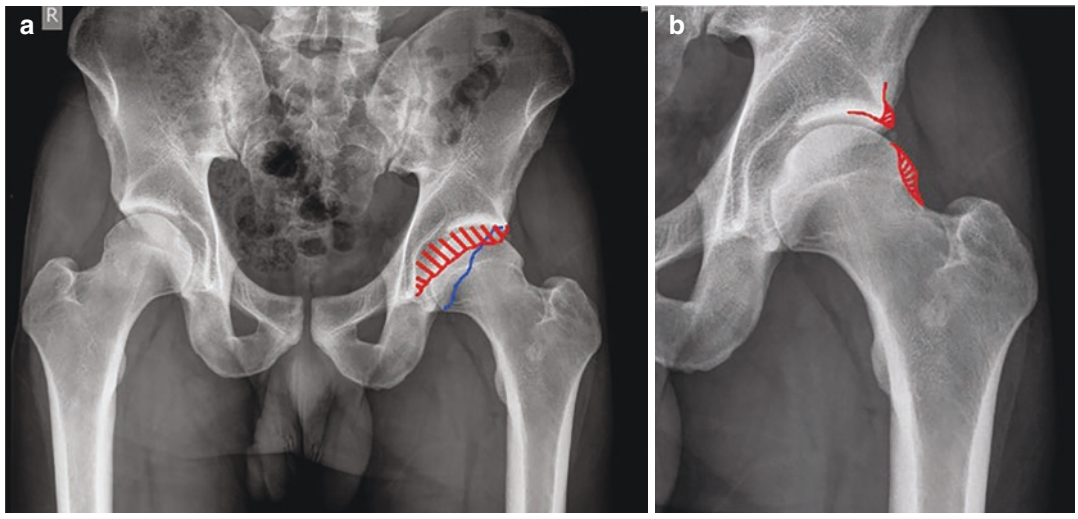

Fig. 47.1 X-ray images demonstrating combined pincer and cam deformity. The anterior-to-posterior (AP) view of the pelvis (**a**) demonstrates bilateral combined FAI with a crossover sign of the acetabulum as demonstrated by the anterior acetabular wall (red line with dashes) crossing over the posterior acetabular wall (blue line). X-ray view of the left hip (**b**) illustrating combined FAI (red) with the pincer deformity of the acetabulum and cam deformity of the femoral head and neck junction

Table 47.2 Summary of the basic hip examination in athletes organized by patient positioning

Standing examination	Supine examination
Gait Trendelenburg/abductor strength Leg length assessment	Range of motion • Flexion • Internal and external rotation • Abduction and adduction Thomas test Adductor strength Hamstring tightness Straight leg raise Scour/labral stress test FADIR/impingement test Stinchfield test McCarthy test Patrick FABER test Log roll Axial loading and foveal distraction test Hyperextension external rotation test Beighton score Palpation: • Abdomen • Pubic symphysis • Adductor and iliopsoas Hesselbach's test
Sitting examination	
Internal and external rotation Iliopsoas strength Neurovascular examination: • Pulses (DP and PT) • Sensation • Motor strength • Deep tendon reflexes	
Lateral examination	*Prone examination*
Palpation: • Trochanteric bursa • Gluteus medius muscle • Piriformis tendon Ober test Guanche instability test	Range of motion: • Internal rotation • External rotation Domb instability test Glute-hamstring dominance

DP dorsalis pedis, *PT* posterior tibialis, *FADIR* flexion adduction internal rotation, *FABER* flexion abduction external rotation

impingement test can be performed by placing the patient supine with their hip flexed to 90°, adducted, and internally rotated (FADIR)—a positive test results in pain. However, despite the test being called the impingement test, it is not pathognomonic for FAI. Patients with FAI may have obligatory abduction and external rotation with flexion, known as Drehmann's sign. The labral stress test, also known as the scour maneuver, starts with the patient supine with the hip flexed, abducted, and externally rotated and then adducted, internally rotated, and extended, stressing the labrum and causing pain in the presence of a labral tear.

Imaging is an important adjunct to the history and physical examination and should begin with an anterior-to-posterior (AP) X-ray of the pelvis. Additional X-ray views can be performed including a cross-table or frog-leg lateral, among others. The plain radiographs can help identify a cross-over sign, measure over-coverage with the lateral center-edge angle, identify a cam deformity, and measure the alpha angle. Magnetic resonance imaging (MRI) is used to identify labral tears and chondral injuries as well as further quantify the cam deformity. However, it is important to note the high prevalence of labral tear on MRI in the asymptomatic general population. Magnetic resonance arthrography uses an intra-articular injection of contrast to more clearly evaluate the labrum, and if the addition of an anesthetic with the injection resolves the patient's pain, even for a short period of time, it confirms that the source of pain is intra-articular such as FAI or labral tear.

47.1.6 Treatment

As with many musculoskeletal pathologies, the initial management consists of rest, nonsteroidal anti-inflammatory drugs (NSAIDs), and physical therapy. Patients with symptoms refractory to nonoperative management or those with mechanical symptoms of the hip are indicated for surgical intervention. Cam and pincer deformities, labral tears, and chondral injuries can be addressed with hip arthroscopy. Osteoplasty of the femoral neck and acetabulum, to remove excessive bone resulting in impingement, can address and normalize cam and pincer deformities, respectively. Labral tears can be debrided or repaired with suture anchors, and chondral flaps can be debrided and treated with marrow stimulation techniques such as microfracture.

47.2 Tendinopathies, Snapping Hip, and Bursitis

47.2.1 Definition

Tendinopathy is the term used to describe the degenerative and often inflamed state of a tendon that typically presents as pain and/or weakness of the affected muscle. Common tendinopathies about the hip include the adductors, iliopsoas, rectus femoris, gluteus medius, hamstring, and piriformis tendons, which can be separated based on the location of the patient's pain as described below. Snapping hip, also known as coxa saltans, is a snapping sensation that occurs as a muscle or tendon slides over a bony prominence around the hip. Bursitis is the irritation and inflammation of the fluid-filled bursal sac that facilitates smooth sliding of one tissue over another, such as the iliotibial band over the greater trochanter.

Extra-articular anterior groin pain is often due to adductor, iliopsoas, or rectus femoris tendinopathy. Iliopsoas tendinopathy is often accompanied by iliopsoas bursitis and can create a snapping sensation known as internal snapping hip.

Lateral hip pain is often due to a constellation of pathologies known as "greater trochanteric pain syndrome," which includes gluteus medius tendinopathy, iliotibial band tightness and snapping (external snapping hip), and trochanteric bursitis.

Posterior hip pain may come from a number of sources including the lumbar spine and sacroiliac joint; however, common tendinopathies that cause posterior hip pain include the proximal hamstrings and the piriformis tendon. Piriformis tendinopathy can lead to piriformis syndrome, which is a compression of the sciatic nerve as it runs deep to a tight piriformis tendon.

47.2.2 Epidemiology

Tendinopathies about the hip joint are quite common and can be a significant cause of time lost from work or sports. Snapping hip and bursitis are also fairly common in athletes and other patients that perform repetitive flexion and extension motions of the hip. Tendinopathies about the hip commonly occur in athletes, particularly runners, cyclists, hockey, and soccer players. Adductor strains are the most common cause of anterior hip and groin pain, also known as a groin pull.

Internal snapping hip is more common than external snapping hip, although both types of snapping occur commonly in young athletes, especially runners and dancers. Associated bursitis can occur in conjunction with snapping hip or in isolation in patients of all ages.

47.2.3 Etiology/Pathogenesis

Tendinopathies occur either as a result of chronic overuse or acute injury. In the case of chronic overuse, the tendon undergoes repetitive stress, resulting in microinjuries to the collagen fibers that weaken the tendon and cause inflammation and pain. These chronic overuse injuries make the tendon more susceptible to acute injury. Acute injury occurs as the tendon is rapidly stretched by a force vector that pulls in the opposite direction to that of the tendon. Tendinopathies about the hip occur in repetitive endurance sports such as running and cycling as well as high-impact sports that require cutting, pivoting, jumping, and kicking.

Internal snapping occurs as the iliopsoas tendon slides over the femoral head, the iliopectineal ridge, or an exostosis present at the lesser trochanter. External snapping hip is associated with iliotibial band tightness, and the snapping occurs as the iliotibial band passes over the greater trochanter (Fig. 47.2). In both locations, a bursitis can develop either with the snapping or in isolation, as the bursal sacs associated with the iliopsoas (iliopsoas bursitis) or the iliotibial band (trochanteric bursitis) become inflamed and irritated. As with tendinopathy, bursitis also occurs in patients that perform activities that require frequent, repetitive motions of the hip joint.

47.2.4 Classifications

There are various classification systems for tendinopathy in general. An early classification from Blazina et al. in 1973 described four stages of

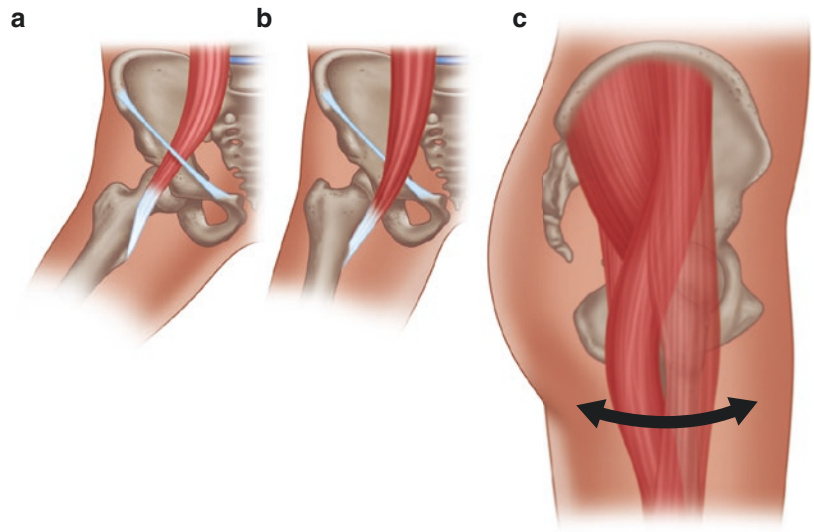

Fig. 47.2 Illustration of internal (**a** and **b**) and external (**c**) snapping hip. Internal snapping hip involves the iliopsoas tendon snapping anteriorly over the femoral head and iliopectineal ridge. With the hip in flexion, abduction, and external rotation, the iliopsoas tendon lies laterally (**a**), and as the hip moves into extension, adduction, and internal rotation, the tendon snaps medially (**b**). External snapping hip involves the iliotibial band snapping over the lateral aspect of the greater trochanter as the hip moves from flexion to extension

tendinopathy: (1) pain after sports activity; (2) pain at the beginning of sports activity that disappears with warm-up and reappears with fatigue; (3) pain at rest and with activity; and (4) tendon rupture. More recently, Nirschl et al. created a classification system for tendinopathy based on tennis elbow. Their classification system has four stages: (1) temporary irritation; (2) permanent tendinosis involving less than 50% of the tendon; (3) permanent tendinosis involving greater than 50% of the tendon; and (4) partial or total rupture of tendon, and seven phases ranging from mild pain after exercise that lasts less than 24 h to constant pain at rest that disturbs sleep.

As mentioned above, snapping hip can be categorized as either internal or external snapping hip determined by the location of the snapping and the tendons involved. Internal snapping hip involves the iliopsoas tendon snapping anteriorly in the groin, while external snapping hip describes the iliotibial band snapping laterally.

Bursitis is typically separated into two types: septic versus aseptic. Aseptic bursitis describes bursal inflammation without infection, while septic bursitis describes an inflamed bursa with a concomitant.

47.2.5 Diagnosis

Patients will often complain of pain and possibly weakness when suffering from a tendinopathy. In tendinopathies about the hip, the location of pain is important. Patients with anterior groin pain may have adductor, iliopsoas, or rectus femoris tendinopathy; lateral hip pain may have gluteus medius tendinopathy; and posterior hip pain may have hamstring or piriformis tendinopathy. Patients with piriformis syndrome will complain of deep posterolateral hip or buttock pain. As mentioned above, it is important to delineate between intra-articular and extra-articular hip pathology. Tendinopathies are categorized as extra-articular pathologies, so passive motions of the hip will typically not cause pain, while active, resisted motions and palpation of the irritated tendon will cause pain. Patients will often describe a change or increase in the amount or type of activity or a specific sudden injury that triggered the pain.

On physical examination, patients with tendinopathy and bursitis will likely have tenderness to palpation along the involved tendon and bursa,

and thus a good understanding of the anatomy of the hip and the surrounding structures is important.

Adductor tendons can be palpated proximally at their origin along the ischium and pubis of the pelvis. Adductor strength can be assessed via resisted hip adduction with the patient sitting at the edge of the examination table or supine with the hips and knees flexed so the feet are flat on the table. Muscle strength is graded on a 0 to 5 scale: 5 is full strength; 4 is decrease from full but able to resist examiner; 3 is movement against gravity; 2 is movement, but not against gravity; 1 is muscle firing but no movement; and 0 is no muscle contraction.

The rectus femoris tendon can be palpated proximally at its origin on the anterior inferior iliac spine. The examiner can stress the patient's rectus femoris by asking them to perform a straight leg raise while lying supine and palpating the rectus femoris tendon.

The iliopsoas tendon is more difficult to palpate due to its deep nature. The patient should externally rotate and flex their hip about 30°. In this position, the sartorius tendon is the most superficial anterior hip muscle in this position, and the iliopsoas can be palpated medial to the sartorius and distal to the inguinal crease. The examiner palpates the iliopsoas, and the patient brings their extremity back to neutral and then performs a straight leg raise of 30°. Pain with this maneuver is consistent with iliopsoas tendinopathy or bursitis. Iliopsoas strength can be tested with the patient sitting on the edge of the examination table and flexing their hip against resistance. Internal snapping hip can be elicited by several active maneuvers, one of which starts with the patient supine with their hip flexed, abducted, and externally rotated and then returning their hip to neutral by extending, internally rotating, and adducting the hip reproducing an audible snap or clunk (Fig. 47.2a and b).

Patients with gluteus medius tendinopathy will have tenderness to palpation at the proximal aspect of the greater trochanter where the gluteus medius inserts on the femur, while tenderness 2 or more centimeters proximal to its insertion may be consistent with gluteus medius syndrome. This is best assessed with the patient lying lateral on the examination table with the injured side up. Tenderness of the greater trochanter is consistent with a trochanteric bursitis. Iliotibial band tightness can be assessed using the Ober test. Weakness of the gluteus medius can result in a positive Trendelenburg sign. In this test, the patient is standing and asked to lift the contralateral leg off of the ground; if the pelvis drops or if the patient shifts their trunk towards the affected side, the test is positive, signaling abductor weakness. External snapping hip can typically be seen and palpated along the lateral hip as the patient flexes and extends their hip (Fig. 47.2c) while walking across the examination room or lying on their side and moving the leg that is away from the examination table in a bicycle motion.

The hamstring origin at the ischial tuberosity is best palpated with the patient in the prone position. Additionally, hamstring muscle strength can be tested by instructing the patient to flex their knee against resistance. If the patient has a complete rupture of the proximal hamstrings, a palpable gap may be present.

The piriformis can be palpated between the ischial tuberosity and greater trochanter with the patient in the lateral position with their hip flexed and adducted so that their knee is resting on the examination table. Additionally, piriformis strength can be tested by starting in this flexed and adducted position, while the patient lifts their knee off the table against resistance. Pain with resisted strength testing is often found with piriformis syndrome.

Patients being evaluated for hip pain should undergo plain X-ray evaluation of their pelvis as described above. While the tendons and bursae cannot be seen on plain X-ray, there may be ossifications consistent with osteophytes or enthesophytes at the tendinous attachments to bone and other bony causes of hip pain can be ruled out.

Other imaging studies that are beneficial include ultrasound, which can help to show tendon quality, bursal inflammation, or, by using dynamic ultrasound when snapping hip is suspected, iliopsoas or iliotibial band snapping. MRI can be used to demonstrate if there is fluid and inflammation surrounding the involved tendon and quantify the extent of tendon degeneration and tearing.

47.2.6 Treatment

Similar to FAI and labral tears, the initial management of tendinopathies, snapping hip, and bursitis consists of rest, NSAIDs, and physical therapy. Physical therapy should focus on stretching and strengthening of the affected muscle and establishing a balance in the antagonist muscle pairs. Injections can be particularly effective for tendinopathy and bursitis. Ultrasound-guided corticosteroid injections to the sheath of the affected tendon or into the inflamed bursa can provide the patient pain relief and allow them to complete physical therapy.

Patients with symptoms refractory to nonoperative management may be indicated for surgical intervention. For tendinopathies refractory to nonoperative management, specifically piriformis and iliopsoas tendinopathy, or internal or external snapping hip, surgical debridement and release or lengthening of the tendon can be performed. Partial tendon tears refractory to nonoperative management or complete tears with retraction are indicated for surgical repair. Patients with bursitis that fail nonoperative management can be indicated for arthroscopic versus open debridement of the inflamed bursa.

Take-Home Message

- Hip pathologies can be a major source of pain causing time lost from work and sports.
- Accurate diagnosis and appropriate treatment rely on a detailed understanding of the hip joint and surrounding anatomy supplemented by a thorough history, physical examination, and focused imaging studies.
- Delineation between intra-articular (FAI and labral tears) and extra-articular pathologies (tendinopathies, snapping hip, and bursitis) is the first step when evaluating a patient with hip pain.
- Nonoperative treatment with rest, activity modification, NSAIDs, physical therapy, and selective use of injections can be effective in the treatment of FAI, acetabular labral tears, tendinopathies, snapping hip, and bursitis.

Summary

Hip pain can be caused by a number of pathologies, both intra-articular and extra-articular. Proper diagnosis requires a detailed understanding of hip anatomy followed by a thorough history, physical examination, and select imaging studies. Treatment for most hip pathologies begins with activity modification, anti-inflammatory medications and modalities, physical therapy, and select injections, while surgical intervention may be indicated for cases refractory to nonoperative management or those with mechanical symptoms or a complete tendon rupture with retraction.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. What is the most common type of FAI?
   (a) Cam
   (b) Pincer
   (c) Combined
   (d) They are equal
2. Internal snapping hip is associated with this tendon:
   (a) Piriformis
   (b) Iliopsoas
   (c) Iliotibial band
   (d) Adductor

3. External snapping hip is associated with this tendon:
   (a) Piriformis
   (b) Iliopsoas
   (c) Iliotibial band
   (d) Adductor
4. Hip joint pain is:
   (a) Characterized by tenderness to palpation
   (b) Often associated with limping
   (c) Never requires surgical treatment
   (d) A disease that requires immediate MRI assessment
5. Hip tendinopaty:
   (a) Hurts with resisted muscolar contraction
   (b) Is ever associated with ecchimosis
   (c) Requires immediate ultrasound assessment
   (d) Requires immediate MRI assessment

Further Reading

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Hip Fractures

48

Mark E. Cinque and Marc R. Safran

Overview

Hip fractures represent a significant public health challenge worldwide and are projected to triple in number in the next 50 years.

48.1 Definitions

Hip fractures represent a significant public health challenge worldwide and are projected to triple in number in the next 50 years. Hip fractures can be broadly categorized into two categories based on the location of the fracture. Hip fractures are described as either being a femoral neck fracture (FNF) or an intertrochanteric (IT) femur fracture. An alternative description is whether the fracture is intracapsular (which by anatomy are generally femoral neck fractures) or extracapsular (intertrochanteric femur) fractures. Identifying and understanding the differences between these fracture patterns are critical, as surgical decision-making begins based on the anatomy of the fracture and its ability to heal without developing avascular necrosis as a complication. Intracapsular fractures are most commonly treatment with open reduction and internal fixation (ORIF) or arthroplasty, depending on the age and functional status of the patient. On the contrary, extracapsular fractures are most commonly treated with open or closed reduction with intramedullary or extramedullary fixation. The aim of this chapter is to describe the background, diagnosis, and treatment strategies for both femoral neck fractures and intertrochanteric femur fractures.

48.2 Epidemiology

Nearly 30 years ago, the international osteoporosis foundation (IOF) estimated that 18% of women and 6% of men will be affected by hip fractures. However, more recent data projects the prevalence of hip fractures to be 4.5 million by 2050. One-year mortality following hip fractures has been reported to be between 14% and 36%. With such a high mortality rate and a projected sharp increase in hip fracture occurrence, hip fractures pose a true global public health crisis. Patient gender has emerged as an important factor in hip fractures, as 1/3 of women living to age 80 will sustain a hip fracture, compared to 15% of their male counterparts. Patient race has also been shown to influence the epidemiology of hip fractures. Moreover, white women over the age of 50 years of age have nearly doubled risk of hip fracture compared to men.

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48.3 Etiology/Pathogenesis

Femoral neck fractures occur in a bimodal age distribution: high-energy mechanisms often result in younger patients sustaining femoral neck fractures, whereas low energy, often ground-level falls, generates femoral neck fractures in elderly patients. Intertrochanteric femur fractures occur in a similar bimodal distribution based on injury energy and patient age.

48.3.1 Femoral Neck Fractures

Femoral neck fractures most commonly occur in elderly patients, which has been attributed to the relatively lower density bone stock seen in older patients. Studies have suggested that femoral neck fractures are actually a pathologic fracture through an area of preexisting osteomalacia. However, more recent studies have elucidated the relationship between preexisting osteoporosis and femoral neck fractures, leading many to classify these fractures as "fragility fractures." Approximately 90% of femoral neck fractures in the elderly occur after a mechanical fall onto the lateral hip. The force of the fall is transmitted to the femoral neck through the greater trochanter. A second mechanism of femoral neck fracture in elderly patients is one that occurs through twisting through a fixed foot causing a rotational force to be transmitted resulting in a fracture. Posterior femoral neck comminution is commonly seen in the twisting pattern of injury. The most common location of fracture in femoral neck fractures is at the weakest segment of the femoral neck, located just below the articular surface. In the younger patient population, femoral neck fractures often occur after high-energy traumatic events, such as motor vehicle accidents or falls from height. Contrary to geriatric femoral neck fractures, younger patients with femoral neck fractures tend to be male.

48.3.2 Intertrochanteric Femur Fractures

Intertrochanteric femur fractures also tend to occur in a bimodal age distribution: young patients sustain these injuries after high-energy trauma, while elderly patients sustain these fractures from simple falls. Biomechanical studies have shown that a ground-level fall generates at least 16 times the energy necessary to fracture the proximal femur. However, not all mechanical falls lead to intertrochanteric fractures, as only 5–10% of falls in older white women result in fracture, indicating that the mechanics of the fall are an important factor in the risk for fracture. The most predictable fall pattern for sustaining an intertrochanteric fracture is a fall directly onto or near the lateral aspect of the hip, at the level of the greater trochanter.

48.4 Classification

48.4.1 Femoral Neck Fractures

Several classifications have been created to describe femoral neck fractures. The first classification system describes the anatomic region of the femoral neck where the fracture occurred: subcapital, transcervical, or basicervical (Fig. 48.1). The most commonly used classification systems that impact surgical decision-making include the Garden and the Pauwels classification systems. The Garden classification system is based on the degree of displacement of the femoral neck, as judged on the anterior-posterior (AP) radiograph. Garden I

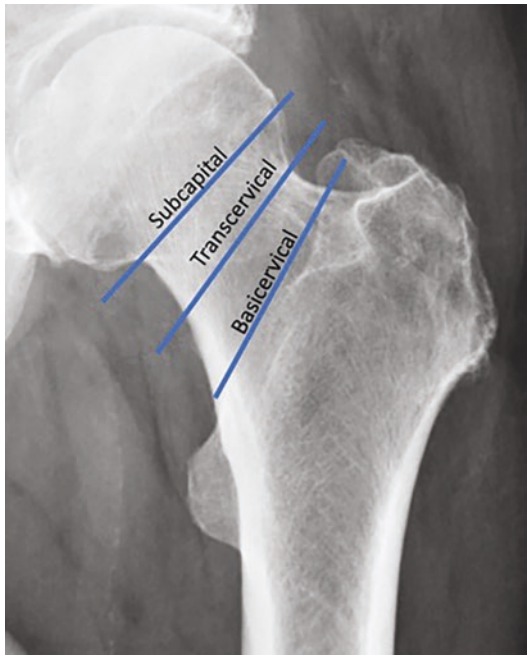

Fig. 48.1 AP radiograph of a left hip demonstrating the three anatomic zones of femoral neck fracture

represents a valgus impacted, subcapital femoral neck fracture; Garden II represents a completed but nondisplaced fracture; Garden III represents an incompletely displaced fracture; and Grade IV represents a completely displaced fracture (Fig. 48.2a). This classification confers some advantage by allowing the surgeon to communicate the degree of displacement and the risk of disruption of the blood supply to the femoral head resulting in avascular necrosis. However, the inter- and intra-rater reliability have been demonstrated to be relatively low, with one study demonstrating only 22% agreement across all fracture types. The second classification system used was described by Pauwels. This classification system is based on the angle of the fracture plane: vertical, oblique, or transverse (Fig. 48.2b). The advantage of the Pauwels classification is that it lends itself to making surgical decisions. With increasing angle of fracture, nonunion and fixation failure are more likely. Type 1 fractures are 30° or less (thus more horizontal fracture line), type II fractures are between 30 and 50° and type III fractures are greater than 50° (more vertical fracture line). However, similar to the Garden Classification, the Pauwels classification has been shown to have a poor inter-rater agreement of 0.31.

48.4.2 Intertrochanteric Femur Fractures

The classification system of intertrochanteric fractures is dictated by the underlying stability of the fracture. The anatomic factors that dictate fracture stability include the fracture obliquity, lateral femoral wall integrity, and posteromedial cortex. Stable fracture patterns have minimal posteromedial comminution and a standard obliquity fracture line. Standard fracture obliquity refers to a fracture line that extends from superolateral to inferomedial along the intertrochanteric line (Fig. 48.3a). Unstable fractures are characterized by significant posteromedial comminution, incompetent lateral wall, or a reverse obliquity fracture line. A reverse obliquity fracture line extends from superomedial to inferolateral (Fig. 48.3b). The integrity of the lateral wall is an important consideration because it can dictate the type of implant that can be utilized for fixation. For example, a lateral buttress device like a sliding hip screw (SHS) cannot be utilized in fracture patterns without an intact lateral wall. Similarly, reverse obliquity patterns are not amenable to SHS constructs as the compression of the lag screw within the barrel will cause displacement of the fracture. Finally, posteromedial comminution is an important consideration as it gives a clue about the degree of fracture instability and the potential need for a percutaneous or open reduction prior to implant placement.

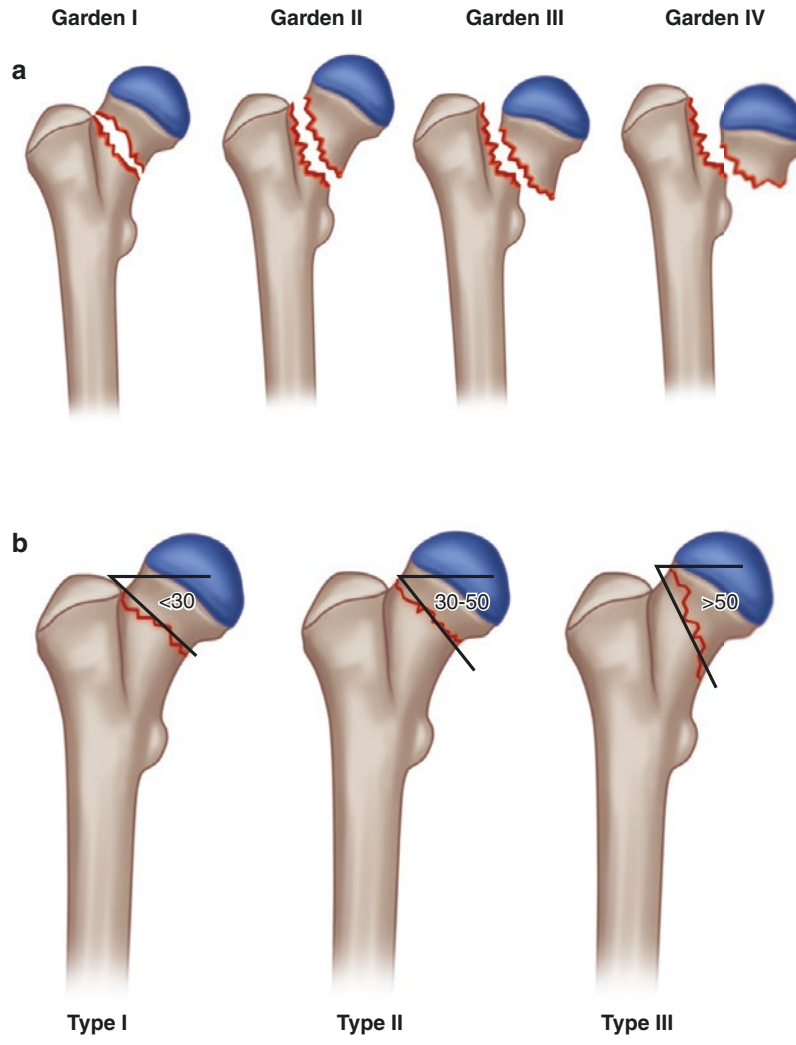

Fig. 48.2 (**a**) Schematic representation of Garden classification of femoral neck fracture. There is increased fracture instability from type I to type IV. (**b**) Schematic demonstrating Pauwels classification; increasing Pauwels angle indicates greater fracture instability and guides implant selection

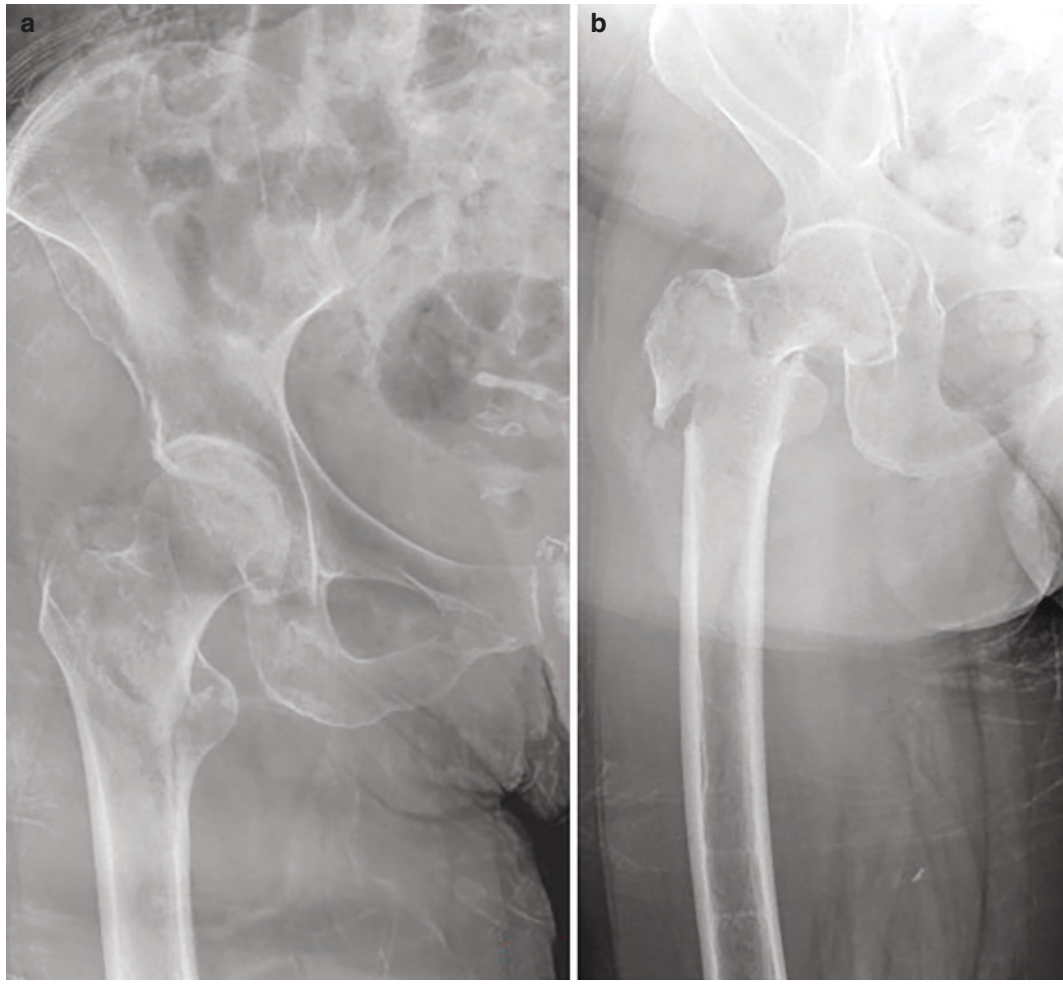

Fig. 48.3 (**a**) AP radiograph of a right hip demonstrating a standard obliquity intertrochanteric femur fracture. The primary fracture traverses superolateral to inferomedial. (**b**) AP radiograph of a right hip demonstrating a reverse obliquity fracture pattern. The primary fracture line begins superomedial and exits inferolateral

48.5 Diagnosis

The workup of both femoral neck fractures and intertrochanteric fractures begins with the physical examination. For high-energy mechanisms, the patient should first be evaluated using the Advanced Trauma and Life Support System (ATLS). Following a multisystem trauma evaluation, the patient should be examined for signs of open or occult fracture. If a hip fracture is suspected, the patient's lower extremities should be examined for flexion, shortening, and external rotation, the usual positioning of the lower extremity due to muscle pull on the displaced hip fracture. Logroll or range of motion of the affected extremity should be avoided as this causes unnecessary pain, may further displace the fracture, and cause damage to surrounding soft tissues.

Following patient stabilization and primary orthopedic survey, radiographic evaluation should commence. This begins with an AP pelvis radiograph and AP hip and cross-table lateral hip radiographs. In some fracture patterns, it can be challenging to determine if a patient has sustained a basicervical femoral neck fracture or an

intertrochanteric femur fracture. In this situation, the surgeon can perform a traction-internal rotation AP view of the affected hip. This is performed by pulling axial traction on the affected limb and internally rotating 10–15°. This view can help match the X-ray beam with the native femoral anteversion and allow for accurate fracture classification.

Advanced imaging, such as computed tomography (CT) and magnetic resonance imaging (MRI), is rarely utilized to evaluate for hip fractures. While CT scan can confirm a femoral neck fracture if there is clinical suspicion for a nondisplaced fracture, it is not the best imaging modality. Similarly, MRI can be used to diagnose femoral stress fractures but has less utility in the trauma setting given the level of soft-tissue edema associated with trauma, which detracts from the utility of the study.

48.6 Treatment

Hip fractures are painful, and there is known significant morbidity associated with prolonged bedrest (with or without traction) in nonoperatively treated hip fracture patients. The risks of prolonged bed rest and lack of mobility while the fracture is healing include atelectasis, pneumonia, and deep venous thrombosis with pulmonary embolism, in addition to bedsores. Thus, the general philosophy of the management of hip fractures is to make the patient comfortable to allow mobilization, at least sitting up, to reduce the pulmonary risks. An additional consideration for the management of hip fractures includes the risk of avascular necrosis due to the disrupted blood supply to the femoral head, which can lead to collapse of the femoral head and arthritis, and the risk of the fracture not healing. Avascular necrosis more frequently occurs with femoral neck fractures (as compared to intertrochanteric fractures), and the risk of AVN increases with displacement of the fracture.

After making the diagnosis of a hip fracture, a thorough review of the patient's medical history, perioperative risk factors, baseline functional status, and preoperative hip pain status are critical. These factors are crucial components to surgical decision-making for any patient with a hip fracture. In general, for hip fractures in the elderly, it is felt that these patients lose one level of ambulation status after they break their hip—for example, a person who walks in the community with a cane, but no cane at home, will use a cane at home after and may not walk much in the community without a walker.

48.6.1 Nondisplaced Femoral Neck Fractures

There still exists some disagreement with regard to the optimal management of nondisplaced femoral neck fractures. Fractures that are valgus impacted and do not exhibit apex anterior alignment (where the fracture apex is pointing anteriorly) on the lateral hip radiograph may merit a trial of nonoperative management. While there are no level I or II studies comparing operative and nonoperative management of nondisplaced femoral neck fractures, one study reported an 86% union rate in a study of 170 consecutive valgus-impacted femoral neck fractures treated with early weight-bearing. This study went on to highlight that the patients that were most likely to have subsequent fracture displacement were patients >70 years of age and those in poor general health. But these patients need to have minimal pain to be able to weight-bear early. If not, then internal fixation may provide enough stability to control the pain for adequate mobilization. Cost-benefit research has demonstrated a 30% 1-year mortality rate for subcapital femoral neck fractures treated conservatively, highlighting that while many of these fractures may go onto union, these patients are still subject to significant morbidity and mortality.

48.6.2 Displaced Femoral Neck Fracture

Patients with displaced femoral neck fractures are likely to have disruption of their retinacular vasculature, resulting in a poor fracture-healing

environment. Furthermore, disruption of the femoral head blood supply can lead to avascular necrosis of the femoral head. Surgical treatment is the primary option for patients without absolute contraindications to surgery. There are a range of treatment modalities ranging across closed reduction with internal fixation, open reduction and internal fixation, hemiarthroplasty, and total hip arthroplasty (THA).

The type of surgical approach used is primarily dictated by the patient's "physiologic age." Younger patients with displaced femoral neck fractures would undergo expedited and open approach with an anatomic reduction with internal fixation to optimize the chances of fracture healing. Older patients are more likely to be indicated for a hemiarthroplasty or THA, as it leads to early stabilization and early weight-bearing, obviating the concern of developing avascular necrosis.

There are two main options for internal fixation implants for these fractures: cannulated screws and sliding hip screw (SHS) construct. Closed reduction and treatment with cannulated screws involve the surgeon using manual traction to reduce the fracture with the aid of intraoperative fluoroscopy. Following fracture reduction, cannulated screws are inserted over Kirschner wires placed into the head-neck segment of the femur (Fig. 48.4a). The screws are most often inserted in an inverted triangle pattern, with emphasis placed on engaging the inferomedial femoral neck cortical bone with the threads of cannulated screws. Care must be taken not to penetrate the femoral neck cortex or the femoral head articular surface. The SHS construct consists of a lateral femoral plate with a cylindrical barrel that allows fracture collapse (allowing compression to stimulate healing) during weight-bearing. In order for the SHS to function in a biomechanically advantageous way, the patient must have an intact lateral femoral cortex. Furthermore, SHS constructs should not be used in basicervical femoral neck fractures as collapse often leads to displacement across the fracture site.

Studies comparing cannulated screws and SHS have not demonstrated a significant difference with regard to union or mortality between the two groups. In a meta-analysis of nearly 5000 patients, Parker and Blundell demonstrate no superiority of either device with regard to fracture union or displacement. However, a more recent growing body of evidence demonstrated fracture displacement, fixation failure, and imperfect reduction following treatment with cannulated screws. Taken together, the best available evidence does not demonstrate superiority of one technique; however, fractures with greater displacement and patients with higher demand may require a stouter implant and thus require treatment with an SHS construct.

If open reduction and internal fixation of the fracture is not selected, the next determination that needs to be made involves evaluating patient factors. The patient's physiologic age and activity level must be assessed, along with preexisting hip pain. Patients with relatively great physiologic age, low demand status, and perioperative risk factors are often indicated for a hemiarthroplasty. A hemiarthroplasty involves replacing the femoral head with a metallic ball, which is connected to a metallic body that is either press-fit or cemented into a prepared femoral canal (Fig. 48.4b). A hemiarthroplasty leaves the native acetabulum intact. There have been a series of studies comparing hemiarthroplasty to internal fixation, with hemiarthroplasty being shown to have fewer significant problems like fracture nonunion and revision (need for further surgery) with modestly increased operative times and blood loss. These are important risks to consider when treating older, higher risk patients when the surgeon is trying to minimize surgical risk (including reoperation) to the patient. One limitation with the existing body of evidence is that there is no age stratification made between the ORIF and hemiarthroplasty groups. This limitation reinforces that the surgeon must evaluate all patient factors on a case-by-case basis.

Total hip arthroplasty involves resurfacing of both the femoral head in a similar fashion as a hemiarthroplasty but with concomitant resurfacing of the acetabulum. Indications for THA following femoral neck fracture include high demand, low physiologic age patients, rheumatoid arthritis, and preexisting osteoarthritis of the

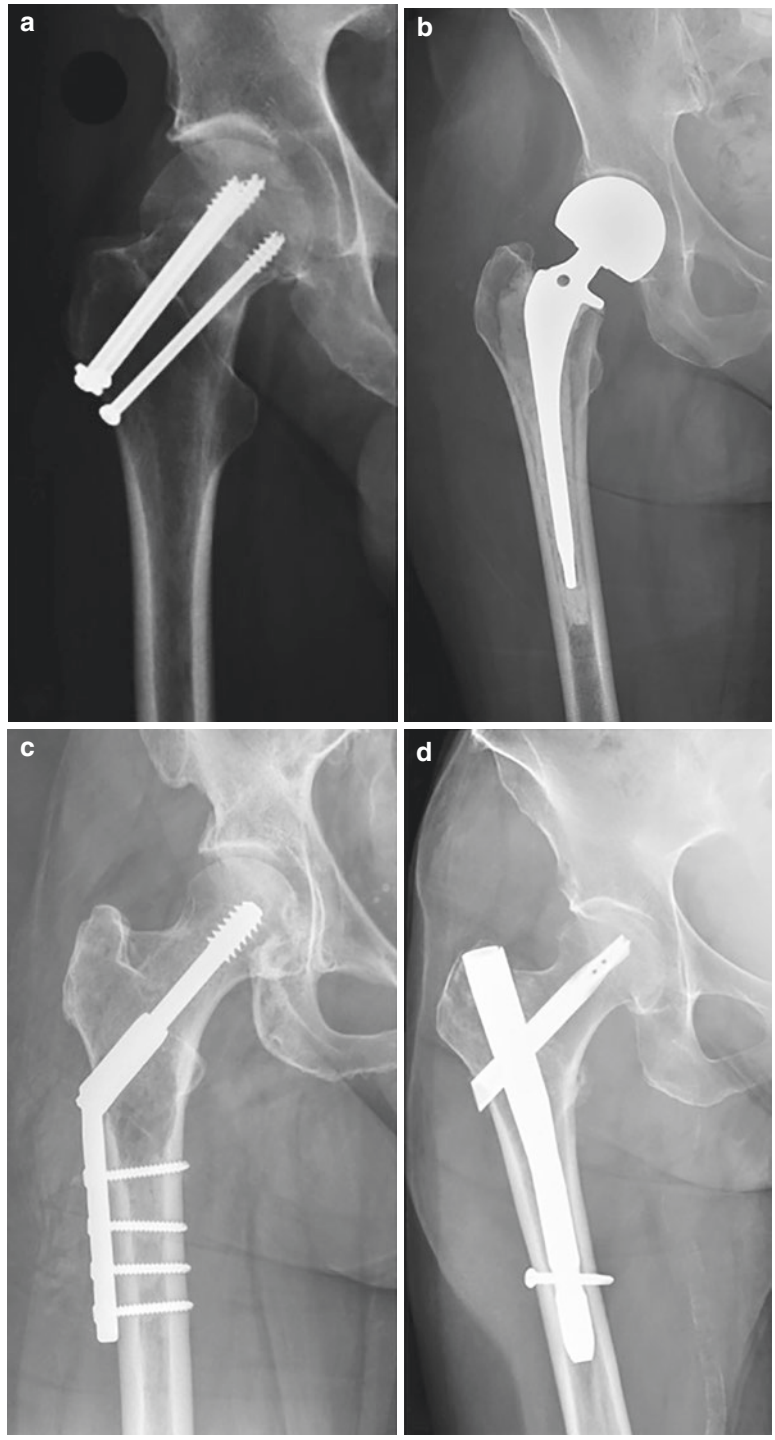

Fig. 48.4 (**a**) AP radiograph of right hip demonstrating three cannulated screws for treatment of a femoral neck fracture; (**b**) AP radiograph of right hip demonstrating a cemented, unipolar hemiarthroplasty; (**c**) AP radiograph of a right hip demonstrating sliding hip screw (extramedullary) treatment of an intertrochanteric femur fracture; (**d**) AP radiograph of a right hip demonstrating short cephalomedullary nail fixation (intramedullary) of an intertrochanteric femur fracture

affected hip. Current evidence demonstrates that THA has better outcomes than internal fixation when evaluated by functional hip scores and need for revision surgery. THA should be strongly considered in physiologically young patients with significantly displaced fractures.

48.6.3 Intertrochanteric Femur Fractures

Intertrochanteric fractures are inherently unstable fractures and are associated with significant morbidity and mortality. As a result, there are very narrow indications for nonoperative treatment of these fractures. The risks stemming from nonoperative treatment are not directly related to the broken bone but rather the sequelae from prolonged immobility, as noted above. Development of pneumonia, deep vein thrombosis, pulmonary embolus, and pressure sores have all been described following prolonged immobilization after intertrochanteric fractures. Jain et al. performed a retrospective review to compare mortality rates in intertrochanteric fractures treated both nonoperatively and operatively and found that the 30-day mortality was lower in patients treated surgically. At present, the only two considerations for nonoperative treatment for these fractures are 1) elderly patients whose comorbidities pose an extreme risk of mortality from anesthesia and 2) nonambulatory patients who have minimal discomfort following fracture.

Operative treatment allows for early mobilization and decreases the morbidity associated with prolonged immobilization. The mechanical and anatomic environment of the pertrochanteric region of the femur, as compared to the femoral neck region, has a decreased risk of osteonecrosis. Therefore, the need for prosthetic replacement is greatly reduced. Given this, there are two main categories of treatment for intertrochanteric fractures: extramedullary and intramedullary fixation. Prior to fixation, a reduction must be performed. This can be performed in a closed, percutaneous, or open fashion. The reduction approach is made based on the fracture pattern, stability of the fracture, and patient-related factors (age, comorbidities, demand level).

Intertrochanteric femur fractures are most commonly treated with either an SHS construct (extramedullary) or a cephalomedullary (intramedullary) nail (Fig. 48.4). Implant choice is driven by the fracture pattern and stability. Fractures with an intact lateral femoral cortex and a minimal amount of displacement may be treated with an SHS construct. However, unstable fractures, or fractures with an incompetent lateral femoral cortex, must be treated with an intramedullary device. Without an intact lateral wall, the SHS construct does not have a pillar of bone to provide a lateral buttress effect. Furthermore, in reverse obliquity patterns, the collapse allowed by SHS constructs causes sheering across the fracture site (as opposed to compression), and thus does not provide a good fracture healing environment. Intramedullary devices are load-sharing devices in that they share the force transmitted through the femur when the patient places weight on the operative extremity, which allows patients to partake in early weight-bearing and rehabilitation. The intramedullary component of the device allows for buttressing against fracture collapse, which is crucial in unstable or reverse obliquity fracture patterns. IM devices also allow for percutaneous entry, which decreases the amount of surgical trauma the patient is exposed to. The majority of level I evidence has not demonstrated a significant difference in blood loss, operating room time, wound complication, length of stay, mobility, reduction maintenance, or mortality when comparing SHS to IM fixation. However, there is a growing body of evidence that reports earlier return to preoperative ambulation, operating room time, and blood loss in patients with unstable fractures.

Take-Home Message

- Hip fractures represent a significant public health challenge worldwide and are projected to triple in number in the next 50 years.
- Femoral neck fractures occur in a bimodal age distribution: high-energy mechanisms often result in younger patients sustaining femoral neck fractures, whereas low energy, often ground-level falls, generates femoral neck fractures in elderly patients. Intertrochanteric femur fractures occur in a similar bimodal distribution based on injury energy and age.
- Stable, nondisplaced femoral neck fractures are typically treated with open reduction and internal fixation using a sliding hip screw construct, whereas displaced fractures are more commonly treated with arthroplasty.
- Intertrochanteric femur fractures are most often treated with cephalomedullary nails, although certain fracture patterns may be amenable to extramedullary fixation.
- Regardless of fracture pattern, the goal of treatment is to allow for early rehabilitation to decrease the morbidity and mortality associated with prolonged immobilization.

Summary

Hip fractures are a very frequently encountered orthopedic injury, one that is a large source of morbidity and mortality for patients. The best evidence available indicates that the frequency of hip fractures and the costs associated with treatment are going to continue to rise. Taken together, these indicate a strong need for all physicians to have a strong understanding of hip fractures. The two most common types of hip fractures are femoral neck and intertrochanteric femur fractures. The former may be treated with internal fixation or arthroplasty and, in rare cases, nonoperative treatment. The latter may be treated with intramedullary or extramedullary fixation, depending on the fracture pattern. When performing internal fixation, the quality of reduction prior to implant placement is of upmost important. The overarching goal of the treatment of hip fractures is to allow for early mobilization and prevent patients from suffering the deleterious effects of prolonged immobility.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. By what multiple are hip fractures projected to increase by over the next 50 years?
   (a) 3×
   (b) 2×
   (c) 10×
   (d) An increase in hip fractures not expected
2. Which fractures are the least stable in the Garden and Pauwels classification systems?
   (a) Garden IV, Pauwels III
   (b) Garden I, Pauwels III
   (c) Garden IV, Pauwels I
   (d) Garden II, Pauwels I
3. Why is a femoral neck fracture in a young person a surgical emergency?
   (a) Disruption of the blood flow to the femoral head occurs with high-energy trauma in young patients with femoral neck fractures
   (b) Because there is a high risk of DVT
   (c) Because there is a high risk of pulmonary fatty embolism
   (d) Because belated surgical treatment could lead to poor bone healing with low function
4. What is the optimal treatment for a displaced femoral neck fracture in an 89-year-old female with extensive cardiac history?
   (a) Hemiarthroplasty
   (b) Total hip replacement
   (c) Hip resurfacing
   (d) External fixation
5. What fracture characteristics would lead a surgeon to select a cephalomedullary nail over a sliding hip screw for the treatment of an intertrochanteric fracture?

(a) Intramedullary fixation is indicated in intertrochanteric fractures with an incompetent lateral wall or reverse obliquity fracture patterns.
(b) Intramedullary fixation is indicated in intertrochanteric fractures with an incompetent medial wall.
(c) Intramedullary fixation is ever indicated in intertrochanteric fracture.
(d) Intramedullary fixation is never indicated in intertrochanteric fracture.

Further Reading

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Fractures of the Femoral Diaphysis

49

Bilgehan Tosun

Overview

Femoral diaphyseal fractures occur in the middle third of the femur. Femoral diaphyseal fractures can be the result of either high- or low-energy injury. The nature of the injury, character of fracture pattern, and presence of associated injuries are the factors that influence the treatment methods. Comminuted fractures are mostly seen after high-energy mechanism. Complications such as malalignment and limb shortening are seen after the treatment of comminuted fractures. Once the life-threatening injuries are dealt with, the focus transforms into the treatment of the fractures by avoiding the potential complications that may occur after the fracture treatment.

49.1 Definition

From the lesser trochanter to 7 cm proximal to the knee joint is called femoral diaphysis. Diaphysis begins at the top of the femoral isthmus and extends to the distal metaphyseal junction. Fractures of the femoral diaphysis are mostly resulted from high-energy trauma. There is also increasing incidence of femoral diaphyseal fracture in low-energy injuries in the elderly. As these injuries can be a part of multiple trauma, physician should not underestimate the amount of blood loss and adequate fluid resuscitation is essential. Since the fractures are mostly the result of high-energy trauma, associated injuries are common. Ipsilateral fractures of the femoral neck and knee, ligamentous injuries of the knee, neurovascular injuries, and injuries to other organs should be investigated. The optimal treatment of femoral diaphyseal fracture is intramedullary nailing, which restores alignment of the lower limb and proper rotation with prevention of shortening. It also allows early weight-bearing and return to early daily activities.

49.2 Epidemiology

Femoral shaft fractures occur in 10–37/100,000 patients per year with a peak incidence occurring in young adult males. Bilateral femur fractures are less common; however, they have a mortality rate of up to 25%. Young male patients are more affected than elderly and female patients. Transverse and oblique fracture patterns are the most common configurations in young patients. Spiral patterns are commonly seen after low-energy mechanism in older patients. A diaphyseal femur fracture is accompanied by a femoral neck fracture in 2.5–9%. The femoral neck frac-

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ture is often nondisplaced and is missed in 30–50% of cases.

Ipsilateral knee ligamentous and meniscal injuries occur in association with femoral diaphyseal fractures. Ligamentous laxity is seen in 50% of patients and meniscal tear in 30% of patients, respectively.

Femoral fractures with arterial injury requiring vascular repair are uncommon severe injuries, accounting for 0.3–0.4% of fractures. It represents a surgical emergency.

Isolated nerve injuries are more likely following penetrating trauma. On the other hand, nerve injuries are usually associated with vascular injury in blunt traumas.

49.3 Etiology/Pathogenesis

Femoral diaphyseal fractures are observed in all age groups. High-energy blunt mechanism is the most common and seen in younger patients. High-energy trauma generally leads to multiple injuries. The most common causes are road traffic accidents and direct fall on the affected limb. The majority of cases are seen in young male patients. Younger patients suffer from high-energy blunt trauma, whereas elderly patients suffer from low-energy mechanism such as simple fall from a standing height.

High-energy trauma due to direct mechanism leads to simple diaphyseal fractures with transverse and oblique fracture pattern. Spiral, segmental, or wedge-type fractures are mostly observed after an indirect trauma mechanism that is usually seen in elder patients. Not surprisingly, extensive soft-tissue damage is accompanied with high-energy trauma. Large segmental bone defects are the consequences of gunshot injuries.

Common associated injuries include pelvis and acetabular fractures, femoral head, distal femur, patella, and tibial plateau fractures.

In the absence of trauma, femoral fractures should be investigated for possible underlying diseases, which can cause pathological fracture. Although it is common in the proximal femur, pathological fractures may also be seen in the femoral diaphysis. Tumors and metabolic bone diseases can lead to this condition.

49.4 Classifications

Femoral diaphyseal fractures are classified according to the level at which they occur, fracture configuration, and associated soft-tissue injury. Location is typically described as proximal, middle, or distal diaphysis. Fracture configuration can be spiral, transverse, oblique, segmental, and comminuted. Comminution at the fracture site is the main factor that affects the clinical outcome. Therefore, a classification concerning fracture comminution would be more useful by predicting the clinical outcome. Although many classification systems have been described for femoral diaphyseal fractures, most of them were developed on the basis of Winquist-Hansen classification. Winquist-Hansen classification system was established on the amount of fracture comminution (Fig. 49.1). It has a therapeutic implication. Four types are defined:

- Type 0: No comminution
- Type I: Minimal comminution, less than 25% of the bony circumference
- Type II: Comminution involves up to 50% of the circumference of the femoral diaphysis
- Type III: Comminution more than 50% of the circumference of the femoral diaphysis
- Type IV: No contact between the fragments after reduction

According to this classification, types I and II have a stable bone contact at the fracture site. On the other hand, types II and IV have limited or no contact between the fragments. Therefore, type I and II fractures are considered as stable, whereas types III and IV are unstable, respectively. Static locking screws were indicated for unstable type III and IV femoral diaphyseal fractures to maintain limb length and correct rotation.

AO/ASIF classification describes fractures based on morphology and suggests the mechanism of the applied force. It uses alphanumeric

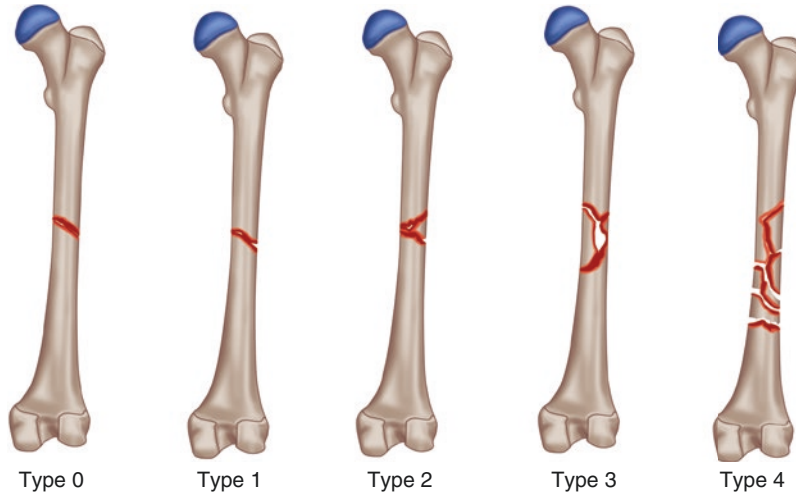

Fig. 49.1 Winquist classification of femoral diaphyseal fractures

codes. The femur has the code of 3, whereas diaphyseal involvement has the code of 2. Fracture configuration is coded alphabetically. A is a simple fracture, B is wedge shaped, and C is for complex fractures. Thus, the code for femoral diaphyseal fracture is 32A/B/C.

49.5 Diagnosis (Clinical and Imaging)

Associated injuries are common after high-energy traumas. Therefore, comprehensive examination of the whole body is essential. Airway, breathing, and circulation should be assessed and managed according to the guidelines. In the cases of fractures with low-energy mechanism, pathologic fractures should be investigated.

The thigh on the fractured side is usually painful and swollen, and the deformity due to instability is obvious. Typically, the affected limb will present shortening, malrotation, and translation. The distal extremity should be evaluated for pulses, sensation, and motor function. If there are no signs of neurological injury, the patient is able to actively plantar- and dorsiflex both ankles and toes. However, leg lifting and knee flexing are difficult due to pain that was created by instability. Clinical evaluation should include soft-tissue condition and neurovascular examination. The affected leg should be handled with care and splinted in order to prevent additional soft-tissue damage.

The clinician should also seek any other orthopedic injuries that could associate with a femoral diaphyseal fracture. The presence of ecchymosis, crepitus, and deformity may be the signs of additional injuries and indicate a further examination. Acetabular fractures, ipsilateral femoral neck fractures, and ligamentous knee injuries are the most common accompanying injuries. Associated fractures can usually be identified on the radiographs, which is typically obtained in the trauma bay. However, knee injuries often stay undetected. It is difficult to examine the stability of the ipsilateral knee in the presence of acute femoral diaphyseal fracture. Therefore, the knee should be examined immediately after the stabilization of the fracture. It was stated that mean time from femoral fracture to documentation of knee instability is greater than 1 year. Half of the ligamentous injuries are anterior cruciate ligament (ACL) tears.

Accompanying femoral neck fractures must also not be ignored. These femoral neck fractures have often vertical orientation and tendency to displace. Thus, failure to diagnose these fractures has devastating consequences including femoral neck nonunion and avascular necrosis.

After the stabilization of the femoral fractures, the hip should be examined and any abnormal finding should be noted. The pain during the hip motion should be investigated by radiological examination.

The physician should seek symmetrical pulses, namely, the ankle-brachial index should be equal in both extremities. If a vascular injury is suspicious (ankle-brachial index of less than 0.9), Doppler ultrasound scanning is necessary. Angiography is reserved for patients with abnormal vascular examination and uncertain level of arterial injury.

Radiological examination begins with plain radiographs. Anteroposterior and lateral radiographs are ordered. The femur with ipsilateral hip and knee joints should entirely be in visualization. Thus, ipsilateral hip and knee fractures can be diagnosed. Although the femoral diaphyseal fractures are easily seen in plain radiographs, nondisplaced femoral neck fractures may easily be overlooked and further evaluation of suspected hip and knee fractures is then done with computerized tomography (CT). CT scans is performed for further surgical planning in complex fracture patterns. After the determination of fracture localization and fracture pattern, anatomic features of the femur in terms of femoral bowing, canal diameter, and bone quality are investigated. The amount of shortening can be expected from the radiographic evaluation of contralateral femur.

49.6 Treatment

Management strategy should be based on the identification and treatment of all life-threatening injuries. Patients are optimally resuscitated before the definitive treatment of such fractures including femoral diaphysis. Open wounds are irrigated, debrided, and covered with a sterile dressing. Administration of antibiotics and tetanus prophylaxis is necessary in open fractures. Immobilization of the limb will control the pain, bleeding, and further soft-tissue injuries.

The optimal treatment of femoral diaphyseal fractures should restore limb length and rotation, stabilize the fracture, and allow early ambulation of the patient and return to daily activities as soon as possible. For this purpose, the diaphyseal femoral fractures are treated surgically. Conservative treatment with skeletal traction as a definitive treatment by means of distal femoral or proximal tibial traction has been used historically and showed no clear benefits. It has several complications such as pin-site infection, pin loosening, neurovascular insult, pulmonary complications, and knee stiffness. Skeletal traction may be used temporarily and may have a role in preventing the motion on fracture site, which can lead to additional soft-tissue injury. Initial realignment of the injured extremity will also decrease pain.

Three kinds of surgical fixation method are available for diaphyseal femoral fractures (Table 49.1):

- Intramedullary nailing
- Plate fixation
- External fixation

The standard treatment for all diaphyseal femoral fractures is intramedullary nailing. It has many advantages compared with other treatment modalities. It can be typically performed in a closed fashion, thereby maintaining the fracture hematoma and the associated soft-tissue envelope. The implant is placed into the femoral canal. Thus, it provides a predictable realignment. As the implant acts as a load-sharing device, it allows early weight-bearing. Intramedullary nails are locked to prevent malrotation and shortening. Locking can be done either static or dynamic. In general, all diaphyseal fractures are statically locked to provide axial and rotational stability to the fracture. In fractures without comminution, dynamic locking may be used for acceleration of fracture union. Locking intramedullary nailing allows immediate joint mobilization and weight-bearing even in the setting of significant comminution.

The diaphyseal femoral fractures treated by intramedullary nailing demonstrated a union rate of 98% and less than 1% of infection rate. Intramedullary nails have several options according to the entry point, reaming, and direction of nailing. According to the direction, fixation can be performed either anterograde or retrograde. Traditionally, anterograde nailing is the treatment of choice. Piriformis or trochanteric entry is the starting point for anterograde nailing. Entry

Table 49.1 Suggested treatment methods for femoral diaphyseal fractures

Injury	Treatment
Closed femoral diaphyseal fractures *Proximal third* *Middle third* *Distal third*	Anterograde or retrograde nailing *Anterograde nailing* *Anterograde nailing* *Retrograde nailing*
Open femoral diaphyseal fractures *Grade I* *Grade II* *Grade III*	Intramedullary nailing of external fixation *Intramedullary nailing* *Intramedullary nailing* *External fixation followed by intramedullary nailing*
Femoral diaphyseal fractures in multiple injured patient	Primary external fixation followed by secondary intramedullary nailing
Femoral diaphyseal fractures with additional skeletal injuries *With ipsilateral femoral neck fracture* *With tibial fracture*	Cephalomedullary or retrograde nailing *Cephalomedullary or retrograde nailing with cannulated screws or sliding hip screw for femoral neck fracture* *Retrograde nailing of femur and standard intramedullary nailing for tibial fracture*
Pathological fractures of the diaphysis	Intramedullary nailing
Femoral diaphyseal fractures in association with vascular injury	*External fixation following vascular repair* *or* *Vascular repair following plate fixation*
Femoral diaphyseal fractures in obese or pregnant patient *Obese* *Pregnant*	Intramedullary nailing *Retrograde nailing* *Retrograde nailing*

points demonstrated to have no effect on union rates, complication rates, and functional outcomes. In the setting of multiple trauma, pregnant, or obese patients, retrograde nail fixation has its own advantages. The technique requires less radiation exposure and less surgical time as compared to anterograde nailing. In the cases of ipsilateral fractures of femoral neck and diaphysis, diaphyseal fracture is treated by retrograde nailing, whereas femoral neck fracture can be fixed with plates or lag screws. Retrograde nailing may also be considered in ipsilateral femoral

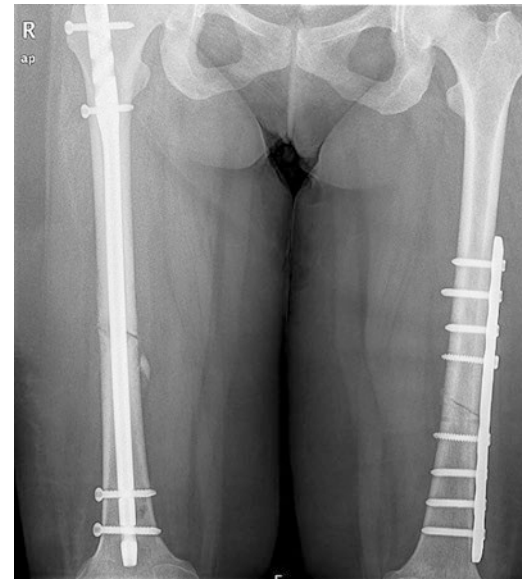

Fig. 49.2 Bilateral femoral diaphyseal fractures in a 29-year-old female patient. Right femur was treated by intramedullary nailing, whereas plate fixation was preferred for the left femur. Although both of the fractures were suitable for intramedullary nailing, the reason for performing plate fixation was to avoid possible pulmonary complications

or tibial diaphyseal fractures. Both of the fractures can be stabilized from the same surgical approach. Reaming before the insertion of the nail increases the union rate. Although reaming showed to be safe in multiple trauma patients, it should be used cautiously in patients with pulmonary contusion. Patients with multiple injuries including open femur fractures should be managed with caution. These patients have a tendency to develop pulmonary complications (Fig. 49.2).

Although newer generation of intramedullary nailing has extended the zone of indication to more proximal and distal diaphyseal fractures, in some circumstances, plate osteosynthesis is preferable instead of intramedullary nailing. Patients with an excessively narrow femoral intramedullary canal or those with preexisting deformities, proximal or distal localized diaphyseal fractures, and femoral fractures associated vascular injury are the candidates of plate-screw osteosynthesis. Although some reports support the use of plate osteosynthesis with subsequent high union and

low infection rates, several studies reveal increased complications including longer union and high infection rates than intramedullary nails. Delayed weight-bearing coupled with high reported complication rates puts plate osteosynthesis into disfavor.

External fixation, which is sometimes used in pediatric age group, appears to be a useful fixation method in systemically compromised adult patients and hemodynamically stable patients with open fractures. In the initial scenario, fractures are stabilized with external fixation temporarily and converted to internal fixation. Open femoral diaphyseal fractures are also managed with external fixation. Risk of infection that has emerged with the soft-tissue stripping at the fracture site during internal fixation will be minimized by external fixation. Thus, wound care can safely be ensured. The literature supports safe conversion to intramedullary nailing within the first 2 weeks to minimize the risk for infection.

Definitive treatment of femoral diaphyseal fractures with external fixation results with high rates of complications including nonunion, malunion, knee stiffness, and pin-tract infection. High complication rates with the use of external fixation make the procedure less desirable option for definitive bony stabilization. Therefore, external fixation has to be followed by internal fixation.

Femoral diaphyseal fracture is also seen in the pediatric population. Although it occurs as a result of direct trauma, pathologic fractures due to cystic bone lesions and osteogenesis imperfecta should be ruled out. Traction, in terms of skeletal or skin, continues to have a place in the closed treatment of femoral diaphyseal fractures. In most instances, traction is used as a bridge to hip spica casting. Traction followed by delayed hip spica casting is used for prevention of limb-length discrepancy. The vast majority of these fractures heal uneventfully with nonsurgical treatment consisting of hip spica cast until 6 years of age. Limb-length inequality of 2 cm, which may be observed after the treatment, usually equalizes as the child grows. In the age group of 6–12, traction and hip spica casting are rarely pursued due to the risk of arthrofibrosis that is caused by immobilization of the knee in a fixed position. Stronger deforming forces in older children group make the maintaining of the fracture reduction difficult. Therefore, surgical treatment is considered as a definitive treatment. Elastic nailing, external fixation, and submuscular plating are the potential treatment options. As the adolescents still have growth potency, protecting the open growth plates has a significant role in the treatment of femoral diaphyseal fractures. Rigid intramedullary nails have potential harm to the growth plates. It may also jeopardize the blood supply to the femoral head, especially if they have a piriformis entry. Flexible intramedullary nails, which are inserted retrograde proximal to the distal femoral growth plate, are more suitable in patients with open growth plates. It is preferred over spica casting and external fixation by providing better personal care and early weight-bearing. Spiral and comminuted fractures present the risk of shortening over flexible nails. These fracture patterns are better treated by submuscular plating.

Take-Home Message

- Advanced Trauma Life Support principles should be initiated in patients with femoral diaphyseal fractures.
- Patients with femur fractures should closely be examined for potential associated injuries.
- Comminuted femur fractures are at risk for malunion, nonunion, and leg-length discrepancy.
- Statically locked intramedullary nailing is the standard of care nearly for all femoral diaphyseal fractures.

Summary

Femoral diaphyseal fractures are reasonably common and can be seen in isolation or associated to other injuries. As the conservative treatment has several complications and take a long period of immobilization, femoral diaphyseal fractures are treated surgically. In adults, internal fixation by intramedullary nailing is the treatment of choice.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Which type of nail is used for the adult femoral diaphyseal fractures with transverse fracture pattern?
   (a) Anterograde reamed nails are mostly preferred.
   (b) Cephalomedullary nails are the standart treatment modality for transverse femoral diaphysial fractures.
   (c) Retrograde nails are the most common IM adopted.
   (d) Titanium elastic nails provide the most stable fixation for adult diaphysial femoral fractures.
2. Do I have to lock the nail with screws? Dynamic or static locking?
   (a) Static locking is preferred in old patients, while dynamic locking is used in young patients.
   (b) Locking has no place in intramedullary nailing.
   (c) Static locking is used for stable fractures; whereas dynamic locking is reserved for comminuted diaphysial fractures.
   (d) Fractures in comminution are statically locked to provide axial and rotational stability to the fracture. Dynamic locking is used in noncomminuted fracture patterns, such as transverse and oblique fractures.
3. Are the patients allowed for weight-bearing after the fracture treatment?
   (a) No, patients are not allowed to weight-bear for the first 3 months. As the intramedullary nailing is a load-sparing implant, these patients are discouraged for early weight-bearing.
   (b) Screw plating procedure allows early weight-bearing.
   (c) Most patients are allowed to bear weight to varying degrees just after the fracture fixation, even in patients with unstable fracture patterns.
   (d) Yes, patients are ever encouraged to weight-bear after 1 month.
4. How long time is enough for conversion from external fixation to internal fixation device?
   (a) There is no need for conversion from external fixation to internal fixation device.
   (b) Even in open fractures, fractures could be well managed by delayed intramedullary nailing within 2 weeks with initial temporarily external fixation.
   (c) In fact, it is necessary to convert internal fixation to external fixation.
   (d) In order to change the fixation method, it is necessary to see the signs of union radiologically.
5. If an associated femoral neck fracture can easily be overlooked, how can I diagnose a nondisplaced femoral neck fracture?
   (a) CT evaluation of the hip to detect associated nondisplaced femoral neck fractures is recommended in trauma patients.
   (b) Every femoral diaphysial fracture requires an MRI evaluation of the hip.
   (c) X-ray (anterolateral and frog-view) evaluation of the hip is always sufficient to establish the diagnosis of associated nondisplaced femoral neck fractures.
   (d) Joint arthrography could be helpful.

Further Reading

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Part X

Knee

50 Treatment Options of Cartilaginous Lesions of the Knee

Caroline Struijk and Peter Verdonk

Overview
Cartilaginous lesions of the joint (most common knee and ankle) cover a broad group of structural defects of the smooth joint surface responsible for progressive pain and functional limitations.

50.1 Definition

Cartilage acts as a natural shock absorber through its firm and elastic properties comprising an extracellular matrix with only single-cell-type "chondrocytes." The matrix forms a highly organized and multilayered network containing collagen fibers and proteoglycans. Cartilaginous lesions of the joint (most common knee and ankle) cover a broad group of structural defects of the smooth joint surface responsible for progressive pain and functional limitations.

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50.2 Epidemiology

With a growing popularity of recreational sports and higher physical demands, the incidence of focal cartilage lesions is increasing across the whole population. A cohort study on the epidemiology of knee articular lesions found that chondral lesions were present in 60% of all patients undergoing arthroscopy with most common affected areas being the medial femoral condyle and the patellar articular surface.

50.3 Etiology/Pathogenesis

The young and active population is often prone to cartilage defects secondary to traumatic events, while defects in the older population are subject to slow degenerative deterioration processes. Focal lesions are well demarcated and typically caused by trauma, osteochondritis dissecans, or osteonecrosis. Degenerative cartilage failure is usually secondary to excessive load, in particular with the presence of concomitant abnormalities of the knee such as malalignment, ligament instability, meniscal injury, and morphotype.

Cartilage homeostasis is mainly driven by paracrine communication of chondrocytes and established through various biochemical, biomechanical, and matrix-related pathways, adapting to functional needs of the patient. Upon dysfunction, fibers become disaggregated

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and cartilage tissue loses its composition and mechanical properties. Cartilage has little intrinsic capacity for repair secondary to its avascular properties. Therefore, damage to the functional unit of the joint (cartilage and subchondral bone) is responsible for insidious onset of pain and mechanical dysfunction of the knee joint.

50.4 Classification

Defects present across a broad scale of severity ranging from an asymptomatic accidental finding on imaging to physically severely disabling and manifest in a variety of forms and types (e.g., superficial vs. full thickness, uni- vs. multifocal lesions). Table 50.1 gives an overview of two grading systems: the ICRS and Outerbridge classification. Importantly, the size of the lesion is not taken into account for both grading systems.

Table 50.1 Classification of osteochondral lesions

ICRS grading system		
Grade 0	Normal	
Grade 1	Almost normal	1a—Superficial lesion 1b—Superficial cracks and fissures
Grade 2	Abnormal	Extent <50% cartilage depth
Grade 3	Severe lesion	3a—Extent >50% cartilage depth 3b—Extent to calcified cartilage 3c—Extent to the surface of subchondral bone without penetration 3d—Cartilage bulging around lesion site
Grade 4	Very severe lesion	4a—Defect penetration to subchondral bone, not across full diameter 4b—Defect penetration to subchondral bone across full diameter
Outerbridge system		
Grade I	Softening and swelling of cartilage	
Grade II	Defect with fissures—Does not extend to subchondral bone or area <1.5 cm in diameter	
Grade III	Defect with fissures—Extent to subchondral bone in area >1.5 cm in diameter	
Grade IV	Defect exposed to subchondral bone	

50.5 Diagnosis

Routine assessment of patients presenting with symptoms caused by cartilage lesions consists of history-taking, physical examination, and imaging. Patients often present with insidious onset of pain, effusion, and/or mechanical dysfunction. Ligament instability and meniscus injury should be evaluated during physical examination. Radiography, including bipodal standing full-leg X-ray, is often performed to rule out tibiofemoral malalignment. Advanced imaging such as MRI and arthro-CT scan provides a noninvasive tool to assess the damaged area and concomitant lesions in detail. MRI is generally considered the golden standard; however, the senior author considers arthro-CT scan as an advantageous tool to assess in detail lesion size and involvement of subchondral bone.

50.6 Treatment

Management of cartilage lesions is highly challenging in clinical practice. The patient with symptomatic chondral injury has a wide extent of treatment options ranging from conservative management through various types of established surgical treatment options.

Each patient's lesion needs an individualized approach taking into account patient's factors (functional loss assigned to defect, comorbidities) and lesion's factors (size, location, depth). Along with the patient's physical goals and commitment to rehabilitation, the surgeon will design the treatment plan. Despite the assumption that early treatment of lesions may reduce further exposure to degenerative progression of the knee joint, a step-by-step conservative approach is preferred if the lesion is not compromising athletic performances.

50.7 Conservative Management

The primary purpose of conservative management is symptomatic relief, improvement of function, and re-establishing a new homeostasis of the knee joint, therefore leaving the root cause of discomfort untouched. Conservative management includes a broad variety of options such as physical therapy, knee bracing, non-steroidal anti-inflammatory medications, pain relievers, and different types of intra-articular injections. The use of bisphosphonates is being investigated in the presence of an associated bone marrow lesion.

State-of-the-art physical therapy includes activity modification and muscle group strengthening. Knee bracing is based on the same principle as both reduce the intra-articular joint load. Oral non-steroidal noninflammatory medication and intra-articular corticosteroids calm down inflammation and alleviate discomfort. Another approach is that of viscosupplemental agents such as hyaluronic acid injections that provide nutrition and lubrification. Finally, biological agents such as platelet-rich plasma (PRP) and m-fat (micro-fragmented adipose tissue) therapy (Lipogems) have gained widespread interest over the years as some investigators claim anti-inflammatory and anabolic benefits and therefore promotion of healing and regeneration. It is generally suggested that inflammatory cytokines and other catabolic processes may disrupt joint homeostasis and therefore contribute to cartilage failure.

50.8 Surgical Management

Surgical approaches address damaged tissue restoration and repair and may be indicated upon failure of nonoperative management.

50.9 Arthroscopic Joint Lavage and Debridement

In this surgical approach, the knee joint is debrided of unstable or loose bodies which alleviates discomfort and restores joint dynamics, while defect repair is not addressed with this intervention. A wide scope of arthroscopic probes is available with applications ranging from mechanical debridement to radiofrequency energy (RFE) probes (Arthrocare®) that generate high energetic plasma fields. The main goal of these applications is cartilage surface smoothening through removal of fibrillations and fissures and stabilization of loosened edges. In recent years, a shift has taken place back from reparative surgery towards joint debridement and stabilization, especially in a young and still active, athletic population. Reparative surgery is only indicated when the athlete can no longer participate in sports at his/her expected level.

50.10 Reparative Management

50.10.1 Microfracture

Microfracture surgery is based on bone marrow stimulation resulting in influx of beneficial factors such as mesenchymal stem cells (MSCs), growth factors, and platelets to regenerate the defect. More specifically, reparation is pursued by penetrating several holes in the subchondral bone, which stimulates a fibrin clot containing these marrow products (Fig. 50.1). Consequently, attachment of the fibrin clot at the damaged site enhances organization and remodeling of the tissue resulting in defect filling with fibrocartilage, a product inferior to the traditional hyaline cartilage. Microfracture is a straightforward one-step procedure and well established worldwide and is therefore often considered as the first choice of treatment for smaller (<2–3 cm^2) chondral lesions.

Despite its encouraging short-term clinical outcomes, patients tend to evolve unfavorable due to degeneration of the repaired tissue and ingrowth of intralesional osteophytes.

50.10.2 Microfracture Plus

Several advanced techniques have arisen from the traditional microfracture. These off-the-

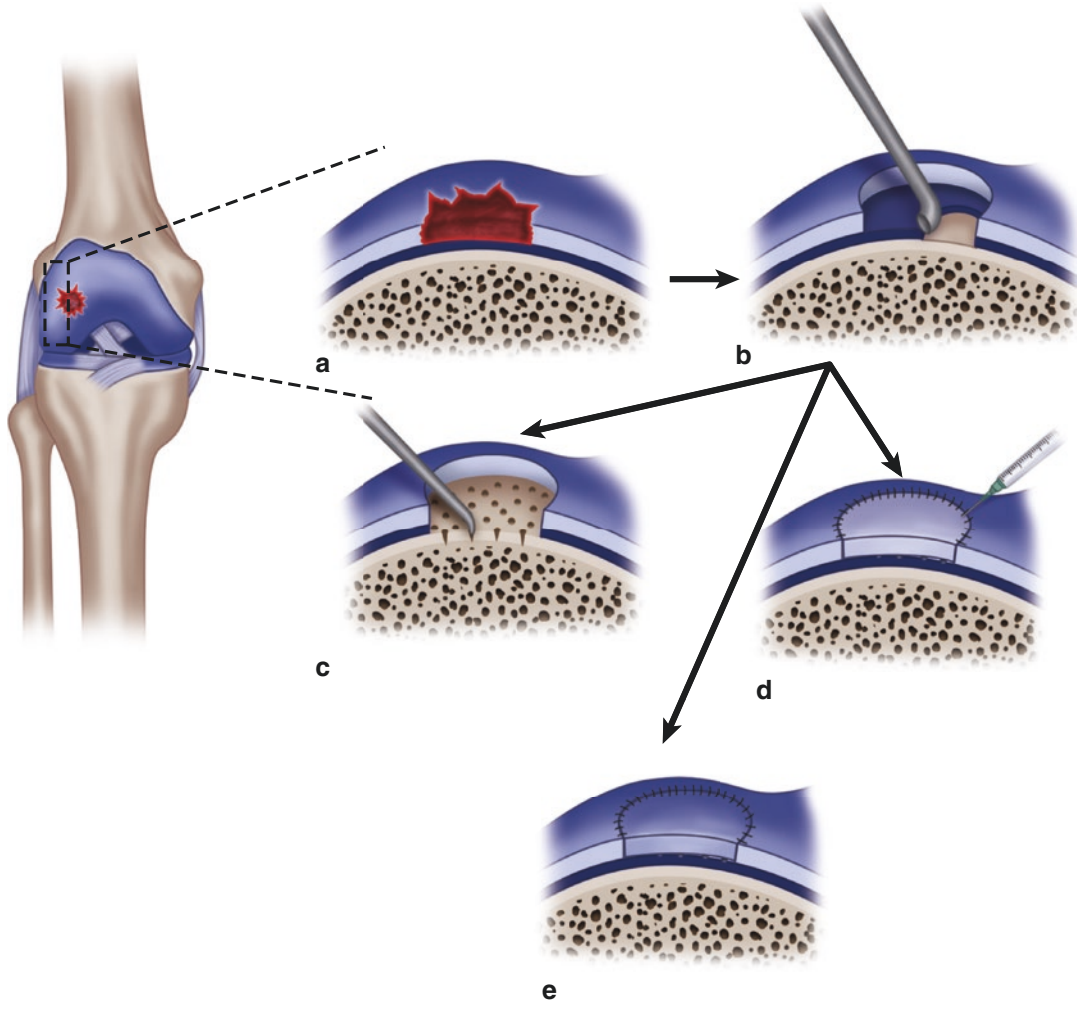

Fig. 50.1 Cartilage regeneration techniques. (**a**) Focal chondral lesion. (**b**) Debridement of lesion. (c) Microfracture. (**d**) Autologous chondrocyte implantation (ACI). (**e**) Matrix-induced autologous chondrocyte implantation (MACI)

shelf available products optimize conventional bone marrow stimulation by addressing the intrinsic healing of patients. *BST-CarGel*® (Smith & Nephew) is a chitosan-based substitute used in the mixture with the patient's whole blood and is responsible for accelerated clot formation and therefore a better attachment and filling of the defect. Several newer treatment techniques combine marrow stimulation in conjunction with acellular scaffolds and will be discussed under the scope of the subsection "acellular scaffolds."

50.11 Osteochondral Transplantation

Osteochondral transplantation is a restorative technique in which hyaline cartilage grafts are transplanted to the damaged area. These grafts may originate from the patient's own healthy cartilage in nonbearing regions of the joint (autologous) or from cadaveric donors (allograft).

Osteochondral autograft transplantation (OAT) is a surgical technique in which osteochondral plugs containing both cartilage and sub-

chondral bone, are harvested from non-load-bearing regions in the knee. The amount of harvested plugs depends on the defect diameter; however, the source is not endless considering donor-site comorbidities, which makes this technique unfeasible for larger lesions. An alternative to using one large graft is opting for multiple, smaller grafts to rebuild the defect site and is referred to as mosaicplasty.

According to the senior author's experience and preference, no more than three plugs are used for mosaicplasty, taking the patient's size into consideration.

Optimal indications for this technique are a young, high-demanding patient with small-to-medium focal lesions (<2–3 cm^2).

Donor-site morbidity and a limitation in lesion size associated with mosaicplasty drove experts to osteochondral allograft transplantation (OCA), a surgical technique wherein cadaveric donor plugs are used to restore the articular joint surface. OCA has shown reproducible clinical success in treating a variety of complex defects and ensures the biomechanical stability of mature hyaline cartilage, which makes OCA seem to be the most favorable treatment option. Major limitations that allogeneic donor tissue carries along are graft availability and costs, which is a major challenge in Europe. However, cost-effectiveness for osteochondral allograft transplantation is very high as expensive initial treatment costs are counterbalanced by excellent long-term survival rates.

50.12 Cell-Based Approaches

Autologous chondrocyte implantation is a two-staged restoration procedure based on healthy cartilage harvesting, in vitro chondrocyte expansion, and subsequent introduction of the cell suspension in the injured defect covered with a membrane of either periosteum, collagen patch, or matrices (Fig. 50.1). ACI is a durable and effective solution for younger individuals with larger (>2–4 cm^2) chondral lesions, without damage to the subchondral base. Disadvantages are costs and procedure complexity, which led to matrix-induced autologous chondrocyte implantation (MACI) (Fig. 50.1). In this second-generation ACI, expanded chondrocytes are introduced to a porcine-derived collagen-based scaffold, cultured and implemented at the defect site. Superiority of this cell-seeded scaffold over ACI in terms of patient-reported clinical outcomes remains unproven.

ACI using spheroids (Co.Don AG) is another approach available in which autologous chondrocytes are expanded, condensed into spheres, and introduced to the lesion site without the need for a membrane. These second-generation ACI techniques allow for controlled defect filling and guided matrix development, presuming to lead to faster recovery times. Yet, it carries the same drawbacks as ACI in terms of multistep procedure, cell expansion, and associated costs.

Therefore, several one-step approaches using (co-)cultures with different (stem) cell sources in conjunction with biodegradable scaffolds are under investigation by cell engineering initiatives.

Carti-ONE is a single-stage ACI approach in which autologous chondrocytes obtained from the knee joint cartilage are mixed intraoperatively with autologous bone marrow aspirate. Consequently, the surgeon introduces this mixture with a membrane carrier to the defect site where enhanced regeneration of articular cartilage takes place. Similar to this approach, Saris et al. designed an enhanced single-stage cell transplantation approach in which co-cultures of allogeneic MSCs and defect-derived autologous chondrons (chondrocytes with pericellular matrix) are used for tissue repair. The clinical trial showed superior clinical outcomes in comparison to ACI and microfracture.

50.13 Cell-Free Scaffolds

A more recent approach in the treatment of osteochondral defects involves acellular scaffolds that may facilitate local cell ingrowth often after mobilization with bone marrow stimulation.

This 3D biodegradable template allows homogeneous organization and remodeling of the microenvironment of hyaline cartilage matrix.

Off-the-shelf availability and single-stage procedures are major assets associated with implementing these cell-free scaffolds, which reduces costs and allows flexibility in procedure planning and on-the-spot decision-making. However, since numerous different materials are used and investigated these days, studying clinical and functional outcomes becomes a challenging task.

Hyalofast™ (Anika Therapeutics) is a hyaluronic acid-based scaffold that upon implantation supports mesenchymal cell attachment, differentiation, and proliferation in (osteo)chondral lesions. Entrapment of cells in combination with hyaluronic acid release by the biodegradable scaffold creates a beneficial microenvironment for hyaline-like cartilage defect repair.

Gelrin C© (Regentis Biomaterials), a cell-free hydrogel scaffold, is implanted at the chondral defect after microfracture as a liquid suspension. Upon exposure to UV light, its composition becomes solid resulting in an advantageous incorporation of the scaffold at the defect site.

Amic® (Autologous Collagen-induced Chondrogenesis) is an advanced technique derived from standard microfracture in which a porcine collagen membrane is implanted at the osteochondral lesion site after bone marrow stimulation.

Osteochondral cartilage lesions require multiphasic scaffolds that address both the cartilage ("chondral") and bone ("osteo") layer simultaneously.

BioMatrix CRD (Arthrex®) is a bi-layered scaffold composed of bovine collagen for the chondral aspect and ß-tricalcium phosphate (ß-TCP) suspended with polylactic acid (PLA) for the subchondral bone region. ***MaioRegen*** (Finceramica) is an osteochondral device with a multiphase gradient which provides different support for regeneration of both the cartilage as the bone. ***Agili-C***® (Cartiheal Ltd.), an aragonite-based porous scaffold, was inspired by natural coralline for the bony integration of the scaffold and has shown excellent potential in recent clinical trials.

50.14 Future Trends

The nature of repair tissue is predominantly fibrocartilaginous, which behaves poorly biomechanically, prompting for cartilage engineering initiatives to seek a hyaline(-like) cartilage product that is durable and easily introduced in clinical practice. In recent years, different treatment technologies have started to fuse, resulting in cell-based strategies delivered in smart scaffolds augmented with biologically active factors.

In addition, more research is currently performed to better profile patients based on phenotype (i.e., imaging) and genotype technology with a goal to personalize conservative, medical, and surgical treatment options.

Take-Home Message

Cartilaginous lesions of the knee present across a broad scale of severity ranging from an asymptomatic accidental finding to physically severely disabling. Treatment decision-making is based on all patient- and lesion-related parameters and includes conservative and surgical procedures. Most often, a stepwise approach is warranted and concomitant problems should be addressed. Nevertheless, the outcome remains variable. Hence, further research is conducted on combinations of yet existing approaches and patient profiling.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Classifications such as the ICRS and Outerbridge systems grade lesions:
   (a) According to the length in MRI
   (b) According to the size in CT scans
   (c) According to their aspect in X-Ray
   (d) According to their extent in depth and size

2. The treatment outcome
   (a) Depends on genetic factors
   (b) Is indirectly proportionate to appropriate patient selection and providing sufficient information regarding the procedure and demanding rehabilitation process
   (c) Is directly proportionate to appropriate patient selection and providing sufficient information regarding the procedure and demanding rehabilitation process
   (d) Depends on the surgical technique
3. Arthroscopic evaluation of cartilage lesions
   (a) Is useful only for therapeutic purposes
   (b) Provides the most information and is considered the best diagnostic tool
   (c) Is useful only for diagnostic purposes
   (d) Is the first treatment after a clinical suspicion
4. Osteochondral allograft transplantation (OCA)
   (a) Is an excellent treatment technique for all the lesions
   (b) Is an excellent treatment technique for challenging lesions, i.e., the type of lesions that are less feasible for microfracture or mosaicplasty.
   (c) Could be used only in <40 years old patients
   (d) Could be used only in elderly patients
5. Early surgical treatment of cartilage lesions
   (a) Is always preferred as it may reduce further exposure to degenerative progression of the knee joint
   (b) Is never preferred as it is too invasive
   (c) Could be performed as an alternative of MRI
   (d) Is mandatory in degenerative knee lesions

Further Reading

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Meniscal Injuries of the Knee

51

Simone Perelli, Veronica Montiel Terrón, and Juan Carlos Monllau

Overview

Meniscal tears remain a daily challenge in orthopedics as they are frequent injuries, and the incorrect treatment can lead to deleterious consequences for the knee joint. The tear can be either traumatic or degenerative. The former is frequent in the young active population and can occur in the context of ligamentous lesions. Degenerative lesions are detected more in older patients without a history of trauma and frequently in the context of cartilage degeneration. In the case of a complete traumatic injury and symptomatic patient, the surgical solution is recommended. In the case of degenerative lesions, a conservative treatment should always be tried before considering a surgical approach. Conservative treatment is based on muscular strengthening, weight loss, anti-inflammatory drugs, and eventual intra-articular infiltrations. If 6 months of conservative treatment is not effective in resolving the symptoms, the surgical option can be considered. When the surgical approach is chosen, the goal is always to preserve as much meniscal tissue as possible. It means the less tissue extirpation as possible and carrying out meniscal repair when it is indicated. Meniscal repair is indicated when the lesion is acute (within the first 4 months of the lesion), the meniscal tissue is not degenerated, and the lesion is in a vascularized portion of the meniscus.

51.1 Definition

Traumatic or degenerative lesions of the meniscus can be isolated or associated with other traumatic or degenerative lesions (e.g., traumatic lesions of the anterior cruciate ligament, degenerative lesions of the articular cartilage).

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51.2 Epidemiology

Meniscal ruptures are a very frequent injury in the young population. It affects 9 in 10,000 men and has a peak incidence between 31 and 40 years old. In women, the rate is 4.2 in 10,000, and there is peak incidence between 11 and 20 years old. Meniscal ruptures are most frequently found in the medial meniscus (74%) and on the dominant side (52%).

Degenerative lesions are even more frequent even though it is not possible to calculate an exact incidence since only a small percentage of them produce symptoms.

51.3 Pathogenesis

The menisci are important knee structures that are fundamental to increasing knee articular congruence due to the geometry of the femoral condyle and tibial plateau. Their role is to transform axial load into circumferential tension or hoop-stress to preserve articular cartilage viability and function. Both menisci move from the anterior area of the tibial plateau to the posterior area with knee flexion. However, they both move in different ways due to their distinct morphologies, insertion sites, and stability. The lateral meniscus transfers around 70% on the weight placed on the lateral compartment, while the medial meniscus transfers only around 50% of the weight. These weight transmissions can be as high as 85–90% in flexion. These basic biomechanical concepts may explain both the pathogenesis of a meniscal lesion and the consequence of meniscal extirpation.

Younger patients suffering a meniscal tear usually have a history of a weight-bearing, twisting, or hyperflexion injury. With these movements, shear forces are rapidly discharged on the meniscus, causing a lesion of the meniscus body or a detachment of its roots. The femur suffers rapid and exaggerated displacement over the tibia if a ligament lesion occurs, thereby increasing the possibility of a traumatic meniscal lesion.

The pathogenesis of the degenerative meniscal lesion is multifactorial and not yet fully understood. A genetic predisposition to collagen softening may play a role. Moreover, the pathogenesis is most often correlated with joint overload. It may be because of overweight, joint deformity, chronic ligament lesions, or a sport or work overload. Considering the biomechanical properties of the menisci, it is easy to understand how degenerative changes of the meniscus cause a decrease in cartilage protection by the menisci themselves. For this reason, a degenerative meniscal lesion is almost always associated with a certain degree of degeneration of the entire joint.

51.4 Classification

51.4.1 Etiological Classification of Meniscal Ruptures

Meniscal ruptures can be classified according to their etiology as either degenerative or traumatic.

Traumatic injuries can be associated with ligament and osteochondral injuries. Some 50% of these kinds of injuries take place while practicing sports like football or ski. They are mostly found in the least mobile meniscal areas. The posterior horn of the medial meniscus is a common site of injury. The rupture usually occurs in a healthy meniscus, meaning the tissue quality is neither pathologic nor degenerated.

On the contrary, degenerative injuries are usually found in degenerative knees with some degree of cartilage damage. In those cases, patients do not remember a clear trauma incident.

51.4.2 Meniscal Rupture Classification According to the Irrigation (Fig. 51.1)

Meniscal ruptures can also be classified according to the area in which they occur. Given that not all the areas in the meniscus receive the same blood supply, this is an important classification because it can determine the healing potential of

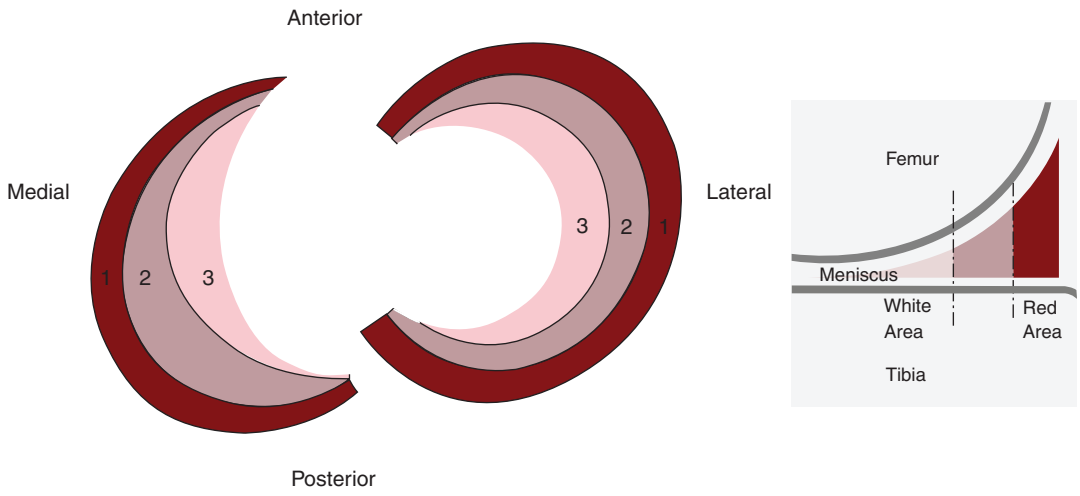

Fig. 51.1 Scheme of classification of meniscal lesions according to vascularization. Zone 1 (red), zone 2 (white-red), and zone 3 (white)

the rupture. Well-irrigated areas will heal better than those that are poorly irrigated. Therefore, three areas can be found in the meniscus:

1. The white area is the more central part of the meniscus, which is avascular. Therefore, white-on-white meniscal repair is not recommended because of the scarce potential for healing. For white-white lesions, meniscectomy is the treatment suggested.
2. The red-white area refers to the zone between the red and the white area. It has partial irrigation and thus a higher healing potential. Repairs in this area are least likely to lead to healing than in the red-to-red repair, but the repair is still recommended if the tissue quality is good.
3. The red area of the meniscus is the most peripheral and closest to the joint capsule. Therefore, it is the one that receives a better blood supply. Therefore, red-on-red repairs have the highest healing potential and the lowest risk of failure. As such, it is mandatory to try a meniscal repair on those lesions.

51.4.3 Morphological Classification of Meniscal Ruptures (Fig. 51.2)

Lastly, meniscal injuries can be classified according to rupture geometry. They can be oblique ruptures (or parrot beak), horizontal ruptures (or cleavage), vertical ruptures (longitudinal or radial), and complex ruptures (a combination of two or three of the other types, mostly found in degenerative joints). In lengthy vertical longitudinal ruptures, the central fragment can separate from the lateral one resulting in a so-called bucket handle tear. This mobile fragment can go in and out of place and cause knee locking. The mobile fragment is frequently found in the trochlear notch. Vertical radial ruptures result in a significant disruption of meniscus biomechanics and force transmission because they interrupt the circumferential meniscal fibers.

Meniscal root ruptures are a special kind of rupture which also cause an important disruption of meniscal biomechanics due to the interruption of the circumferential meniscus fibers. They can result in complete extrusion of the

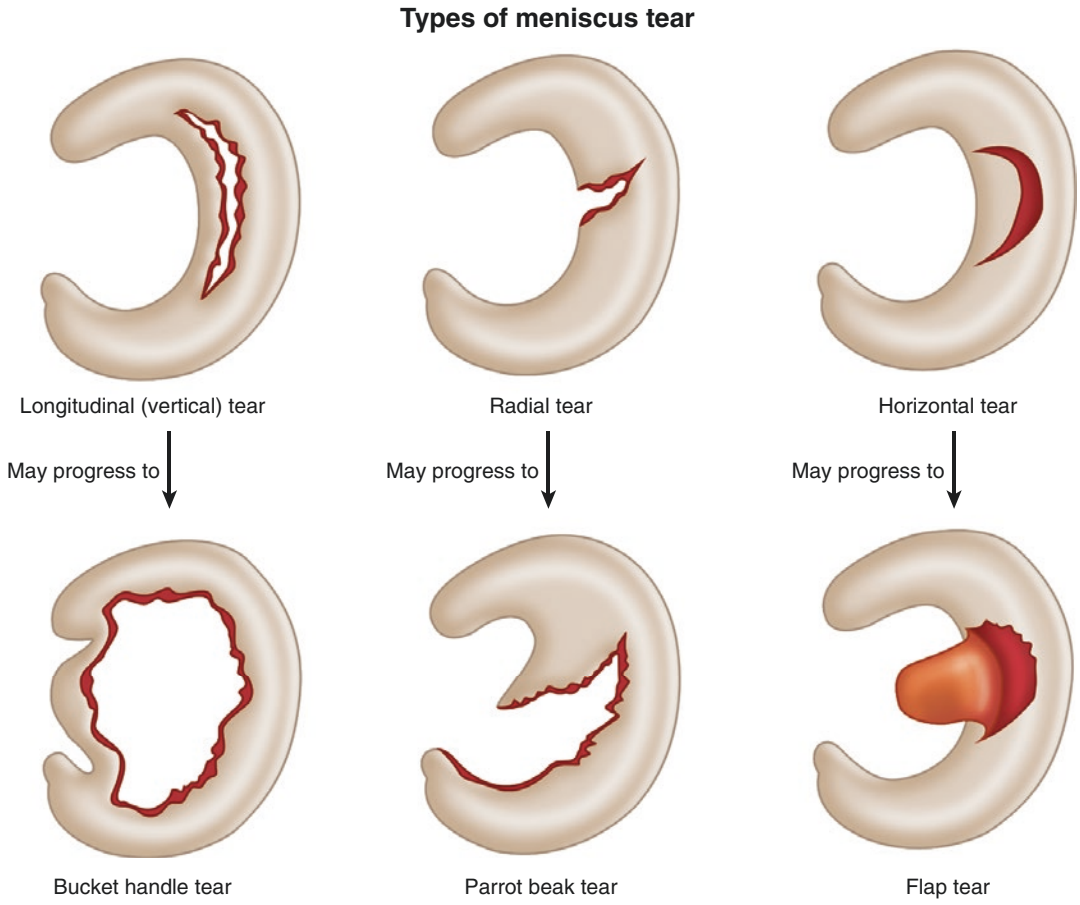

Fig. 51.2 Scheme of the classification of meniscal lesions according to morphology

menisci because of the absence of solid meniscal structure bone insertion. Meniscal extrusion is biomechanically equivalent to a complete meniscectomy and so has severe consequences on knee biomechanics. A posterolateral root lesion is usually associated with an ACL lesion with a rotatory mechanism. On the other hand, posteromedial root lesion is frequently observed in varus knee with mild degeneration in middle-age patients. The meniscal root tear is classified separately into five different types. They depend on the location and morphology of the lesion.

The most frequent location, injury mechanism, and type of patient in which each meniscal injury is found have been detailed in Table 51.1.

Table 51.1 The most frequent location, injury mechanism, and type of patient in which each meniscal injury is found

Type of meniscal rupture	Most frequent location	Injury mechanism	Type of patient
Vertical longitudinal tears (incomplete or complete depending on whether they involve part or the entire thickness of the meniscus)	Posterior horn of the medial meniscus because it is less mobile	Shearing forces	Young patients (usually associated with ACL ruptures)
Vertical radial tears	Area between the posterior horn and meniscal body	Axial load and rotation	Young patients (usually associated with a traumatic event)
Horizontal or cleavage ruptures	Start at the free edge and press towards the peripheral wall of the meniscus	Shearing forces	More frequent in degenerative knees and often associated with parameniscal cyst
Complex ruptures	Complex geometry and are most frequent in the posterior horn of both the medial and lateral meniscus		More frequent in degenerative knees (most frequent in patients over 40 years old)

51.5 Diagnosis

The diagnosis should include a clinical evaluation with a thorough physical examination and diagnostic imaging.

51.5.1 Medical History

Traumatic ruptures, which occur usually in younger patients, present with swelling and acute joint-line pain. Catching, "popping," or locking may also be present, suggesting a bucket handle tear that is displaced. The patients can always remember a precise trauma and complains about mechanical symptoms like pain during hyperflexion and twisting. On the other hand, degenerative meniscal tears, occurring in patients over 40 years old, usually present a history of mild swelling and joint-line pain without any clear trauma in their anamnesis. These ruptures are often associated with some degree of chondral degeneration. In this case, the patients always complain about both mechanical and inflammatory symptoms.

51.5.2 Physical Examination

Examination of the knee should begin with inspection of the skin and surrounding tissues. The knee should be examined for evidence of effusion. Range of motion (ROM) should be assessed and compared to the opposite side to evaluate possible mechanical locking. The ligamentous structures should be tested to determine whether concomitant ligament lesions are present. There are two specific clinical tests that have been described to assess meniscal lesions. The first is the McMurray test. To perform it, the examiner holds the knee with the patient supine and palpates the joint line with one hand. The thumb is on one side and fingers on the other, while the other hand holds the sole of the foot and acts to support the limb and provide the required movement through range. From a position of maximal flexion, extend the knee with internal rotation of the tibia and a varus stress. Then, return to maximal flexion and extend the knee with external rotation of the tibia and a valgus stress. With internal rotation of the tibia followed by extension, the examiner can test the entire

posterior horn to the middle segment of the meniscus. The anterior portion of the meniscus is not easily tested because the pressure to that part of the meniscus is not as great. The second specific test is the Apley grinding test. Here, the knee is flexed to 90 degrees with the patient in the prone position. The examiner holds the thigh fixed to the examining table with the examiner's knee. First off, the examiner rotates the tibia laterally and medially in combination with distraction while noting any excessive movement, restriction, or discomfort. Then, the process is then repeated using compression instead of distraction. If rotation plus distraction is more painful or shows increased rotation relative to the normal side, the lesion is most likely ligamentous. If rotation plus compression is more painful or shows decreased rotation relative to the normal side, the lesion is most likely a meniscus injury.

51.5.3 Diagnostic Imaging

51.5.3.1 Radiography

Standard knee radiographic films should include a PA weight-bearing view at 30° of flexion (Rosenberg view), a lateral view, a merchant or skyline view, and a weight-bearing long leg X-ray. These should be used to assess the alignment of the lower limb and osteoarthritic signs. The Rosenberg view assesses the posterior femoral condyles and will detect early osteoarthritic changes or joint collapse. The detection of joint degeneration or malalignment is important to detecting degenerative meniscal lesion and to directing towards a therapeutic choice.

51.5.3.2 Magnetic Resonance Imaging

MRI is a powerful tool in the assessment of knee pathology, but it should be analyzed in the context of the patient's history and examination. Some patients may show meniscal tears on MRI scans but are asymptomatic. It has been shown that 5.6% of the patients between 18 and 39 years of age with no knee complaints and a normal examination had a meniscus rupture on the MRI. The MRI can provide us with the precise location of the meniscal lesion and show us associated lesions (i.e., cartilage, ligaments). In a few cases, the MRI can miss small or peripheric meniscal lesions.

51.6 Treatment

51.6.1 Conservative Treatment

The conservative management of meniscus injuries includes symptomatic treatment with cryotherapy and NSAIDs as well as physiotherapy with muscle strengthening and working on maintaining adequate range of motion. Moreover, a loss of weight is suggested in cases of overweight, and a temporary correction of the alignment (braces, insoles) is useful in patients with varus or valgus knee. That has proven to be especially useful in patients with degenerative injuries. When considering treatment for degenerative meniscal tears, surgery should only be considered if the patient presents with knee locking and pain with rotation or with hyperflexion after a minimum of 6 months of conservative treatment. We can define these three as mechanical meniscal symptoms. Conservative treatment could also be employed for isolated traumatic lesions when they are incomplete or partial ruptures (<1 cm) and even in cases of meniscal root tears with intact meniscofemoral ligaments. In these cases, conservative treatment should include avoiding sports activity and deep knee flexion and symptomatic treatment.

51.6.2 Surgical Treatment

51.6.2.1 Meniscectomy

When removal of meniscal tissue (meniscectomy) is necessary, it must be limited to removing the unstable as well as highly degenerated tissue. Nowadays, the meniscectomy is performed arthroscopically through sharp forceps of different sizes and shapes (basket clamp). The arthroscopic shaver and radiofrequency are other

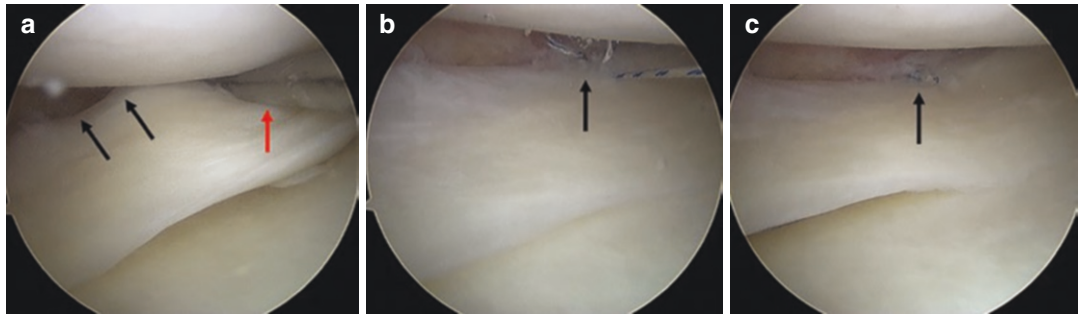

Fig. 51.3 (**a**) Longitudinal medial meniscal tear (showed by black arrows). A probe (red arrow) is evaluating the stability of the lesion and the quality of the tissue. (**b**) All-inside suture has been performed, the suture is retrieved, and the knot is pushed. (**c**) Vertical stitch completed

arthroscopic tools that can be used for a fine-tuning of the meniscal remnant once the unstable tissue is removed.

51.6.2.2 Meniscal Suture

There are different arthroscopic techniques and tools for meniscal suturing that are used, depending on the type and location of the lesion. In general, all-inside stitches are used for the posterior horn and for the posterior part of the meniscal body while the inside-out or outside-in suture is the preferred technique for the anterior part of the body and the anterior horn. The all-inside suture is so named because it allows to perform a suture without the need to pierce the patient's skin by using specific instruments and disposable implants through the arthroscopic portals. Implants penetrate the meniscus thanks to a needle and consist of a loop with a sliding knot. Through the needle, at the peripheral edge of the meniscus, we place a device made of rigid material, while the loop with its knot remains at the level of the articular surface of the meniscus. Once the suture is made, the knot can be tightened as needed and the suture cut with arthroscopic instruments. In the outside-in and inside-out techniques, needles and cannulas that allow the sutures to be passed in two different directions are used. They can be introduced, always under arthroscopic guidance, either through the skin towards the meniscus or from the meniscal surface towards the skin. In both techniques, there is a need for small cutaneous access points to retrieve and tighten the suture knots.

Regardless of the technique used, the sutures can be placed in three different ways according to the type of lesion being dealt with. They are vertical, horizontal, and crossed stitches (Fig. 51.3).

As regards the meniscal roots, special repair techniques must be used, given the anatomical peculiarity of this structure. It is necessary to place suture wires at the end of the root under repair with special arthroscopic instrumentation and then create a tunnel at the level of the anatomical insertion of the root itself. In this tunnel, the suture wires will be retrieved and passed through this tunnel in order to fix the root.

51.6.2.3 Meniscal Substitution

More recently, new treatments have been introduced to deal with postmeniscectomized painful knee. The aim is to replace the extirpated meniscal tissue in order to improve knee function and prevent progressive deterioration of the joint. The first possibility is to use meniscal implants that have been designed to replace partial meniscal defects. They serve as a scaffold for the ingrowths of new meniscal tissue, which eventually leads to a regeneration of the meniscal tissue lacking. The second possibility is to use meniscal allografts that are intended to replace a whole meniscus lost.

Take-Home Message

- Meniscal tears are frequent and can lead to biomechanical problems and consequently joint degeneration.
- Meniscal injuries can be caused by a knee trauma or developed because of joint degeneration.
- Meniscal lesion can be classified based on the pathogenesis (traumatic or degenerative), on the localization of the rupture (from red zone more vascularized to the white zone less vascularized), and on the morphology of the lesion (vertical, horizontal, oblique, root tear).
- A precise clinical evaluation and an MRI are enough for a correct diagnosis and to check the joint for associated lesions. An X-ray can be used to further investigate the joint status.
- Conservative treatment is suggested in cases of partial traumatic lesions or degenerative lesions.
- In cases of traumatic complete lesions or degenerative lesions associated with mechanical symptoms, a surgical approach is indicated.
- The surgical treatment aims to conserve as much meniscal tissue as possible. The meniscectomy should be restricted to unstable tissue. When the lesion is reparable, a meniscal suturing should be carried out.
- In case of symptomatic meniscal deficiency, meniscal substitution can be considered.

Summary

Meniscal tears can be roughly divided into traumatic and degenerative. Traumatic tears are common in young patients and are often associated with ligament injuries. The degenerative tears are found in older patients and are associated with joint degeneration. An MRI makes it possible to evaluate the location and type of lesion and evaluate any associated lesions at the same time. When joint degeneration is present, the surgical approach is not recommended. Therefore, a nonoperative approach is the best option. When faced with a traumatic tear or a degenerative tear with mechanical symptoms, the surgical approach should be considered. Unstable meniscal tissue located in a poorly vascularized area is suitable for removal. When the meniscal tear is in a well-vascularized area of the meniscus and the meniscal tissue is not degenerated, a suture is recommended. Independently of the technique of suturing chosen, the aim of a repair is to stabilize the meniscal rupture to allow for proper cicatrization.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Traumatic meniscal lesions are frequently associated with:
   (a) Genetic predisposition
   (b) Inflammatory diseases
   (c) Ligamentous knee lesions
   (d) Osteochondritis dissecans
2. Which is the initially recommended treatment for degenerative meniscal lesions?
   (a) Arthroscopy
   (b) Meniscectomy
   (c) Meniscal suture
   (d) Nonoperative treatment
3. Meniscal lesions may be classified by:
   (a) Morphology
   (b) The age of the patients
   (c) The shape of the meniscus
   (d) None of the above is correct
4. Which is not a meniscal suturing technique?
   (a) All-inside technique
   (b) Outside-in technique
   (c) Inside-out technique
   (d) Root repair technique
   (e) Single-stitch technique
5. Which meniscal area has the best chances of repair after a suture:
   (a) Red-white zone
   (b) Anterior meniscal root
   (c) Posterior meniscal root
   (d) Red-red zone

Further Reading

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Tendon Injuries

52

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Overview

This chapter explores the common presentations, pathoanatomy, classification, and management of tendon injuries around the knee joint. These injuries generally require a significant tensile force but can also occur as a result of chronic inflammation or systemic diseases causing weakened tendons. Timely diagnosis is critical to achieving best outcomes, which relies upon on good history taking, examination, and appropriate investigations.

52.1 Introduction

The main tendons around the knee are the quadriceps tendon (QT) and patella tendon (PT). The quadriceps tendon has contributions from four main muscles: rectus femoris, vastus lateralis, vastus medialis and vastus intermedius. It inserts into the superior pole of patella (Fig. 52.1). The patella tendon runs from the inferior pole of patella to its insertion at the tibial tuberosity. Together, QT, PT and patella form the extensor mechanism of the knee that allows for straight leg

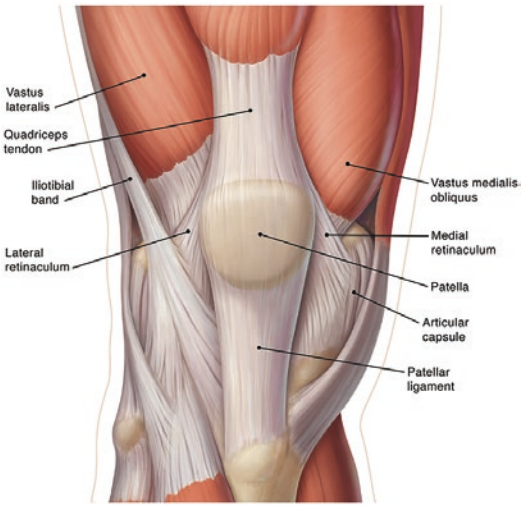

Fig. 52.1 The knee extensor mechanism is composed of the quadriceps muscle, quadriceps tendon, medial and lateral patellar retinaculum, patella, patellar tendon, and tibial tubercle. The quadriceps muscle is composed of four separate muscles with different origins but a common insertion point on the patella through the quadriceps tendon. The patellar tendon, by definition, is a ligament as it connects bone (patella) to bone (tibial tubercle). Source: © Todd Buck Illustration LLC.

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raise. These two tendons are also among the most commonly injured tendons in the body.

Hamstrings are a group of muscles located within the posterior compartment of the thigh. They consist of the biceps femoris (long and short heads) and semimembranosus and semitendinosus muscles, which form prominent tendons medially and laterally at the back of the knee. As a group, these muscles act to extend the hip and flex the knee joint.

Although both muscle groups cross the knee and hip, tendon injuries to the knee extensor mechanism (quadriceps) occur mostly distal about the knee, and injuries to the knee flexors (hamstring) occur proximally about the hip.

52.2 Extensor Mechanism Injuries

Injuries to the extensor mechanism are either usually a result of direct trauma in younger patients such as falls and lacerations or secondary to repetitive trauma resulting in tendon inflammation also termed tendonitis. However, in older patients, eccentric contraction of the extensor mechanism can cause tendon rupture, for example when the knee is flexed while the foot is firmly planted on the ground resulting in a fall.

52.2.1 Patella Tendon

A common term used to describe chronic PT injury is ‘jumper’s knee’. As the name implies, it affects athletes that participate in jumping activities with repetitive motion such as basketball, volleyball or track and field and can occur in up to 20% of the athletic population. This exerts continuous stress on the PT especially at its proximal end near its attachment on the patella. There is a slightly higher incidence in males than females and in patients under the age of 40. Jumper’s knee has been thought of as a pre-injury level state in patients who go on to rupture their PT.

52.2.2 Quadriceps Tendon

QT ruptures are more common than PT, and although they can happen in any age group, there is a higher incidence in patients older than 40 years. These are usually unilateral but can also be bilateral in rare cases. The incidence of QT rupture is up to eight times higher in males than females. The point of rupture is normally at the bone-tendon junction.

Complete rupture of the QT is common in older patients, which usually occurs as a result of tendon degeneration and as such is not seen commonly in athletic population. This is attributed to a change in collagen fibrils, as well as a poor vascular supply to these tendons in older population due to chronic condition such as diabetes. However, studies have shown that the vast majority of tendon ruptures have degenerative changes on histological examination. There are a number of risk factors that can lead to increased risk of rupture in certain patients with:

- Drugs:
  - Anabolic steroid use
  - Oral glucocorticoid treatment
  - Steroid injections
  - Antibiotics (fluoroquinolones)
- Chronic medical conditions
  - Diabetes mellitus
  - Rheumatoid arthritis
  - Systemic lupus erythematosus
  - Gout
  - Renal failure
  - Hyperparathyroidism
- Inherited disorders
  - Connective tissue disease
- Infection
- Tumours
- High BMI
- Chronic immobility

Less common causes include ruptures following total knee replacements or following patella or quadricep tendon autograft harvest for anterior cruciate ligament reconstruction.

52.2.3 Diagnosis

Patients with extensor mechanism injuries present with anterior knee pain with or without preceding trauma. There may be bruising, swelling and local tenderness. For both PT and QT, patients are unable to straight leg raise with a complete rupture and when asked to extend against resistance are unable to do so.

Primary investigations that are helpful in establishing the diagnosis include radiographs of affected knee in anteroposterior and lateral views. In PT rupture, the patella in lateral view would be sitting high—also termed patella alta. This can be interpreted when patella is sitting above the level of intercondylar notch. In QT rupture, patella would sit lower than normally, also termed patella baja. In some cases, taking a radiograph of contralateral knee can be helpful for comparison if diagnosis is unclear and magnetic resonance imaging (MRI) is not readily available. Ultrasound (US) scan is useful in establishing diagnosis but may be challenging in patients with elevated body mass index (BMI). MRI is very helpful in determining whether the tear is complete or incomplete especially in QT rupture.

52.2.4 Classification

There have been a number of classifications proposed for patella tendon rupture, but the most commonly used classification is the one that involves timing of rupture and repair by Siweck and Rao in 1981. If the injury and repair are within 2 weeks from the time of injury, then it is termed immediate, and if it occurs more than 2 weeks from the injury, it is termed chronic. Other classifications describe it based on the pattern at the time of surgery such as an inverted U shape, transverse and Z type, or it is based on the level of tendon rupture such as at tibial tubercle, mid-level or distal pole of patella. Quadriceps tendon rupture can be described as either partial or complete.

52.2.5 Treatment

Partial tendon tears with intact extensor mechanism can be treated non-operatively in a splint or plaster cast that prevents knee flexion followed by physiotherapy, which helps to restore muscle strength and joint function. Complete QT and PT tears on the other hand require early tendon repair followed by physiotherapy-led rehabilitation. Numerous techniques have been described in the literature for repair of acute rupture of the quadriceps and patellar tendons, which range from simple suture repair to wire-reinforced repairs or suture anchors. Patients who present with missed or neglected tendon ruptures sometimes require complex surgical reconstruction with biological or synthetic grafts to overcome tendon retraction and bridge any gap (Fig. 52.2). Acute repairs usually yield good outcomes, with most patients regaining full power, a good range of movement and little or no extensor lag, whereas late repairs can be less predictable.

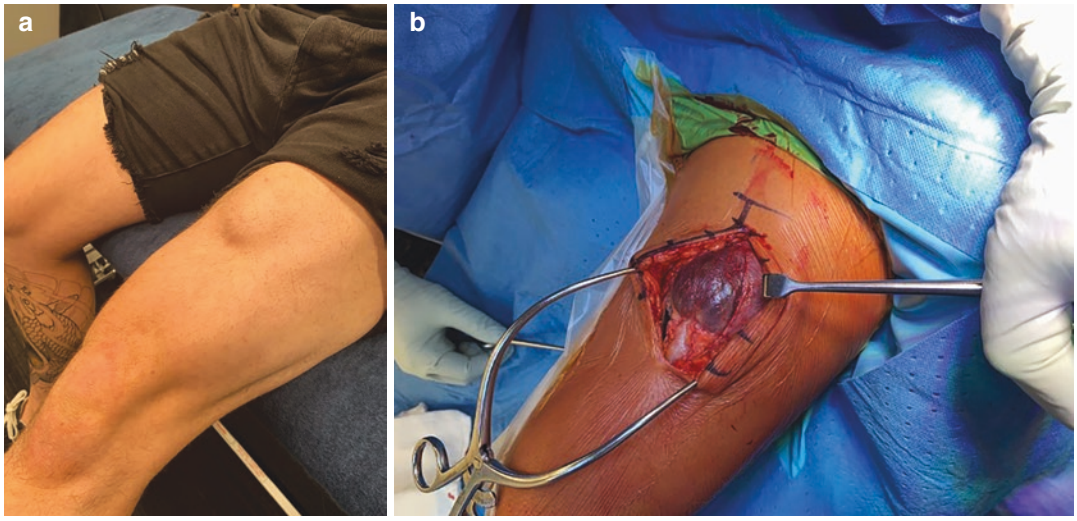

Fig. 52.2 The appearance of chronic quadriceps tendon rupture (**a**) on clinical examination and (**b**) intra-operatively, courtesy of Dr Peter D'Alessandro. 'Chronic' ruptures (usually the result of delayed presentations or missed diagnoses) are difficult to repair because the ends have retracted. The gap can be closed and residual defects covered with turn-down techniques, or augmentation techniques can be used, most commonly using autologous hamstring tendons

52.3 Hamstring Injuries

Injuries to the hamstring muscles can range from minor sprains and strains to complete tears with tendon retraction. Proximal hamstring tears are among the most common athletic injuries that occur as a result of eccentric muscle contraction caused by forceful hip flexion with the knee held in extension. Literature suggests that more than 90% of these injuries occur without any contact, for example in water skiers who experience sudden and unanticipated pull from the boat during a water start (Fig. 52.3a). In adults, the most commonly injured point is the proximal muscle-tendon junction. In adolescents, an avulsion fracture of the ischial tuberosity (the site where the hamstring tendons attach) is more likely.

52.3.1 Diagnosis

Patients usually present with disabling pain in the posterior region of the thigh. Those with tendinopathy report a gradual onset of pain in the

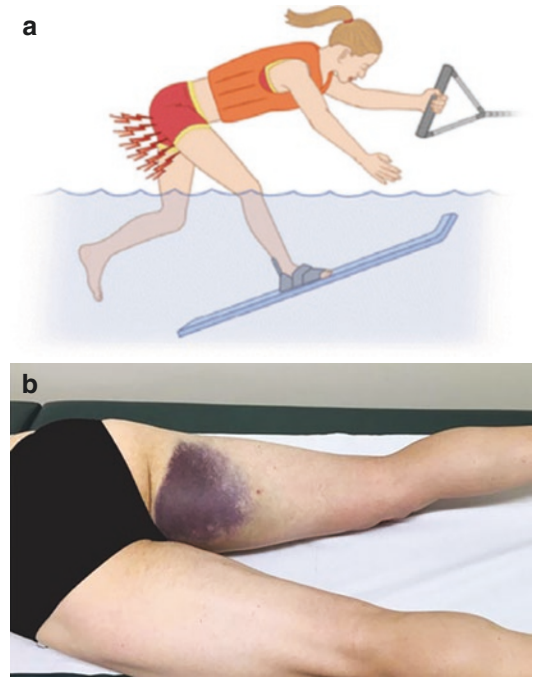

Fig. 52.3 (**a**) Possible genesis of trauma. (**b**) The typical appearance of posterior thigh bruising on clinical examination in a patient with acute hamstring rupture. Image courtesy of Dr Peter D'Alessandro

lower gluteal region, radiating along the hamstrings, which is exacerbated by repetitive hip flexion or even long periods of sitting. Examining the patient in prone position may reveal bruising and swelling in the posterior thigh (Fig. 52.3b) along with weakness of knee flexion and hip extension against resistance. In cases of tendon rupture, there may also be a palpable defect in the mid-thigh hamstring musculature. Sciatic nerve irritation symptoms may be present. Often, the diagnosis can be made clinically; however, MRI is very useful in determining the level of injury and number of tendons involved. If a high level of suspicion exists for a proximal hamstring injury, plain radiographs of the pelvis and a lateral radiograph of the affected hip should be obtained to rule out any osseous avulsions, particularly to the ischial apophysis in adolescent athletes.

52.3.2 Classification

Injuries to the hamstring muscle complex can be broadly separated into injuries of the muscle belly and those of the proximal tendon. Proximal tendon injuries can be further classified (Wood et al. 2008) according to the anatomical location of the injury (osseous or musculotendinous), the degree of avulsion (incomplete or complete) and the degree of muscle retraction. This classification is useful as it can assist in determining the indications for surgical management.

52.3.3 Treatment

Most hamstring tendon injuries are either strains or partial tears at the proximal muscle-tendon junction that can be managed conservatively and generally result in full recovery. Treatment is guided by the number of tendons involved, presence or amount of tendon retraction and other patient-related factors (i.e. general medical health and activity level). Proximal avulsion of the hamstring origin from the ischial tuberosity occurs less frequently, and the available literature supports early operative treatment in order to avoid long-term functional disability and to allow a return to sporting activity.

Take-Home Message

- Quadriceps tendon rupture is more common than patellar tendon rupture and usually occurs in patients >40 years of age.
- Patellar tendinitis, sometimes referred to as ‘jumper’s knee’, is most common in people who participate in activities that require running or jumping.
- Hamstring injuries usually occur in running athletes as a result of sudden hip flexion and knee extension.
- Small tears may be treated with rest and splinting, followed by physiotherapy. Larger tears typically require surgery within a couple of weeks.

Summary

Tendons are strong cords of fibrous tissue that attach muscles to bones.

The main tendons around the knee are the quadriceps tendon (QT) and patella tendon (PT).

Injuries to the extensor mechanism are either usually a result of direct trauma in younger patients such as falls and lacerations or secondary to repetitive trauma resulting in tendon inflammation also termed tendonitis.

A common term used to describe chronic PT injury is ‘jumper’s knee’.

Primary investigations that are helpful in establishing the diagnosis include radiographs of affected knee in anteroposterior and lateral views.

Partial tendon tears with intact extensor mechanism can be treated non-operatively in a splint or plaster cast that prevents knee flexion followed by physiotherapy, which helps to restore muscle strength and joint function. Complete QT and PT tears on the other hand require early tendon repair followed by physiotherapy-led rehabilitation.

Injuries to the hamstring muscles can range from minor sprains and strains to complete tears with tendon retraction.

Patients usually present with disabling pain in the posterior region of the thigh. Those with tendinopathy report a gradual onset of pain in the lower gluteal region, radiating along the hamstrings, which is exacerbated by repetitive hip flexion or even long periods of sitting.

Most hamstring tendon injuries are either strains or partial tears at the proximal muscle-tendon junction that can be managed conservatively and generally result in full recovery. Treatment is guided by the number of tendons involved, presence or amount of tendon retraction and other patient-related factors (i.e. general medical health and activity level).

Questions Multiple correct answers are possible. Answers available in the book back matter.

1. Which of the following is not a part of the quadriceps muscle?
   (a) Rectus femoris
   (b) Biceps femoris
   (c) Vastus intermedius
   (d) Vastus medialis
2. The following conditions are all risk factors for patella tendon rupture except
   (a) Ehlers-Danlos syndrome
   (b) Diabetes mellitus
   (c) Hyperlipidaemia
   (d) Rheumatoid arthritis
3. Which of the following radiological features is a hallmark of a complete quadriceps tendon rupture?
   (a) Patella alta
   (b) Patella baja
   (c) Lipohaemarthrosis
   (d) Segond fracture
4. Which of the following clinical findings is not suggestive of an acute quadriceps rupture?
   (a) Inability to extend the knee against gravity
   (b) Swelling
   (c) Ecchymosis
   (d) Palpable gap in the tendon, distal to the inferior pole of patella
5. Which of the following statements regarding the hamstring muscles is false?
   (a) All arise from the ischial tuberosity
   (b) They flex the knee joint and extend the hip joint
   (c) The popliteal tendon is an extension of the biceps femoris
   (d) In adults, the most commonly injured point is the musculotendinous junction

Further Reading

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Knee Ligament Injuries

53

Anna Lundeen, Robert F. LaPrade,
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Overview
This chapter provides an overview of knee ligament injuries, an understanding of which is crucial for proper evaluation and treatment of both the general public and athletic population.

53.1 Isolated Ligament Injury

53.1.1 Anterior Cruciate Ligament

53.1.1.1 Anatomy

The anterior cruciate ligament (ACL) consists of two bundles, anteromedial (AM) and posterolateral (PL), which originate on the medial surface of the lateral femoral condyle (LFC) and insert on the tibia immediately posterior to the ACL tubercle and ridge. There is significant overlap of the ACL and the anterolateral meniscal root (Fig. 53.1). More specifically, the AM bundle attaches just proximal to the bifurcate ridge and posterior to the lateral intercondylar ridge (LIR), both of which are useful arthroscopic landmarks on the femur. The tibial attachment of the AM bundle constitutes the entire anterior boundary of the ACL, inserting between the retro-eminence and ACL ridge. The PL bundle originates distally beyond the bifurcate ridge and posterior to the LIR of the femur and inserts medial to the anterior and posterior horns of the lateral meniscus.

53.1.1.2 Function and Injury

The ACL prevents anterior translation and internal rotation of the tibia. Noncontact mechanisms of injury (MOI) are most common (70%) and include pivoting with the foot planted or landing from a jump with the knee in an extended position. Intrinsic risk factors for ACL tears include femoral intercondylar notch stenosis, large lateral tibial slope, and large beta angle (angle between the long axis of the femur and Blumensaat line). Poor lower extremity biomechanics also increase the risk of ACL tear, highlighting the importance of quality prophylactic and perioperative ROM and strengthening exercises. The Lachman test assesses the integrity of the ACL (Fig. 53.2).

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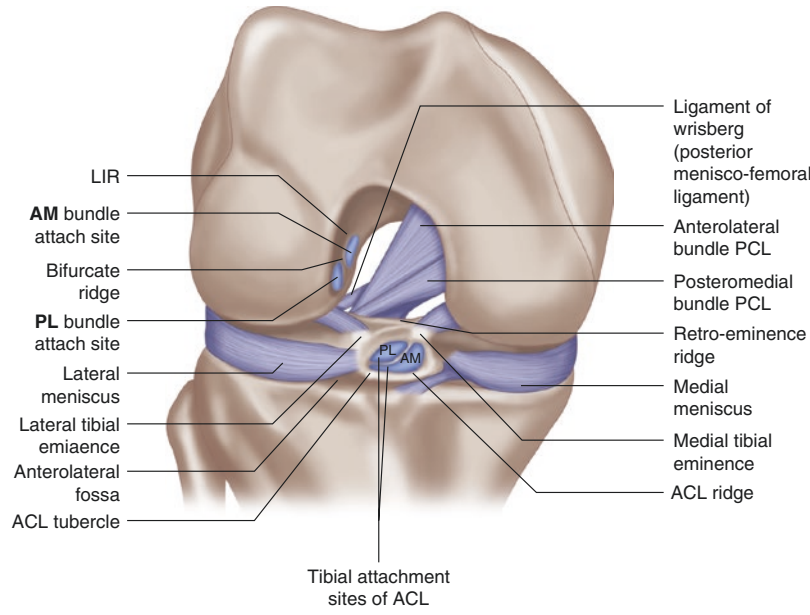

Fig. 53.1 AM and PL bundle attachment of the ACL. *AM* anteromedial, *PL* posterolateral, *ACL* anterior cruciate ligament, *PCL* posterior cruciate ligament, *LIR* lateral intercondylar ridge

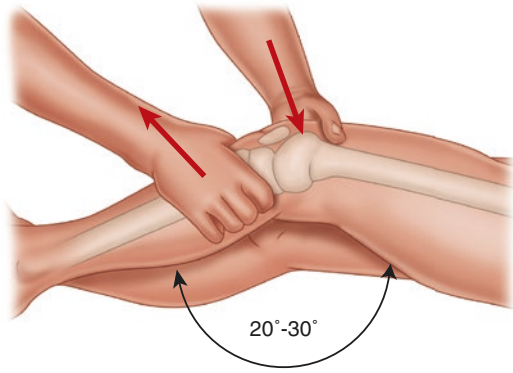

Fig. 53.2 Lachman test for ACL

53.1.1.3 Treatment

Returning to cutting and pivoting sports with a partial ACL tear coincides with a significantly increased risk of a subsequent complete ACL tear. Complete ACL tears warrant a discussion regarding operative treatment, which improves symptomatology and quality of life for those who aim to return to jumping, pivoting, and cutting activities. A delay in surgery can lead to increased cartilage and meniscus damage, resulting in an increased risk of post-traumatic osteoarthritis.

Nonoperative treatment for isolated ACL tear is an option for those planning to return exclusively to straight plane activities; however, current research is lacking in long-term clinical outcomes, and where concurrent meniscus and collateral ligament injuries are involved. There has been increased interest in ACL repairs due to quicker return to previous activity level; however, there is a paucity of reliable data as to its effectiveness.

Bone-patellar tendon-bone (BTB) autograft is the gold standard ACL graft option. Harvesting a portion of patellar tendon can result in increased anterior knee pain, although patients with BTB autografts have significantly improved pivot shift outcomes and increased rates of return to preinjury level. Another option is the hamstring tendon (HT) autograft, which is reserved for carefully selected patients. The quadriceps tendon (QT) autograft is a third option as it supplies ample viable tissue. Soft-tissue allografts are not desired due to high graft failure rates in young and active populations.

Five-year ACL reconstruction (ACLR) survival rates are above 95 percent. ACLR significantly improves the Knee injury and Osteoarthritis Outcome Score (KOOS), with smokers reporting significantly inferior KOOS than nonsmokers. Although ACL grafts have high survival rates,

grafts may fail and need to be revised. The following factors are associated with increased revision rates: HT autografts, younger age at surgery, and use of allografts in patients younger than 25 years of age.

53.1.2 Pediatric ACL Injury

53.1.2.1 Diagnosis

Three basics of pediatric ACL assessment include evaluation of hemarthrosis, comparison to the contralateral knee due to increased joint laxity in children, and recognition of pediatric-specific injuries. Plain radiographs are a good starting point, followed by confirmation with magnetic resonance imaging (MRI).

53.1.2.2 Management

Consistent and frequent preventative exercises are important and simple to integrate into training. Proper body position of fundamental movements, such as cutting and landing techniques, is paramount for avoiding ACL injury. A presurgical rehabilitation phase with bracing is acceptable and should be aimed at improving ROM, strength, and inflammation.

Bracing may continue for a short time postoperatively, as gaining early postoperative neuromuscular control of the lower extremity is important. Postponing surgical treatment of pediatric ACL tears can result in an increased risk of meniscus and cartilage damage and is not generally recommended. Indications for surgery include the presence of concomitant knee injury, recurrent instability episodes, and an unacceptable modified activity level. It is important to consider open physes when planning the surgical approach. Autografts are preferable to allografts for pediatric ACLRs due to poor allograft outcomes. Grafts with a diameter of 7 mm or smaller are at greater risk for graft rupture, and HT autografts fail at a significantly greater rate than BTB autografts. Regardless, HT autografts remain the most commonly used graft in pediatric ACLR, since harvesting BTB autografts may disrupt open tibial tubercle growth plates and bone plugs cannot cross open growth plates.

53.1.3 Posterior Cruciate Ligament

53.1.3.1 Anatomy

The posterior cruciate ligament (PCL) consists of two bundles, the anterolateral bundle (ALB) and posteromedial bundle (PMB). The ALB is tightest at 90 degrees of knee flexion, attaching to the femur between the trochlear point, medial arch point, and medial bifurcate prominence. The PMB is most tense at end-range knee flexion and extension and originates on the femur distal to the medial intercondylar ridge and posterior to the femoral attachment of the ALB. The ALB inserts on the tibia below the articular surface, between the posterior portions of the medial and lateral tibial plateaus and anterior to the PMB insertion. Arthroscopically, the distal border of the PMB is marked by the champagne-glass drop-off, where the joint capsule separates the popliteus muscle and intra-articular structures.

53.1.3.2 Function and Injury

As the largest and strongest ligament of the knee, the PCL functions as the primary posterior stabilizer and restricts internal rotation of the tibia. Common mechanisms of injury include motor vehicle accidents, landing directly on the flexed knee, hyperflexion, and dislocations of the tibiofemoral joint. The PCL is commonly injured concomitant to the medial collateral ligament (MCL), ACL, or posterolateral corner. While MRI has high diagnostic accuracy for a suspected acute PCL injury, posterior stress radiographs are fundamental for objective evaluation of PCL tears. Complete PCL tears usually have 8 mm or more of increased posterior tibial translation compared to the normal contralateral knee. Posterior stress radiographs involve kneeling on a horizontal support, leaving the femoral condyles and patella unsupported, and prove to be a reliable and reproducible test.

53.1.3.3 Treatment

Grade I and II PCL injuries can be treated nonoperatively with dynamic force bracing, which alters the applied force based on the knee angle

in attempts to match anatomic forces. Grade III PCL tears, a rupture of both bundles, require surgical reconstruction. The goal of the single-bundle PCL reconstruction (SB PCLR) is to reconstruct the ALB fibers. The double-bundle PCLR (DB PCLR) is superior to SB PCLR, resulting in improved objective stability with the goal of restoring the native ligament function.

Outcomes for the DB PCLR technique are comparable to ACLR outcomes. The anatomic double-bundle technique results in significant subjective and objective improvement compared to the preoperative state. Compared to chronic DB PCLRs, acute DB PCLRs result in significantly improved activity and satisfaction scores.

53.2 Multiligament Injury

53.2.1 Knee Dislocations

Some define knee dislocations (KDs) as tearing of both the ACL and PCL, with or without involvement of posteromedial and posterolateral corner structures. Knee dislocations are ranked by the Schenck classification system:

- KD I: Injury to a single cruciate ligament plus collaterals
- KD II: Injury to ACL, PCL, intact collaterals
- KD III M: Injury to ACL, PCL, and MCL
- KD III L: Injury to ACL, PCL, and fibular collateral ligament (FCL)
- KD IV: Injury to ACL, PCL, MCL, and FCL
- KD V: Dislocation plus fracture

Common mechanisms of injury include skiing, motor vehicle accidents, falls from significant heights, and other recreational activities. Meniscus and cartilage injuries are associated with knee dislocations. The risk of cartilage injury increases significantly with meniscus injury and chronic dislocations. The common peroneal nerve and popliteal artery may also be compromised with these injuries.

Limb vascular status should be assessed immediately. If compromised, emergent treatment involves reduction and splinting. Emergent surgery is performed in cases of open injury, vascular compromise, irreducible dislocation, or compartment syndrome.

Knee joint reconstruction/repair is initiated once vascular status has been found to be normal or improved. The optimal surgical treatment window is far enough from the date of injury to allow for decreased inflammation, partial ROM recovery, continued vascular monitoring, and improvement of quadriceps muscle tone (ideally <14 days). Acute treatment is common and results in improved subjective and objective outcomes. Delaying such a procedure greater than 3 weeks may result in increased scarring.

The goal of a multiple-ligament knee surgery is to fix all pathology within a single surgery, allowing for 9–12 months of postoperative rehabilitation before resuming full activity. Long-term outcomes of knee dislocation include significantly increased risk of osteoarthritis, especially for patients who were of increased age at the time of injury. Concurrent cartilage injury results in significantly worse function and symptomatology.

53.2.2 ACL- and PCL-Based Multiligament Reconstruction

These reconstructions treat injuries resulting from grade III tears of at least two out of four major knee ligaments without a knee dislocation (ACL, PCL, FCL, superficial MCL (sMCL), or a complete posterolateral knee injury). Single-stage reconstruction of all injured knee ligaments paired with immediate postoperative rehabilitation significantly improves outcomes and function, achieving low complication rates. Early postoperative rehabilitation improves knee ROM without stretching the fresh grafts.

53.2.3 Posterolateral Corner

53.2.3.1 Anatomy

The posterolateral corner (PLC) of the knee consists of three primary static structures: the FCL,

popliteus tendon, and popliteofibular ligament (PFL). The FCL originates on the femur, proximal and posterior to the lateral epicondyle, and inserts on the fibular head, posterior to the anterior margin and distal to the fibular styloid process apex (Fig. 53.3). The popliteus tendon courses laterally from the popliteus muscle and inserts on the lateral femur anterior to the FCL attachment. The PFL originates from the myotendinous junction of the popliteus and runs distolaterally splitting into anterior and posterior divisions, which insert on the anteromedial and posteromedial downslope of the fibular styloid process, respectively.

53.2.3.2 Function and Injury

The FCL, popliteus tendon, and PFL work together to resist varus stress, external rotation, and posterolateral translational joint movement. The FCL is the primary stabilizer against varus forces at low degrees of knee flexion, and the popliteus tendon and PFL are responsible for stabilizing the knee at higher degrees of flexion. Most commonly, PLC injuries are combined with ACL and/or PCL injuries, resulting from a direct blow to the anteromedial knee causing varus stress or hyperextension.

Varus stress radiographs play an essential role in the vital recognition of PLC injuries. This method of evaluation measures gapping between the distal-most portion of the LFC subchondral surface and the lateral tibial plateau. Varus stress radiographs are repeatable and reproducible, and the extent of suspected injured tissue correlates with the amount of lateral joint gapping compared to the healthy knee. Suspected injured tissue and the corresponding increase in gapping compared to the intact state are as follows: isolated FCL tear (2.7 mm), grade III PLC injury (4.0 mm), combined PLC and ACL injury (6.6 mm), and combined PLC, ACL, and PCL injury (7.8 mm).

Bone bruising occurs frequently with PLC injury and is commonly found on the anteromedial femoral condyle and anteromedial tibial plateau. These patterns may fluctuate based on the

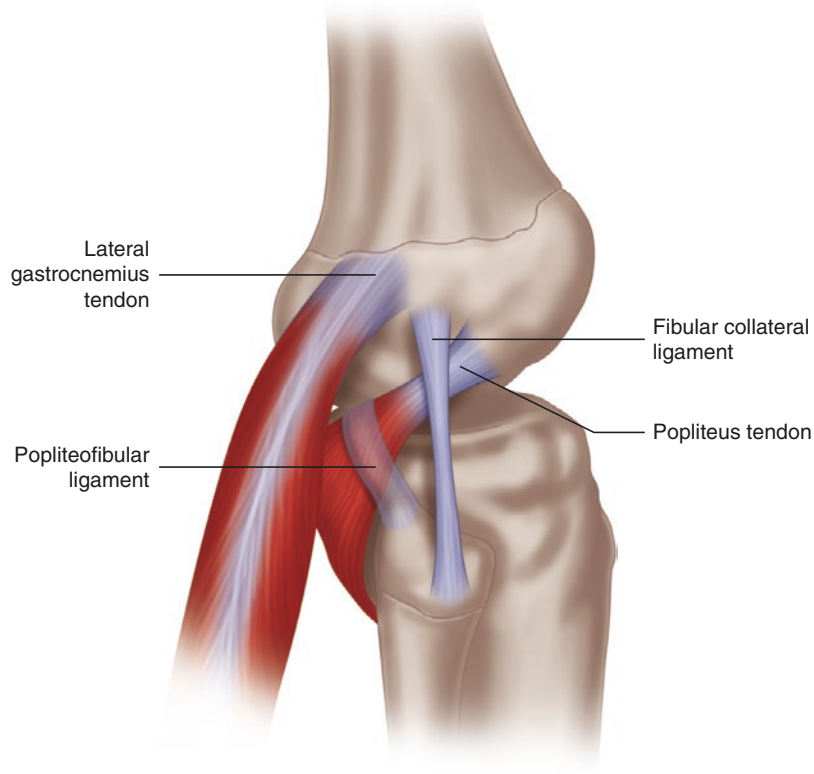

Fig. 53.3 Illustration of the posterolateral corner of the knee

structures injured. If the PLC injury is combined with an ACL tear, a bone bruise may present on the posteromedial tibial plateau.

53.2.3.3 Treatment

Grade III PLC injuries are treated operatively and acutely (<2 weeks) if possible. Lack of surgical correction results in poor biomechanics and outcomes. Structures should be assessed to determine if a repair or reconstruction is most appropriate. A combination of repair and reconstruction of posterolateral avulsed and midsubstance tears, respectively, results in significant objective improvement in stability.

For the first 2 weeks, rehabilitation is guided by a "safe zone" of ROM established intraoperatively (normally 0–90 °). A combination of repair and reconstruction paired with early protected range of motion improves objective outcomes of PLC treatment.

Table 53.1 Quantified medial joint line gapping compared to the intact state at 20° knee flexion with clinician-applied valgus stress

Average medial gapping increase compared with intact state	
	Medial joint line gapping
Ligament state	Increase (mm)
Intact	–
Proximal sMCL tear	3.2
Complete medial knee tear	9.8
Complete medial knee and ACL tears	13.8
Complete medial knee, ACL, and PCL tears	20.4

Adapted from LaPrade RF, Bernhardson AS, Griffith CJ, Macalena JA, Wijdicks CA. Correlation of Valgus Stress Radiographs with Medial Knee Ligament Injuries: An in Vitro Biomechanical Study. *Am J Sports Med*. 2010;38(2):330-338

sMCL superficial medial collateral ligament, *PCL* posterior cruciate ligament, *ACL* anterior cruciate ligament

53.2.4 Posteromedial Corner

53.2.4.1 Anatomy

The three main static stabilizers of the medial knee are the sMCL, deep MCL (dMCL), and posterior oblique ligament (POL). The sMCL is the largest and primary medial stabilizing ligament. It attaches proximal and posterior to the medial femoral epicondyle and inserts on the tibia proximally on the anterior arm of the semimembranosus tendon and distally on bone along the floor of the pes anserine bursa. The dMCL is a thickening of the medial joint capsule and has meniscotibial and meniscofemoral components. The dMCL originates distal to the sMCL femoral attachment and inserts distal to the articular surface of the tibia. The POL is a thickening of the posteromedial joint capsule and consists of three arms: superficial, central, and capsular. The central arm is the primary POL structure and should be repaired or reconstructed with a posteromedial corner (PMC) injury.

53.2.4.2 Function and Injury

The sMCL is the primary stabilizer against valgus and external rotation forces, while the POL resists internal rotation near terminal extension. When the PMC is damaged, strain on the ACL and PCL increases due to anteromedial rotatory instability. Common mechanisms of injury include valgus stress of the knee and forced tibial external rotation. Complete medial knee injury is defined as combined sMCL, dMCL, and POL tears. Grade III PMC injuries commonly result in concomitant cruciate ligament injury. Injury to the medial knee structures may present with a lateral compartment bone bruise.

Valgus stress radiographs are a reproducible technique used to detect PMC injury. Increased medial joint gapping is present at 20 degrees of knee flexion with a complete MCL tear (an increase of 3.2 mm or more) and gaps to a greater degree with complete medial knee injuries (9.8 mm increase compared to the intact state) (Table 53.1).

Grading medial knee injuries has been historically based on the amount of subjective joint gapping with valgus force. However, transected structures and their corresponding instability warrant suspicion of medial knee injury at lower degrees of joint gapping, highlighting importance of objective stress radiographs and grading based on the number of structures torn.

53.2.4.3 Treatment

Isolated PMC injuries are treated nonoperatively with 5–7 weeks of bracing and physical therapy. Surgical repair is indicated for grade III multiligament PMC injuries and sMCL injury with severe rotational instability or valgus gapping in knee extension. Reconstruction of the sMCL and POL is indicated in acute, midsubstance nonrepairable injury, and chronic injury with increased medial gapping, side-to-side instability, and external rotation on both the dial and anteromedial drawer tests. Valgus alignment should be evaluated and corrected in chronic PMC injuries to avoid PMC graft failure. Recreating the anatomic insertion points and the two sMCL functional arms involves use of two separate grafts for the sMCL and POL. Anatomic MCL reconstruction improves side-to-side instability, medial joint gapping, and rotatory instability. Arthrofibrosis and quadricep atrophy are prevented with immediate postoperative ROM guided by a "safe zone" established intraoperatively.

Take-Home Messages

- Reconstruction of the ACL is especially important for those planning to return to pivoting sports. Prevention exercises are important to implement early and often.
- PCL integrity can be reliably assessed with posterior stress radiographs, and DB PCLR successfully restores objective stability.
- It is important to address multiligament knee injuries with acute, single-stage procedures and early postoperative ROM.
- Varus stress radiographs are crucial for objective identification of PLC injuries. A combination of repair and reconstruction of PLC structures leads to improved objective outcomes.
- The PMC can be objectively evaluated with valgus stress radiographs. Surgical correction of high-grade multiligament PMC injuries results in improved objective outcomes.

Summary

Prompt and accurate identification of knee ligament injury is important for the long-term health and well-being of patients. Knowledge of ligament anatomy and function is necessary for proper treatment and rehabilitation.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Which factors put an adult at increased risk for an ACL tear?
   (a) Increased lateral tibial slope
   (b) Wide femoral intercondylar notch
   (c) Poor lower extremity biomechanics
   (d) A and C
   (e) All of the above
2. A patient presents to the clinic after slipping on a dock the past weekend. Patient states that she felt immediate pain on the medial side of her knee. She reports that her knee feels "unstable" and falls in toward her contralateral knee while weight-bearing. Upon examination, there is a contusion present with moderate edema and trace effusion. The patient is point tender to palpation over the proximal medial tibia, medial joint line, and just proximal to the medial femoral epicondyle. Initial set of radiographs are negative for fracture. What should be the clinician's next course of action?
   (a) Obtain a computed tomography (CT) scan
   (b) Obtain valgus stress radiographs
   (c) Obtain magnetic resonance imaging (MRI)
   (d) Put the patient in a long leg splint
3. Which ACL graft type is most commonly used for pediatric ACL reconstruction?
   (a) Soft-tissue allograft
   (b) Hamstring tendon autograft
   (c) Quadriceps tendon autograft
   (d) Patellar tendon autograft
4. In regard to ACL reconstruction, the soft-tissue allograft:
   (a) Is appropriate for use in ACL reconstruction for college-aged soccer players

(b) Is associated with increased postoperative anterior knee pain
(c) Is associated with high graft failure rate in the young and active population
(d) Is the most reliable, gold standard treatment for ACL reconstruction

5. Which of the following should be considered with a multiligament knee reconstruction?
   (a) Aim to repair and reconstruct all structures in a single-stage procedure
   (b) Early controlled postoperative range-of-motion exercises
   (c) Wait at least 4 weeks following the injury to operate to ensure that swelling has decreased
   (d) Both A and B
   (e) All of the above
6. In the posterolateral corner of the knee, which structure(s) is/are most restrictive against varus stress at near-terminal knee extension?
   (a) Popliteus tendon
   (b) Popliteofibular ligament (PFL)
   (c) Fibular collateral ligament (FCL)
   (d) All three (PFL, FCL, and popliteus tendon) provide the same amount of restriction to varus stress at this degree of knee extension
7. Which knee ligament(s) can withstand the greatest force?
   (a) Posterior cruciate ligament (PCL)
   (b) Anterior cruciate ligament (ACL)
   (c) Superficial medial collateral ligament (sMCL)
   (d) The posterolateral bundle of the ACL
   (e) A and B are equally strong

Further Reading

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Patellofemoral Pathology

54

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Overview

The patellofemoral joint is a unique and complex structure consisting of static elements (bones and ligaments) and dynamic elements (neuromuscular system). This implies that alterations at any of this level can cause changes in joint function. Anatomical factors (e.g., trochlear dysplasia), functional factors (e.g., muscle-tendon alterations), or traumatic events (e.g., medial patellofemoral ligament tears) can lead to the appearance of pathological changes.

54.1 Definition

It is a group of pathological disorders of the patellofemoral joint consisting of pain, subjective and objective instability, and consequent chondral lesions and functional impotence.

54.2 Epidemiology

Patellar dislocation accounts for approximately 2–3% of all knee injuries. This injury tends to affect young active individuals, with adolescent females and athletes at a higher risk. The incidence of patellofemoral instability is reported as 5.8 per 100,000 but could be as high as 29 per 100,000 in the adolescent population. Patellofemoral pain (PFP) syndrome is one of the most common causes of knee pain. It affects approximately 25% of the physically active population, with females being two to three times more likely to develop PFP compared to their male counterparts. PFP syndrome is diagnosed in up to 7.3% of all patients seeking medical care within the United States. Cartilage overload is an important issue even though it is asymptomatic most of the time. Up to 18% of all patients undergoing knee arthroscopy have cartilage lesions at the patellofemoral joint. This percentage doubles if we only consider professional sports patients.

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54.3 Etiology-Pathogenesis

Patellofemoral instability—It appears when the static and dynamic joint stabilizers cannot keep the patella centered on the trochlear groove during flexion and extension of the knee. In most cases, this happens due to a congenital predisposition-defined dysplasia of the extensor apparatus. This consists of the presence of anatomical alterations of different degrees in the entire control system of the patella sliding on the distal femur. The most frequent are dysplasia of the trochlear groove and incorrect positioning of the patella with respect to the groove itself. There are also less frequent alterations such as axial or rotational deformities of the femur and tibia that can contribute to the genesis of instability. These factors inherently increase patellar instability by generating continual stress on the static stabilizers of the patella that try to counteract the tendency of the patella to dislocate. In particular, the medial patellofemoral ligament is subjected to significant stress until its rupture leads to the definitive lack of stability control and to the genesis of permanent recurrent instability. Finally, there are patella instabilities in the absence of congenital predispositions due to a traumatic rupture of the medial patellofemoral ligament, the most important static stabilizer at the level of the patellofemoral joint. These are more uncommon and always linked to displacement trauma. In all cases of instability, a contribution also comes from the dynamic stabilizers given that the quadriceps and the gluteal muscles substantially contribute to the stabilization of the patellofemoral joint. In the event of minimal alterations predisposing to instability, good muscle control may be sufficient to effectively control patellofemoral tracking. On the contrary, patients without anatomical predispositions to instability but with a marked muscular hypotrophy can show symptoms of micro-instability.

PFP syndrome—Anterior knee pain is a complex pathology to be analyzed as its pathogenesis is multifactorial and still not fully understood. Once again, a fundamental role is played by the anatomical alterations that can cause modifications in patellofemoral tracking. That means the way the patella slides on the femur. If the patella is not perfectly centered in the trochlear groove like a train on a rail, the distribution of tangential forces at the level of the cartilage layer will not be homogeneous. This can cause pain due to overload on the cartilage surface, inflammatory states of the cartilage (chondritis), or eventual degeneration of the cartilage layers in the chronic phase of the disease. Like for the instability, the patella in high and lateralized positions with respect to the center of the trochlear groove are the two most frequently encountered alterations in the case of anterior knee pain. In some cases, no anatomical variations are found. In those cases, the pain can be considered as resulting from functional alterations due to biomechanical variations. Those variations can be correlated with altered load distribution at the level of the knee as in the case of a valgus knee or pronated foot but also with muscle weakness in the quadriceps and buttocks, important dynamic stabilizers of the joint.

54.4 Classification

Patellofemoral disorders are classified as follows:

- Patellofemoral pain: anterior knee pain without primary anatomical predisposing factors, usually linked to joint overload or muscular weakness associated or not with secondary anatomical predisposing factors
- Potential patellar instability: anterior knee pain associated or not with subjective instability without any episode of patellar dislocation but in the presence of at least one of the primary predisposing factors
- Objective patellar instability: at least one episode of patellar dislocation associated with at least one of the primary predisposing factors

Primary predisposing factors: trochlear dysplasia, high patella, lateralized patella (in relation to the trochlear groove) measured by the tibial tuberosity-trochlear groove (TT-TG) distance, and patellar tilting (exaggerated external rotation on the axial plane).

Secondary predisposing factors: excessive femoral anteversion, excessive tibial torsion, genu recurvatum, and static or dynamic (linked to foot pronation) valgus lower limb axis.

Trochlear dysplasia: classified by Dejour in 4° (A–D) based on the shape of the trochlear groove evaluated in an axial and sagittal view (any kind of radiological imaging). The degree of dysplasia is inversely proportional to the congruence of the trochlear groove with the patella.

Patellar height: there is no classification, but a normal range, which the value is considered pathological, has been identified above. Considering the Caton-Deschamps index, we consider a high patella value greater than 1.2.

Tibial tuberosity-trochlear groove (TT-TG) distance: as for the patellar height, the normal reference values are indicated. The average distance in the general population is 11 mm, 11–20 is considered paraphysiological, and above 20 mm is most certainly pathological.

54.5 Diagnosis (Clinical and Imaging)

Clinical—The patient should be examined carefully, being on the lookout for any pathological signs at the knee (swelling, lack of ROM, instability, etc.) and any kind of deformity of the lower limbs both rotational and in the axis of the lower limb. It is possible to evaluate evident patellofemoral maltracking with abnormal sliding of the patella on the distal femur by checking for the J sign. It is the rapid movement of the patella during an active extension that resembles this letter. Upon trying to move the patient's patella laterally, we might provoke the apprehension test, with the patient reporting a sensation of imminent dislocation of the patella during this gesture. This indicates a tendency of the patella to dislocation. Finally, it is possible to bring on patellofemoral pain. To evaluate that, the tests are used to increase the pressure of the patella over the femoral trochlea and to assess whether the patient reports pain or not. It can be easily performed with the patient supine by applying continuous pressure with the palm of the hand against the bony surface of the patella and first moving the patella in a cranio-caudal direction and then in a mediolateral direction (Ribot test). The patella can also be subluxed for direct palpation of both its facet joints to determine the origin of the pain.

Imaging—Plain and weight-bearing radiographs are essential to assess knee or load axis deformities of the lower limb. By performing a precise lateral radiograph, it is possible to evaluate the shape of the femoral trochlea and classify it according to the Dejour classification (Fig. 54.1). Again, in a lateral view, it is possible to evaluate the height of the patella. Different evaluation methods have been described. The three most famous are the Insall-Salvati index, the Caton-Deschamps index, and the Blackburne-Peel ratio. The most reliable and simple to perform is the Caton-Deschamps index. It consists of the ratio between the measurement of the distance from the most anterior edge of the tibial plateau to the point where the articular cartilage of the patella ends and the measurement of the length of the articular surface of the patella itself (Fig. 54.2).

A CT scan will be necessary to evaluate torsional alterations of the femur and tibia, the tibial tuberosity-trochlear groove (TT-TG) distance, and the patellar tilt. It is always performed on both lower limbs in order to evaluate differences between the two. The TT-TG distance is calculated by superimposing the axial view where the deepest point of the trochlear sulcus is visible, the view where the anterior tibial tuberosity is visible and measuring the distance between these two points in millimeters (Fig. 54.3).

Finally, magnetic resonance imaging must be performed for a correct evaluation of any associated injuries, any injuries of the cartilage layer, and medial patellofemoral ligament.

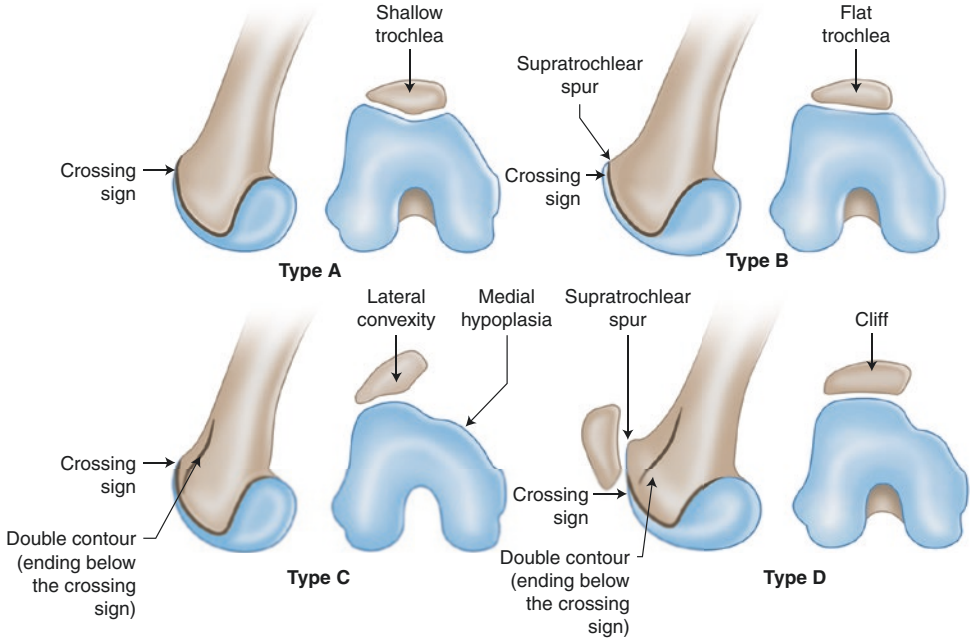

Fig. 54.1 Dejour's classification of trochlear dysplasia

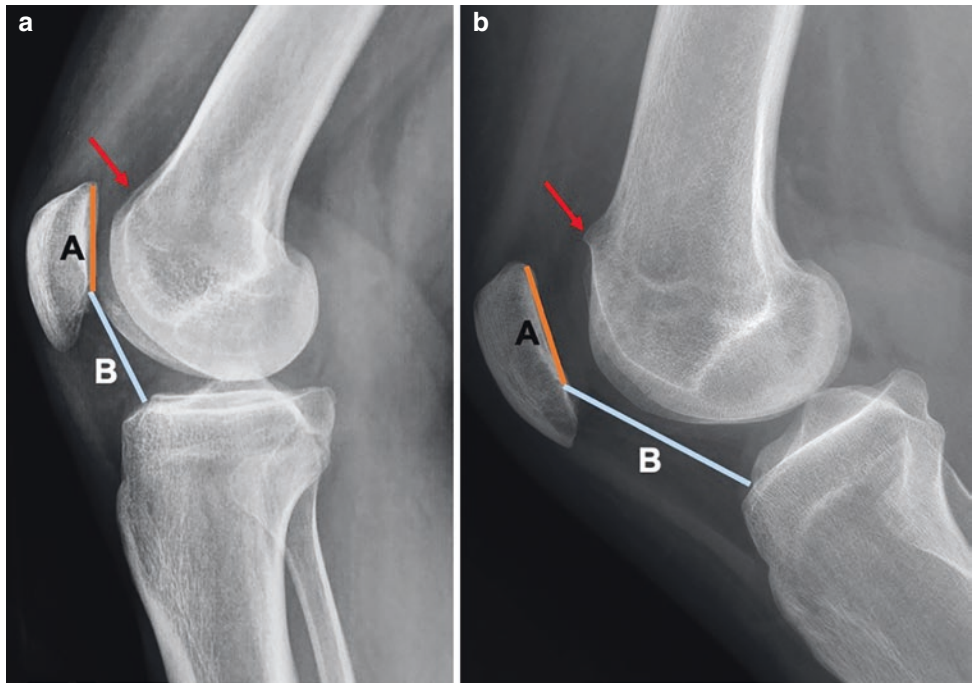

Fig. 54.2 (**a**) Lateral view of a normal right knee. (**b**) Lateral view of pathological right knee. The patient complains about patellofemoral instability. Red arrows show a normal trochlea in "a" and a spur indicating a trochlear dysplasia in "b." Caton-Deschamps index (CDI) is used to evaluate the patellar height. It consists of the ratio between the distance from the most anterior edge of the tibial plateau and the point where the articular patellar cartilage of the patella ends (B) and the measurement of the length of the articular surface of the patella itself (A). In "a," the CDI results to be 1.09, and in "b" results to be 1.31

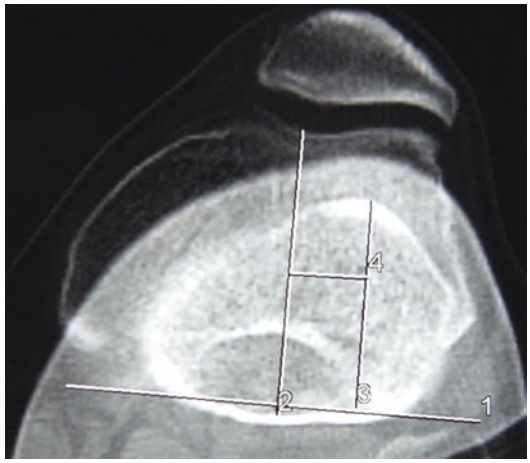

Fig. 54.3 CT axial views of a lateral knee superimposed to calculate the TT-TG distance. A line is drawn along the posterior femoral condyles (1). A line perpendicular to line 1 bisecting the trochlear groove at its deepest point is drawn (2). A line perpendicular to line 1 bisecting the anterior tibial tuberosity is drawn (3). The distance between 2 and 3 is the TT-TG distance

54.6 Treatment

In the case of patellofemoral pain in the absence of dislocations, the treatment will always be bloodless in the first instance. Functional rehabilitation must aim to improve patellofemoral tracking and stabilize the patella by reducing micro-instability and the tendency to sublux externally. That makes for improved distribution of tangential forces over the entire joint surface to avoid focal overloads. From a rehabilitation point of view, these objectives will be achieved by trying to increase hip rotations and through muscle strengthening of the core, quadriceps, and buttocks. Furthermore, the correction of secondary risk factors must be the aim where possible. For example, it can be done through the use of orthotics that reduces foot pronation and knee valgus. If the patient reports major pain that does not support following the correct rehabilitation process, an intra-articular infiltrative treatment can be considered with the aim of temporarily reducing local inflammation and the resulting pain. If pain persists despite 6 months of correct functional rehabilitation, surgery should be considered. As far as instability is concerned, the first episode of dislocation is always treated conservatively unless particular occurrences coexist, e.g., acute osteochondral avulsions secondary to dislocation or avulsion of the medial femoral patellar ligament with a bone fragment. If the patient has two or more episodes of dislocation or has experienced one episode and suffers from anterior pain resistant to nonoperative treatments, surgery may be indicated.

The surgical treatment of this joint disorder is based on the correction of predisposing factors for maltracking. Therefore, in the case of a high or lateralized patella, osteotomies of the anterior tuberosity as well as the consequent fixation of the same in a more favorable point from the biomechanical point of view is performed. If the external tilt is increased, a release of the lateral capsule-ligamentous structures of the patella may be called for. In the case of trochlear dysplasia, trochleoplasty can be performed to increase the concavity of the trochlear groove and therefore its congruence with the patella. Finally, severe valgus deformity or aberrant rotations can be addressed with specific corrective osteotomies of either the femur or the tibia. In instances of instability, in addition to the correction of the predisposing factors, a reconstruction of the medial patellofemoral ligament should be carried out since it ruptures in all cases of relapsing dislocation.

Take-Home Message

- Patellofemoral disorders are divisible into patellofemoral pain and patellofemoral instability.
- Some specific anatomical features may underline the development of patellofemoral joint pathologies.
- Neuromuscular control, specifically the muscle tone of the quadriceps and buttocks, contributes to the correct biomechanical functioning of the patellofemoral joint.
- In the event of a failure of functional treatment based on the development of dynamic joint stabilizers, surgery can be chosen.
- The surgical approach is based on the correction of the anatomical factors that can generate patellar femoral pain or instability.

Summary

Patellofemoral pain and patellar instability are frequent pathologies in young active population. Among numerous anatomical features that affect both the genesis of pain and patellar instability, the most frequently encountered are the patella in a high position, an increased distance between the trochlear groove and the tibial tuberosity, and trochlear dysplasia. A careful radiological analysis can easily determine the anatomical alterations present while the clinical evaluation must analyze neuromuscular alterations that may play a role in the development of symptoms at the patellofemoral joint. Both for patellofemoral pain and in the case of the first episode of patellar luxation, the initial treatment is conservative. If the symptoms continue after at least 6 months of conservative treatment, surgical treatment should be proposed to improve joint mechanics by correcting the anatomical alterations present. A key point in the surgical treatment of patellar instability is the reconstruction of the medial femoral-patellar ligament, an important stabilizer of the patella that ruptures when the patella dislocates.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Which muscular complex contributes most to patellofemoral tracking?
   (a) Quadriceps and abductors
   (b) Psoas and adductors
   (c) Quadriceps and gluteus
   (d) Psoas and quadriceps
2. Dejour's classification of trochlear dysplasia is based on:
   (a) The width of the trochlea in a sagittal view
   (b) The height of the trochlea in an axial view
   (c) The shape of the trochlea in sagittal and axial views
   (d) The position of the patella on sagittal and axial views
3. How is potential patella instability defined?
   (a) A dislocation associated with at least one primary predisposing factor
   (b) Patellar subluxation with one or more secondary predisposing factors
   (c) Anterior knee pain associated with one primary predisposing factor
   (d) None of the previous answers
4. Choose the correct answer from the following:
   (a) Patellar dislocation accounts for approximately 6–8% of knee injuries
   (b) Patellofemoral pathologies are more frequent in the young active population
   (c) Patellofemoral pathologies are more frequent in the middle-aged population
   (d) Chondral injuries to the patellofemoral joint are not frequent
5. As to the treatment of patellofemoral pathology, choose the correct one:
   (a) Surgical treatment is the first option in most cases.
   (b) Intra-articular injections are always the first step in conservative treatment.
   (c) If 2 months of conservative treatment fails to reduce pain or instability, surgical treatment is needed.
   (d) The surgical approach aims to correct anatomical features that predispose to instability in or overload on the patellofemoral joint.

Further Reading

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Osteoarthritis, Osteotomies, and Knee Arthroplasty

55

Elliot Sappey-Marinier, Cécile Batailler, and Sébastien Lustig

Overview

Osteoarthritis is one of the most common causes of disability in adults.

Osteoarthritis in turn manifests initially as abnormal joint tissue metabolism and subsequently by anatomic and physiologic derangements.

55.1 Introduction

Osteoarthritis (OA) is one of the most common causes of disability in adults. The prevalence increases with age, with a surprising 13.9% of the population over 25 years old being affected and 33.6% of the population over 65 years old affected. Osteoarthritis in turn manifests initially as abnormal joint tissue metabolism and subsequently by anatomic and physiologic derangements.

Classification strategies include:

1. Classification via radiographic imaging: Ahlbäck classification
2. Classification utilizing advanced imaging
3. Classification emphasizing clinical symptoms, including stiffness, swelling, knee range of motion, and knee crepitus
4. Combination of symptoms and imaging

For the purposes of this chapter, osteoarthritis, arthrosis, and arthritis will be used interchangeably. The most limiting aspect of radiologic classification is that it often does not detect arthritis until a more advanced stage. Classification strategies for radiographic imaging have emphasized joint space narrowing, subchondral sclerosis, and osteophyte formation. By advanced imaging (MRI), the most common features that indicate osteoarthritis are cartilage thinning and subchondral bone edema.

55.2 Etiology

One factor that is consistent in all studies of arthritis is its association with the aging process. The etiology of osteoarthritis has long been thought to be cartilage driven. Imaging definitions of osteoarthritis have as a main factor some inclusion of changes in the subchondral bone. Osteophyte formation, bone remodeling, subchondral sclerosis, and bone attrition are crucial for radiographic diagnosis; several of these bone changes take place not only during the final stages of the disease but sometimes at the onset of the disease, before carti-

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lage degradation is apparent. Recent evidence shows an additional and integrated role of bone and synovial tissue. Synovial inflammation corresponds to clinical symptoms such as joint swelling and inflammatory pain and is thought to be secondary to cartilage debris and catabolic mediators entering the synovial cavity.

55.2.1 Impairments in Muscle Function

Muscle function in the lower extremity, including weakness, altered muscle activation patterns, and proprioceptive deficits, is commonly found in association with knee OA. Improvement of muscle strength is a key component of conservative management of knee OA and has been found to be effective in symptom reduction. Whether exercise influences disease development and potentially stalls progression needs more study.

55.2.2 OA and Knee Injury

The risk of developing knee OA is strongly associated with BMI (kg/m^2) (adjusted for age, sex, and other covariates), as well as the heaviest category of physical stress at work (compared with the lightest category), and past knee injury.

55.2.3 Sex

Knee OA has a strong female sex preponderance. Women develop more knee OA than men, based on the rate of knee arthroplasty surgery. Obesity is a stronger risk factor for knee OA in women than in men. Independent of the effects of obesity, altered metabolism is related to knee OA, and these relations differ for men and women.

55.2.4 Heavy Physical Work

Of note, vibration, repetitive movement, and long hours of kneeling and squatting, standing, and solitary standing are associated with an increased risk of development of OA.

55.2.5 Obesity

Not all obesities are equivalent to the development of knee osteoarthritis; development appears to be strongly related to the coexistence of disordered glucose and lipid metabolism. Metabolic risk factors including obesity, hypertension, dyslipidemia, and impaired glucose tolerance raise not only the risk of occurrence of OA but also its progression. This risk rises with the increasing number of metabolic risk factors present.

55.3 Osteotomies

55.3.1 Introduction

In valgus and varus knee malalignment in relatively young and active patients, osteotomy has long been recognized as an appropriate option in the management of knee osteoarthritis. Historically, the first high tibial osteotomy (HTO) was performed by Jackson in 1958 with a ball-and-socket osteotomy below the anterior tibial tuberosity and osteotomy at the middle third of the fibula. In the same period, opening wedge technique of HTO was developed in France with medial approach, using allograft or autograft bone and plates that allowed stable fixation. After years of popularity, between the 1960s and 1980s, HTO had a slow decline after good results were demonstrated with unicompartmental and total knee arthroplasty and rising surgeon preferences for these techniques. Currently, knee osteotomy is undergoing a revival, particularly in younger more active patients, due to the desire to preserve the native knee, bone stock, and proprioception and also the possibility to allow physical activities that may not well be tolerated with a unicompartmental knee arthroplasty (UKA). In addition, knee osteotomy has become a better option due to new hardware: plates that work like an “internal fixation” allowing a very stable osteosynthesis and periosteal vascular supply preservation. The first aim of knee osteotomy is to eliminate or reduce pain, translating loads to the contralateral femorotibial compartment by correcting deformity.

55.3.2 Indications and Contraindications

Physical indications include age between 30 and 70 years; well-localized pain at the femorotibial joint line; flexion more than 90° and, if present, a lack of extension <15°; normal or correctable ligamentous status (but anterior cruciate ligament [ACL] or posterior cruciate ligament [PCL] insufficiency is not a contraindication); nonreducible deformity; and patients with an active lifestyle. Physical contraindications include obesity, inflammatory disease, smoking, osteoarthritis or meniscectomy in the contralateral compartment, and tibial subluxation more than 1 cm. Radiological indications include partial or complete joint space width narrowing in one compartment, no contralateral femorotibial joint space width narrowing or patellofemoral joint space width narrowing, and extra-articular deformity more than 5°. MRI can also be used to more accurately assess the contralateral compartment. Disputable contraindications include patellofemoral arthritis, flexion less than 100° or fixed flexion deformity, severe extra-articular deformity, being older than 70 years, and obese females.

55.3.3 Surgical Techniques for Medial Arthritis

Two techniques have been used for the treatment of medial compartment arthritis: medial opening wedge high tibial osteotomy (OWHTO) and lateral closing wedge high tibial osteotomy (CWHTO). It is important to consider the preoperative length of the limb. This could help to choose which technique to perform. Indeed, CWHTO will slightly shorten the limb, whereas OWHTO will slightly lengthen the limb.

55.3.3.1 Surgical Planning

Preoperatively, a complete radiological evaluation of the limb is mandatory for accurate planning. This is to determine the mechanical axis and calculate the amount of correction required. The standard X-ray series shows the osteoarthritis grade and the tibial slope, including X-rays done in Rosenberg view (45° of flexion). The weight-bearing anteroposterior long-leg X-ray allows measurement of the HKA angle to plan the correction. The axial patellar X-ray assesses involvement of the patellofemoral joint. The alignment goal of correction for osteoarthritis is usually 2–3° of mechanical valgus.

55.3.3.2 Opening Wedge High Tibial Osteotomy

The osteotomy is performed just proximal to the tibial tubercle, having elevated the superficial medial collateral ligament. The plane of the osteotomy is horizontal, slightly different from the medial closing wedge HTO, which is more oblique. In order to maintain the tibial slope, the opening of the osteotomy at the posteromedial cortex should be approximately twice that at the tibial tubercle. Options for fixation include staples, external fixators, and plates (conventional, blade plates, locking plates).

55.3.3.3 Closing Wedge High Tibial Osteotomy

Once the fibular osteotomy is performed, the distal cut of the closing wedge osteotomy is performed. An angled cutting guide (6, 8, or 10°) is introduced in the distal cut of the osteotomy, and the proximal cut is then made using this angle. The wedge is closed, and using a long metal bar positioned on the center of the femur head and in the middle of the ankle joint, the mechanical femorotibial axis is evaluated. The metal bar should pass just laterally to the lateral tibial spine. The osteotomy can be fixed with staples, blade plate, or locking plates.

55.3.3.4 Results of CWHTO

Good results have been reported regarding survival rates, survivorship at 5 years of follow-up from 73% to 98%, at 10 years of follow-up from 51% to 92%, and more than 15 years of follow-up from 39% to 71%. Koshino et al. reported a satisfaction rate at the final follow-up for excellent/good results of 98% at 15–28 years of follow-up. Conversion rates included for conversion to total knee arthroplasty or unicompartmental arthroplasty are from 3% to 39%.

55.3.3.5 Results of OWHTO

Hernigou et al. reported good results regarding survival rates, survivorship at 5 years of follow-up from 89% to 94%, at 10 years of follow-up from 74% to 85%, and more than 15 years of follow-up around 68%. Hernigou et al. mentioned a satisfaction rate at 10 years of follow-up for excellent/good results of 81%. At 10 years, conversion rates included for conversion to total knee arthroplasty or unicompartmental arthroplasty are 2–35%.

55.3.3.6 Complications

For CWHTO complication rates, the average is from 3.3% to 28%. The most frequent complication reported in this group is peroneal nerve palsy with rates from 2% to 43%, followed by delayed union with an average of 2–23%. Other important complications are deep vein thrombosis, pulmonary embolism, superficial infection, skin necrosis, and sympathetic dystrophies.

OWHTO complication rates are 3–22%, mainly due to tibial plateau fracture in 10%, nerve palsy in 10%, and delayed union in 1%. Other important complications are superficial infection and vascular problems.

55.3.4 Surgical Techniques for Lateral Arthritis

Valgus deformity of the knee is much less common than varus deformity and is caused most often by primary arthrosis. A varus-producing osteotomy can be performed on either the proximal tibia or the distal femur. Contraindications are similar to those for a valgus osteotomy, including an extension deficit >15° and a flexion arc <90°. The type of osteotomy performed depends on the degree of the deformity. The alignment goal of correction for lateral osteoarthritis is usually neutral mechanical axis to avoid recurrence.

55.3.4.1 Proximal Tibial Closing Wedge

For tibiofemoral deformities <10°, a proximal tibial closing wedge varus-producing osteotomy can be performed. Any greater deformity will be difficult to correct because of the anatomical valgus of the femur. A larger correction will necessitate an excessive wedge size and result in obliquity of the joint line.

55.3.4.2 Distal Femoral Osteotomy (DFO) by Lateral Approach

Among the advantages of the lateral opening wedge tibial osteotomy for the valgus knee are a familiar exposure, tightening of secondary lateral ligamentous laxity, and preserving the pes anserinus on the medial side. An oblique fibular diaphyseal osteotomy is also performed, and the wedge is filled with tricortical iliac crest bone graft or a bone substitute.

55.3.4.3 Distal Femoral Osteotomy (DFO) by Medial Approach

Actually, the most commonly performed is the medial closing DFO as reported in multiple studies. The femoral vessels must be protected. The osteotomy is performed above the adductor tubercle. The rigid internal fixation with the locking plate allows for early range of motion and partial weight-bearing.

55.3.4.4 Outcomes

Medial femoral closing wedge is the most commonly used technique for the correction of valgus alignment, since McDermott et al. described this technique. They showed good results in 92% of 24 patients treated with DFO. Backstein et al. described a DFO survivorship of 82% at 10-year follow-up and 45% at 15-year follow-up of 38 knees.

55.3.4.5 Complications

Complications involving the two techniques are not infrequent, and they are represented mostly by delayed union and nonunion, stiffness, and hardware failure that are frequently associated with lateral opening wedge osteotomy. In addition, in lateral DFO, the majority of patients complained about iliotibial band pain because of plate irritation (21–86%). This technique is simpler than the closing DFO since the lateral approach avoids the risk of neurovascular complications and is easier to do and the correction will be more accurate.

55.4 Unicompartmental Knee Arthroplasty (UKA)

55.4.1 Introduction

Isolated unicompartmental knee arthritis is a challenging problem. Surgical management of unicompartmental knee arthritis includes non-prosthetic treatments such as arthroscopic debridement and high tibial osteotomy (HTO) or prosthetic options including unicompartmental knee arthroplasty (UKA) and conventional total knee arthroplasty (TKA). These procedures, however, have a finite life span in active patients, and concerns regarding functional recovery and ability to return to sporting activities should be considered. The risk of failure after HTO for patients whose osteoarthritis is rated Ahlbäck grade two or higher has led surgeons to reduce the number of HTOs performed with an increase in arthroplasty even in younger patients. UKA in a patient in whom only one compartment of the knee is affected may provide better kinematics and quicker recovery with better conservation of bone stock than TKA. It should be emphasized that UKA is still a form of joint arthroplasty and not a biological, joint-preserving procedure. Revision is often to TKA, and as such, careful patient selection is of paramount importance.

55.4.2 Patient Selection and Indications

The indications for UKA are painful osteoarthritis (OA) or osteonecrosis limited to one compartment of the knee associated with significant loss of joint space on the radiographs. In fact, results after UKA for osteonecrosis limited to one compartment of the knee are comparable to those observed for OA at a mean of 12 years. Any type of inflammatory arthropathy, such as rheumatoid arthritis, is recognized as a formal contraindication for UKA because this can be a cause of rapid degeneration of the unreplaced compartments. Preoperative physical exam should ensure a range of knee flexion >100°, full knee extension, and a stable knee in the anteroposterior (AP) and sagittal planes. The patellofemoral joint should be clinically asymptomatic.

55.4.3 Radiological Evaluation

The radiological analysis should include full-length X-rays, AP, and ML view of the knee, skyline views, and stress radiographs. The radiographic analysis should ensure that there is no patellofemoral loss of joint space on skyline views at 30°, 60°, and 90° of flexion and confirm full-thickness articular cartilage in the uninvolved compartment. A varus or valgus deformity of the lower limb greater than 15° may represent a contraindication for UKA as the correction of such deformation may require soft-tissue release, which should not be performed when doing UKA. If the ACL status is unclear during the clinical exam, magnetic resonance imaging may be useful to confirm that the ACL is intact. Performing an isolated UKA on a deficient ACL knee is possible when the patient is not reporting instability.

55.4.4 Age and Weight

Age and weight may still represent debatable issues for UKA as the procedure is often presented as an alternative to either osteotomy or TKA. Very good survivorship was reported in a group of patients under 50, despite greater polyethylene wear than seen in older patients which is also seen in this group following TKA. Early reports of UKA considered obesity as a relative contraindication for UKA, but recent studies found no correlation between body weight and outcome, and we concur with the idea that wear is related to activity rather than weight. Obesity itself is therefore not deemed a contraindication.

55.4.5 Results of UKA

Recent studies demonstrate very good survival for modern UKA with 10-year survivorship at 90% or greater, and it would be relatively easier to revise in case of failure. Revision of a failed UKA had better results with more satisfied patient than revision TKA.

55.4.6 Complications

The main causes of revision UKA are progression of osteoarthritis in the other compartments, aseptic loosening of the components and polyethylene (PE) wear, impingement, bearing dislocation in mobile-bearing implants, periprosthetic fracture, infection, stiffness, and unexplained pain.

55.5 Total Knee Arthroplasty

55.5.1 Introduction

The primary indication for total knee arthroplasty (TKA) is to relieve pain caused by severe tricompartmental arthritis, with or without significant deformity. Other sources of knee and leg pain must be sought and systematically excluded. These include radicular pain from spinal disease, referred pain from the ipsilateral hip, peripheral vascular disease, meniscal pathology, and bursitis of the knee. Radiographic finding must correlate with a clear clinical impression of knee arthritis. Patients who do not have complete cartilage space loss before surgery tend to be less satisfied with their clinical result after TKA. Before surgery is considered, conservative treatment measures should be exhausted.

55.5.2 Patient Selection and Contraindications

Because knee replacement has a finite expected survival that is adversely affected by activity level, it generally is indicated in older patients with more sedentary lifestyles. It is also clearly indicated in younger patients who have limited function because of systemic arthritis with multiple joint involvement. Osteonecrosis with subchondral collapse of a femoral condyle can be an indication for arthroplasty.

Deformity can become the principal indication for arthroplasty in patients with moderate arthritis and variable levels of pain when the progression of deformity begins to threaten the expected outcome of an anticipated arthroplasty. As flexion contracture progresses beyond 20 degrees, gait is significantly hampered, and difficulty with regaining extension may warrant surgical intervention. Similarly, as varus or valgus laxity becomes severe, a constrained condylar type of prosthesis becomes necessary to prevent subsequent coronal plane instability. Intervening before this degree of laxity is present allows the use of a prosthesis that lacks coronal plane constraint and has a more favorable expected survival.

Absolute contraindications to TKA include recent or current knee sepsis, a remote source of ongoing infection, extensor mechanism discontinuity, or severe dysfunction. Relative contraindications include significant atherosclerotic disease of the operative leg, venous stasis disease with recurrent cellulitis, morbid obesity (BMI >40), and recurrent urinary tract infection.

55.5.3 Results of TKA

Long-term series by Ranawat et al., Front-Rodriguez et al., and Pavone et al. have documented the longevity of the original total condylar prosthesis to be 95% at 15 years and 91% at 21 and 23 years. Multiple studies of recent cruciate-retaining and cruciate-substituting designs have documented 10-year survivorship of 95% or greater.

55.5.4 Complications After TKA

A recent study reveals that infection is now the primary cause of failure in both the short and long terms (30.6%), followed by aseptic loosen-

ing (16.8%), stiffness (14.9%), extensor mechanism pathology (14.2%), pain (11.7%), instability (6.4%), periprosthetic fracture (3.1%), wound pathology (1.8%), and metal sensitivity (0.8%).

Take-Home Message

- Osteoarthritis is one of the most common causes of disability in adults.
- Osteophyte formation, bone remodeling, subchondral sclerosis, and bone attrition are crucial for radiographic diagnosis.
- The risk factors with the most frequent association with knee OA are age, obesity, female sex, prior joint trauma including repetitive workload, and metabolic syndrome.
- Conservative treatment such as osteotomy around the knee joint is valuable in the management of OA in the younger patient, as it allows a significant improvement in pain and function without resorting to the irreversible arthroplasty option.
- Unicompartmental knee arthroplasty has become the standard of treatment for patients with full loss of cartilage limited to one femorotibial compartment.
- Total knee arthroplasty should be considered for patients with severe arthritis.

Summary

Osteoarthritis

There is no universal definition of arthritis. Patients suffer pain, and joint space narrowing is observed on radiographs at an advanced stage of knee OA. The risk factors with the most frequent association with knee OA are age, obesity, female sex, prior joint trauma including repetitive workload, and metabolic syndrome.

Osteotomies Around the Knee

Osteotomy around the knee joint is a particularly valuable procedure for a specific group of patients, as discussed in detail in this chapter. It is especially valuable in the management of OA in the younger patient, as it allows a significant improvement in pain and function without resorting to the irreversible arthroplasty option. Achieving success with osteotomy relies on careful patient selection and precise surgical technique.

Unicompartmental Knee Arthroplasty (UKA)

Unicompartmental knee arthroplasty has become the standard of treatment for patients with full loss of cartilage limited to one femorotibial compartment. When the indications are satisfied and the surgical principles respected, it is possible to restore full knee function after UKA. The recent evolution in surgical technique and instrumentation has allowed the surgeon to extend the indication to lateral compartment osteoarthritis, osteonecrosis, or post-traumatic disorders.

Total Knee Arthroplasty

Total knee arthroplasty should be considered for patients with severe arthritis. It should only be offered after nonoperative treatments fail. It is essential to set expectations for pain relief, improvement in function, and durability. Patients must understand that it is a nonconservative treatment. In case of complications such as infection and aseptic loosening, revision surgeries are needed with often poorer outcomes than after primary TKA.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Radiographic elements crucial for OA diagnosis are:
   (a) Osteophyte formation, bone remodeling, subchondral sclerosis, and bone attrition
   (b) Osteophyte formation, cysts, bone edema
   (c) Bone edema and subchondral sclerosis
   (d) Cysts and subchondral sclerosis
2. Indications for knee osteotomies are:
   (a) B + C + D
   (b) Age between 30 and 70 years
   (c) Well-localized pain at the femorotibial joint line

(d) Flexion more than 90° and, if present, a lack of extension <15°; normal or correctable ligamentous status; nonreducible deformity; and patients with an active lifestyle

3. Indications for UKA are:
   (a) B + C
   (b) Significant loss of joint space on the radiographs
   (c) Painful osteoarthritis or osteonecrosis limited to one compartment of the knee
   (d) Early stages of knee OA
4. The main cause of UKA revision is:
   (a) Progression of osteoarthritis in the other compartments
   (b) Periprosthetic fracture
   (c) Unexplained pain
   (d) Infection
5. Which is the primary cause of TKA failure in short and long terms?
   (a) Infection
   (b) Metal sensitivity
   (c) Periprosthetic fracture
   (d) Pain

Further Reading

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Knee Fractures

56

Nicholas Mannering and Joo Sunwoo

Overview

Distal femur fractures are a traumatic injury that extends from the distal metaphyseal-diaphyseal junction to the femoral condyle articular surface. An understanding of the distal femur anatomy is necessary both to achieve anatomical reduction and restore function.

56.1 Distal Femur Fractures

56.1.1 Introduction

Distal femur fractures are a traumatic injury that extends from the distal metaphyseal-diaphyseal junction to the femoral condyle articular surface. An understanding of the distal femur anatomy is necessary both to achieve anatomical reduction and restore function. Anatomical landmarks can be divided into the three anatomical planes. In the sagittal plane, the anterior femoral cortex aligns with the front half of the femoral condyles. In the coronal plane, anatomical axis of the distal femur forms a valgus angle of 7–10° with the knee joint. In the axial plane, the lateral femur cortex slopes 10°, compared to the medial cortex which slopes 25°.

The majority of the key deforming forces on the distal femur involve the quadriceps muscle and the hamstrings (causing limb shortening and flexion or extension deformity), adductor magnus (causing varus deformity), and gastrocnemius (which may cause rotational deformity of the condyles).

56.1.1.1 Epidemiology and History

Distal femur fractures account for 3% of all femur fractures and 0.4% of all fractures combined. The demographic of patients reveals a bimodal distribution. Young healthy males suffer a high-energy mechanism of injury with significant displacement, whereas elderly females encounter a low-energy mechanism of injury with less fracture displacement.

Classification System

Several classification systems have been described, including a descriptive and AO/OTA (Orthopaedic Trauma Association) classification system (Fig. 56.1). The descriptive classification system identifies either supracondylar fractures or intercondylar fractures. The AO/OTA further describes extra-articular fractures as avulsions

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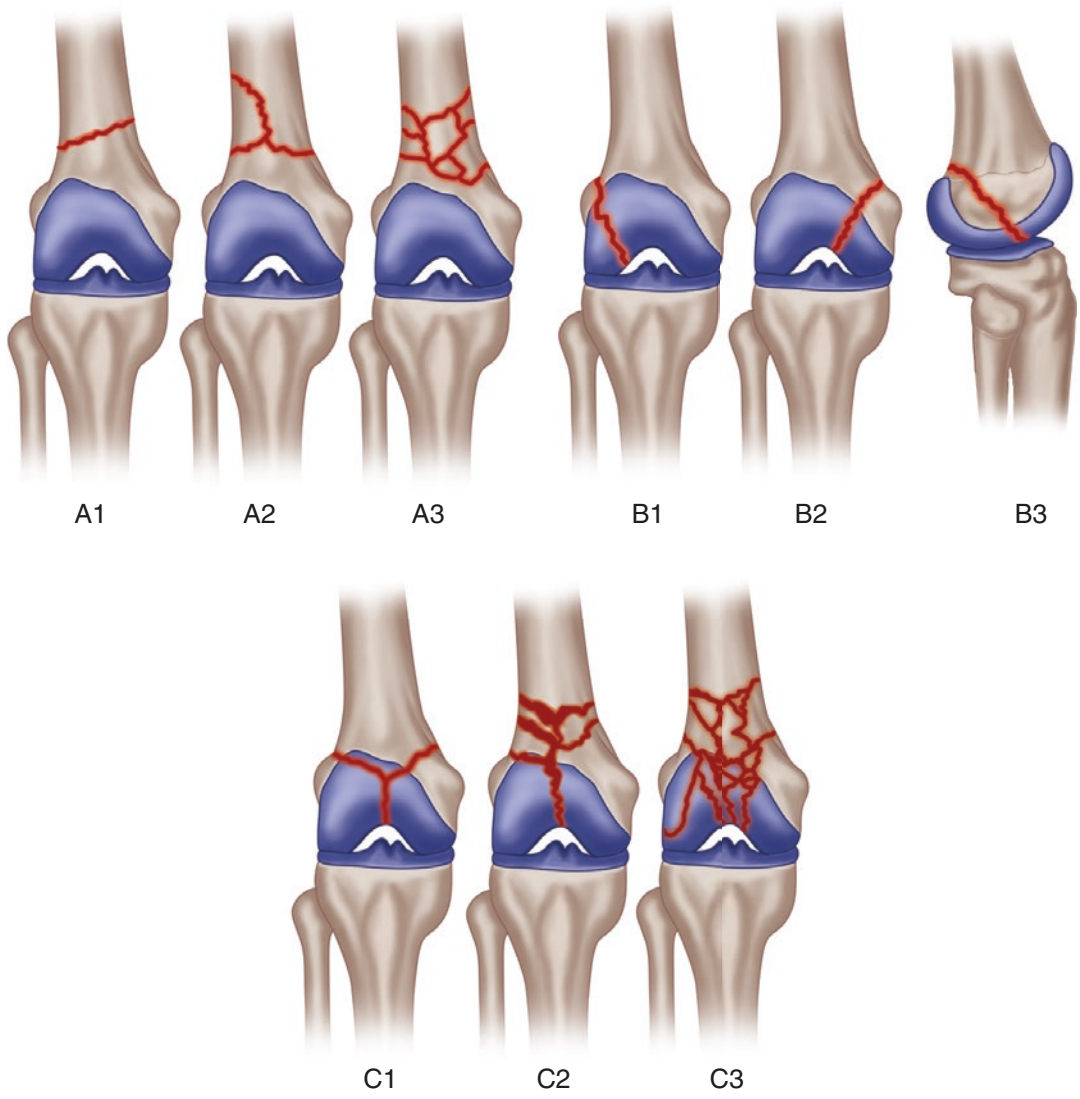

Fig. 56.1 AO/OTA (Orthopaedic Trauma Association) classification system

(A1), simple fractures (A2), or multifragmentary fractures (A3). Partial articular fractures can be divided into the lateral condyle (B1), medial condyle (B2), or involving the coronal plane (B3, also known as a Hoffa fragment). Type C fractures are more complex, involving both condyles and complete articular surface, as a T-shape or extensive comminution. Fractures of any type may be associated with skin, soft tissue, vascular, or nerve injuries.

Assessment

Assessment of distal femur fractures should involve a standardized approach in accordance with ATLS (Advanced Trauma Life Support®) principles, considering the mechanism of injury, patient risk factors, and search for possible other associated injuries. A thorough examination of the affected limb must be performed, including neurological and vascular assessment. This can involve measurement of an ankle brachial index

(ABI), Doppler ultrasound, and/or CT angiogram. There should be a high index of suspicion for vascular injury, given that early detection and treatment directly affect the outcome.

Plain-film radiographs may be taken either in the emergency department with mobile X-ray or in the radiology department if the patient is clinically stable. Adjacent joint X-rays are important to rule out associated injuries. Furthermore, contralateral femur X-rays may be useful in preoperative planning, templating, and identifying leg-length discrepancies.

Whilst X-ray alone can characterize the fracture, computed tomography (CT) is often necessary to fully understand the articular fracture pattern. A Hoffa fragment (type B3) can be missed in plain-film radiographs and can be as prevalent as 38% of distal femur fractures.

Management

Principles of management for distal femur fractures should include minimizing iatrogenic soft-tissue trauma, restoration of limb length, alignment, and rotation, and preservation of the normal knee joint function, by achieving articular congruity of the knee with absolute stability. These principles can be achieved using multiple surgical modalities, each one with advantages and disadvantages that should be used appropriately according to the fracture pattern and severity of the injury.

Briefly, a nonoperative approach is rarely indicated and a hinged knee brace may be used in this setting. This should be strictly limited to nondisplaced fractures, nonambulatory patients, and trauma patients that present an unacceptably high surgical or anesthetic risk.

Immediate Stabilization

As part of ATLS principles, having first stabilized the cardiorespiratory status of the patient according to the ABCD method, the fracture should also be stabilized. In the emergency room setting, this can be achieved, with the patient adequately sedated, by closed reduction of the affected limb to relatively normal alignment and an above-knee plaster of Paris backslab applied or skin traction. This will maximize pain management and reduce disruption of the fracture site when being transferred for essential imaging and to the operating room.

External Fixation

External fixation is a key aspect of damage control orthopedics, when the patient's clinical condition does not allow for definitive internal fixation. This achieves the main treatment goals of relative stability, minimizing operative time, minimizing blood loss and surgical exposure, reducing the risk of surgical site infection, and reducing pain.

Various types of external fixation can be used to treat distal femur fractures. The Taylor Spatial Frame (TSF) has been used as a type of external fixation for the treatment of both supra- and intra-condylar femur fractures, showing effective definitive fixation and fracture healing, of 19 out of 19 fractures, with no reported cases of infection.

However, whilst the Ilizarov fixator, a type of external fixation, can be an attractive option for complex C2 and C3 fractures, poor outcomes with higher infection rates have been reported with a limited open approach and external tensioned wire fixation. It is likely that these infection rates are linked to the severity of the initial injury with associated soft-tissue trauma, rather than a criticism of the technique itself. These methods have the advantage of minimizing soft-tissue disruption and are well-known treatment modalities when combined with debridement and irrigation for open fractures.

Other studies strongly support the provisional use of external fixation for extensively compounded distal femur fractures, with early conversion to internal fixation (one study within a mean of 5 days, compared to others which quoted within a mean of 4.7 weeks). Of note, the group with intended definitive external fixation achieved active flexion of 62.3° at 1-year follow-up, compared to 101° for the group with early conversion to internal fixation.

Minimally Invasive Plate Osteosynthesis (MIPO)

The MIPO technique is an attractive option for minimally displaced distal femur fractures in

elderly patients suffering a low-energy injury. The hallmarks of this technique include traditional exposure of the fracture site and internal fixation of intra-articular fracture components, and closed reduction of metaphyseal/diaphyseal fracture components and fixation by submuscular plating. Relative fracture-site alignment is achieved with closed reduction of metaphyseal/diaphyseal fractures under image intensifier (II) guidance and can be performed on a traction table. The technique lends itself to minimal periosteal stripping, maintenance of tissue viability, and quicker operative time. Various locking plates have been developed, including the Synthes LISS (Less Invasive Stabilization System) plate or the Zimmer Periarticular plate.

MIPO allows early mobilization and stimulates the healing process from loading at the fracture site generating compression. The risk of hardware failure due to loading in some cases may be reduced using longer plates, which spreads the load over a larger surface area, creating a longer working length. Despite early mobilization, DVT rates were still reported to be as much as 25% in one study, although routine chemoprophylaxis was not utilized in this patient cohort.

Multiple complications can arise from the MIPO technique. Due to the minimally invasive nature of this method, the surgeon's percutaneous approach for metaphyseal/diaphyseal fixation may result in unforeseen neurovascular injury. Multiple studies outline a safe zone to prevent neurovascular injury, on both the medial side and the lateral side. The median distance from screw tip to superficial femoral artery was 21 mm, in particular for the distal sixth to tenth holes. Hardware malpositioning can also be the source of surgical related complications. Follow-up radiographs of distal femur fractures treated with LISS plate identified common errors including valgus-varus mal-reduction, malrotation, sagittal malalignment of the plate, screw penetration into the knee joint, and drilling holes for unicortical screws. The authors also identify that the use of Kirschner wires should be avoided.

Open Reduction Internal Fixation

Open reduction and internal fixation aims to anatomically restore the articular surface of the distal femur. To restore congruency of the knee articulation, the intra-articular fracture site can be exposed, reduced under direct vision, and held with Kirschner wires. This requires adequate exposure within an operative window whilst minimizing soft-tissue trauma, as outlined by Hoppenfeld et al. A direct lateral approach to the distal femur is traditionally utilized with the knee in flexion. The swashbuckler approach has been described as a modified anterior approach, by making an incision on the lateral edge of the tibial tubercle to the superolateral corner of the patella with the ability to perform a lateral parapatellar arthrotomy. Studies show the swashbuckler approach to be comparable to the standard lateral approach for patients managed with locking compression plate (LCP) for distal femur fractures.

It should be noted that achieving a congruent knee joint may not be possible with severe C3-type distal femur fractures due to bone loss, gross instability, or lack of suitable proximal bone. Studies indicate that a double-plating technique with an extended medial parapatellar anterior approach can achieve good outcomes for severe C3-type fractures. This can be appropriate for multiple reasons, giving the surgeon excellent exposure through one incision for anatomical reduction and stable fixation, with 68.75% achieving well-to-excellent functional outcomes. Lab models have also demonstrated that double plating is a stronger construct compared to single LISS plate, with less bend angles and reduced fracture gap.

Due to the more invasive nature of open reduction and internal fixation techniques for distal femur fractures, operative time can be longer compared to external fixation and MIPO. This is important to consider, as the technique may lead to complications such as surgical site infection (4% in one multicenter study, 3.6% in another observational case-control study).

Femoral Nail

The femoral nail can be performed via either an anterograde or a retrograde technique. There are a relatively small range of indications, including extra-articular fractures or simple intra-articular fractures. This technique has the benefit of being a closed technique and having a shorter operative time. An interfragmentary screw should first be utilized for simple intra-articular fractures to avoid fracture propagation during nail insertion.

The anterograde nail technique should be spared for metaphyseal/diaphyseal fractures, given that adequate distal fixation is paramount to the integrity of this relative stability construct. To highlight this further, the best fixation has been found to be an oblique proximal screw with two distal screws in the coronal plane. A proof-of-concept three-dimensional finite element model study identified anterograde nail to be the more stable construct for supracondylar distal femur fractures.

Conversely, the retrograde nail technique requires a parapatellar approach to sublux or evert the patella to allow retrograde femur reaming. Multiple complications can arise, including septic arthritis, knee stiffness, impingement, and hardware malpositioning or mal-fixation. Whilst studies show slightly less knee functional range postoperatively for retrograde nail compared to LISS plate, the infection rate, malalignment complications, and pain profiles postoperatively were comparable.

Distal Femur Replacement

Distal femur replacement is a final treatment option for comminuted intra-articular distal femur fractures in low-demand patients. It has the theoretical advantage of achieving early mobilization, which has been demonstrated at 1-year follow-up compared to ORIF.

56.2 Patellar Fractures

56.2.1 Introduction

The patella is the largest sesamoid bone in the body with the quadriceps tendon and fascia lata attaching to its anterosuperior margin and the patella tendon attaching to its anteroinferior margin. The patella functions as a pulley helping to shift the contractile force of the quadriceps muscle anteriorly.

The posterior articular surface of the patella is divided into medial and lateral facets. A thick layer of articular cartilage covers the superior three-quarters of the posterior surface whilst the inferior quarter is non-articulating. An anastomotic ring supplied by the geniculate arteries provides the vascular supply to the patella in a centripetal fashion.

56.2.1.1 Epidemiology and History

Patellar fractures make up 1% of all fractures, occurring most commonly in the 20–50-year age group. The incidence of patellar fractures in males is twice as high as the incidence in females.

Patellar fractures are caused by two main mechanisms of injury from direct and indirect forces. Direct trauma to the anterior knee commonly occurs from a fall onto the knee or a dashboard injury during a motor vehicle accident. This causes a failure of the patella bone in compression commonly resulting in a comminuted fracture pattern, usually with minimal displacement, and damage to the articular cartilage.

Indirect trauma to the patella occurs when an eccentric load is applied on the extensor mechanism of the knee. This most characteristically occurs when the knee is rapidly flexed against a maximally contracted quadriceps muscle. The patella fails under tension commonly resulting in a displaced transverse/avulsion-type fracture, usually with less damage to the articular cartilage compared to patellar fractures from direct trauma.

In the pediatric population (age 8–12), indirect trauma may result in a patella sleeve fracture, a rare injury where the cartilage "sleeve" of the patella separates from the ossified patella. A high index of suspicion is required in this patient cohort, and further imaging such as magnetic resonance imaging (MRI) scans or ultrasound may be indicated when clinical and radiographic findings are equivocal.

56.2.2 Classification System

Patella fractures present with a high degree of diversity and variability in fracture type and fracture pattern. Several classification systems have been described in the literature for patellar fractures. A holistic classification system linking the distinct fracture types with specific management options is currently still lacking.

Patellar fractures are classified descriptively based on the fracture morphology. Fractures with multiple fragments are considered comminuted. Fractures are classified as displaced when there is a separation of fragments greater than 3 mm or an articular step-off greater than 2 mm.

Fractures running in a medial-lateral direction are described as transverse, whereas fractures in a superior-inferior direction are described as vertical. Stellate fractures describe comminuted fractures in which the fracture lines radiate from a single point. Fractures involving the articular cartilage and underlying subchondral bone can be described as osteochondral.

Patellar fractures can also be classified according to the AO/OTA classification system based on the degree of articular involvement and comminution of the fracture. Type A fractures describe extra-articular fractures, type B fractures describe partial-articular vertical fractures, and type C describe complete articular, transverse fractures of the patella.

56.2.3 Assessment

Once again, assessment should involve a standardized approach in accordance with the ATLS principles to rule out any more immediate life-threatening injuries and confirm an isolated injury.

A thorough history and examination of the patient must be obtained, considering the mechanism of injury, patient risk factors, and detection of any other associated injuries. Suspicion for a patellar fracture should be raised when either direct trauma or an eccentric load involves the knee. High-energy mechanisms, such as dashboard, should raise concern for other associated injuries including femoral fractures, acetabular fractures, knee dislocations, ligamentous injuries, and neurovascular injuries.

Examination of a knee with patellar fracture may reveal a hematoma, hemarthrosis, and pain on palpation or palpable defect in the patella. Examination should involve a thorough inspection of the integrity of surrounding soft tissue and identify any open injury that may complicate surgical management. Straight leg raise examination should be performed to evaluate the integrity of the extensor mechanism.

Plain radiographs of the knee including standard anterior-posterior, lateral, and skyline views are usually sufficient. More sophisticated imaging modalities, such as CT or MRI scans, are rarely required but can be useful in the assessment of comminuted fractures and any associated ligamentous, meniscal, or osteochondral injuries.

Importantly, bipartite patella is present in 2–3% of the population. It occurs when a second ossification center for the patella fails to unite with the primary nucleus. They occur most commonly at the superolateral edge and are bilateral in 50% of patients. Bipartite patella should not be confused with an acute patella fracture.

56.2.4 Management

56.2.4.1 Nonoperative Management

Nonoperative management is indicated in nondisplaced and minimally displaced patellar fractures with an intact extensor mechanism and articular congruency. Around 30% of the force of extending the knee joint is transmitted through the medial and lateral retinaculum and ligamentous sheath overlying the patella. In subaponeurotic patellar fractures where the retinaculum is intact, these structures can provide fracture stability. Nonsurgical management may be considered in patients with displaced patellar fractures that have significant comorbidities or are poor surgical candidates.

Nonoperative management for patellar fractures involves early weight-bearing usually in a range-of-motion brace locked in extension or

near extension. Active or active-assisted range of motion is started at 7–14 days and gradually increased to full range of motion over 4–6 weeks. Exercises against resistance typically begin at the 6-week mark. Delayed weight-bearing is rarely indicated except in patients unable to follow instructions or who have a high-fall risk (i.e., geriatric population). Early mobilization and range of motion are encouraged to prevent joint stiffness. A repeat radiograph of the patella 1 week after beginning range of motion can be used to evaluate the stability of the fracture. A further radiograph of the patella at 6 weeks can be used to confirm bone healing.

Nonoperative management of non-displaced patellar fractures with an intact extensor mechanism has shown good outcomes in terms of pain and function in 90–99% of patients. For displaced patellar fractures, outcomes of nonoperative management are poorer. Two-year follow-up of patients with nonoperatively managed displaced patellar fractures showed an extensor lag of >20% for all patients (n = 12), with 25% of patients considering their outcome to be poor.

56.2.4.2 Operative Management

Operative management is indicated in patellar fractures with associated extensor mechanism incompetence. Displacement of fracture segments, incongruence of the articular surface, intra-articular loose bodies, or osteochondral fractures are other relative indications for operative management. The principles of operative management for patellar fractures include a restoration of the extensor mechanism, articular congruency, and preservation of patellar bone. Ideally, operative methods should aim to achieve a stable fixation to allow for early mobility and rehabilitation.

56.2.4.3 Open Reduction and Internal Fixation

The preferred operative treatment for displaced patellar fractures is open reduction and internal fixation to provide stability, restoration of normal patellar anatomy, and function to the extensor mechanism. Numerous operative techniques using a variety of different materials have been described in the literature.

The patella fracture is approached commonly through a vertical midline incision. Soft tissue is dissected to achieve good exposure of the fracture. Small medial or lateral parapatellar arthrotomies can be performed for palpation or direct visualization of the posterior articular surface. Anatomic reduction is achieved, and provisional stabilization can be provided with the aid of Kirschner wires or bone reduction forceps. Definitive fixation can be achieved through the use of a combination of different materials including screws, plates, and wires, most commonly in a tension band construct.

56.2.4.4 Tension Band Wiring

Tension band wiring is most ideally suitable for transverse patellar fractures with minimal comminution. Comminuted portions of the fracture may first need to be secured with lag screw fixation to convert the fracture into a simple transverse pattern for more optimal results. The modified anterior tension band (MATB) construct is currently the most widely accepted technique. This technique involves two longitudinal Kirschner wires through the fracture reinforced by an 18-gauge wire in a figure-of-eight pattern over the anterior surface of the patella. The tension band construct converts the anterior force generated on the patella, by the extensor mechanisms and knee flexion, into a compressive force on the patella. Biomechanical studies have shown that screws offer more biomechanical stability than Kirschner wires when used in tension band constructs. In this technique, a tension band wire is passed through longitudinal cannulated screws in a tension band construct. Additional interfragmentary screws, small plates, and circumferential cerclage wiring can be useful in supporting fixation in comminuted fractures (Fig. 56.2).

Nonabsorbable sutures have been used as an alternative material to steel wire for use in the tension band construct. Outcomes of biomechanical and a limited number of small clinical studies have shown braided polyester sutures to be comparable to steel wire when used for tension band wiring.

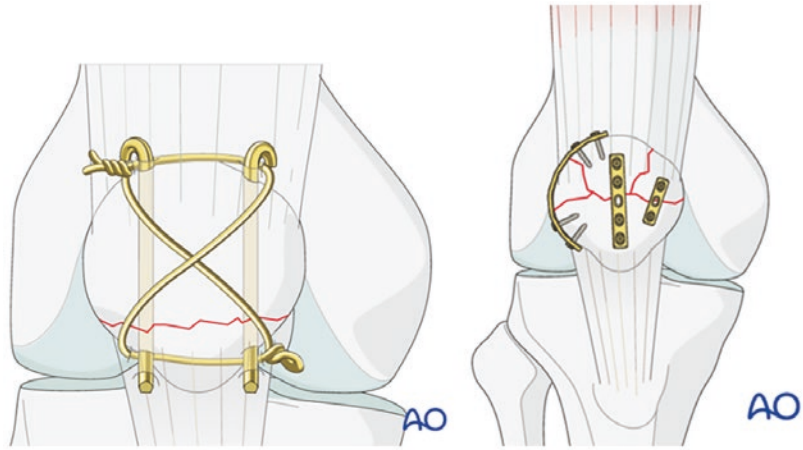

Fig. 56.2 Tension band wire construct for transverse patellar fracture, and fragment specific fixation with small plates for comminuted patellar fracture

56.2.4.5 Arthroscopic Assisted Percutaneous Fixation

More recently, less invasive procedures have been designed utilizing percutaneous fixation assisted by knee arthroscopy. Knee arthroscopy allows for the assessment of cartilage, ligaments, and fracture reduction from within the joint itself. Arthroscopic assisted percutaneous fixation techniques have shown promising results in simple, mildly displaced transverse fractures. Percutaneous fixation is also useful in the presence of soft-tissue injury in association with a patellar fracture.

56.2.4.6 Patellectomy

Patellectomy or partial patellectomy is reserved for rare cases of patellar fractures that are unable to be fixed with open reduction and internal fixation. The indications for partial or complete patellectomies have significantly diminished with improvements in surgical techniques and understanding of patella biomechanics and anatomy.

Partial patellectomy may be indicated in cases with significant cartilage loss and severe comminution. Even in these cases, all effort is made to preserve as much patella bone as possible. Preservation of even a portion of the patella can help to retain some patellar moment arm and improve strength of the knee. Severely comminuted fractures of the inferior pole or lateral margins of the patellar may be most amenable to partial patellectomy. After the excision of fragments, some fractures involving the inferior pole of the patella can be managed as patella avulsions and reattached to the patella body.

Complete patellectomy is rarely indicated except in rare cases of failed internal fixation, infection, tumor, or patellofemoral arthritis. Complete patellectomy is largely avoided due to poor outcomes and loss of quadriceps strength.

Take-Home Message

- The AO/OTA classification systems utilized are useful in succinctly communicating the degree of injury and the level of intervention thus required.
- Fracture of the knee can range from very minor injuries with minimal treatment required to potentially limb-threatening injuries requiring immediate and invasive intervention.
- Goals of treatment should involve restoration of knee function and stability, by either nonoperative or operative means, and pain control, to return patients to their premorbid level of function.

Summary

Orthopedic surgeons are constantly re-evaluating the approaches, techniques, and prostheses available for the treatment of distal femur fractures and patella fractures. Large randomized controlled trials are important to objectively determine the effectiveness of these techniques, yet the financial and ethical feasibility of these studies is a significant barrier. The growing availability of distal femur replacements and improvements in replication of the anatomical knee function will create exciting opportunities for current and future knee arthroplasty surgeons. All techniques will need to be further studied and utilized appropriately, given that the aging population as well as the high-energy motor vehicle patient cohorts will continue to present knee fracture challenges to the orthopedic community.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Distal femur fractures account for:
   (a) 3% of all femur fractures
   (b) 50% of all femur fractures
   (c) 0–1% of all femur fractures
   (d) 30% of all femur fractures
2. The AO/OTA type B1 is:
   (a) Partial articular fracture of the lateral condyle
   (b) Partial articular fracture of the medial condyle
   (c) Simple extra-articular fracture
   (d) Multifragmentary extra-articular fracture
3. Management of distal femur fractures includes:
   (a) Minimizing iatrogenic soft-tissue trauma, achieving articular congruity of the knee, absolute stability, restoration of limb length, and preservation of the normal knee joint function
   (b) Urgent reduction with external fixation
   (c) Urgent reduction with ORIF
   (d) Restoration of limb length and preservation of the normal knee joint function through percutaneous pinning, to avoid infections
4. Bipartite patella:
   (a) Is present in 2–3% of population and does not require treatment
   (b) Is present in 2–3% of population and requires treatment
   (c) Is present in 10% of population and is a risk factor for fractures
   (d) Requires urgent fixation with K-wires
5. Patellectomy:
   (a) Is reserved for rare cases of patellar fractures that are unable to be fixed with open reduction and internal fixation
   (b) Is required in case of transverse fractures
   (c) Is required in case of oblique fractures
   (d) Is never performed

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Part XI

Leg, Foot and Ankle

Fractures of the Leg

57

Enrico Cautero and Alessandro Mazzola

Overview

The tibial plateau articulates directly with the femoral condyles: a fracture at this level is generally caused by a compressive trauma, such as a fall from above or a knee sprain in varus or valgus.

57.1 Introduction

The tibial plateau articulates directly with the femoral condyles: a fracture at this level is generally caused by a compressive trauma, such as a fall from above or a knee sprain in varus or valgus. Adult males are more frequently involved in high-energy traumas, whereas falls are a leading cause of fracture in adult women. The lateral part of the knee is the most frequently affected by knee fractures. It is due to the anatomy of the knee: the medial plateau, that bears 60% of the knee load, is more distal than the lateral one, so there is a less risk for fracture. As regards the diagnosis of knee fractures, it should be suspected in case of aspiration of blood during knee arthrocentesis. Moreover, this technique helps to relieve the patient's pain. Conventional radiology is mandatory to define the fracture site and to define the best treatment choice. Schatzker classification of tibial plateau fractures is widely used by orthopedic surgeons to assess the initial injury, plan management, and predict prognosis. CT scan can better evaluate joint integrity in case of comminuted fractures. In case of fractures that do not require surgical treatment (not displaced or minimally displaced and stable fractures), immobilization in a long knee brace with partial knee joint load and free joint range of motion (ROM), for a period of 4–8 weeks, is necessary to achieve good bone healing. For articular fractures with more than 3 mm step-off, condylar widening more than 5 mm, varus/valgus instability, and all medial plateau or bicondylar ones, surgical treatment is mandatory (Table 57.1). When definitive fixation is not possible (in case of polytrauma or severe soft-tissue injures), a bridging therapy with external fixation is recommended: two 5 mm half-pins in the distal anterior femur and two in the distal anterior tibia are inserted, and axial traction is applied to the external fixation device. It is used to stabilize bone and soft tissues at a distance from the operative focus. The goal of the definitive surgical treatment is to refill the metaphyseal void with autologous or allogenic bone graft, restoring the joint surface. The arthroscopically assisted reduction of the articular frag-

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Table 57.1 Tibial plateau fractures surgical criteria

>3 mm articular step-off
>5 mm Condylar widening
Varus/valgus instability
Medial plateau fractures
Bicondylar fractures

Table 57.2 Indications for nonsurgical treatment in diaphyseal tibial fractures

<5° of varus/valgus
<10° Flexion/extension
<10° Malrotation
<1 cm of shortening

ments is indicated in case of unicondylar fractures, mainly of the lateral plateau and that are minimally displaced; direct articular visualization allows to evaluate, in addition to any meniscus or ligament injures of the knee, the correct reduction of fragments on the joint surface. Through an anterior access, the plateau is raised, and with a lateral incision, the plate or screws are introduced. In case of complex unicondylar fractures, open reduction is required: surgical incisions are dependent on the fracture site and the need to place one or more plates. A lateral incision is the most commonly used. However, a posterior incision can be useful for posterior shearing fractures. Dual-surgical incisions with dual-plate fixation are indicated for bicondylar tibial plateau fractures. In this case, attention should be paid to the distance between the two incisions: postoperative skin necrosis can cause fixation device exposure, with infective sequelae.

57.2 Diaphyseal Tibial Fractures

They represent the most common long-bone fractures worldwide and are often associated with soft-tissue injuries or compartment syndrome; the mechanism of injury is generally a high-energy trauma, with a direct force that fractures the fibula at the same level and provokes severe soft-tissue damage. Also, low-energy patterns may cause diaphyseal tibial fractures: they are due to the torsion of the leg that fractures the fibula at a different level, resulting in a spiral tibial fracture. An X-ray of the entire leg, including the knee and ankle, should be performed urgently as well as a careful clinical evaluation for open lesions and potential vascular nerve damage. Indications for nonsurgical treatment are the same as a good surgical reduction (Table 57.2). Fractures of the proximal third of the tibia are unstable, because of intrinsic deforming forces: the patellar tendon displaces the proximal fragment in extension while the gastrocnemius pulls the distal one into flexion. Anterior muscles deform the fracture in valgus while the pes anserinus deforms in varus. Surgical treatment of displaced shinbone fractures represents an orthopedic urgency: damage control and temporary stabilization utilizing an external fixator are excellent options when definitive fixation is not possible: it is mandatory in case of polytrauma, extensive soft-tissue damage, or an open injury with increased risk of infection. The most accurate method for open fracture grading is by intraoperative examination: Gustilo and Anderson produced their classification based on a study cohort performed on 1025 exposed fractures that were distinguished initially into three types: I, II, and III. Subsequently, clinical practice led Gustilo to extend and divide type III lesions into the subgroups A, B, and C.

57.2.1 Type I

This type of fractures present a clean wound less than 1 cm in size, with poor or absent contamination.

The wound is generally caused by a fracture fragment breaking through the skin at the moment of the injury. Type I fractures are simple fractures, such as spiral or short oblique ones.

57.2.2 Type II

These lesions have a skin involvement greater than 1 cm in diameter, but the surrounding tissues show minimal or absent signs of contusion.

57.2.3 Type III

Fractures are exposed and exhibit extensive soft-tissue damage, often with impaired vascularization, with or without severe contamination of the wound, with pronounced fracture instability due to comminuted fragments or bone defects. Given that many different factors contribute to identifying this group, Gustilo later decided to create the subgroups III A, III B, and III C.

57.2.3.1 Type III A

Generally, the injury is caused by a high-energy trauma. In this group of fractures, there is still an adequate coverage of the injured bone, despite the presence of large lacerations or flaps in the soft tissues.

57.2.3.2 Type III B

In contrast to type III A, lesions present extensive periosteal stripping, severe soft-tissue damage, and exposure of the bone. The wound requires soft-tissue coverage (rotational or free flap). These injuries are associated with significant wound contamination.

57.2.3.3 Type III C

All exposed fractures with severe arterial lesions that require prompt surgical repair, regardless of the type of fracture, are included here. Depending on the site and type of fracture, external fixators present different designs: in these type of injures, they allow the anatomical reduction of the fracture, the access to the wound in order to perform the debridement, and to prevent any compartment syndrome. Compartment syndrome is a condition in which increased pressure in an enclosed osteofascial space inhibits capillary perfusion necessary for tissue blood supply: compartment syndrome following tibial fractures is frequent in case of closed fractures (up to 20%) but may also occur with open ones. The first sign of a compartment syndrome is an excessive pain not relieved by rest or drugs in an at-risk patient. If untreated within 6 h, paresthesia and paralysis rapidly occur, followed by permanent muscle or nerve damage. Treatment generally requires a prompt surgical fasciotomy to decompress the compartment. Type III C fractures are treated with surgical internal fixation: plate and screws are used in case of proximal and distal tibial diaphyseal fractures that cannot be reduced with intramedullary nailing. The latter represents the best choice in tibial diaphyseal fractures: compared to the open reduction and fixation, it shortens the healing time and the need for unloaded position. The nail can be introduced through either an infra- or a supra-patellar access and is stabilized proximally and distally with screws. Contraindications for intramedullary nailing are a preexisting tibial shaft deformity that may preclude the passage of the nail, previous total knee arthroplasty (TKA), or previous tibial plateau open reduction and internal fixation (ORIF) that cannot be removed.

57.3 Ankle Fractures

Ankle fractures are the most common types of fractures worldwide: the treatment target is to achieve an anatomical consolidation of the fracture fragments and to restore the original ROM without pain. Typically, ankle fractures result from low-energy rotational traumas but, due to an increase in traffic accidents, the severity of fractures and the energy of traumas have significantly grown. As a result of a better understanding in the biomechanics of the ankle, there has been a gradual improvement in the effective treatment strategies of ankle fractures. Before starting the ankle fracture management, a risk-benefit and a cost-benefit analysis should be performed. The primary risk associated with closed treatment is an inadequate restoration of the biomechanics of the ankle, which may potentially lead to a poor outcome. Conversely, while open reduction and internal fixation is an excellent method for the restoration of the normal anatomy of the joint, it is accompanied by the costs and risks of a surgical intervention. A prompt evaluation of the patient with ankle fracture always includes a conventional radiographic investigation, often performed with frontal, lateral, and ankle mortise (15 degree of internal rotation of the leg) view. The reliability of the radiographic evaluation of an ankle fracture is limited, as all of the measure-

ments are subject to interobserver variability: furthermore, the fracture cannot be evaluated with a weight-bearing X-ray and, as a consequence, it does not provide essential information on the residual stability of the ankle joint. Despite all, treatment of ankle fractures is based on the radiographic evaluation: the Weber ankle fracture classification is a simple system for classification of lateral malleolar fractures, relating to the level of the fracture in relation to the ankle joint, specifically the distal tibiofibular syndesmosis: type A fractures occur distal to the level of the tibial plafond, type B fractures start at the level of the plafond and frequently continue as a spiral fracture proximally, and type C fractures originate proximal to the level of the plafond and are associated with a variable severity of syndesmotic injury. The AO classification system, more complete, is a modification of the Weber system in which the A, B, and C fracture types are subdivided on the basis of the presence of medial and posterior injury.

57.4 Isolated Lateral Malleolus Fractures

Isolated fractures of the lateral malleolus are the most common fractures of the ankle and are not associated with appreciable medial injury of either the deltoid ligament or the medial malleolus. The treatment for undisplaced fractures of the lateral malleolus consists of plaster immobilization with a protected load; treatment for displaced fractures is the open reduction and internal fixation. On the other hand, intermediate cases are still debated, with several studies showing that a displacement up to 3 mm may achieve a good healing also without surgery.

57.5 Isolated Medial Malleolus Fractures

These fractures are less common than other ankle fractures, such as lateral malleolar, bimalleolar, and trimalleolar fractures: it may be due to the deltoid ligament complex that provides stability to the medial side of the joint. If displaced, a restoration with internal fixation using either cancellous lag screws or tension band wiring is necessary.

57.6 Bimalleolar and Bimalleolar-Equivalent Fractures

A lateral malleolar fracture associated with a complete injury of the deltoid ligament is biomechanically equivalent to a bimalleolar fracture. These injuries are considered unstable, and surgery is generally recommended: nonsurgical treatment should be considered just in case surgery is contraindicated. Lateral and medial malleolus fractures are treated with the same surgical techniques as written above for each fracture listed. Deltoid ligament injuries are treated with side-to-side sutures or ligament reattachment with anchors to the bone.

57.7 Posterior Malleolar Fractures

The dynamics of this type of fracture can be caused by an external rotation associated with abduction of the lower limb: the mechanism of injury is generally an avulsion force acting through the posterior syndesmotic ligaments on the posterolateral part of the tibia. Less commonly, the mechanism is the impaction of the externally rotating talus on the posterior lip of the tibia. The primary issues regarding the treatment of a posterior malleolar fracture are the effect of the fracture fragment on the posterior stability of the ankle and what criteria should be used to determine when internal fixation is needed. Typically, being often associated with fractures of the lateral malleolus, the posterior malleolar fracture is reduced once the realignment of the latter is obtained. Most current texts recommend internal fixation with open or percutaneous reduction and fixation of the posterior malleolus if the reduced fragment involves more than one-fourth to one-third of the articular surface, or if intra-articular displacement persists with a step-off or gap of more than 2–3 mm. Fixation can be achieved by using a screw placed either posterior-to-anterior or anterior-to-posterior, depending on the size of the posterior fragment.

57.8 Trimalleolar Fractures

Trimalleolar ankle fractures are complex injuries. They are relatively rare, and their prognosis is worse than bimalleolar fracture. In addition to a reduced articular surface area of interaction, complex fractures involving both the lateral and posterior malleoli result in clinically significant posterior instability of the ankle (more severe in case of posterior fragments of 30–40% of the total articulating surface). They could be associated with ankle dislocation, and this typically happens in motor vehicle accidents; it may also happen while playing sports, especially those including jumping. Physical examination should comprise identification of soft-tissue changes such as swelling, skin tenting, open wounds, and blanching. An assessment of the neurovascular status should also be performed before the initial reduction attempts. Immediate closed reduction and immobilization should be performed to decrease the risk of neurovascular and skin complications, release soft-tissue tension, reduce the time of cartilaginous impingement, and procrastinate surgery if indicated; up to one-third of ankle fracture-dislocations present as open injuries and should be treated with external fixation. Closed reduction is generally successful; however, soft tissue may hinder reduction attempts. In posterolateral dislocations, a common block to reduction is the tibialis posterior tendon, which may need to be manually reduced before reducing the foot. A unique irreducible fracture variant was firstly described by Bosworth in 1947, in which the proximal fibular shaft fragment becomes incarcerated on the osseous posterolateral ridge of the distal part of the tibia. Clinical evaluation and postreduction attempt imaging guide the nonoperative or operative treatment of ankle fracture-dislocations. Surgical treatment aims to restore native ankle anatomy but may require special considerations to manage concomitant soft-tissue injuries, posterior malleolar involvement, osteochondral lesions, intra-articular loose bodies, and malreduction.

57.9 Syndesmotic Injuries

Stability of the ankle results from contributions of the medial complex (the medial malleolus and the deltoid ligament), the lateral complex (the lateral malleolus and the lateral ligament complex), and the syndesmotic complex; the latter lies between the distal tibia and the fibula and it is composed of the interosseous ligament (IOL), the anterior-inferior tibiofibular ligament (AITFL), the posterior-inferior tibiofibular ligament (PITFL), and the transverse tibiofibular ligament (TTFL). The instability of the syndesmosis is primarily based on the mechanism of injury and the fracture pattern. Clinical tests such as the squeeze test (manual medial-lateral compression across the syndesmosis) and the external-rotation stress test may elicit pain, but these tests are not reliable in the acute setting. A radiographic clear tibiofibular space lesser than 5 mm and widening of the medial clear space more than 4 mm are highly suspected for a syndesmotic injury. Recent studies have suggested that the need for trans-syndesmotic fixation may be lower than previously assumed: if the medial and lateral complexes are intact or can be restored anatomically and stabilized with internal fixation, the syndesmosis will be stable regardless of the degree of injury. Internal fixation of the syndesmosis may be needed if there is a fracture of the fibula that extends more than 3–4 cm proximal to the joint line as well as an associated medial-side injury that cannot be fixed or repaired (even if the fibular fracture has been already fixed anatomically). Intraoperative evaluation should determine if there is excessive lateral displacement, indicative of a syndesmotic injury after the internal fixation of the malleoli: many methods of fixation, including suture and use of synthetic grafts, have been reported, but fixation with screws is the technique most commonly adopted. The screw can be used independently or in conjunction with a plate, depending on the site and type of the fibular injury. The screw is inserted in the tibia at the top of the fibular sulcus, generally about three to four centimeters proximal and parallel to the ankle joint, and is angled approximately 30° anteriorly so that it is perpendicular to the tibiofibular joint. The foot should be in minimal dorsiflexion to avoid overtightening of the mortise and loss of dorsiflexion postoperatively. Screws incorrectly placed too much proximally may deform the fibula and widen the mortise. Moreover, screws that are not parallel to the ankle joint or not perpendicular to the tibiofibular joint may lead the fibula to shift proximally

or laterally. Non-weight-bearing in a cast or a fracture brace for 6–8 weeks is strongly recommended after surgery. The removal of the inter-syndesmotic wires, if needed, should be performed before weight-bearing, in order to avoid their breakage.

57.10 Maisonneuve Fracture

Jules Germain Francois Maisonneuve was a French surgeon who demonstrated the relationship between extrarotation and ankle fractures: in 1840, he described, using cadaveric studies, how an external rotation force applied to the foot could result in a fracture of the proximal third of the fibula. The Maisonneuve fracture is defined as a spiral fracture of the upper third of the fibula associated with rupture of the deep deltoid ligament or a fracture of the medial malleolus with a tear of the distal tibiofibular syndesmosis and the interosseous membrane. Fracture of the proximal fibula as a result of a pronation external rotation or supination external rotation mechanism of the foot and ankle should be differentiated from an isolated fibular fracture caused by direct force transmission from the lateral aspect. A proximal fibular fracture should always be investigated in the presence of an isolated medial malleolar fracture or a medial ankle ligament rupture: Maisonneuve fractures should be suspected whenever there is lateral talar displacement or tibiofibular widening without distal fibular fracture. Stress radiographs and full-length tibiofibular radiographs should always be obtained. Recommendations for these fractures include the medial malleolus fixation (the torn deltoid ligament does not need to be repaired). Syndesmotic instability can be treated with screws, and the proximal fibular fracture does not require direct internal fixation.

57.11 Tibial Pilon Fractures

"Pilon" is the French word standing for pestle, which means a tool like a small club, used for pounding things to powder, especially in a mortar. These fractures involve the weight-bearing surface of the ankle joint and typically occur as a result of a high-energy trauma, such as a car collision or falls from height. In most cases, surgery is needed to restore the injured bone to its normal position. Because of the energy required to cause a pilon fracture, patients may often present other severe injuries that require treatment as well. In addition to conventional radiology (that allows the patient to be assessed and treated urgently with a possible immediate closed reduction and temporary stabilization), a CT scan can provide valuable information about the severity of the fracture influencing the choice of treatment. Nowadays, the Rüedi and Allgöwer classification, integrated by Ovadia and Beals, is widely adopted: type I is an undisplaced, T-shaped fracture of the distal end of the tibia that extends into the joint; type II is the same as type I but with a displacement of the intra-articular components; type III is a complex, intra-articular, multifragmentary fracture. Types IV and V include fractures that extend to the metaphyseal and diaphyseal regions with severe comminution. As a result of the potential damage to the soft tissues surrounding this anatomical area, a careful evaluation of the lack of skin coverage that could arise consequently to surgery should be done. The emergency treatment in displaced fractures aims at the realignment of the limb, the recovery of the physiological axes of the lower limb, and the reduction of the swelling of the soft tissues. This can be achieved through trans-skeletal traction with a trans-calcaneal pin: when this solution is not feasible or in case of open fractures, the use of an external fixator becomes the best choice. At the same time, the fibula can be fixed. If the external fixation cannot be the definitive treatment, a careful planning of the surgical accesses and the internal devices to use should be performed: low-profile plates are used where the soft-tissue coverage is poor. Skin necrosis and superficial and deep infection with loss of fixation are the earlier postoperative complications. Late complications are delayed union or nonunion, loss of reduction of the articular surface, and varus or valgus deformities. Post-traumatic osteoarthritis may occur as a result of damage of the articular cartilage at the time of injury. It may also be a consequence of a nonanatomical reconstruction of the tibial pilon (fractures in which a congruous articular surface was not restored or maintained over time).

Take-Home Message

- Fractures of the leg are potentially serious injuries since they damage not only the bone but also the surrounding soft tissues, ligaments, and vascular/nervous structures.
- The patient should be carefully evaluated for initial signs of compartment syndrome, especially in case of fractures of the one-third middle leg, even if there is an open fracture.
- The radiographic projections available in the emergency setting are often not reliable for quantifying the true extent of the damage; therefore, knowing the dynamics of the trauma may help to suspect any associated injuries.

Summary

As a result of their incremental incidence rate, leg fractures (and in particular ankle fractures) are a debated and interesting topic. Thanks to the evidences produced by the researchers and the evolution of the internal fixation systems, the results of surgical treatment have greatly improved. Nowadays, it is possible to obtain immediate stability and early loading. The goal of the orthopedic surgeon is to promptly suspect potentially fatal complications associated with leg fractures and to treat them before they develop. Conventional radiological investigation should always be completed by clinical evaluation in order to exclude dangerous lesions and Maisonneuve fractures. In case of open fractures, in case of severely comminuted fractures with soft-tissue damage, or whenever definitive fixation is not possible, early stabilization and damage control are the first choice. It is necessary in order to reduce the risk of complications and plan the definitive surgery. The treatment target is to achieve an anatomical consolidation of the fracture fragments and to restore the original ROM without pain.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. What is the correct treatment approach in a patient with a tibial plateau fracture?
   (a) Firstly, soft-tissue damage should be excluded and an arthrocentesis should be performed in order to reduce joint pressure and therefore pain. The knee should be immobilized in minimal flexion with a long brace and heparin prophylaxis initiated, when not contraindicated. If necessary, an external bridge fixator can be positioned to maintain alignment while waiting for definitive treatment.
   (b) Firstly, the knee should be immobilized in minimal flexion with a long brace and heparin prophylaxis initiated, when not contraindicated. Then, if necessary, an external bridge fixator can be positioned to maintain alignment while waiting for definitive treatment.
   (c) Firstly, soft-tissue damage should be excluded and an arthrocentesis should be performed in order to reduce joint pressure and therefore pain. The knee should be immobilized in maximal flexion with a short brace and heparin prophylaxis initiated, when not contraindicated. If necessary, an internal fixation device can be considered for definitive treatment.
   (d) Firstly, the knee should be immobilized in maximal flexion with a long brace and heparin prophylaxis initiated, when not contraindicated. Then, if necessary, an internal fixation device can be considered for definitive treatment.
2. What are the most frequent complications of an intramedullary nailing of the tibia?
   (a) About 50% of patients complain of anterior knee pain if the nail is introduced with an infrapatellar approach. Malrotation, cubitus valgus or varus, and pseudoarthrosis are less frequent complications.
   (b) Malrotation, cubitus valgus or varus, and pseudoarthrosis are frequent complications. About 20% of patients complain of posterior knee pain if the nail is introduced with an infrapatellar approach.
   (c) Malrotation, cubitus valgus or varus, and pseudoarthrosis are common complications. About 30% of patients complain of anterior knee pain if the nail is introduced with an infrapatellar approach.

(d) Malrotation, cubitus valgus or varus, and pseudoarthrosis are prevalent complications. About 10% of patients complain of posterior knee pain if the nail is introduced with an infrapatellar approach.

3. In the surgical treatment of trimalleolar fractures, in addition to the medial access for fixation of the internal malleolus, which other incision is useful in the simultaneous treatment of the lateral and posterior malleolus?
   (a) Posterolateral approach to the fibula with patient in lateral decubitus allows to treat both the fracture of the external malleolus and the posterior malleolus, if the latter is not reducible with the ankle in dorsiflexion.
   (b) Anterolateral approach to the fibula with the patient in anterior decubitus allows to treat both the fracture of the external malleolus and the posterior malleolus, if the latter is not reducible with the ankle in dorsiflexion.
   (c) Anteromedial approach to the fibula with the patient in prone position allows to treat both the fracture of the external malleolus and the posterior malleolus, if the latter is not reducible with the ankle in dorsiflexion.
   (d) Posteromedial approach to the fibula with the patient in supine position allows to treat both the fracture of the external malleolus and the posterior malleolus, if the latter is not reducible with the ankle in dorsiflexion.
4. What is the only dislocation of the ankle that cannot be reduced with external maneuvers?
   (a) It is the Bosworth fracture/dislocation of the ankle.
   (b) It is the Maisonneuve fracture/dislocation of the ankle.
   (c) It is the Monteggia fracture/dislocation of the ankle.
   (d) It is the Galeazzi fracture/dislocation of the ankle.
5. What is the postoperative treatment after a trans-syndesmotic fixation?
   (a) The aim of surgically stabilizing the syndesmosis is to allow the healing of the structures that have experienced a laceration with the trauma. Its stabilization is done after fixing any coexisting malleolar fractures. It is a surgeon's choice to remove the screws before giving the load to the patient, after about 4–6 weeks postoperatively, or to leave them in place with the possibility that they break over time.
   (b) The aim of surgically stabilizing the syndesmosis is to allow the healing of the structures that have experienced a laceration with the trauma. Its stabilization is done after fixing any coexisting malleolar fractures. It is a surgeon's choice to remove the screws before giving the load to the patient, after about 1–2 weeks postoperatively, or leave them in place with the possibility that they break over time.
   (c) The aim of surgically stabilizing the syndesmosis is to allow the healing of the structures that have experienced a laceration with the trauma. Its stabilization is done after fixing any coexisting malleolar fractures. It is a surgeon's choice to remove the screws before giving the load to the patient, after about 8 weeks postoperatively, or leave them in place with the possibility that they break over time.
   (d) The aim of surgically stabilizing the syndesmosis is to allow the healing of the structures that have experienced a laceration with the trauma. Its stabilization is done after fixing any coexisting malleolar fractures. It is a surgeon's choice to remove the screws before giving the load to the patient, after about 12 weeks postoperatively, or leave them in place with the possibility that they break over time.

Further Reading

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Forefoot Deformities

58

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Overview
Forefoot deformities comprise a wide spectrum of pathologic conditions with a high prevalence in the worldwide population. This chapter aims to focus on the most prevalent diseases.

58.1 Hallux Valgus

58.1.1 Introduction

The term hallux valgus was originally introduced by Carl Hueter to define a subluxation of the first MTP joint characterized by lateral deviation of the great toe and medial deviation of the first metatarsal. Currently, it is understood that hallux valgus deformity can occur because of lateral deviation of the articular surface of the metatarsal head without any subluxation of the first MTP joint.

Hallux valgus deformity can also be associated with abnormal foot mechanics, such as a pes planus, shortened Achilles tendon, generalized neuromuscular diseases, or secondary to a hindfoot deformity. In addition, it can occur with various inflammatory arthritic conditions, such as rheumatoid arthritis.

58.1.1.1 Pathoanatomy and Pathophysiology

A congruent MTP joint is inherently more stable than an incongruent or subluxated joint. A congruent joint tends to remain stable, whereas once a joint has begun to subluxate, the deformity tends to progress with the passing of time.

Incongruent or subluxated hallux valgus deformities are usually progressive. Movement of the proximal phalanx laterally over the metatarsal head causes a medially directed pressure on the head pushing it medially and causing an increased intermetatarsal (IM) angle. This causes an attenuated medial capsule and a contracted lateral joint capsule. Simultaneously, the sesamoid sling, which is anchored laterally by the insertion of the adductor hallucis muscle and the transverse metatarsal ligament, remains in place as the metatarsal head moves medially, creating added pressure on the medial joint capsule. The medial joint capsule is weakest just superior to the abductor hallucis muscle where it ultimately gives way, allowing the abductor hallucis muscle to gradually slide beneath the medially deviating metatarsal head. This will thereby result in causing the intrinsic muscles to become a deforming force and become

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unable to support the MTP joint. Also due to the fact that this abductor hallucis muscle is attached to the phalanx, it will spin the proximal phalanx into pronation. It has been shown that as the hallux valgus deformity progresses, so does this degree of pronation. This pronation will cause calluses to develop on the medial side of the IP joint. Eventually, as the MTP joint becomes less stable, the hallux carries less weight, body weight is transferred laterally in the forefoot, and callus may also develop beneath the second, third, or both metatarsal heads. Increased pressure may lead to capsulitis, instability, or deviation of the second MTP joint as well.

With more severe degrees of this hallux valgus deformity, the extensor hallucis longus tendon is also displaced laterally, and this also leads to it not only extending the toe but also adducting it, further worsening the deformity. The flexor hallucis longus tendon, which retains its relationship to the sesamoids, moves laterally and also becomes a dynamic deforming force (Fig. 58.1). In rare circumstances, if the progressive deformity of the MTP joint continues unabated, dislocation of the MTP joint may occur over time, with the fibular and tibial sesamoids becoming dislocated into the first IM space.

Hallux valgus deformities are also associated with a splayed appearance of the foot. Initially, this might occur because the first metatarsal head is no longer contained within the sesamoid sling and is displaced in a medially deviated position. The middle metatarsals do not splay because of the stable articulation at their tarsometatarsal joints. Sometimes, the fifth metatarsal lacks stability and drifts laterally, thereby completing the appearance of a splayed foot.

With further drifting of the hallux laterally, the other lesser toes come under increasing pressure, especially the second toe. If the second MTP joint remains stable, the great toe may drift beneath it or occasionally on top of it. In other instances, subluxation or complete dislocation of the second MTP joint occurs. Additionally, occasionally, all the lesser toes are pushed into lateral deviation or a "wind-swept" appearance.

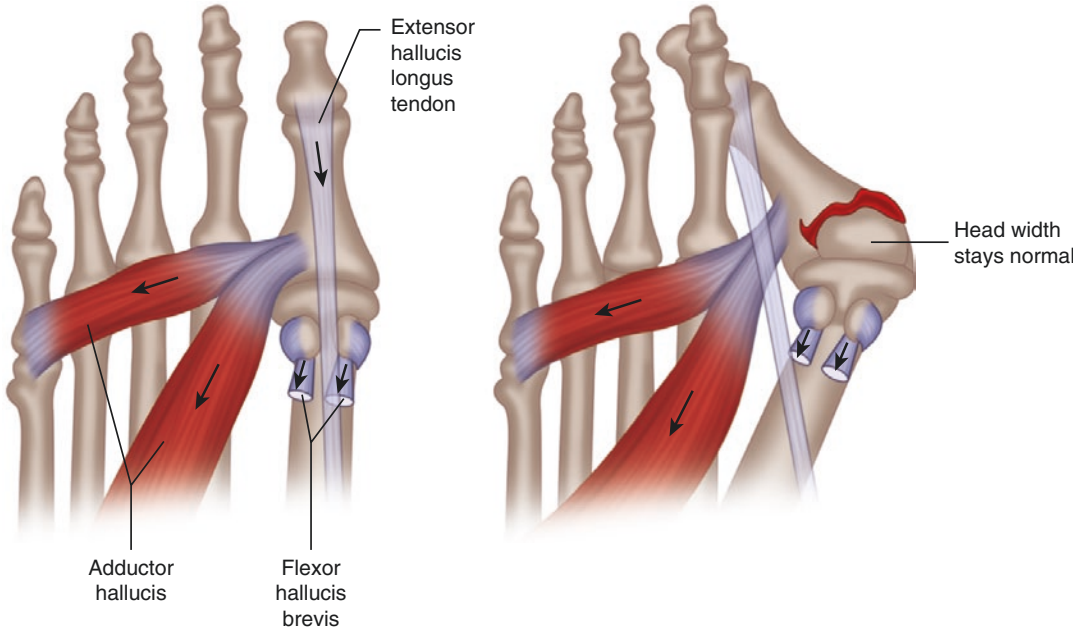

Fig. 58.1 Pathophysiology of hallux valgus deformity. With lateral deviation of the proximal phalanx, medially directed pressure is applied on the metatarsal head, which deviates medially. This results in attenuation of the medial joint capsule and contracture of the lateral joint capsule. Additionally, the extensor hallucis longus and flexor hallucis brevis tendons become deforming forces by their displacement medially to the metatarsal head

58.1.2 Demographics

Myerson has suggested that bunions occur in 2–4% of the population. Almost 46% of bunions have been reported to occur before the age of 20 years. The mean age at which patients had surgery for the deformity was 60 years of age.

Regarding gender distribution, several studies have found a female preponderance of up to 90%.

Although it has been reported that almost 84% of patients had bilateral hallux deformities, yet only 18% had both sides corrected.

58.1.3 Etiology

The hallux valgus deformity can be caused by extrinsic or intrinsic factors. The most common extrinsic factor is reportedly *shoe wear*. Some authors have reported *trauma* with rupture of medial joint capsule as a causative factor for the development of the deformity.

The most commonly cited intrinsic factor is a *hereditary factor*. On the other hand, the association of hallux valgus with *pes planus* is controversial. Another factor is *pronation of the foot*. This causes more stress on the first MTP joint, which assumes a more oblique orientation with the ground resulting in more pressure being exerted on the medial joint capsule and thereby causing a progression of a hallux deformity. Another intrinsic factor is *hypermobility of the first metatarsocuneiform joint*. Some authors have also described *ligamentous laxity* as a causative factor in the development of hallux valgus deformity. Some authors have suggested that an *Achilles contracture* might occur with hallux valgus. Others noted no such correlation. There are other miscellaneous causes of hallux valgus. *Second toe amputation* has been shown to be a cause of the deformity. *Syndactylization* of first and second toes has also been shown to be a cause.

58.1.4 Radiographic Parameters and Specific Anatomic Implications

The classical radiographic studies have been described as weight-bearing standard AP, lateral, and oblique radiographic views. The AP view has been classically described as taken at 1 m tube-to-film distance centered on the tarsometatarsal joints and angled 15° towards the ankle joint. On this weight-bearing AP view, some important angular measurements should be assessed (Fig. 58.2).

The *intermetatarsal angle* is the line formed between the longitudinal axis of the first and second metatarsal bones. It is normally usually less than 9°. Mild deformity has been described as between 9° and 11°, while moderate is defined between 11° and 16° and any degree of deviation above 16° is considered severe.

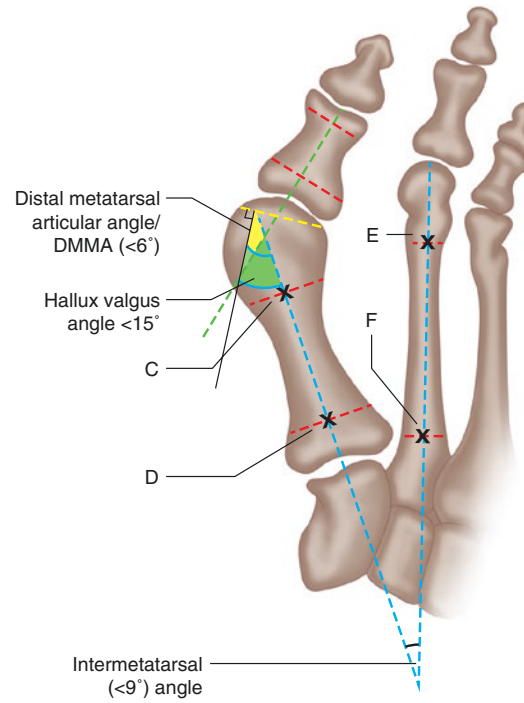

Fig. 58.2 Schematic representation of angular measurements on weight-bearing radiograph in a case of hallux valgus deformity. Points C and D are mid-diaphyseal points along the axis of the first metatarsal. Points E and F are the mid-diaphyseal points along the axis of the second metatarsal. *Intermetatarsal angle* (red colored angle) is formed between the axis of the first and second metatarsal bones (lines CD and EF). *Distal metatarsal articular angle* (yellow-colored angle) is formed between the axis of the first metatarsal (line CD) and a line that is drawn perpendicular to the line joining the most medial and most lateral points of the first distal metatarsal articular surface (yellow dotted line). *Hallux valgus angle* (green-colored angle) is the angle formed between the axis of the first metatarsal (line CD) and the axis of the proximal phalanx (green dotted line)

The *distal metatarsal articular angle* is the angle of lateral slope of the articular surface on the first metatarsal axis. It is formed between a line perpendicular to another line connecting the most medial and most lateral points on the articular surface and between the first metatarsal axis. Normally, it has been defined as being less than 6°.

Hallux valgus angle is the angle of deviation of the proximal phalanx on the first metatarsal and is formed between the lines representing the longitudinal axis of both the first metatarsal and the first proximal phalanx. Normally, it has been defined at 15°. Mild deformities are less than 20° moderate between 20° and 40°, and severe are defined as larger than 40°.

Similarly, the *hallux valgus interphalangeal angle* is formed between the longitudinal axis of the distal and proximal phalanx.

An important point of assessment is the *congruency of the first metatarsal joint surfaces*, between the respective articular surfaces of the first metatarsal head and the proximal phalanx base. It should be distinguished from mere subluxation where corresponding points on the articular surface of the base of the proximal phalanx migrate laterally in relation to first metatarsal head articular surface (Fig. 58.3). It has been stated that no lateral shift of the proximal phalanx will occur with a congruent deformity.

Another point of interest is the *medial eminence*. Authors have described the medial eminence as being new bone formation and a bunion. Others believe that it is not a new bone but rather a part of the metatarsal that becomes exposed with lateral deviation of the proximal phalanx. Although medial eminence resection has become a standard part of hallux valgus corrective surgery, it should be stated that basing this resection on the sagittal sulcus may result in excessive bone resection and may be a cause for hallux varus deformity.

Metatarsus primus varus has also a close association with hallux valgus. While some authors report that it in itself is a cause for the development of the deformity, others believe that it might actually be a cause of the deformity. It is still a matter of debate.

The *first metatarsocuneiform joint* (MTC joint) is also thought to play an important role in hallux valgus deformities. The shape of this joint has been described as flat, curved, or oblique. Some authors have observed that with increasing "medial obliquity" of this joint, a hallux valgus deformity would be more likely to occur. Other authors have questioned the fact that this orientation is actually an apparent orientation on the radiographs. Coughlin and Anderson have observed in cadaveric dissections that the first MTC joint at the cuneiform articulation has a superior dorsal convex surface and a flatter concave plantar and inferior surface. This might in itself allow for the variable appearance of the joint on radiographic projections. Also, as previously stated, stability of this first MTC joint is of importance in hallux valgus deformities. Some authors have found a decrease in the intermetatarsal angle following distal soft-tissue releases only or following concomitant metatarsal osteotomies. This might lead one to believe that sufficient mobility is present at the first MTC joint to allow for surgical correction of an increased intermetatarsal angle.

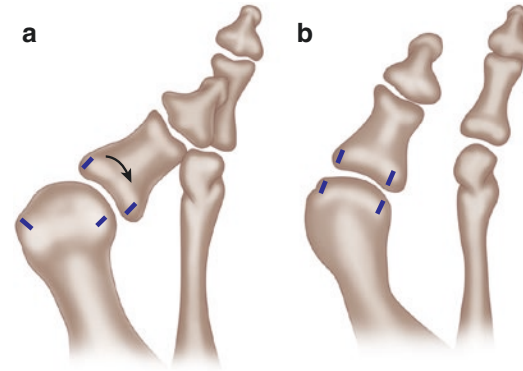

Fig. 58.3 Difference between (**a**) subluxated (incongruent) and (**b**) congruent articular surfaces of first metatarsal joint articular surfaces

The blood supply of the metatarsal head is of particular anatomical importance. It usually arises from a nutrient artery that traverses from lateral to the midshaft of the first metatarsal in a distal direction where it divides into distal and proximal branches. Shereff et al. have shown that the primary blood supply arises from the first dorsal metatarsal artery, first plantar metatarsal artery, and superficial branch of the medial plantar artery. Most of this blood supply penetrates the joint capsule through the dorsal and lateral aspects. The clinical implication of this is that

while performing a capsulorrhaphy or a metatarsal osteotomy, the blood supply is dependent on the remaining vessels and thus a wide soft-tissue dissection might result in avascular necrosis of the metatarsal head.

58.1.5 Classification

The purpose of any classification system should be to ultimately dictate management options. However, with that in mind, one should acknowledge that classification systems for hallux valgus deformities are not rigidly established and should be used as a general guide (Table 58.1).

58.1.6 Patient Evaluation

Similar conditions in the family should also be investigated. The onset of symptoms whether adolescent or later and also the patient's level of activity, occupation, sporting participation, type of shoe wear preference, and consistency with symptoms should be investigated. Additionally, one should thoroughly investigate the reason for surgery and the degree of intensity of symptoms. Some points of consideration would be to inquire about transfer metatarsalgia, callosities or corns, and lesser toe deformities.

As with most foot and ankle disorders, physical examination of the patient should include the gait observation, and foot examination while standing and while sitting. One should document the degree of the hallux deformity, hallux pronation, lesser toe deformity, as well as state of the medial longitudinal arch and the hind foot position.

While examining the first MTP joint range of motion, one should note any crepitus or painful motion. Passive ROM of the first MTP has been reported to be almost 87°.

Examination of the mobility of the first MTC joint should also be performed. This is usually performed with the patient sitting knee flexed, with ankle in neutral and forefoot stabilized with one of the examiner's hand while the thumb and index finger of the other hand hold the first metatarsal and move it from a dorsomedial to plantar-lateral direction. It is the opinion of some authors that even if instability is present, merely realigning the hallux restores stability to the first MTC joint.

58.1.7 Management

58.1.7.1 Conservative Management

Shoe wear with wide toe box and soft soles may give significant relief from patients' symptoms. The use of custom or prefabricated orthotics has not been found to be of value to these patients. Actually, some authors have concluded that these orthotics do not prevent progression of the deformity and even in some cases might increase the hallux valgus angle. Conservative care might perhaps be considered more in patients with hyperlaxity, or those with neuromuscular disorders due to the high recurrence rates.

58.1.7.2 Surgical Management

The patient must be informed that perhaps some residual stiffness or minor reduction in their daily activities might occur.

Broadly when one is faced with a hallux valgus deformity, the congruency and condition of the first MTP joint will play a pivotal role in the procedure selection. If from the offset the joint is degenerative, then a fusion or MTP joint replacement should be considered. On the other hand, if this

Table 58.1 Hallux valgus classification

Hallux degree	HV angle	IM angle	MTP joint	Hallux position	Sesamoid position
Mild	>20°	>11°	Congruent	Might be mild pronation	Normal or 50% subluxation of lateral sesamoid
Moderate	20°–40°	11°–16°	Usually subluxated joint	Pronated	75–100% subluxation of lateral sesamoid
Severe	>40°	>16°–18°	Significant subluxated joint	Moderate or severe pronation	100% subluxation of lateral sesamoid

joint is with normal cartilage, one should examine congruency: if it is congruent or subluxed (proximal phalanx is subluxed on the MT head or not). A working algorithm has been proposed for such deformities. Although several authors might propose different methods of management, this algorithm sets general broad lines of management and one can use it as a general rule of thumb.

Generally, the concept behind this algorithm is that when the proximal phalanx is congruent, then the procedures utilized should be mainly extracapsular to maintain normal relationship between the phalanx and metatarsal head. Conversely, when the phalanx is subluxed on the metatarsal head, a soft-tissue release would be required to restore normal joint congruency. Additionally, the procedure selected might be based on the degree of the deformity present whether mild, moderate, or severe (Fig. 58.4).

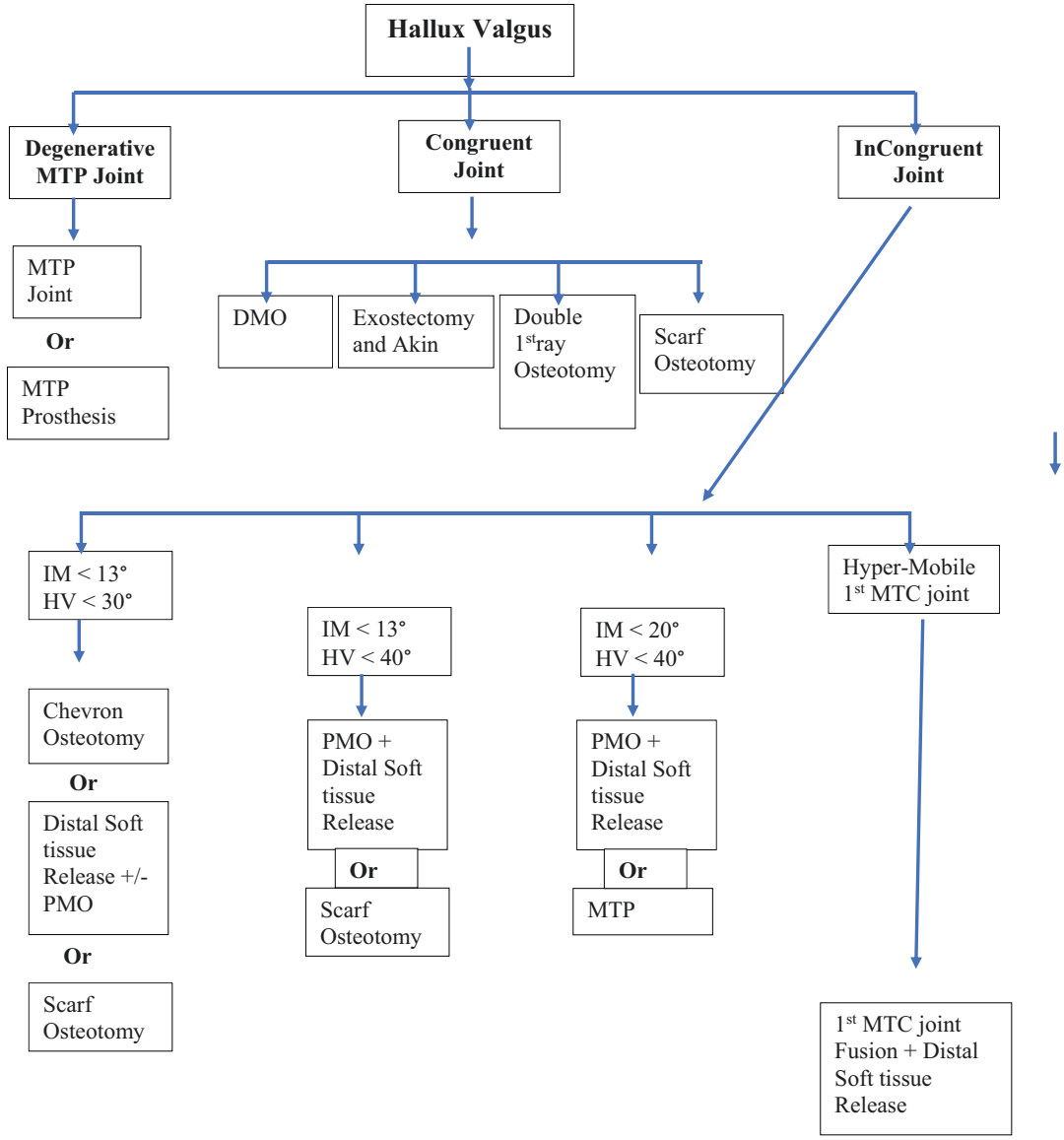

Fig. 58.4 Proposed algorithm for management of hallux valgus deformities. *MTP* metatarsophalangeal, *DMO* distal metatarsal osteotomy, *PMO* proximal metatarsal osteotomy, *IM* intermetatarsal angle, *HV* hallux valgus angle, *MTC* metatarso-cuneiform

When performing a lateral soft-tissue release, the basic concept is the release of the constrained lateral structures, namely the lateral joint capsule, the transverse metatarsal ligament, and the adductor hallucis, thereby reallowing the proximal phalanx to be realigned on the metatarsal head. A medial incision might be performed separately for the removal of the medial eminence and a medial capsulorrhaphy or plication. An Akin procedure is a minimal medial closing wedge resection osteotomy of the proximal phalanx. A chevron osteotomy was so-named by the chevron or V-shaped osteotomy that is performed in the distal metatarsal head. Various modifications and methods of its fixation have since been introduced. This is one of the distal metatarsal osteotomies; others are the Bosch or the Mitchell distal osteotomies. Among the most common metatarsal osteotomies is the scarf osteotomy. It was originally described as a longitudinal Z-type of osteotomy of almost the whole length of the first metatarsal bone.

58.2 Hallux Rigidus

It has been reported that in 2.5% of patients over 50 years, a degenerative arthritis of the first metatarsophalangeal joint (MTPJ) shall be present. This is termed "hallux rigidus" and is the most common arthritic condition that might affect the foot. This condition is associated with pain and loss of motion of the big toe. Usually, conservative management is the first line of treatment, and later surgical management might be warranted with options being removal of excess osteophytes, osteotomies, or fusion of the first MTPJ.

58.2.1 Evaluation

Normal first MTPJ range of motion is usually 75° dorsiflexion and 35° plantar flexion. Pain and joint stiffness that will both worsen with activity are the main complaints of patients with hallux rigidus.

On examination, one might find dorsal osteophytes and tenderness localized to the dorsal joint. Some authors have described a "grind test," which is pain on compression of the first MTPJ.

Radiographs are classic standing anteroposterior oblique and lateral projections. Joint space narrowing, flattening, and widening of the metatarsal head with subchondral sclerosis will be evident. Dorsal osteophytes of the proximal phalanx and metatarsal head might also be evident. Other diagnostic modalities are usually not required. Coughlin and Shurnas described a classification system based on both radiographic and clinical findings.

58.2.2 Management

58.2.2.1 Nonsurgical Management

Management of the condition should start with nonoperative treatment. Nonsteroidal anti-inflammatory drugs, activity modification, orthotics designed to limit motion across the MTPJ, and shoe modifications with high and wide toe boxes are the usual options available. Some authors have suggested steroid injections or hyaluronic acid injections into the first MTPJ.

58.2.2.2 Operative Management

Surgical management options are removal of excess osteophytes, osteotomies, or fusion of the first MTPJ.

Cheilectomy

This was originally described by DuVries in 1959; it involves resection of the dorsal one-third of the articular cartilage of the metatarsal head along with the associated osteophytes on the dorsal surface of both the head and the phalanx.

Moberg Osteotomy

Described as a closing wedge dorsiflexion osteotomy of the proximal phalanx by Boney and Macnab in 1952, it was designed to shift the arc of motion of the first MTPJ to more dorsiflexion.

Keller Resection Arthroplasty

This involves the resection of the proximal base of the phalanx to increase dorsiflexion but at the

expense of stability of the MTPJ. This procedure has been associated with several complications as cock-up deformities of the big toe, weakness at toe-off, and transfer metatarsalgia.

Interpositional Arthroplasty

This is essentially a Keller resection but with the addition of a biological spacer into the joint. Various modifications have been described for the biological spacers, preservation of the flexor hallucis brevis insertion, use of gracilis tendon as interposition, and various other modifications.

MTP Arthroplasty

Historically, the arthroplasty has progressed from silastic implants to all metal implants and then synthetic cartilage implants. The results for each of these types have not been very promising with high failure and loosening rates reported.

First MTPJ Arthrodesis

This is still considered the "gold standard" of treatment for cases with MTPJ arthritis.

58.3 Bunionette Deformity

This is a lateral bony prominence at the fifth metatarsal head that is sometimes painful, especially with constricting shoe wear. The condition was originally described in tailors and hence the name "tailor's bunionette," and this was attributed to the fact that they would be seated working cross-legged causing pressure and friction on the lateral aspect of the foot.

58.3.1 Clinical Evaluation

More commonly, patients will describe pain on the lateral aspect of the foot with pressure from shoe wear. With examination, one should assess hindfoot alignment and pes planus deformities. Hallux valgus and increased width of the foot may be noticed. While the patient is seated, one should examine for lateral eminence swelling, redness ulceration, or keratosis. In particular, location of any callosity is of importance; for example, if the callosity is plantar, one should plan for a correction that involves not only translation but also elevation of the metatarsal head.

Normal radiographic projections and standing anteroposterior, lateral, and oblique views are usually obtained. Most measurements are made on the anteroposterior views similar to hallux valgus deformities. These measurements are metatarsal head width, intermetatarsal angle, and fifth metatarsophalangeal angle.

58.3.2 Management

58.3.2.1 Nonsurgical Management

Very good results have been reported with the use of conservative management for treatment of bunionette deformities although strong high-grade evidence is still lacking. Wide shoes, custom-made orthotics, and pads can alleviate pain.

58.3.2.2 Surgical Management

Surgical options are reserved for failure of conservative treatment. They range from lateral eminence resection to different osteotomies. Adequate randomized controlled studies are still lacking.

58.3.2.3 Lateral Eminence Resection

This might be reserved for patients who exhibit an isolated prominent lateral condyle as this only reduces the mass effect.

58.3.2.4 Distal Fifth Metatarsal Head Osteotomy

Transverse, oblique, and chevron-type osteotomies have been described.

58.3.2.5 Diaphyseal Fifth Metatarsal Osteotomy

This technique is probably reserved for patients with increased four to five intermetatarsal angle or with significant lateral metatarsal shaft bowing.

58.3.2.6 Proximal Fifth Metatarsal Osteotomy

This osteotomy was designed to correct increased four to five intermetatarsal angles but is associated with higher rates of nonunion due to impaired blood supply in the region.

58.3.2.7 Metatarsal Head Resection

This may be considered as a salvage procedure for painful bunionettes, failed osteotomies, rheumatoid forefoot, infections, and neuropathies with ulcers.

58.4 Lesser Toe Deformities

Of all foot problems, those involving the lesser toes are more common with almost 20% incidence. Also, they seem to occur more in females and more in older individuals.

Coughlin and Mann defined the principal deformity to be occurring at the metatarsophalangeal joint (MTPJ). So, a *hammertoe deformity* will have no deformity at the MTPJ but flexion of the middle and possibly the distal phalanges. *Claw toe deformities* will show hyperextension at the MTPJ and flexion of the proximal interphalangeal joint (PIPJ) and the distal interphalangeal joint. However, in most cases, the distinction is not that evident and both deformities can coexist together.

58.4.1 Etiology

Some authors agree that an imbalance between the intrinsic (flexors) and the long extensors plays a role. Others have implicated tight shoe wear causing a crowding of the toes and high heels that might also result in hyperextension at the MTPJ. Another common association is with hallux valgus deformity. Other etiological causes are inflammatory conditions (as rheumatoid or psoriasis) or diabetes or other neurological conditions. In particular, claw toes are associated with the latter condition.

58.4.2 Clinical Examination

As with most foot and ankle conditions, the patient should be examined standing, seated, and during gait or walking. While standing, examination should include assessment of cavus deformities, hallux valgus, or hindfoot malalignments. With the patient seated, one should also examine both the plantar surface of the foot and the dorsal aspects of the toes for callosities or ulcers and try to assess whether the toe deformities are fixed or mobile at both the MTPJ and the interphalangeal joints.

Standard weight-bearing radiographs of the feet with anteroposterior, lateral, and oblique views should be requested.

58.4.3 Management

58.4.3.1 Nonsurgical Management

Obviously, a change of shoe wear is warranted. Usually, the patient is advised wide toe box shoes to accommodate the digits. Tapping and strapping of the toes might be effective in flexible deformities. Additionally, silicon pads or felt pads might be useful for the painful callosities. Nonsteroidal anti-inflammatory medications might be used for pain alleviation.

58.4.3.2 Surgical Management

Generally, if the MTPJ becomes subluxed or dislocated, the management would be usually surgical. Multiple procedures have been described for lesser toe deformity management. These can be broadly classified into soft tissue, bony procedures, or joint destruction procedures.

Soft-Tissue Procedures

Dhukaram and colleagues evaluated a sequential soft-tissue release in the management of lesser toe deformities. The sequence suggested was extensor digitorum longus (or Z-lengthening), extensor digitorum brevis release, dorsal capsule release, collateral ligament release, and reduction of the plantar plate. The authors added a Weil osteotomy plus a flexor to extensor transfer if MTPJ extension remained after those sequential releases.

58.4.3.3 Bony Procedures

If deformity of the MTPJ is severe with dislocation of the proximal phalanx, soft-tissue release may be insufficient and a bony "decompression" might be necessary. Such options as shortening osteotomy of metatarsal head, partial hemiphalangectomy, or metatarsal head arthroplasty might be used.

Metatarsal Shortening Osteotomy

The concept is to relief plantar pressure and reduce the hyperextended or dislocated MTPJ.

Partial Proximal Hemiphalangectomy

This involves excision of the proximal phalanx base.

PIP Joint Arthrodesis

PIP joint arthrodesis may provide a stable pain-free construct.

Take-Home Message

- The term hallux valgus defines a subluxation of the first MTP joint characterized by lateral deviation of the great toe and medial deviation of the first metatarsal.
- Myerson has suggested that bunions occur in 2–4% of the population. The mean age at which patients had surgery for the deformity was 60 years of age.
- The most common extrinsic factor is reportedly shoe wear.
- Hallux rigidus has been reported in 2.5% of patients over 50 years and a degenerative arthritis of the first metatarsophalangeal joint (MTPJ) shall be present.
- Bunionette deformity is a lateral bony prominence at the fifth metatarsal head that is sometimes painful, especially with constricting shoe wear.
- Very good results have been reported with the use of conservative management for treatment of bunionette deformities, although strong high-grade evidence is still lacking.

Summary

The term hallux valgus was originally introduced by Carl Hueter to define a subluxation of the first MTP joint characterized by lateral deviation of the great toe and medial deviation of the first metatarsal. Currently, it is understood that hallux valgus deformity can occur because of lateral deviation of the articular surface of the metatarsal head without any subluxation of the first MTP joint. Incongruent or subluxated hallux valgus deformities are usually progressive.

Myerson has suggested that bunions occur in 2–4% of the population. Almost 46% of bunions have been reported to occur before the age of 20 years. The mean age at which patients had surgery for the deformity was 60 years. The hallux valgus deformity can be caused by extrinsic or intrinsic factors. The most common extrinsic factor is reportedly shoe wear. Patient's level of activity, occupation, sporting participation, type of shoe wear preference, and consistency with symptoms should be investigated. Shoe wear with wide toe box and soft soles may give significant relief from patients' symptoms. The patient must be informed that perhaps some residual stiffness or minor reduction in their daily activities might occur.

Hallux rigidus has been reported in 2.5% of patients over 50 years, and a degenerative arthritis of the first metatarsophalangeal joint (MTPJ) shall be present. Normal first MTPJ range of motion is usually 75° dorsiflexion and 35° plantar flexion. Pain and joint stiffness that will both worsen with activity are the main complaints of patients with hallux rigidus. Management of the condition should start with nonoperative treatment. Surgical management options are removal of excess osteophytes, osteotomies, or fusion of the first MTPJ.

Bunionette deformity is a lateral bony prominence at the fifth metatarsal head that is sometimes painful, especially with constricting shoe wear. More commonly, patients will describe pain on the lateral aspect of the foot with pressure from shoe wear. Very good results have been reported with the use of conservative management for the treatment of bunionette deformities, although strong high-grade evidence is still lacking. Surgical options are reserved for failure of conservative treatment. They range from lateral eminence resection to different osteotomies.

Of all foot problems, those involving the lesser toes are more common with almost 20% incidence. So, a hammertoe deformity will have no

deformity at the MTPJ but flexion of the middle and possibly the distal phalanges. Claw toe deformities will show hyperextension at the MTPJ and flexion of the proximal interphalangeal joint (PIPJ) and the distal interphalangeal joint.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Hallux valgus is frequently associated with:
   (a) Transverse flat foot
   (b) Hollow foot
   (c) Flat foot
   (d) Club foot
2. Hallux valgus occurs in:
   (a) 2–4% of the population
   (b) 5–10% of the population
   (c) 10–20% of the population
   (d) 20–30% of the population
3. The most common extrinsic factor causing hallux valgus is:
   (a) Shoe wear
   (b) BMI
   (c) Age
   (d) Gender
4. The intermetatarsal angle is the line formed between the longitudinal axis:
   (a) Of the first and second metatarsal bones
   (b) Of the second and third metatarsal bones
   (c) Of the third and fourth metatarsal bones
   (d) Of the fourth and fifth metatarsal bones
5. Normal first MTPJ range of motion is usually:
   (a) 75° dorsiflexion and 35° plantar flexion
   (b) 35° dorsiflexion and 75° plantar flexion
   (c) 15° dorsiflexion and 25° plantar flexion
   (d) 5° dorsiflexion and 55° plantar flexion

Further Reading

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Foot and Ankle Tendinopathies

59

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Overview
Tendinopathies in general continue to represent a challenging clinical topic for all health professionals involved in dealing with patients (global population, athletes, more often those who perform repetitive actions).

59.1 Biology and Biomechanics

The etiology of tendinopathy is considered to be multifactorial, and its pathogenesis remains unclear, surrounded by doubts and blank spots. Tendons act as structures which transfer forces from muscles to bones. Their key component is a complex hierarchical collagen structure, which is interlaced with several non-fibrillar proteins. This structure is essential to the capacity of tendons to sustain load transfer while preserving its integrity and stability. The key resident cells of tendons are called "tenocytes." These cells are capable to "sense" force transmission from the extracellular matrix (ECM), and in return they act by regulating the ECM. Considering the abovementioned factors, loading is indispensable for tendon homeostasis, but it can promote either remodeling or degeneration. The complete knowledge of the pathway by which loading can have a "healthy" or deteriorating effect in any specific tendons remains unclear.

Mechanical loading and changes to regular loading patterns have been suggested to play a critical role in tendon's response to stimulus, possibly leading to tissue degeneration. Once tendons are composed of collagen fibrils, these are prepared for energy storage and have progressed/adapted mainly to function as tensile load transmitters. Thus, they reduce the pattern of force transmission to the attached muscle. The mechanical behavior of tendons is typically presented as stress-strain curves, which include three typical phases: (1) the toe region where 2% of strain is applied (enough to remove any resting fiber crimp, but unable to cause injury to the tendon's structure), (2) the linear region where around 4–8% of strain loading is applied and causes elongation of fibers that might lead to some fiber damage (microscopic failure), and (3) the failure region characterized by complete breakage of fibers (macroscopic failure) at loads superior to 12%.

Fatigue has been registered as a possible response to certain patterns of mechanical loading. Such changes are considered to make tendons more prone to degeneration and disease.

The occurrence of tendinopathies has been correlated to aging (less effective biological and mechanical response to "aggression"). Similarly, "disuse" has been presented as another risk factor once it induces changes in tendon properties. Denervation and immobilization have also been suggested to decline tendon's stiffness and strength. On the other hand, an increase in physical activity is recognized to improve tendon's mechanical properties such as by inducing changes to the collagen physiology.

Under some conditions (such as "healthy exercise"), repetitive loading promotes tendon's remodeling inducing long-term structural and functional benefits. Tendon remodeling comprises a balance between synthesis and degradation of collagen. Immediately after exercise, collagen net degradation begins and afterwards changes to a net synthesis. Adult tendon healing is characterized by scar formation with disorganized tissue and diminished mechanical properties. In contrast, regeneration, as seen in fetal healing and nonmammalian vertebrates, is characterized by restoration of the native structural and functional properties of the tissue, without scar. While therapeutic interventions to improve scar-mediated healing are an advancement to the field, the ultimate unmet goal is to promote regenerative healing. Differences between normal versus tendon tissue in every location and specificity must be acknowledged, given the objective to achieve full tissue repair (biological and biomechanical) in future (Fig. 59.1).

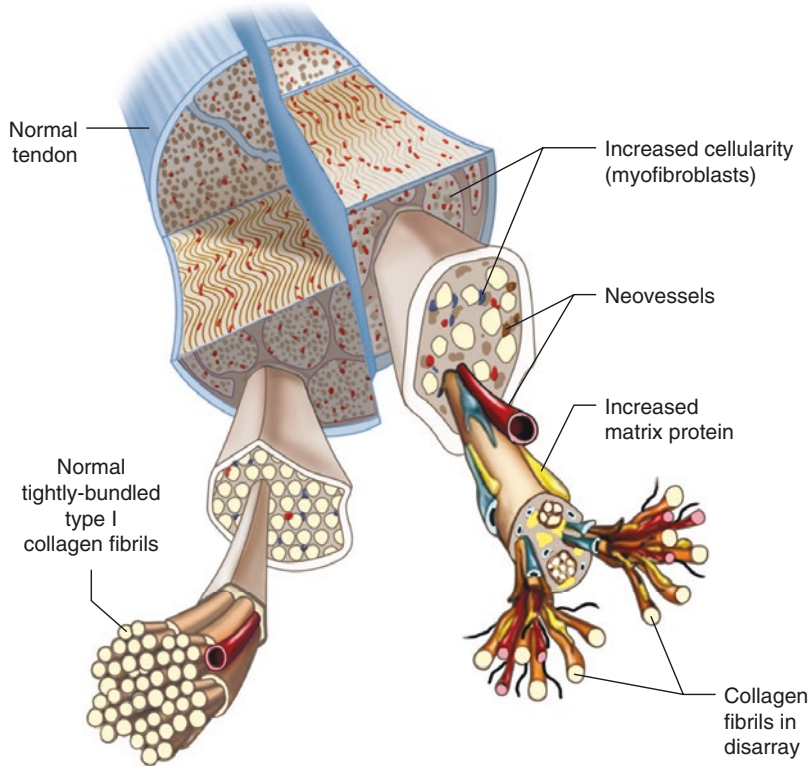

Fig. 59.1 Normal tendon versus tendinopathy

59.2 Overall Concepts on Treatment for Tendon Pathologies

The treatment of tendinopathies aims to achieve the control of symptoms, stimulate the regeneration of the tendon, and restore normal biomechanics. Tenocytes have a low mitotic rate, which must be considered in any therapeutical approach. Moreover, the response pathways of tendons to injury, micro-repeated trauma, and degeneration are not fully understood. The use of systems enabling controlled release of medicines (e.g., nanoparticles) might bring new solutions for future.

Percutaneous injection therapy (Fig. 59.2) has been tried including "stem cells," growth factors, and several combinations of the former with biomaterials. However, further studies are required in order to establish the outcome and indication for each of these uprising possibilities.

Conventional conservative treatments have been used aiming to reduce pain and inflammation. These options include rest and/or activity modification, cold, stretching, braces, physiotherapy, and biomechanical correction. The treatment is often provided in the acute setting. However, these techniques do not restore the changes in the ultrastructure of the tendon, once the structuring and maturation of collagen fibers require mechanical stress, adequate physical and biological environment, and time.

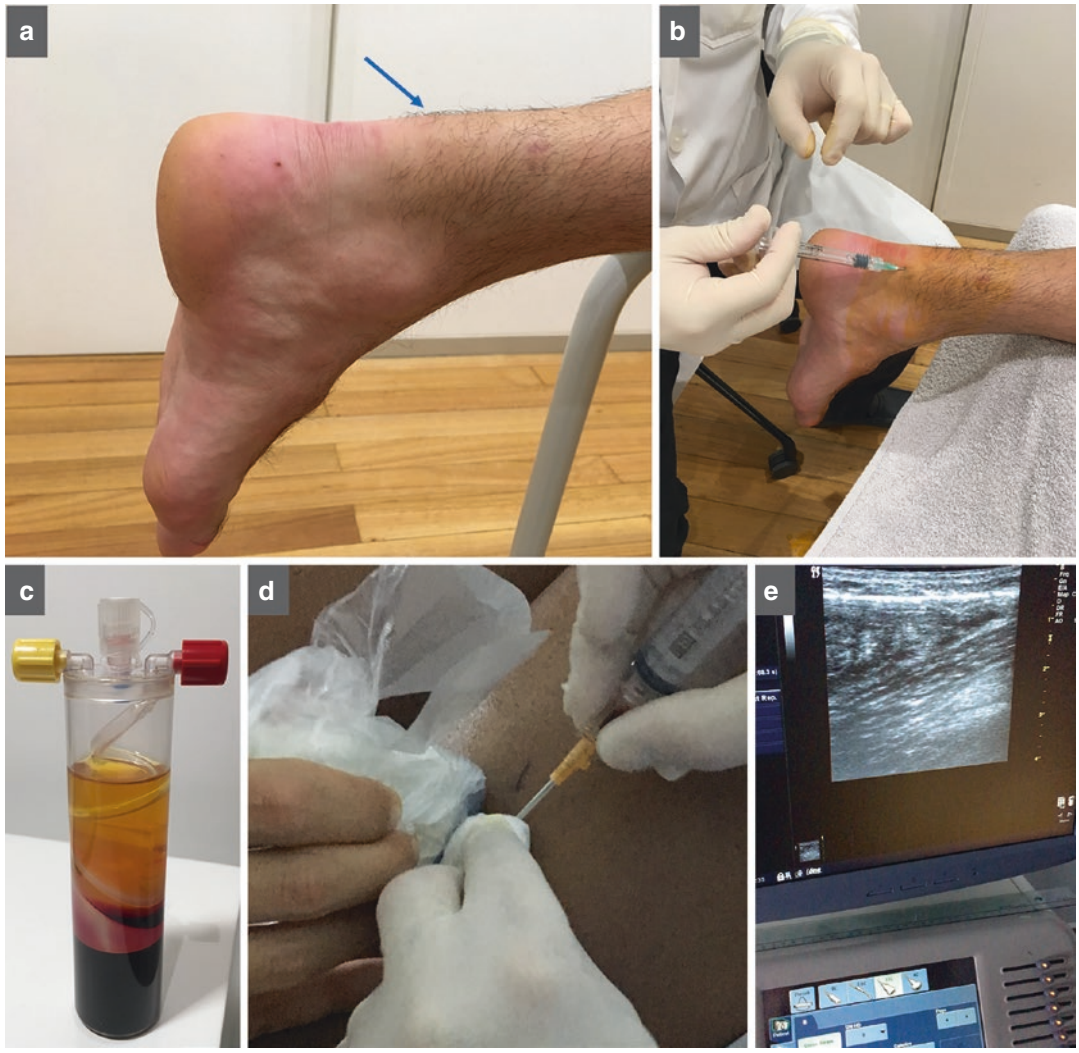

Fig. 59.2 Blinded injection of hyaluronic acid in the Achilles tendon sheath (**a** and **b**); platelet-rich plasma (PRP) growth factors (**c**); ultrasound-guided injection (**d** and **e**)

59.3 Achilles Tendinopathies

Injuries related to the Achilles tendon are common problems and some of the most researched topics regarding the foot and ankle. These can involve the tendon, paratenon, or surrounding bursas. Injuries affecting the tendon can be divided into insertional and non-insertional. Since it is difficult to establish a correct histologic diagnosis between tendinitis and tendinosis, the term tendinopathy or paratendinopathy is preferred, depending on whether the tendon or paratenon is affected.

59.3.1 Diagnosis and Treatment

59.3.1.1 Midportion Achilles Tendinopathy

Achilles tendinopathy is one of the most commonly studied disorders of the foot and ankle. The incidence rate of midportion Achilles tendinopathy in the Dutch population is 2.35 per 1000 registered patients with ages between 21 and 60 years, with 35% of the cases being related with sports activity.

This clinical syndrome is characterized by pain and swelling, which can be localized or diffuse, between 2 and 7 cm from the calcaneal insertion, causing impaired performance.

In this disorder, only the tendon proper is affected. Histopathological diagnosis is not limited to tendinosis, and it should be noted that the fundamental lesion of tendinopathy is failed healing response. Treatment for non-insertional Achilles tendinopathy is largely conservative. Initial rest, modification of training regimes, specific exercises, and correction of underlying lower limb alignment issues with orthotics are some of the treatment modalities. Most patients will be able to return to previous activities: in an 8-year follow-up study, only 29% failed to respond adequately to nonoperative management.

Injection therapies can also be effective treatments. Platelet-rich plasma (PRP) injections also produce good overall results, with a stable outcome at medium- and long-term follow-up, but these seem to be less effective on aged patients.

Conventional surgical treatments entail the release of adhesions, which may be combined with resection of the paratenon. If using an open approach, a longitudinal tenotomy can be performed and macroscopic areas of tendinopathic tissue are removed. Additional multiple tenotomies can be performed to initiate vascular ingrowth and a healing process. Augmentation is recommended if more than 50% of the tendon has to be debrided (24). Success rates for open surgery vary widely but are generally reported as being between 75 and 100%. Stripping of the paratenon is thought to be effective by removing the neovascularization and denervate the diseased area of the tendon.

59.3.1.2 Achilles Paratendinopathy

Paratendinopathy is defined as inflammation, either acute or chronic, with or without degeneration, of the thin membrane surrounding the Achilles tendon. Most important clinical features are pain induced by exercise and swelling around the midportion of the tendon. Acute cases will have swelling and crepitation, with the latter not being present in midportion Achilles tendinopathy. Swelling and crepitations will be less pronounced in chronic cases. Typically, in patients who have acute symptoms, the area of swelling and tenderness does not move when the ankle joint is dorsiflexed. It should be noted that Achilles paratendinopathy frequently coexists with midportion tendinopathy.

If surgical treatment is warranted, an endoscopic approach can be used to attain release of adhesions and resection of the paratenon, similarly to what was described above for midportion Achilles tendinopathy.

59.3.1.3 Insertional Achilles Tendinopathy

This is a condition affecting the calcaneal insertion of the Achilles tendon and may be accompanied by the formation of bone spurs and calcifications in this area. Clinically, there is pain on palpation at the tendon insertion, swelling may be present, and a bony spur may be palpable. This disorder tends to affect more active persons, in contrast with non-insertional tendinopathy which tends to affect older, less active, and overweight patients. An epidemiologic study in nonathletes found Achilles tendinopathy in 5.6% of the patients, with 4% being insertional and 1.9% being both insertional and non-insertional.

Several nonoperative treatment modalities have been proposed, including eccentric stretching and strength training and heel lifts. When conservative measures fail, surgical treatment is indicated. This can be achieved through a central incision with complete detachment of the Achilles tendon, debridement of diseased tissue, removal of spurs, and reattachment with the suture bridge technique (Fig. 59.3).

Achilles insertional tendinopathy surgery results in few complications with good functional results if the surgical technique is adapted to the type of tendon injury.

When augmentation is felt to be needed, bone-quadriceps tendon grafting is also a good alternative for the insertional Achilles lesions with partial detachment.

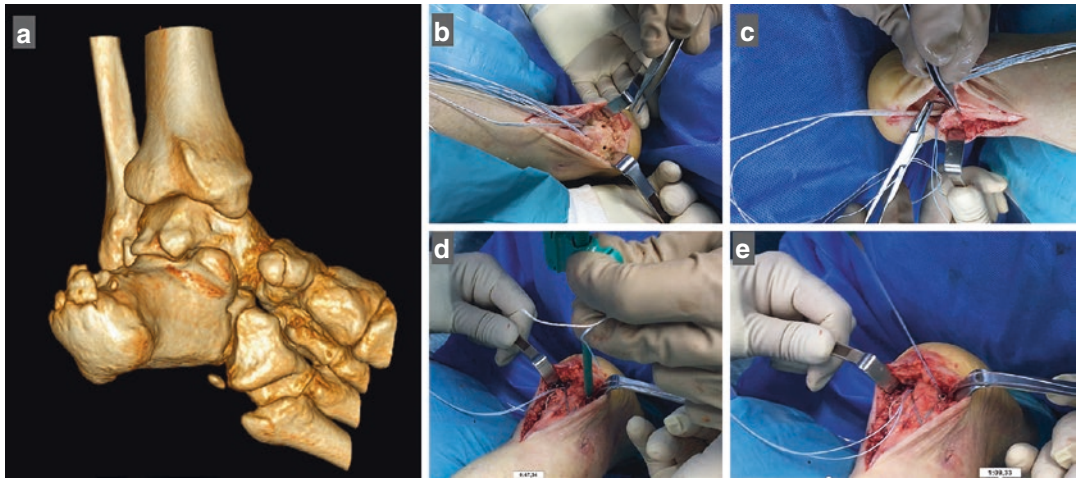

Fig. 59.3 (**a**) CT image demonstrating severe Achilles insertional tendinopathy with multiple calcifications; (**b**–**e**) detachment of the Achilles, removal of calcifications and posterior process of the calcaneus; reinsertion of the Achilles with double-row suture anchors

59.3.1.4 Retrocalcaneal Bursitis

This disorder is defined as an inflammation of the bursa in the retrocalcaneal recess, which is located between the anterior inferior side of the Achilles tendon and the posterosuperior facet of the calcaneus. This manifests as a visible and painful soft-tissue swelling, on the medial and lateral sides of Achilles tendon at the same level of the posterosuperior calcaneus.

It is common to find an increased prominence of the posterior superior tuberosity on plain radiography, but there is no correlation between prominence dimension and symptoms. Patients with prominent posterior superior calcaneal tuberosities can stay asymptomatic all life, and patients with a normal calcaneus can have recalcitrant symptoms. Patients with hindfoot varus and cavus foot have a higher predisposition for this disorder.

Conservative treatment includes multiple physical therapy protocols. Shoe changes or modifications can also produce positive effects, especially in athletes during the competitive season. Corticosteroid injections should be avoided in retrocalcaneal bursitis because of a connection between the retrocalcaneal bursa and the anterior fibers of the Achilles tendon, which puts the tendon at risk of rupture.

59.4 Anterior and Posterior Tibialis Tendinopathies

Besides the needs related to normal gait, several popular sports activities require specific repetitive technical gestures involving foot and ankle.

Fine-tuned neuromuscular control and proprioceptive mechanisms related to foot and ankle are specifically needed for walking in uneven ground and several sports-related actions. The former comprises mechanisms of fine balance between dorsiflexion-plantar flexion and pronation-supination. In brief, the "ankle joint complex" includes the tibiotalar (talocrural), talocalcaneal (subtalar), and transverse-tarsal (talocalcaneonavicular) joints.

Plantar- and dorsiflexion take place mainly at the tibiotalar joint, and pronation-supination mostly occurs at the subtalar joint; however, tibiotalar and talocalcaneonavicular also contribute. Foot pronation is a complex movement which includes abduction of forefoot, eversion of hindfoot, and dorsiflexion. Supination also occurs at tri-dimensions of the space by combining internal ankle rotation, hindfoot adduction, forefoot inversion, and rise of the medial arch. The supination movement creates an increase of foot stability, while pronation promotes higher degree of flexibility.

The *tibialis posterior* (combined with the *flexor digitorum longus* and the *flexor hallucis longus*) causes foot plantar flexion and inversion. The *tibialis anterior* (synergistically with the *extensor hallucis longus*) leads to dorsiflexion as well as some degree of foot's inversion. The *extensor digitorum longus* is only implicated in dorsiflexion. Contrariwise, the peroneal muscles (PMs), including the *peroneus longus*, *brevis*, and *tertius*, play the role of active evertors of the foot. Plantar flexion, as previously mentioned, is mainly provided by the *triceps surae* and *plantaris muscles*. These can be considered "the key muscles controlling ankle motion."

As part of the fundamentals of biomechanics of foot and ankle, one must recognize the central role of the navicular bone providing distal support to the talus. The navicular is the highest structure of the longitudinal medial arch in the standing position. In the static, standing position, the equilibrium of these structures depends on the contiguous bones and the spring ligament (calcaneonavicular). However, while in movement, the activity of the posterior tibial muscle (PTM) holds the superior position of the navicular, consequently assisting in the support of the medial arch.

Considering the abovementioned facts, a debilitated PTM (e.g., neurologic diseases) or an insufficient posterior tibial tendon (PTT) is incompetent to maintain the navicular in its place and the medial arch might "fall" (leading to acquired flatfoot condition) (Fig. 59.4).

Another example of the complex biomechanics of the foot is the dorsiflexion moment at heel strike as the dorsiflexors (e.g., tibialis anterior) eccentrically contract in order to assist the rotation of the foot, thus impairing the foot from smacking into the ground.

Foot and ankle principles of clinical assessment always include clinical examination (including weight-bearing), global assessment, alignment of inferior limbs in standing position, specific tests, and painful spots and gait analysis.

Shoe wear inspection is mandatory in the assessment of foot and ankle pathologies (e.g., focal compression leading to inflammatory response). Moreover, this might assist in raising suspicions of certain conditions once uneven

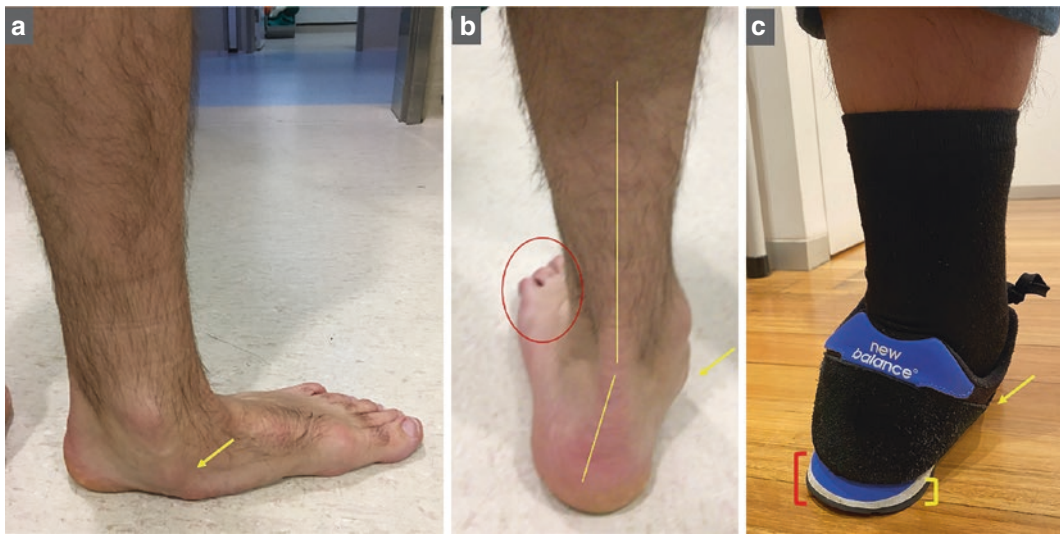

Fig. 59.4 (**a**) Medial arch fall due to posterior tibial tendon (PTT) insufficiency (yellow arrow); (**b**) adult pes planus-valgus with hindfoot valgus (yellow lines); "too many toes sign" (red ellipse) secondary to PTT insufficiency (yellow arrow); (**c**) medial arch fall due to PTT insufficiency (yellow arrow); notice shoe deformity and the difference between yellow and red brackets traducing the deformity of the shoe sole

wear of shoe's soles or form might suggest malalignment.

It is critical to listen to the patient's complaints: related events such as trauma, worsening, and relief factors. Patient's expectations must be considered in the choice of therapeutic options.

The most relevant pathologies affecting anterior tibial tendon (ATT) and PTT will be herein considered. Although infrequent**,** combined disease of both tendons has been described leading to severe flatfoot and drop foot gait.

59.5 Peroneal Tendinopathies

Despite being considered in the past as rare entities, knowledge based on recent studies has ensured that peroneal tendon disorders are a relevant cause of posterolateral ankle symptoms. Pathology may range from tendinopathy to ruptures, tears, and instability/dislocation of the tendons. As chronic peroneal tendinopathy is related to long-term sequelae, correct diagnosis and adequate treatment as soon as possible are of paramount relevance.

59.5.1 Peroneal Tendon Pathology

59.5.1.1 Peroneal Tendinopathy

The term "tendinopathy" generally describes a disease of a tendon with complaint of pain and swelling, as previously explained. Most tendon-related problems, particularly in high-level athletes, are connected to overuse. The second most frequent etiology is traumatic events (contusions, sprains, fractures); however, morphological foot/ankle patterns and sports-specific actions must also be acknowledged. As a general rule, in the presence of lateral foot and ankle pain, it is mandatory to rule out concurrent ankle instability. Clinical examination will elucidate tenderness on palpation, local edema, and aggravation of symptoms during active eversion against resistance. We emphasize that conservative treatment is the first option: rest, ice, massage therapy, eccentric exercise, NSAIDs, ultrasound therapy, electrotherapy, taping, glyceryl trinitrate patches, last-generation (ESWT) extracorporeal shockwave therapy, intra-tissue percutaneous electrolysis (EPI), insoles, and shoe changes.

If conservative approach fails, peroneal tendoscopy (Fig. 59.5) might be considered.

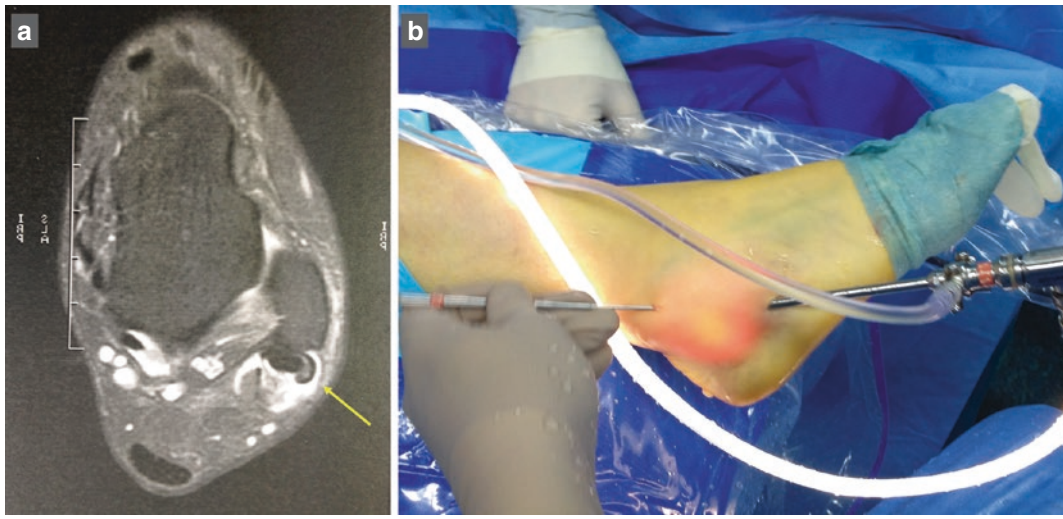

Fig. 59.5 (**a**) MRI demonstrating longitudinal rupture of peroneal brevis (yellow arrow); (**b**) two-portal technique for peroneal tendoscopy

59.5.1.2 Peroneal Tendon Ruptures

In an asymptomatic, low-demanding patient, a complete rupture of a tendon can be treated conservatively. Symptomatic, complete rupture, in demanding/active patients, of one of the two peroneal tendons should usually be treated with repair. If repair is not possible, a single-stage hamstring autograft or tenodesis may be performed.

59.6 Flexor Hallucis Longus Tendinopathies

Disorders of the flexor hallucis longus (FHL) have traditionally been described in the context of injuries afflicting female classical ballet dancers. The forced hyperplantarflexed positioning of the ankle practiced during ballet (particularly *en pointe*) might create direct compression of the FHL while kinking the tendon when it enters the fibro-osseous tunnel posterior to the talus, causing a painful tenosynovitis. There have been sporadic reports of FHL tenosynovitis in nondancers, but this has been termed a "rare" occurrence. The FHL originates from the posterior border of the fibula and interosseous membrane in the leg. Afterwards, it passes through retinacular structures posterior to the ankle (the fibro-osseous tunnel), the plantar midfoot (the knot of Henry), and the sesamoids (the intersesamoid ligament) before its insertion at the base of the distal phalanx of the hallux. In each of the described areas, the FHL is potentially exposed to constriction that can either cause or exacerbate a tenosynovitis. Similarly, the restraint to motion that can occur at each of these sites in the presence of peritendinous swelling can lead to focal symptoms (pain) as well as a change in the biomechanical function of the FHL. As an example, nodular swelling of the proximal FHL has been associated with limitation of hallux dorsiflexion, a condition called a "pseudo" hallux rigidus because hallux motion is restored when the ankle is plantarflexed (which permits the FHL to retract proximally away from the constricting fibro-osseous tunnel).

The FHL has been successfully used for the treatment of other tendinopathies, particularly Achilles chronic or acute ruptures or chronic tendinopathy and insufficiency with success.

Take-Home Message

- Tendinopathies can be either acute or chronic entities even if a "grey area" where such distinction is not so easy has been considered.
- Moreover, a lack of consensus concerning diagnostic tools and treatment modalities remains.
- The clinical management of foot- and ankle-related tendinopathies requires familiarity with the anatomy and biomechanics of the lower limb, besides comprehension of the challenging topic of tendinopathy in general, as well as the specificities related to foot and ankle.

Summary

The clinical management of foot- and ankle-related tendinopathies requires familiarity with the anatomy and biomechanics of the lower limb, besides comprehension of the challenging topic of tendinopathy in general, as well as the specificities related to foot and ankle.

There is no consensus in classification, and there are no straight, flawless, guidelines for management. However, this topic has recently been a focus of growing research, developments, and increased clinical experience from basic science to tissue engineering and regenerative medicine, and from conservative to surgical treatments. Based on the former, a systematic, careful clinical approach, considering the most up-to-date information and clinical knowledge, will most likely lead to improved outcome of those suffering from these pathologies.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Degenerative changes in the posterior tibial tendon are a possible cause of:
   (a) Medial malleolus fracture
   (b) Adult acquired flatfoot
   (c) Recurrent ankle sprains
   (d) Achilles tendon rupture
2. Which of these muscles is not innervated by the external popliteal sciatic nerve?
   (a) Extensor hallucis longus of the big toe
   (b) Tibialis anterior
   (c) Peroneal
   (d) Posterior tibial
3. The diagnosis of subcutaneous rupture of the Achilles tendon is:
   (a) Clinical
   (b) Ultrasound
   (c) Radiological in magnetic resonance imaging
   (d) Clinical radiological in ultrasonography
4. Which of these tendon ruptures occurs most frequently?
   (a) Patellar tendon
   (b) Quadriceps tendon
   (c) Distal head of the biceps brachii
   (d) Achilles tendon
5. Partial ruptures of the Achilles tendon are generally found as a result of the following:
   (a) Malleolar fractures
   (b) Tendinosis
   (c) Hollow foot
   (d) Flatfoot

Further Reading

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Ligament Injuries and Instability

60

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Overview

Ankle ligament injuries are common, and there are several high-level research studies that have studied the epidemiology, injury grading and treatment, both non-surgical and surgical.

60.1 Introduction

It is well known that ankle sprain is the most common ankle injury related to sports. In fact, in the past, it has repeatedly been shown to be the most common sports-related injury overall. Because it is so common, it is costly. It has also been shown that most acute ligament injuries can be treated with non-surgical therapy. The conservative treatment protocol is based on a short period of rest, mobilisation, strength and agility training and a well-guided return to sports, with structured rehabilitation. However, 10–30% (or even more) of all ankle ligament injuries do not experience full functional recovery after non-surgical treatment. The question of which patients will or will not respond to non-surgical treatment has recently been discussed in the literature. One factor that affects the clinical outcome after anterior talo-fibular ligament surgery is cartilage damage, especially on the medial side of the joint. The post-operative period for starting activities depends on cartilage damage and laxity remaining post-operatively. Attention should therefore be paid to the intra-articular damage at the time of the index injury.

As mentioned above, approximately 10–30% of all ankle injuries will have long-term problems and eventually develop chronic lateral ankle instability. Some researchers even claim that as many as 40% of acute ligament injuries will result in chronic or recurrent ankle instability. For most of these patients, non-surgical treatment for 3–6 months is recommended as primary treatment. First after a period of supervised non-surgical treatment of this kind, including an individually designed rehabilitation protocol, surgery should be considered.

The two ligaments that are most commonly injured are the anterior talo-fibular ligament and the calcaneo-fibular ligament. In only very few cases is the posterior talo-fibular ligament damaged as well (major injury). An injury to the anterior tibio-fibular ligament (syndesmosis injury) is a different kind of injury mechanism, and concomitant syndesmosis injuries are not frequent.

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The most common injury mechanism is a combination of supination (inversion) and forefoot adduction, at the same time as the foot is in plantar flexion. The first injury is to the anterior talo-fibular ligament, leading to antero-lateral rotatory instability (laxity) of the ankle. In the event of an additional injury to the calcaneofibular ligament, this commonly leads to the tilting of the talus as well (talar tilt).

Ankle ligament injuries are commonly graded on the basis of their severity, i.e. grades I–III. In this context, grade I (mild) is described as minor stretching/bleeding of the ligaments, but without any macroscopic rupture or increased laxity. This type of injury can always be treated using non-surgical measures. Grade II (moderate) is defined as a partial injury/rupture of the ligament(s), accompanied by moderate swelling and bleeding. Moderate pain is also present. Patients with grade II injury often have problems with weight-bearing and functional impairment, with slight to moderate instability, but, as a rule, they can be treated non-surgically, with good results. A grade III injury (severe/major) is defined by a total ligament rupture, and the pain is marked. On objective investigation, there is marked swelling and haematoma. This injury is always accompanied by an impairment of function, reduced range of motion and functional instability. There is some debate about whether these injuries should be treated by stabilising surgery in the acute phase. Most researchers, however, claim that non-surgical treatment is the first line, with good options for secondary treatment if non-surgical treatment fails and the patient/athlete exhibits functional instability.

The healing phases can be divided into the *inflammatory phase*, up to 10 days after injury; the *proliferation phase*, up to 8 weeks; and the *remodelling/maturation phase*, which may take up to one year or even more. It should therefore be remembered that healing may take time. Due to the fact that the proliferation phase takes months, the ankle is often painful for a long time, and long-term rehabilitation and external ankle support are often needed for a long period.

There are several, mostly well-known, treatment alternatives, such as acute-phase surgery (stabilisation), immobilisation, followed by functional treatment, external support (brace and/or tape), neuromuscular training and strength training in the acute phase. Finally, there are several surgical options when acute, non-surgical treatment is not successful. One common reason is that many ankle sprains are more serious injuries than previously anticipated, as several patients develop symptoms and medium- to long-term impairment, with pain and/or instability and recurrent giving way. The most common problems are chronic/recurrent pain, either chronic or recurrent swelling and chronic instability, which may require surgical correction. The most common reasons for failure after acute-phase ankle sprain treatment are concomitant injuries, such as cartilage and, more uncommonly, syndesmosis injuries. However, the most overwhelming reason for chronic injury is probably inappropriate treatment (or minimal or no treatment) of the acute injury, especially for patients with a higher injury grade.

One problem that should not be neglected is the risk of recurrent injury. In fact, there is evidence showing that athletes run twice the risk of recurrent ankle sprain within one year after the index injury, indicating that first-time injury may lead to a second injury. Preventive measures to avoid a second injury are important. Due to the fact that ankle sprains are so frequent, attempts to avoid them are of great interest. Accordingly, prevention has been studied in great detail in recent years.

Ankle ligament injuries are common, and there are several high-level research studies that have studied the epidemiology, injury grading and treatment, both non-surgical and surgical. In spite of this, there are still unresolved questions, such as the efficacy of mini-invasive treatment (stabilisation) and return to sports.

60.2 Prevention

Can ankle ligament injuries be prevented at all? This question has been raised in several well-performed studies over the last 20 years. Tropp was probably the first to advocate preventive

measures, back in the 1980s. He studied balance boards and a simple ankle brace and found that both were effective in football players in lower divisions in Sweden. After a lateral ankle ligament injury, it is recommended that exercise therapy should always be included and added to regular training activities in order to prevent a recurrence. It clearly appears that prevention is most effective in preventing a recurrence in the event of a previous injury.

More than 50 years ago, Freeman stated that balance and co-ordination training was successfully able to reduce the proprioceptive deficits associated with ligament injury to the ankle. Since then, this type of training has been the mainstay of preventive measures for ankle ligament injuries. The question of how effective this type of training is, however, has been the subject of debate. It is suggested that balance and co-ordination training may have both a central and a local effect on the sensorimotor system. However, there is no consensus on the clinical evidence relating to the effectiveness and efficacy of these interventions. There have been well-powered randomised studies that have reported fewer recurrent sprains after 12 months' follow-up, when a training programme of balance and proprioception has been implemented. Even though some studies have been unable to show differences in the occurrence of re-sprains or subjective recovery, they have shown positive effects in terms of improved ankle function and activity. Researchers have concluded that balance training can be used after an acute ankle sprain in order to reduce future ankle sprains.

When it comes to the prevention of injuries, it has previously been shown that the most important risk factor for an ankle sprain is a previous ankle sprain. This may also be related to impaired proprioceptive function after the injury, such as slower reaction times (peroneal muscle reaction times) and postural sway deficits in ankles that suffer from functional instability. Those that have sustained an acute ankle sprain and those with functional instability will therefore require rehabilitation aimed at improving proprioception, strengthening the calf muscles and restoring peroneal reaction times (if possible). Taken together, there is evidence that supervised balance training is useful in reducing the number (and possibly the magnitude) of future ankle sprains in athletes with a previous injury.

Braces and/or sports tape have been much used in order to prevent ankle ligament injuries. Several studies have shown that the use of lace-up braces is effective in reducing the number of injuries, but not their severity, especially in football, volleyball and basketball players. One study has compared the prophylactic use of a brace and sports tape in football players. Both remedies were found to be effective, while there was no difference in terms of the rate of ankle sprains between the two study groups. A cost analysis has shown that a brace was significantly more cost effective than tape. A recent meta-analysis has also confirmed evidence of the beneficial effect of an ankle brace in preventing ankle sprains during high-risk sports activity, such as football.

Based on the evidence from high-level randomised trials, it can be concluded that the use of a brace (or sports tape) is effective in preventing ankle sprains. It should be borne in mind that most football players prefer sports tape to a brace for practical reasons. This preference is mostly dependent on the individual situation, where a brace is preferable to tape as a result of practical usability and cost-effectiveness. The phasing-out of the brace or tape over time is recommended. No recommendations can be made, however, when it comes to any type of shoe to prevent ankle sprain. Workers should preferably be treated with a brace in order to resume work activity at an earlier stage. It should also be recommended that exercise therapy should be included as much as possible in regular training activities, already at an early stage.

60.3 Chronic Ankle Instability: Treatment

The indications for further (in most cases surgical) treatment are persistent pain, recurrent instability in spite of stabilising treatment and repeated giving way. Chronic lateral ankle instability is a

multifactorial condition that hampers activities in daily life and even more so in terms of sports activities. There may be several predisposing factors for this condition, such as hindfoot varus, cavovarus foot morphology and generalised joint laxity. All these need to be considered when surgical treatment is planned. In particular, it has been shown that generalised joint laxity leads to an inferior outcome after ligament reconstruction, compared with patients with no such laxity. All these entities may play a role in the development of ankle joint laxity, with a combination of mechanical and functional instability. Accordingly, when non-surgical treatment is not effective, surgical reconstruction should be performed in order to maintain the integrity and stability (laxity restoration) of the lateral ankle ligaments and to prevent further sprains/dislocations. The goal is to restore and maintain normal contact mechanics and motion patterns of the ankle and hindfoot.

Different surgical options have been described (and much used) since the original report by Broström in 1966; in fact, there are several earlier reports, like the Evans (1953) and Watson-Jones (1952) reconstructions. The different options can be divided into three categories: i.e. ligament repair, non-anatomic reconstruction and anatomic reconstruction.

1. *Ligament repair*: Broström, Gould et al.'s modified Broström and Karlsson et al.'s modified Broström are the best known procedures. They were introduced at an early stage (between the 1960s and 1980s), and they represent simple, safe surgical options. They also represent anatomic procedures, but they may be limited by the quality of the ligament scar tissue, which is often seen in the event of long-standing ligament insufficiency (Figs. 60.1a–c and 60.2).
2. *Non-anatomic reconstruction*: Evans, Watson-Jones and Chrisman-Snook reconstructions. These procedures were all introduced to overcome the limitations of repair surgery with insufficient tissue quality. They are, however, jeopardised by the high incidence of range-of-motion restriction by not being anatomic, and accordingly ankle biomechanics will be endangered. There is also a risk of potential donor-site morbidity. Because these procedures are non-anatomic, they may lead to an increased risk of the development of osteoarthritis in the medium to long term (Figs. 60.3, 60.4, and 60.5).
3. *Anatomic reconstruction*: These procedures can be performed using either autografts or allografts (no preference in clinical studies, however). These procedures were initially

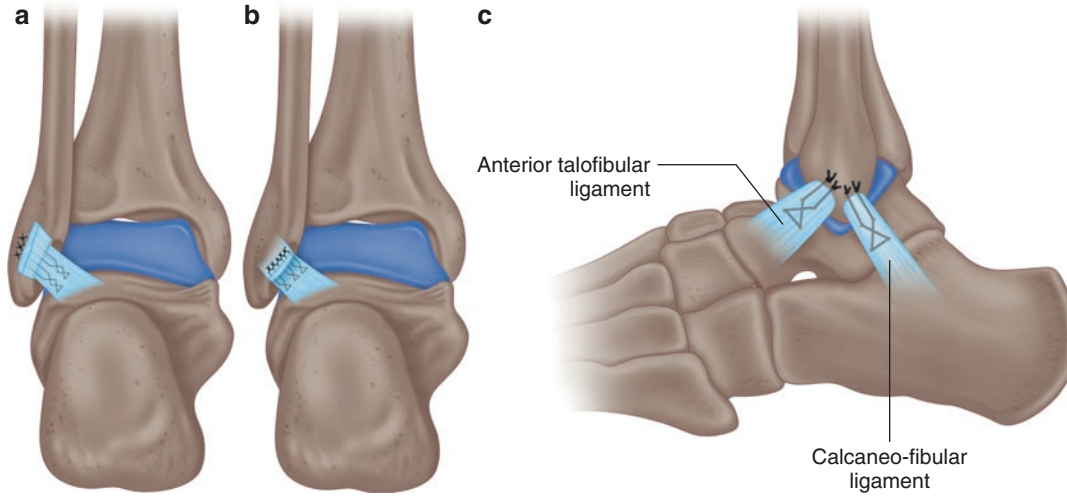

Fig. 60.1 (**a**) Anatomic repair, with the suture and imbrication of the anterior talo-fibular ligament, frontal view. (**b**) Anatomic repair, final stage after imbrication with the proximal end of the ligament, frontal view. (**c**) Anatomic repair, after the imbrication and re-insertion of both the anterior talo-fibular and calcaneo-fibular ligaments, side view

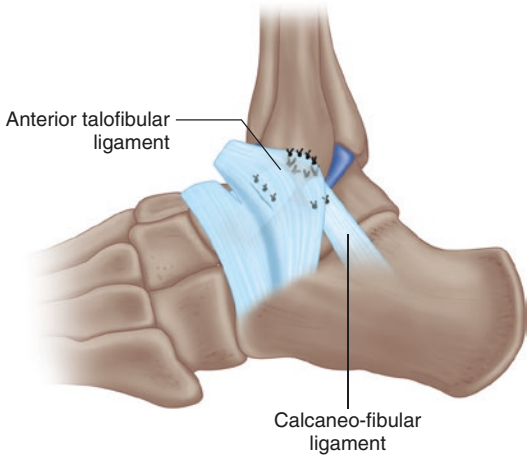

Fig. 60.2 Reinforcement using the extensor retinaculum, on top of the anatomic repair of both the anterior talo-fibular and calcaneo-fibular ligaments, side view

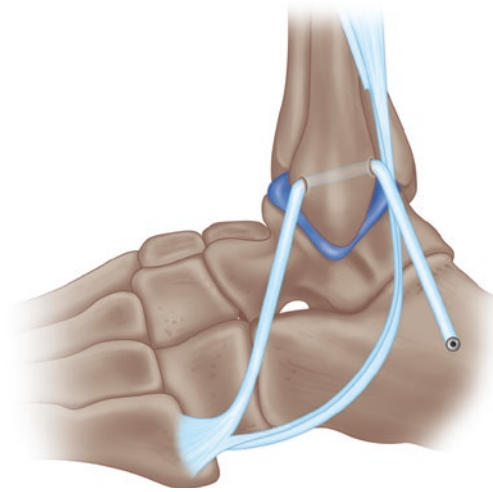

Fig. 60.4 Chrisman-Snook anatomic reconstruction using the split peroneus brevis tendon (tenodesis)

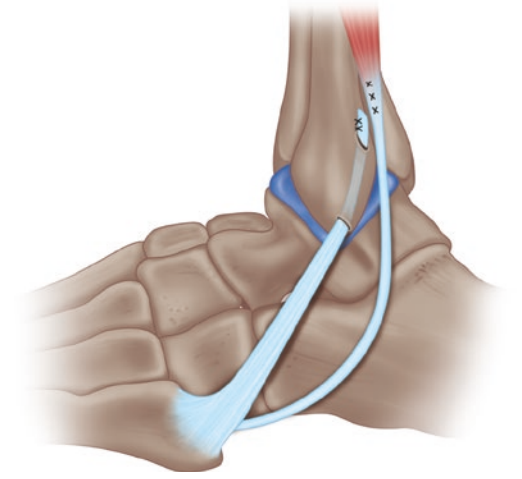

Fig. 60.3 Evans non-anatomic reconstruction, using the peroneus brevis tendon

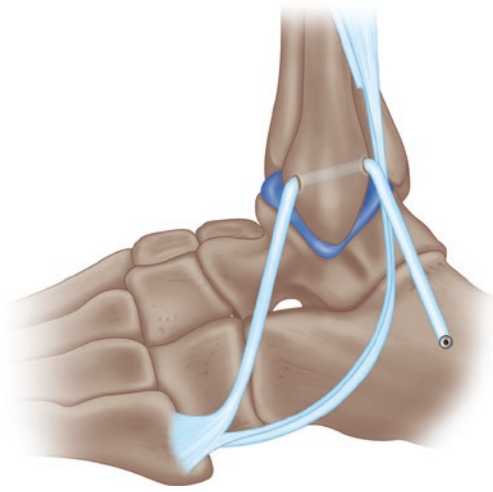

Fig. 60.5 Non-anatomic reconstruction of only the anterior talo-fibular ligament, using a free tendon autograft (an allograft is possible as well); modified Watson-Jones technique

introduced in the 1980s, and they are currently regarded as the primary choice due to the good restoration of ankle laxity, as well as good clinical and radiological outcomes (as they do not predispose to osteoarthritis). The results are independent of the state of the native ligaments, which is contrary to the ligament repair procedure.

The surgical techniques can also be described as either *anatomic repair* or *tenodesis*. One important aspect in all anatomic reconstructions is maintaining the subtalar motion without any restriction, which will lead to osteoarthritis if restricted. The foundation of all anatomic repairs is the original Broström technique. The original technique included the imbrication and suture of the midsubstance of either the anterior talo-fibular ligament alone or both the anterior talo-

fibular and calcaneo-fibular ligaments. This may, however, be technically difficult in the event of several ligament attenuations. For this reason, Gould et al. augmented the original Broström technique with a mobilised lateral portion of the inferior extensor retinaculum, which is also useful for stabilising subtalar instability. Karlsson et al. found that the ligaments were usually elongated and scarred (not disrupted) and accordingly recommended re-attachment to their original attachments through drill holes, with the proximal ends duplicated to reinforce the repair. This type of anatomic repair has been shown to be versatile and effective, in both the short and medium term, with good functional outcome, and in the long term, with a low risk of the development of osteoarthritis.

As early as 1952, Watson-Jones described non-anatomic tenodesis stabilisation, where he used the peroneus tendon that was routed through the calcaneus and talus. This technique was later simplified by Evans, who passed the distally attached peroneus brevis graft through an oblique drill tunnel in the distal fibula and sutured it back to the peroneus muscle proximally. This type of non-anatomic reconstruction has been common in clinical practice, but it has several limitations, as it does not replicate the position of either the anterior talo-fibular or the calcaneo-fibular ligaments but lies between them. This is biomechanically incorrect and will inevitably lead to the restriction of ankle and subtalar motion and, in the medium to long term, to the development of osteoarthritis, as has been shown in several studies. Finally, Chrisman and Snook developed a slightly different technique, where they more closely approximated the location of the two ligaments, thereby reducing the risk of incorrect biomechanics in and around the ankle. They used a split peroneus brevis tendon, in order to maintain at least some peroneus function. A randomised, controlled study by Hennrikus et al. compared the modified Broström and the Chrisman-Snook procedure. At follow-up (2+ years), patients who received the modified Broström repair had better clinical scores and fewer complications than those who were treated with the Chrisman-Snook procedure, (Figs. 60.3, 60.4, 60.5, and 60.6).

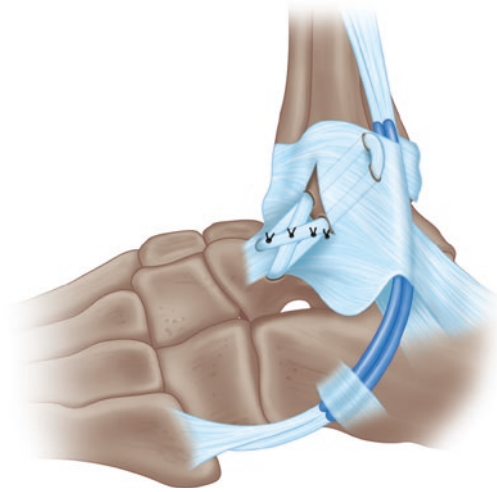

Fig. 60.6 Anatomic reconstruction, using a free tendon graft. Both the anterior talo-fibular and calcaneo-fibular ligaments are reconstructed, through drill holes in the talus, fibula and calcaneus. The direction of the reconstructed ligament is identical to that of the native ligament. Interference screw fixation at both ends. A similar technique can also be performed using an arthroscopic technique

Karlsson et al. reported almost 90% good and excellent functional outcome after anatomic repair, but they concluded that the failure of previous repair, generalised ligamentous laxity and long-standing instability were risk factors and were regarded as relative contraindications to these types of delayed repair. The results were also slightly poorer in women than in men, possibly correlated to joint laxity. Long-term outcomes after the Evans procedure are not favourable, with several patients reporting persistent swelling, reduced range of motion and recurrent instability, especially in the long term. Karlsson et al. reported that 50% of their patients had unsatisfactory results after 14 years using the Evans procedure; patients reported either recurrent instability or that the reconstruction was "too tight." Almost 75% of the patients had signs of osteoarthritis, some with advanced degenerative changes. The same group of researchers compared anatomic repair with Evans reconstruction at a 15- to 30-year follow-up. Patients treated with Evans tenodesis had undergone more surgical revision procedures and had more frequent chronic pain, limited range of motion and, in

many cases, osteoarthritis. The functional outcome was good in 80% of the anatomic repair group, compared with only 33% in the Evans tenodesis group.

Snook et al. reported favourable results (45/48 good or excellent) at the 10-year follow-up. However, 14 patients had sural nerve injuries and 3 had recurrent instability. There were also significantly more complications after the Chrisman-Snook procedure, compared with the modified Broström repair.

Recently, arthroscopic techniques have evolved. In a recent study, no differences in terms of clinical outcome were found after 24 months' follow-up (Fig. 60.7). The arthroscopic technique appears to be more difficult, as reflected by longer surgical times, but the advantage is the mini-invasive approach and thereby the possible lower risk of complications. A recent meta-analysis including four comparative studies comprising a total of 207 ankles found that the short-term AOFAS functional outcome score improved significantly with arthroscopic lateral ankle repair compared with open repair. However, there were no significant differences between arthroscopic and open repair, in terms of the Karlsson functional outcome score, total complication rate and nerve and wound complications. A second systematic review and meta-analysis by Zhi et al. revealed that arthroscopic repair showed excellent clinical results comparable to those of open repair. Moreover, the arthroscopic repair alleviated more pain, due to the minimally invasive procedure. There was a significant difference in favour of the arthroscopic technique in terms of AOFAS scores and VAS scores. However, there was no significant difference in terms of stress radiographic assessments, and the same finding applied to complication rates.

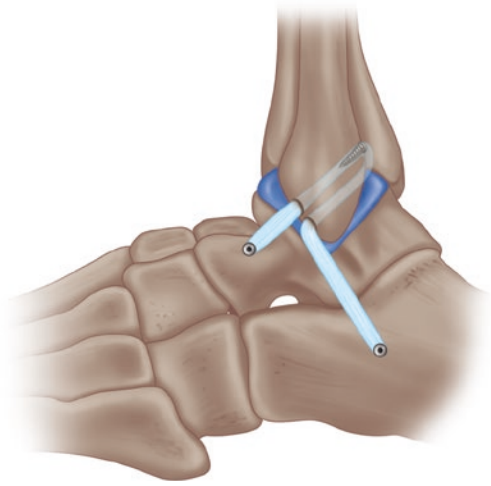

Fig. 60.7 Arthroscopic technique of ligament reconstruction; end result after stabilisation of both the anterior talo-fibular and calcaneo-fibular ligaments

Take-Home Message

Either anatomic repair or anatomic reconstruction is the preferred surgical method for the treatment of functional lateral ankle instability. There is little evidence to enable a choice between the many different surgical techniques, with mini-invasive/arthroscopic techniques still evolving. The choice of technique is mostly the surgeon's individual choice.

The original direct repair was of the anterior talo-fibular ligament only, but there is some evidence that both the anterior talo-fibular and calcaneo-fibular ligaments should be repaired/reconstructed simultaneously. The functional results are universally good after the reconstruction of both the anterior talo-fibular and calcaneo-fibular ligaments, approximately 90% good/excellent in most studies. Anatomic repair (if tissue quality is good) or anatomic reconstruction is the preferred procedure compared with non-anatomic reconstructions or tenodesis. The risk of osteoarthritis after non-anatomic reconstruction is high in the medium to long term.

Summary

It is well known that ankle sprain is the most common ankle injury related to sports. The conservative treatment protocol is based on a short period of rest, mobilisation, strength and agility training and a well-guided return to sports, with structured rehabilitation. However, 10–30% (or

even more) of all ankle ligament injuries do not experience full functional recovery after non-surgical treatment. Some researchers even claim that as many as 40% of acute ligament injuries will result in chronic or recurrent ankle instability. The two ligaments that are most commonly injured are the anterior talo-fibular ligament and the calcaneo-fibular ligament. The most common injury mechanism is a combination of supination (inversion) and forefoot adduction, at the same time as the foot is in plantar flexion. Ankle ligament injuries are commonly graded on the basis of their severity, i.e. grades I–III. In this context, grade I (mild) is described as minor stretching/bleeding of the ligaments, but without any macroscopic rupture or increased laxity. This type of injury can always be treated using non-surgical measures. Grade II (moderate) is defined as a partial injury/rupture of the ligament(s), accompanied by moderate swelling and bleeding. Patients with grade II injury often have problems with weight-bearing and functional impairment, with slight to moderate instability, but, as a rule, they can be treated non-surgically, with good results. The healing phases can be divided into the inflammatory phase, up to 10 days after injury; the proliferation phase, up to 8 weeks; and the remodelling/maturation phase, which may take up to one year or even more. There are several, mostly well-known, treatment alternatives, such as acute-phase surgery (stabilisation), immobilisation, followed by functional treatment, external support (brace and/or tape), neuromuscular training and strength training in the acute phase. One problem that should not be neglected is the risk of recurrent injury. Braces and/or sports tape have been much used in order to prevent ankle ligament injuries. The indications for further (in most cases surgical) treatment are persistent pain, recurrent instability in spite of stabilising treatment and repeated giving way. Chronic lateral ankle instability is a multifactorial condition that hampers activities in daily life and even more so in terms of sports activities. The different surgical options can be divided into three categories: i.e. ligament repair, non-anatomic reconstruction and anatomic reconstruction.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Which of these treatments is not indicated in acute ankle sprains during the acute phase?
   (a) Joint rest
   (b) Limb elevation
   (c) Elastic compression
   (d) Ice
   (e) Massotherapy
2. Which percentage of all ankle injuries will have long-term problems and eventually develop chronic lateral ankle instability?
   (a) 10–30%
   (b) 20–50%
   (c) 50–70%
   (d) 0–10%
3. The indications for surgical treatment after conservative therapy are:
   (a) Persistent pain, recurrent instability in spite of stabilising treatment and repeated giving way
   (b) Pain and inability to weight-bear
   (c) Infections
   (d) Recurrences
4. The functional results are universally good after the reconstruction of:
   (a) Both the anterior talo-fibular and calcaneo-fibular ligaments
   (b) Both the posterior talo-fibular and calcaneo-fibular ligaments
   (c) The anterior talo-fibular ligament
   (d) The posterior calcaneo-fibular ligament

Further Reading

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61 Cartilaginous Lesions, Osteoarthritis, and Arthroplasty in Foot and Ankle

Lew C. Schon and Zijun Zhang

Overview

Articular cartilage in foot and ankle experiences heavy wear and tear in daily activities and sports. This chapter focuses on these cartilaginous conditions, their orthopedic management, and the commonly performed arthroplasty procedures.

61.1 Introduction

Joints in the foot and ankle provide stability and flexibility that are required to support body weight and maintain mobility. While ankle, talonavicular, subtalar, metatarsophalangeal, and interphalangeal joints are notably mobile, the more stable articulations in the mid-foot provide force transmission through the arch and absorb shock stresses. Adapting to the diverse joint functions, articular cartilage in each locations has distinct morphological, biological, and mechanical features, which contribute to the natural history of diseases and injury and ultimately influence the treatment options and outcome of these foot and ankle conditions.

The ankle joint, the largest in the foot and ankle complex, consists of articulations among distal tibia, distal fibula, and talus. The main movement of the ankle is in the sagittal plane, performing dorsiflexion and plantar flexion. Although the ankle often bears a loading force several times greater than the body weight, due to its highly congruent geometry, stresses disperse relatively evenly across the articular cartilage within the ankle. This may explain thinner articular cartilage in the ankle than that in the knee. During physical activities, the articular cartilage on both distal tibia and talar dome experiences less compression strain than the cartilage on the tibial plateau in the knee does. Nevertheless, the peak strain in contact areas could be as much as 35% of the roughly 1.5 mm thick articular cartilage in the ankle. The biomechanics and functionality of ankle cartilage are backed by its biology. Histologically, there are fewer chondrocytes distributed in the superficial zone of ankle cartilage as compared with other articular cartilage. Moreover, the superficial layer of the ankle cartilage contains more type II collagen and aggrecan, the major structural molecules in cartilage matrix, which translates to sustained surface tensile stiffness over the course of aging. Cadaveric studies revealed consistently, across the gender and age groups, far less articular sur-

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face degradation in the ankle than in the knee. Chondrocytes and their immediate territory of matrix, which together form the functional unit of the cartilage, are comparable between the ankle and knee cartilage as assessed by morphometrical measurements. Metabolically, however, ankle cartilage differs from the cartilage in the knee, where osteoarthritis (OA) is more prevalent. For example, compared with knee cartilage, the ankle cartilage is less responsive to the catabolic cytokines, such as interleukin 1β, in degrading matrix molecules. In early OA, ankle cartilage has higher rates of type II collagen and aggrecan syntheses than knee cartilage. The metabolic trait inherited in ankle cartilage may, in part, contribute to a relatively low incidence of OA in the ankle and influence its progression. The biochemical features of ankle cartilage are recognizable by high Tesla magnetic resonance imaging (MRI): its T2 value is lower than the patella. Within the ankle joint, however, cartilages on both distal tibia and talus are not different on T2 mapping.

Articular cartilage in foot and ankle experiences heavy wear and tear in daily activities and sports.

61.2 Cartilaginous Lesions in the Foot and Ankle

The articular cartilage in the foot and ankle joints endures repetitive mechanical strain and is susceptible to physical injury (Fig. 61.1). An osteochondral lesion is an articular cartilage defect and extends into subchondral bone. When a focal defect is confined within articular cartilage or, precisely, its depth does not extend beyond the tidemark between cartilage and subchondral bone, it is a chondral defect. The two types of cartilage lesions have different natural healing capacities and outcomes. While a chondral defect is unable to heal by itself, osteochondral defect may be repaired without intervention, through the migration and differentiation of mesenchymal stem/progenitor cells which originate from bone marrow. Clinically, however, the decision-making of the treatment of a chondral or osteochondral lesion relies more on the length and severity of the symptoms, patient's age, professional demands, ankle stability, and size and location of the lesion. Osteochondral lesions in foot and ankle joints may occur on tibial plafond, talonavicular joint, and metatarsal heads but are most common on the talar dome.

Osteochondral lesion of the talus (OLT): An accurate estimate of the incidence of OLT is not available. It is known, however, that more than 70% of OLT cases have a history of ankle sprains or fracture, both of which are common ankle injuries. For those without a history of ankle injury, the OLT has been referred to as chondritis dissecans. The etiology of OLT is still unclear but generally believed to be in association with repetitive microtrauma to the cartilage resulted from ankle injury or heavy use. For example, injury of ankle lateral ligaments destabilizes the ankle and makes it excessively internally rotate when the ankle is in plantar flexion. This kinematic alteration could elevate the contact strain at the medial and anterior regions of the talar dome and cause cartilage damages. Anatomically, a slight deviation of bony morphology may make the ankle more susceptible to the development of OLT. For example, a wider distal opening angle of the medial malleolus or a larger anterior opening angle of the talus often presents in OLT patients.

The progression of OLT varies among cases. The pathology of OLT is divided into stages based on its imaging features. Although conventional ankle radiography could show signs of compression/depression of the subchondral bone of the talus in early OLT and, later, the detachment and displacement of the osteochondral fragments from the talus, it misses the early signs of cartilage damage during the development of OLT. Based on MRI, the progression of OLT is classified as stage I: cartilage damage only; stage II: cartilage injury with underlining fracture, with/without bony edema; stage III: detached but undisplaced cartilage fragment; stage IV: displaced fragment; and stage V: a cartilage lesion with subchondral cyst formation.

Clinically, OLT may be asymptomatic. For those with symptoms, the most common complaint is deep ankle pain during activities. The

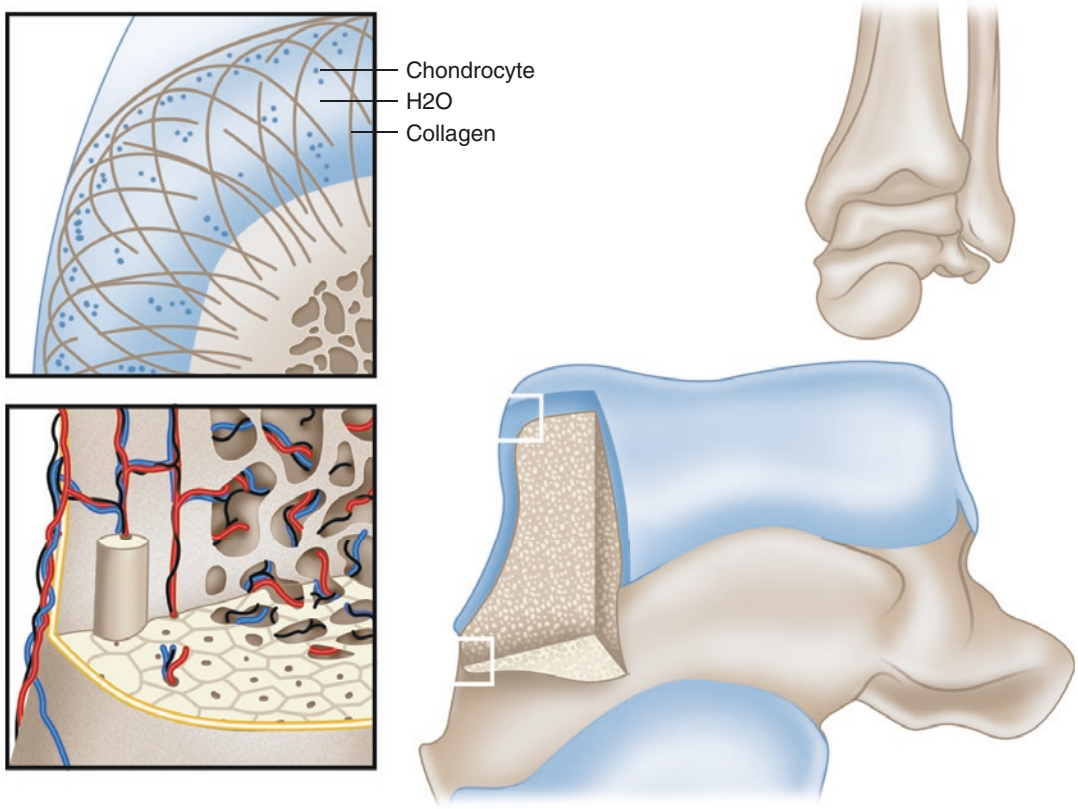

Fig. 61.1 Cartilaginous lesion of the ankle

ankle may be felt as stiff or unstable. There may be locking, catching, or giving way. Patients may have swelling and decreased endurance. On physical examination, there can be tenderness around the joint line and an effusion. The range of motion of the ankle is often limited. For severe and chronic OLT, radiographs may reveal a localized radiolucency on the talus and possibly a narrowing joint space. An ankle MRI is valuable for OLT diagnosis, particularly with either a delayed gadolinium-enhanced technique or T2 mapping techniques. MRI demonstrates the location and size of the OLT as well as associated pathologies, such as bone marrow edema and cysts in the talus, and synovitis in the ankle (Fig. 61.2). Additionally, MRI is useful for the evaluation of surrounding ligaments and tendons, which are critical for ankle stability and long-term outcome of the OLT treatment. Scans of computed tomography (CT) accurately show the bony defect of the OLT, and the size and location of cysts in the talus, which are important for surgical planning and approaches.

Early-stage OLT, with mild symptoms, is managed conservatively to mitigate the reactive pathologies around the lesion and in the ankle, such as synovitis and tendonitis. Conservative therapies include avoidance of the offending activity, reducing ankle loading from limited weight-bearing to total non-weight-bearing, using off-the-shelf cloth or boot ankle braces, custom ankle foot orthosis (AFO), and physical therapy.

The ultimate goal of surgical interventions of OLT is to repair the osteochondral defect, when it becomes a source of persistent ankle pain, inflammation, and instability. For surgical planning, the stage of an OLT is important, but its size and location are more relevant to the selection of procedures. Generally, debridement or microfracture

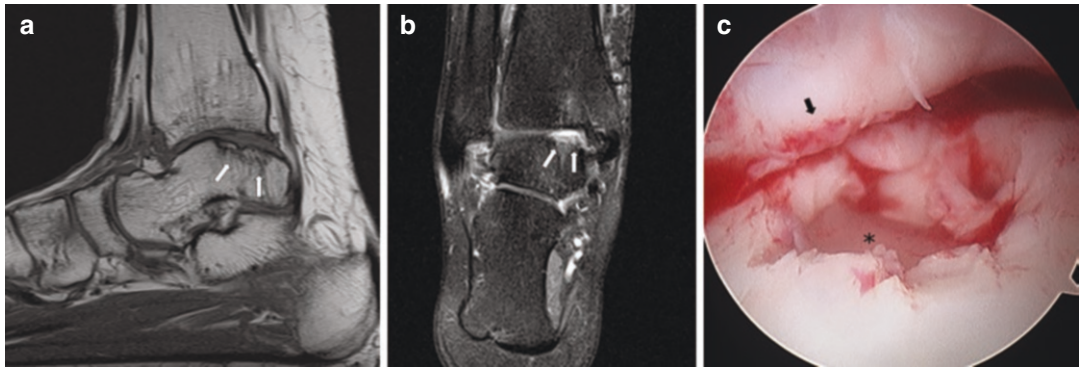

Fig. 61.2 Osteochondral lesion of the talus. (**a**) Sagittal MRI (T1 weighted) shows that an osteochondral lesion (indicated with arrows) occupies medial and posterior sections of the talar dome, including a defect on the surface and detached osteochondral fragments. (**b**) Coronal MRI (T2 weighted) locates an osteochondral lesion (indicated with arrows) on the medial side of the talar dome, with chondral fragments and local accumulation of joint fluid. Regional bone marrow edema extended into the talar body and in the opposite side of the tibia. (**c**) An arthroscopic view of an osteochondral lesion on the talar dome. Note the rough articular surface on the opposite tibial plafond (marked with an arrow). * indicates subchondral bone

is considered for an OLT smaller than 1.5–2.0 cm^2. To repair an OLT larger than 2.0 cm^2, transplantations of (auto/allo)osteochondral grafts, with or without the enhancement of other chondrogenic biologics, may be required.

- Debridement and/or microfracture: The procedure can be performed arthroscopically or through an open incision. The key point of the procedure is that by debriding or drilling through the subchondral bone at the bottom of the osteochondral defect, mesenchymal stromal/progenitor cells in bone marrow migrate into the OLT. In a favorable (biological and mechanical) joint environment, the stromal/progenitor cells differentiate and form fibrocartilage to fill the defect. Additionally, microfractures themselves trigger acute healing responses around the lesion in the talus. In most of the OLT cases, debridement and microfracture relieve ankle pain and improve its function. Because microfracture repairs OLT with fibrocartilage, which is inferior to hyaline articular cartilage mechanically, the repair may not be a long-lasting one.
- Osteochondral transplants: Transplantation of osteochondral graft (autograft or allograft) restores the surface contour of the talus and the congruity of ankle joint approximately. The autografts are osteochondral plugs harvested, with a separate procedure, from a non-weight-bearing area of a joint, such as the trochlear border of the femoral condyle. Allo-osteochondral grafts are cadaveric tissues prepared by a tissue bank. When a cadaveric talus is available, the allografts can be made to match the OLT shape and location for more precise restoration of the talar surfaces. The osteochondral autografts or allografts can be implanted as one piece to repair a defect or multiple plugs (also called mosaic arthroplasty) to cover a large OLT. Unlike microfracture, osteochondral grafts repair the OLT with hyaline cartilage, which better matches the talar cartilage biochemically and mechanically. However, the preparation of the graft, the size of the defect, and other ankle conditions also influence the outcome of osteochondral grafts.
- Transplantation of autologous chondrocytes or particulated juvenile cartilage allografts are other surgical options for repairing OLT. Chondrogenic biologics are increasingly available for OLT treatment, being used alone or in combination with other procedures, and diversify the treatment strategy of OLT. For most of the products, their effectiveness and cost-effectiveness remain to be validated by large, long-term clinical trials.

Currently, OLT is still a challenging condition with uncertain outcomes. When planning surgical interventions, it is important to thoroughly evaluate the entire ankle. Any ankle instability should be investigated and addressed at the time of repairing OLT. Cartilage regenerative therapies and biologics function well only in relatively normal joint conditions. Ankle inflammation, often in the form of synovitis, interferes with the molecular and cellular biology of chondrogenesis. A window of opportunity for repairing OLT exists only before secondary OA is evident in the ankle.

61.3 Osteoarthritis (OA) of Foot and Ankle Joints

The foot and ankle joints are common sites of OA. Of the population aged 50 and over, about one in six (or 15%) experiences osteoarthritic symptoms in the foot and ankle. In the foot, the most common site of OA is the first metatarsophalangeal joint (first MTP), followed by midfoot joints and the ankle.

1. Ankle OA: It is estimated that about 1% of the adult population suffers from ankle OA. Unlike most of the hip and knee OA associated with aging, the majority of ankle OA cases (>70%) are posttraumatic, having previous fracture or repeated ligament injuries in the foot and ankle. In average, the patients of ankle OA are younger than hip and knee OA patients. Consequently, ankle OA more profoundly impacts the patient's quality of life and social productivity.

Symptoms and diagnosis: Activity-aggravated ankle pain is the chief complaint of ankle OA. Another common symptom is ankle stiffness, swelling, and decreased endurance. Some patients complain of progressive deformity of the ankle. On physical examination, ankle is swollen, warm, and tender to palpation and has reduced range of motion. A history of ankle fracture or injury is an important lead of further examination.

Although MRI is advantageous for detection of cartilage lesion, radiography is essential and reliable for diagnosis of ankle OA. On radiographs, preferably taken while in weight-bearing positions, ankle OA presents narrowing joint space, osteophytes, and subchondral sclerosis as indirect signs of cartilage degeneration in the ankle (Fig. 61.3).

Treatment: The stage of pathology, symptoms, and patient's age and physical conditions are taken into consideration in the management of ankle OA. In its early stage, especially when the ankle symptoms are moderate, ankle OA is treated conservatively. To relieve ankle pain, nonoperative treatments shield the degenerative ankle from stresses. By guiding the patients to control body weight and modify daily activities, symptoms can be reduced. Additionally, modified shoes such as adding a soft heel, a rocker-bottom sole, foot orthosis inserts, an ankle brace, and walking aids help shift ankle stress and reduce pain during activities. A variety of physical therapies and rehabilitation programs are beneficial for reducing pain and stiffness. Nonsteroidal anti-inflammatory drugs (NSAIDs) inhibit joint inflammation and can be used to relieve ankle pain.

Intra-articular injection: Hyaluronic acid (or hyaluronate) in synovial fluid lubricates joint surfaces. In an OA joint, hyaluronate is depolymerized and loses the function of lubrication. After supplementation of hyaluronate via intra-articular injection, clinical studies have reported pain relief and function recovery of the OA joints. Many hyaluronate products vary in compositions and delivery vehicles, which complicate their indications, optimal dosages, and application scheme. The therapy's long-term efficacy and impact on OA pathology are still to be investigated by large, well-controlled clinical trials.

Intra-articular injection of corticosteroid reduces ankle pain for most of the ankle OA patients. The pain relief, however, generally lasts less than 6 months.

Surgery: Late-stage ankle OA is traditionally treated with arthrodesis, which includes removal of degenerative cartilage from the ankle and fusion of the tibiotalar articulation. The fused

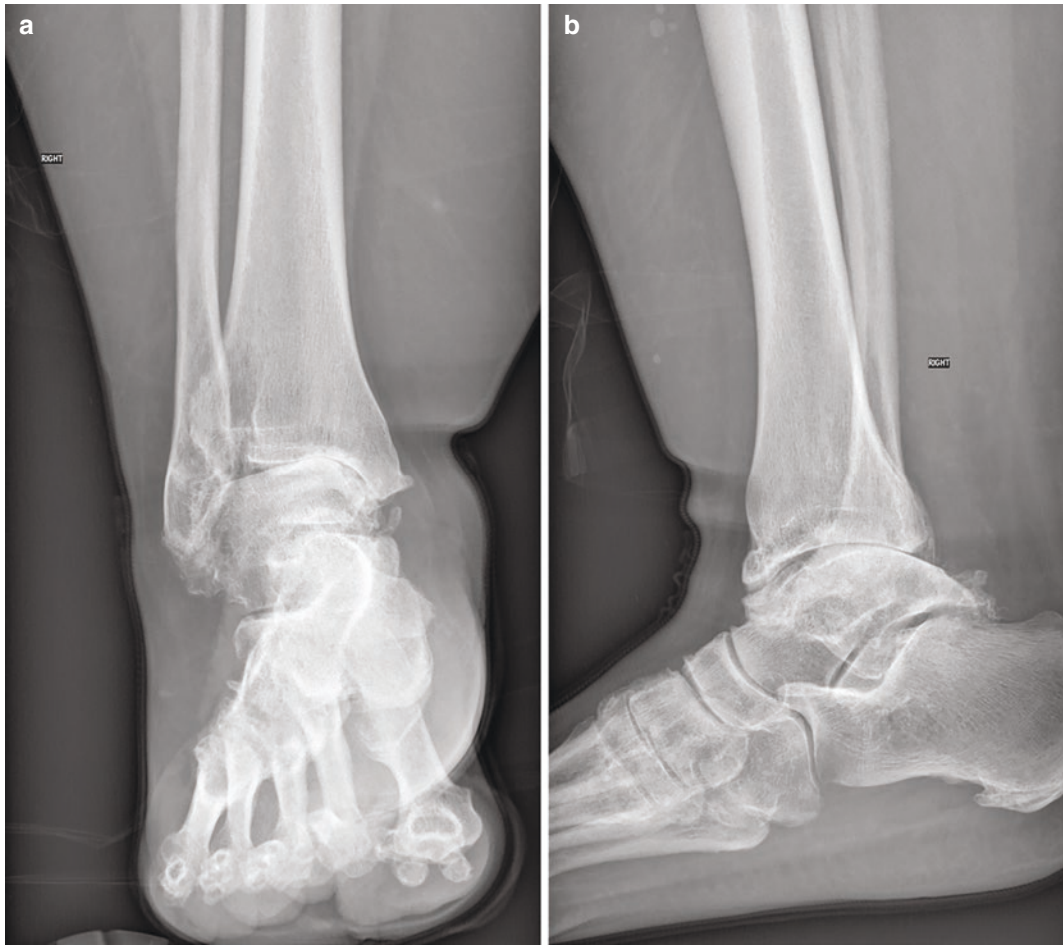

Fig. 61.3 Osteoarthritis of the ankle. (**a**) A radiograph of the anterior-posterior view of the ankle shows the disappearance of a normal joint space, especially on the medial side. The talus is tilted. There is subchondral sclerosis on both sides of joints: the talus and tibia. Osteophytes are formed on the tips of both medial and lateral malleoli. (**b**) A lateral-view radiograph of the ankle shows that the ankle joint surfaces are incongruent. Additionally, there are osteophytes formed on the anterior and posterior lips of the tibia and talus

ankle joint no longer moves and has reduced pain at rest, walking, and standing. But walking up or down a slope can be problematic. Rising up from a seated position, squatting, or operating foot pedals can be challenging. The patient usually tolerates the restricted motion at the fused ankle through compensatory motion of the hindfoot joints (subtalar, talonavicular, and calcaneocuboid joints) and overall has improved physical activity. Unfortunately, the elevated stresses in the hindfoot joints can be the source of foot pain, degeneration, and dysfunction after ankle arthrodesis.

The ankle arthrodesis procedure has improved over the last several decades with the introduction of less invasive approaches, better options for internal fixation, and application of osteogenic biologics. Using ankle arthroscopy, the procedure can be performed with less associated surgical trauma. Optimally designed internal fixation and osteogenic biologics have reduced complications of ankle arthrodesis, such as nonunion, infection, and malalignment. The fusion rate of ankle arthrodesis is greater than 90% but can be compromised by smoking and diabetes.

Total ankle replacement (arthroplasty) has advanced in the last decades with better component design, advanced materials, more bone preservation, and more accurate implantation techniques to become for many the first choice of late-stage ankle OA. The procedure removes osteoarthritic cartilage from the ankle, restores alignment, and preserves/restores ankle motion by providing a synthetic (metal and plastic) surface. Ankle replacement for ankle OA improves ankle pain and swelling and preserves ankle motion.

Hindfoot and mid-foot OA: More than 10% of the population over age 50 have symptomatic OA in the hindfoot and mid-foot joints, which include first and second cuneometatarsal, navicular-first cuneiform, and talonavicular joints. OA in this location is more prevalent in females and significantly increased in the population aged 75 years and over. Its risk factors include obesity, previous foot injury, and pain in other weight-bearing joints.

Hindfoot and mid-foot OA tends to involve in both feet. Foot pain and stiffness, particularly at the push-off position of the foot, are the most common complaints. On physical examination, the foot longitudinal arch may collapse with weight-bearing. The stride length is shorter, and cadence is reduced. Bony prominences may be palpable over the affected joints. On radiographs, osteoarthritic changes, such as narrowing of the joint space, osteophytes, cysts, and erosions, often show in a cluster of the joints. Weight-bearing radiographs, including dorso-plantar, oblique, and lateral views, of the foot are required for the diagnosis.

The conservative treatment of hindfoot and mid-foot OA emphasizes modifying the load distribution by using foot orthoses, wearing thicker cushioned rocker-soled footwear, and at times using an AFO. NSAIDs are useful for pain relief. Intra-articular injection of corticosteroid, however, is not used as common as for OA in other major joints due to the difficulty performing the injections by nonspecialists. With persistent and debilitating symptoms, arthrodesis of the affected joints is indicated. As the osteoarthritic pathology is often clustered among several joints, arthrodesis of multiple hindfoot and mid-foot joints may be necessary. To determine which joints are to be fused, clinical examination correlated with X-rays is needed. At times, an MRI, CT, or SPECT-CT (single photon emission computed tomography combined with CT) offers more detail of the involvement. Alternatively, the symptomatic joints can be identified by injection of local anesthetics under fluoroscopic guidance. Arthrodesis for hindfoot and mid-foot OA has a high union rate and satisfactory outcome.

OA of the first metatarsophalangeal joint (1st MTP-OA): About 35–60% of adults aged 65 years and over have 1st MTP-OA of the great toe. Radiographic anomaly of 1st MTP has been reported in about 20–50% of the people aged 40 years and over. Indeed, 1st MTP-OA is the most common OA in the foot. Two types of skeletal anomaly are relevant to 1st MTP-OA: hallux valgus and hallux rigidus. By definition, hallux valgus is a deformity at the 1st MTP joint, but clinically it may present with joint degeneration. Hallux rigidus is the "primary" OA of the 1st MTP joint. Its pathology as well as clinical manifestation is progressive pain, stiffness, and dorsal bony prominence over the great toe joint. X-rays demonstrate narrowing of the joint space and osteophytes on the dorsal surface of the first metatarsal head and base of the proximal phalanx. On physical examination, the joint may have crepitus and a reduced range of motion.

In the early stage of 1st MTP-OA, activity modification and using a stiffer soled shoe can reduce foot pain. For advanced conditions (sometimes associated with hallux valgus), surgical procedures are performed to correct the deformity and address OA pathology. For hallux rigidus, according to the stages of the pathology, there are three options of surgeries. Cheilectomy includes removal of 1/3 to ½ of the dorsal articular cartilage and removing the dorsal osteophytes. This procedure can be successful if the remaining articular cartilage is healthy. Several other procedures using osteotomy or resection of the osteoarthritic articulation have been shown to improve the pain and mobility of the 1st MTP joint. The disadvantages of the resection procedures can be instability of the 1st MTP joint and weakened big

toe power. Implant arthroplasty replaces the osteoarthritic 1st MTP joint with metallic or silicone prosthesis. There are several prostheses designed for the 1st MTP joint. In general, the procedure restores motion in the 1st MTP joint and can improve pain. With newer implant designs and improved techniques, the survivorship of the implanted prostheses has been extended. Nevertheless, implant failure remains an issue compromising the long-term outcome. Arthrodesis includes resecting the arthritic joint surfaces locking the 1st MTP joint in a neutral position with internal fixation. The procedure is reliable for pain relief and suitable for late-stage OA, by sacrificing the motion of the 1st MTP joint.

61.4 Arthroplasty of Foot and Ankle Joints

Arthroplasty is a collection of surgical procedures that preserve the mobility of a joint using biologic methods—with autogenous or allogenic or synthetic materials. One example of resectional arthroplasty is the Valenti arthroplasty: a V-shape resection of the 1st MTP joint for the treatment of severe hallux rigidus. An interpositional arthroplasty maintains articulation between bones by interposing tissues, such as periosteum, fat, tendon, joint capsule, or allografts.

In modern orthopedic surgery, total joint replacement/arthroplasty removes the diseased articular surfaces and replaces them with artificial prostheses. Total ankle replacement (TAR) is now performed on many ankle conditions that are traditionally treated by arthrodesis. Particularly for late-stage ankle OA, TAR reduces ankle pain and retains ankle mobility (Fig. 61.4). As a result, modern TARs can usually outperform ankle arthrodesis in improving patients' quality of life. Studies show that one year after TAR, the ankle functions measured by the American Orthopaedic Foot and Ankle Society hindfoot scores (normal = 100) are improved from 40 to 80, and this outcome can be sustained for about 10 years. After TAR, patients also improve in gait velocity cadence, stride length, and ankle power. About 90% of the TAR implants survive for at least 10 years.

TAR indications: The most common condition for TAR is late-stage ankle OA. For an optimal outcome, the OA ankle should be stable, with good-quality bone, minimal deformity, and good mobility. Another indication of TAR is nonunion or malunion of a previous arthrodesis. Currently, nearly half of TAR procedures are performed on the ankles with posttraumatic OA, followed by primary OA, rheumatoid arthritis, and postinfection arthrosis.

Contraindications: Charcot neuroarthropathy of the ankle is not suitable for TAR, because its progressive neuropathic pathology may destroy the local bone around the implant or trigger destruction of other foot joints. Ankle infection and compromised limb circulation are the other contraindications of TAR.

Surgical technique: The ankle is accessed through either an anterior approach or a lateral trans-fibular approach. Osteophytes around the joint surfaces are removed, and the anterior and posterior joint capsule is released, to free the ankle from any restriction of motion. Several total ankle designs and instrumentations have been developed, with different engineering principles and prosthesis fixations. For example, the Zimmer Trabecular Metal Total Ankle™ system takes a trans-fibular lateral approach and uses a frame to guide bone cutting, and the installation and alignment of total ankle components. If necessary, the length of the osteotomized fibula is adjusted to correct deformity and address ligamentous imbalance on the coronal plane of the hindfoot. The fibula is fixed with a plate and screw before closing the wound.

Postoperative care includes rehabilitation programs focusing on ankle motion and weight-bearing. According to several national registries (collected in later 1990s and 2000s), TAR revision rate increases significantly after 5 years. TAR complications include aseptic loosening of prosthesis, which is often caused by periprosthetic osteolysis. Wearing particles produced from total ankle components, plastic or metallic,

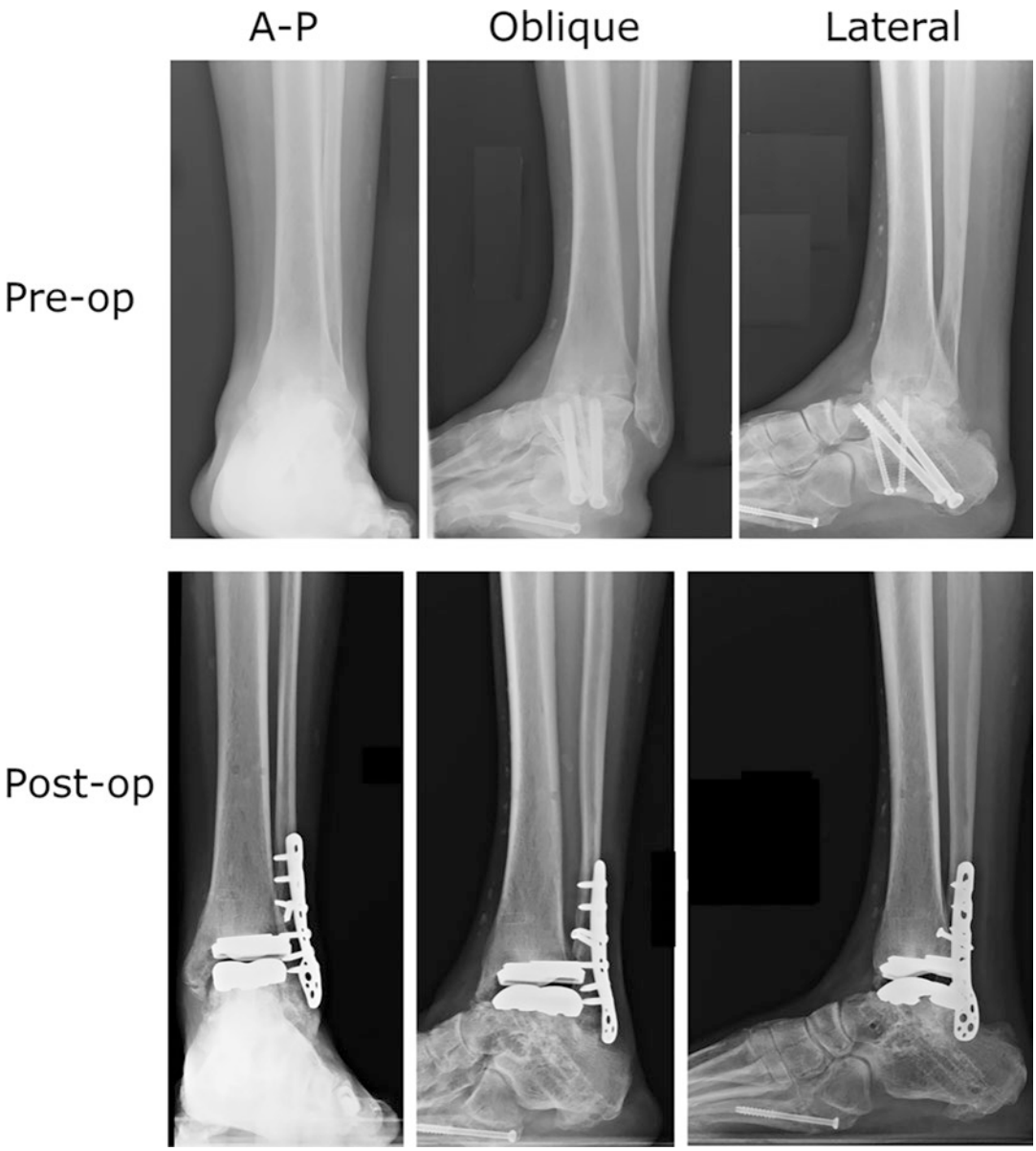

Fig. 61.4 Total ankle replacement for ankle osteoarthritis. The top row (pre-op) shows a posttraumatic ankle osteoarthritis in anterior-posterior (A-P), oblique, and lateral views. The bottom row (post-op) shows an ankle replaced with Zimmer Trabecular Metal Total Ankle™ (anterior-posterior, oblique, and lateral views). Some of the internal fixation hardware for previous foot injury are removed. Two metal components of the total ankle are implanted on the surfaces of the distal tibia and talar dome. This total ankle system includes a highly cross-linked polyethylene liner, which is radio-translucent, on the tibial component. The procedure was performed through a trans-fibular approach. The osteotomized distal fibula is fixed with a plate and screw

may induce inflammation and activate osteoclasts absorbing the bone surrounding the prostheses. Aseptic loosening of TAR is usually managed with surgical revision of the loosened components and grafting bone or its substitutes. If the surrounding bone no longer provides adequate support for the prostheses, an ankle arthrodesis is an option.

Take-Home Message

- The foot and ankle joints are prone to injuries from daily activities and sports.
- The foot and ankle are common sites of osteoarthritis in aging population.
- Ankle arthroplasty is increasingly performed for ankle conditions that are traditionally treated with arthrodesis.

Summary

The repetitive insults the foot and ankle joints endure during daily activities and sports are the main sources of cartilage lesions and joint degeneration. An osteochondral lesion of the talus, depending on its location and size, can be treated with debridement or microfracture procedures to introduce bone marrow stromal cells for cartilage repair, and transplantation of chondrocytes or cartilage grafts. The first metatarsophalangeal joint is a common site of osteoarthritis in aging population. The majority of ankle osteoarthritis cases are posttraumatic at a relatively younger age. It, therefore, has greater impact on social productivity and quality of life. With improved prosthesis design and surgical techniques, total ankle arthroplasty (replacement) has increasingly replaced the traditional arthrodesis for the treatment of late-stage ankle osteoarthritis and other ankle conditions.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Where is the most common site of osteochondral lesion in the ankle?
   (a) Talar dome
   (b) Talus
   (c) Navicular
   (d) Distal tibia

2. What type of cartilage does the microfracture procedure produce?
   (a) Fibrocartilage
   (b) Articular cartilage
   (c) Enchondral cartilage
   (d) Subchondral cartilage

3. Is it aging or injury more often associated with ankle osteoarthritis?
   (a) Injury
   (b) Aging

4. Which joint is the most common site of osteoarthritis in the foot?
   (a) First metatarsophalangeal joint
   (b) Second metatarsophalangeal joint
   (c) Third metatarsophalangeal joint
   (d) Fourth metatarsophalangeal joint

5. What is the most common ankle condition for ankle arthroplasty?
   (a) Posttraumatic ankle osteoarthritis
   (b) Joint degeneration due to aging
   (c) Degeneration due to ankle infection
   (d) Acute traumas

Further Reading

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62 Other Common Foot and Ankle Pathologies (Morton Neuroma, Flatfoot, Dysfunction of the Posterior Tibial Tendon, and Plantar Fasciitis)

Nasef M. N. Abdelatif

Overview
Foot and ankle pathologies constitute a wide spectrum of common diseases that affect people of all ages.

62.1 Morton's Neuroma

62.1.1 Introduction

This condition was first described in 1876 by Thomas George Morton. In spite of the commonly used term Morton's neuroma, it has been shown that the condition is not really a true neuroma, but rather a fibrosis of the interdigital nerve, which might result from pressure or repetitive irritation resulting ultimately in increased nerve thickness. This condition more commonly occurs in the second or third intermetatarsal webspace, with the third intermetatarsal space being the most common location. Histologically, the lesion shows axonal demyelination, neural edema, and perineural fibrosis.

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62.1.2 Clinical Presentation

The condition seems to affect females more commonly. There are usually complaints of burning pain and paresthesia related to the affected nerve. Additionally, a significant number of patients report the sensation of feeling "a small pebble underneath their feet." Some patients have reported that the condition follows a minor foot trauma. Pain usually increases by walking, tight shoes, or pointed high heels. Removal of shoes or rest from activities might alleviate the pain early in the condition. Later on, pain might become constant and might also occur at rest or specifically at night.

Examination of a suspected case of Morton's neuroma should proceed along the general guidelines of any foot and ankle examination as mentioned previously while the patient is standing, sitting, and during gait. One should look for any foot deformities that might be predisposing to the condition as hallux valgus, cavus feet, or pes planovalgus feet. Additionally, one can examine for plantar callosities that might indicate transfer metatarsalgia. There are certain specific clinical tests for Morton's neuroma. The most sensitive clinical test is reported to be the *thumb-index finger test* (Fig. 62.1). To perform this test, the thumb is placed on the plantar aspect of the foot and the index finger is placed on the dorsal aspect of the foot. One should ensure that both are placed correctly at the webspace but not on the

UG. Longo, V. Denaro (eds.), *Textbook of Musculoskeletal Disorders*,
https://doi.org/10.1007/978-3-031-20987-1_62

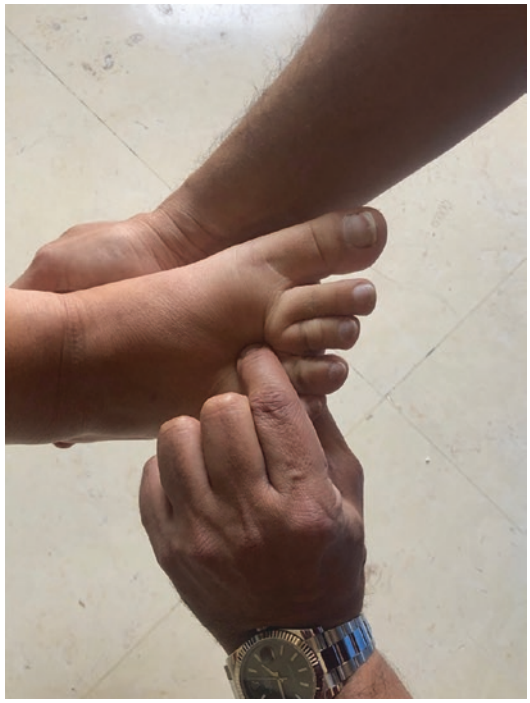

Fig. 62.1 Photograph demonstrating the "thumb-index finger test" for Morton's neuroma of the third webspace of a right foot

metatarsophalangeal joint; usually, splaying of the corresponding two toes might occur with this gentle pressure. A positive test results in pain. *Mulder's click test* is also reported to be of good sensitivity. It is performed by dorsiflexing the foot and squeezing the metatarsals. An audible click is suggestive of Morton's neuroma but has been reported to be more indicative if the neuroma is 1 cm or more. Lastly, the foot squeeze test has also been reported to be significant if reproducible pain occurs on squeezing the forefoot. However, like the Mulder's click test, both do not specify the affected intermetatarsal space.

62.1.2.1 Imaging

Standard weight-bearing radiographs are routinely used to exclude other foot deformities. However, ultrasonography and magnetic resonance imaging (MRI) are considered the better modalities for diagnosis of suspected cases of Morton's neuroma, and some authors have compared the usage of both modalities in diagnosis and deemed that both are comparable. However, MRI is still considered the gold standard for investigation specifically on the axial cuts.

62.1.2.2 Management

Nonoperative Management

Patient education and use of wide toe-box shoes are important aspects of nonoperative methods of management. Orthotics are also one of the more common methods of treatment. In actuality, the metatarsal bar is the commonest form of initial treatment. There has been no evidence shown to support inversion or eversion insoles.

Injections have also been used frequently in the management of neuromas. These injections could be sonographic guided or not. These injections have classically been steroid based. They are postulated to reduce the inflammation surrounding the neuroma and thus reduce the pain and pressure. Randomized controlled trials have shown that corticosteroid injections are more effective than local anesthetics alone.

Another treatment method is radiofrequency ablation although it has not been recommended as routine treatment by the National Institute of Clinical Excellence (NICE).

Surgical Management

Standard management is excision of the neuroma. This can be performed through dorsal or plantar approaches. No studies have proven one method over another. However, advantages and disadvantages have been cited for either approach. With dorsal approach, one can weight bear immediately as opposed to wound complications and scar sensitivity associated with plantar approach. Proponents of the plantar approach argue that perhaps it allows a more direct and accurate visualization of the neuroma.

A high complication rate has been associated with surgical excision with some studies citing ratios as high as 30%. The reasons have been incomplete resection, regional pain syndrome, and recurrence of the neuroma normally labelled "stump neuroma." In the latter case, resection of the neuroma more proximally with muscle implantation has been proposed as a method of management.

62.2 Flatfoot Deformities

62.2.1 Pediatric Flatfoot

Pediatric flatfoot is described in Chap. 11.

62.2.2 Adult Acquired Flatfoot Deformity

In the adult population, one of the more common causes of collapse of the medial longitudinal arch is adult acquired flatfoot deformity (AAFD). Several causes have been proposed for the etiology of AAFD ranging from inflammatory arthropathies to neuromuscular, genetic, and traumatic causes. Tibialis posterior tendon dysfunction still remains the more common pathology. More recently, other medial structures have also been implicated in this deformity as well, such as the spring ligament, the deltoid, and the interosseous talocalcaneal ligaments.

Classically although described as early as 1936, the hindfoot valgus, midfoot abduction, forefoot supination with the pathognomonic "too many toes" sign, and heel-rise test were described later by other authors (Fig. 62.2).

62.2.3 Diagnosis

As with all orthopedic pathologies, a detailed history and physical examination should be carried out by the physician during the evaluation of adult acquired flatfoot deformities. Initially, patients might describe pain along the course of the posterior tibial tendon (PTT), which will appear with standing and progressively increase with activity. Later, the condition may progress to pain at rest and progressive collapse of the medial arch with pain shifting to the lateral aspect of the foot due to sinus tarsi and sub-fibular impingement.

Physical assessment is carried out especially with the patient weight bearing. The apparent hindfoot valgus and "too many toes" sign when viewing the standing patient from behind have become consistent with excessive forefoot abduction. Tenderness and particularly swelling along the course of the PTT can also be elicited. Range of motion and muscle strength of the structures around the foot and ankle should also be assessed. When assessing the strength of the PTT, the foot is better placed in plantar flexion and inversion to isolate the strength of the tendon itself from other plantar flexors of the ankle and foot as the tibialis anterior tendon. Silfverskiold test is also useful in assessing gastrocnemius-soleus complex tightness.

Position of the foot relative to a neutral heel should be assessed, as should the mobility of the subtalar joint itself. Single and bilateral heel rises are performed and repeated to assess the functional ability of the PTT to invert the hindfoot. One should not forget to assess the patient's gait pattern.

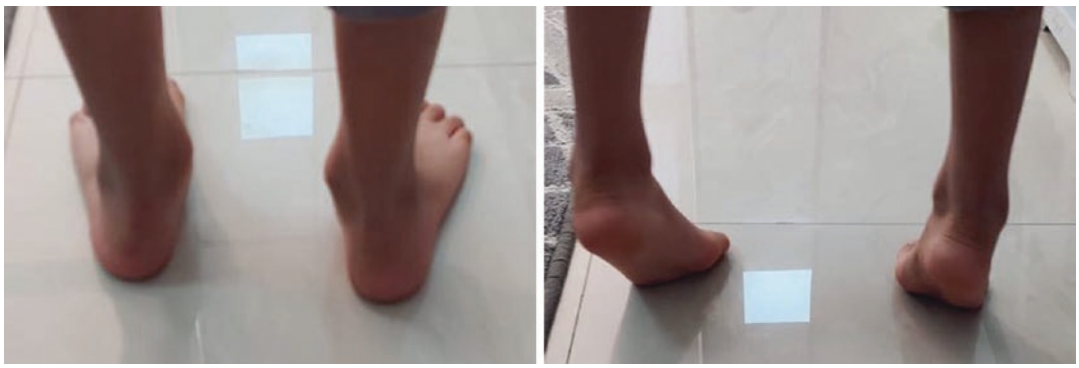

Fig. 62.2 Photograph on the left shows the patient standing on both feet with valgus heel and reduced foot medial arch. Photograph on the right shows the same patient with heel-rise test showing the hindfoot moving into varus

62.2.3.1 Radiological Evaluation

The *standard radiographic evaluations* are weight-bearing anteroposterior, lateral, and hindfoot radiographs of both feet. On the anteroposterior weight-bearing projection, forefoot abduction by the anteroposterior talus-first metatarsal angle and talar head uncoverage can be assessed by various parameters such as talonavicular coverage angle, talonavicular uncoverage angle, and lateral incongruency angle.

On the lateral weight-bearing radiographs, one can assess the lateral talus-first metatarsal angle, the lateral talocalcaneal angle, the calcaneal pitch, and the medial cuneiform height. Weight-bearing ankle radiographs could show valgus orientation of the talus in more pronounced cases.

The hindfoot alignment view will show the relationship between relative axis of the tibial and the most inferior calcaneal point.

Recently, newer modalities as multiplanar *weight-bearing imaging* and even more recently *weight-bearing 3D CT* might have increased our knowledge about the pathomechanics and deformities present in this disease.

MRI evaluation although not mandatory in such situations might be helpful in the evaluation of the degree of initial damage to the tibialis posterior tendon, to assess the spring ligament, and to show the condition of the deltoid ligament or the peroneal tendons to adequately plan for surgical correction when needed.

Ultrasound evaluation has been used and compared to MRI in its efficacy. One study compared the efficiency of ultrasound with MRI in the diagnosis of posterior tibial tendon dysfunction, and the results were comparable. Another study compared the efficiency of ultrasound with MRI in the diagnosis of spring ligament rupture with also similar results.

Tendoscopic evaluation could also be utilized to assess the tendon viability and in perhaps earlier stages of synovitis with a possibly negative MRI diagnosis. Studies have shown tendoscopy to be helpful in not only diagnosis but also treatment at least in short term for early stages.

62.2.4 Classification

The Johnson and Strom classification system originally created in 1989, and modified by Myerson in 1997, remains the most commonly used classification system for adult acquired flatfoot deformity. It was originally a three-stage anatomical and clinical classification system based on the tibialis posterior tendon condition, hindfoot alignment, and forefoot flexibility. Myerson added a fourth stage to describe deltoid ligament insufficiency valgus collapse and ankle degeneration.

62.2.5 Management

62.2.5.1 Conservative Management

This includes immobilization, nonsteroidal anti-inflammatory drugs, bracing, and physical therapy. Conservative methods are almost exclusively left for stage I posterior tibial tendon dysfunction. Also, it can be used for more advanced stages before embarking on the definitive surgical procedures.

62.2.5.2 Operative Management

Operative intervention is generally merited when conservative measures fail. One must take into consideration the general associated medical comorbidities, the functional demands of the patient, and other factors as smoking and alcohol consumption. For the scope of this chapter, we shall discuss surgical options according to the more commonly used modified Johnson and Strom classification, although as has been previously stated lately this classification and even the nomenclature of adult acquired flatfoot deformities have been challenged.

Stage I

Tibialis posterior tendon tenosynovectomy, debridement with or without flexor digitorum longus tendon transfer alone or combined with medial calcaneal displacement osteotomy, or arthroereisis can be utilized as surgical management methods for stage I patients.

Stage II

Flexor Digitorum Longus (FDL) Transfer

Medializing Calcaneal Osteotomy

The procedure relies on medially displacing the posterior aspect of the calcaneus, thus correcting heel valgus, shifting the gastrocnemius-soleus complex line of pull medial to the subtalar joint, thereby producing an inversion action, and also decreasing the stretch on the medial spring ligament and tibialis posterior tendon.

Lateral Column Lengthening

Lateral column lengthening has been described for stage IIB disease with talar head uncoverage more than 30%, to improve the talar coverage and correct forefoot abduction. This lengthening can be performed through the anterior calcaneus or by a distraction arthrodesis of the calcaneocuboid joint. The distraction arthrodesis has been associated with lesser outcomes and decreased range of motion of the ankle and subtalar joints. This procedure has been historically associated with less favorable outcomes such as foot stiffness, decreased eversion, fifth metatarsal stress fractures, and lateral sided foot pain.

Subtalar Arthroereisis

The rationale behind this procedure was discussed briefly with pediatric flatfoot deformities. Its use in the management of adult flatfoot deformities is still controversial, although some studies have shown good intermediate clinical results. However, the procedure is also associated with several complications; the most common is persistent pain in the sinus tarsi. Other complications include implant extrusion, foreign body reaction, and talar cysts.

In addition to these main procedures, some other deformity corrective procedures might be called upon if necessary. Some of these are the following procedures:

Cotton Osteotomy

This is a dorsal opening wedge osteotomy of the medial cuneiform first described in 1936. It can also be performed on the first metatarsal. It can be performed if the foot is judged to have residual first ray elevation to depress it.

First Tarsometatarsal Arthrodesis or Medial Column Fusion

This can be performed if there is marked instability or hypermobility of the first ray, or degenerative changes or a medial column sag/depression at that level.

Isolated Naviculo-Cuneiform Arthrodesis

This likewise can be performed for patients for whom the apex of the deformity is deemed to be at that location. This procedure has the theoretical advantage of preserving the subtalar and Chopart joint motions.

Spring Ligament Reconstruction

This has been advocated by some investigators especially if forefoot abduction remains after lateral column lengthening, or if the adequate correction would necessitate excessively large grafts, but as was stated earlier, the exact size of these grafts is not yet fully investigated or known.

Stage III

This stage shows a fixed deformity with pain and inability to perform the single heel-rise test. Management of this stage is usually by arthrodesis. Both double (subtalar and talonavicular joints) and triple arthrodesis (adding calcaneocuboid joint fusion) have been used successfully in these patients. The ultimate goal is to achieve a stable plantigrade painless foot with neutral to slight (5°) valgus and a neutral forefoot. Either of these arthrodesis might require additional supplementary procedures as medializing calcaneal osteotomy, Cotton osteotomy or fusion, or gastrocnemius or Achilles lengthening. The calcaneocuboid joint-sparing double-arthrodesis approach is thought to avoid the risk of calcaneocuboid nonunion and/or stiffness. Triple arthrodesis has also been associated with complications as inability to adapt to uneven grounds, nonunion, stiffness, and progressive ankle valgus and arthritis. Also, nonunion rates from 10 to 23% have been recorded mostly involving the talonavicular joint.

Stage IV

Here, there is a flatfoot associated with an ankle valgus deformity, which might be rigid or flexible. Some authors have advocated deltoid reconstruction in flexible deformities. Other authors have preferred performing ankle fusion or ankle replacements if the deformity is rigid with ankle replacements gaining more popularity to avoid pantalar fusions.

62.3 Plantar Fasciitis

This disorder is considered the most common cause of inferior heel pain, and it is estimated that it is responsible for around 11–15% of all foot complaints.

62.3.1 Anatomy and Pathophysiology

The plantar fascia is a fibrous tissue band that originates on the anteromedial aspect of the calcaneal tuberosity to insert onto the bases of the proximal phalanges. It is divided by strong vertical septa into central, medial, and lateral parts. These divide into five digital bands at the metatarsophalangeal joints, and then each band divides once more to pass on both sides of the flexor tendon and insert into the periosteum of the base of the proximal phalanges as stated previously. During its course, it becomes superficial and palpable beneath the longitudinal arch on the plantar aspect of the foot. This fascia is inelastic and has shown only maximum elongations of 4%. It will become tense when the toes dorsiflex and exert a tension effect on its calcaneal origin, which also elevates the longitudinal arch. This effect is referred to as "windlass mechanism." At the same time, there is a pull of the gastrocnemius soleus muscle complex, which adds more pressure on the forefoot; this occurs in addition to the ground reaction forces that also increase loads on the foot. Repetitions of these mechanisms might result in overloads, inflammations, microtears, and chronic degenerative changes.

Another important structure is the heel fat pad. Its anatomy was first described in 1921 by Tietze. It is a honeycomb-shaped fibroelastic septum that holds solely fat globules. These septa are arranged in a U-shape around the tuberosity and attached to the skin and the calcaneus bone. The main function is to act as a cushion for the hindfoot. It has been shown that each heel strike can generate 110% of body weight, and that can increase up to 250% of body weight with running. Generally, after the age of approximately 40, this fat pad begins to atrophy, with loss of elastic tissue, collagen, and water. Thus, there is a decrease in height and thinning of the pad with softening, decreased impaction properties, and reduced heel protection.

62.3.1.1 Etiology

Histologically, plantar fasciitis is a fibrofatty degeneration of the origin of the plantar fascia with microtears and collagen necrosis. Thus, it is more a degenerative process than an inflammatory one. Generally, plantar fasciitis can occur due to mechanical causes, spondyloarthropathies, or neural compression causes. Another very common association is heel spurs, and although they have been reported to occur in almost 75% of patients with heel pain, they are not considered to be a cause. It is also worthy to note that some authors have found an increase in fat pad thickness in patients with plantar fasciitis. According to other authors, the elasticity is a more important determinant. Therefore, with decreased elasticity and more accumulation of fat globules in that space, an increase in the pressure on the calcaneal tuberosity might occur.

62.3.2 Patient Evaluation

A thorough history taking from the patient is as always vital to making the correct diagnosis. One should inquire about the patient's general health and ask for constitutional manifestations as weight loss, fever, malaise, night sweats, etc. to exclude systemic conditions. During history taking, one should also inquire about recent changes in activity, weight, or general daily habits.

Additionally, it might be worthy to inquire about the onset of symptoms and its relation to activity as pain with first few steps of walking or activity is different from that which might occur after periods of activity. On the other hand, night pain unrelated to activity should point one to another direction such as infections or tumors. Bilaterality of the condition would usually point to a systemic condition or some spondyloarthropathies.

Moving on to the physical examination, it is always advisable to primarily exclude other sources of referred pain by examining the spine, and extremities to exclude L5–S1 radiculopathies that might itself be a cause of heel pain. Locally, one should examine the foot structure itself. A pes planus or a cavus foot might be causes of the heel pain. Additionally, one should always examine for Achilles tightness.

Specifically, plantar fascia patients will complain of heel pain with the first few steps in the morning and after resting the foot with sitting for extended periods of time. Pain usually starts as a sharp shooting pain that becomes dull aching and with continued walking. Pain is localized at the medial heel and arch. By examination, the most tenderness is located at the medial calcaneal tubercle and might extend along the middle fascial band. This pain will increase with dorsiflexion of the ankle and toes, as that tightens the fascia and the windlass mechanism of the foot. Examination should be continued with the patient standing to assess any hindfoot abnormalities or abnormalities of the longitudinal arch. Additionally, one should obviously examine for Achilles tendon tightness or isolated gastrocnemius tendon tightness.

Some patients will present with a rupture of the plantar fascia itself. In this case, they will recall a pop or tearing sensation on the plantar aspect of their feet, and quite a few will report a decrease of the plantar fascia symptoms after this event. There might be focal ecchymosis on the medial surface of the plantar fascia. They will also typically exhibit inability to do single-leg stance heel rises, and there will be pain with MTPJ extension.

One should be aware of the differential diagnosis of plantar fasciitis, such as tarsal tunnel syndromes, radiculopathies, calcaneal stress fractures, plantar fibromatosis, and flexor hallucis longus tendinopathy.

62.3.2.1 Diagnostic Evaluation

Ankle and foot radiographs are done to exclude other causes of heel pain and rule out calcaneal stress fractures and degenerative conditions of hindfoot. MRI has been shown to be very reliable for the diagnosis of plantar fasciitis and can also detect other conditions as tarsal tunnel space-occupying lesions and stress fractures of calcaneus. Ultrasonography has been shown to be as reliable as MRI for diagnosing plantar fasciitis, but not helpful for other conditions. For diagnosing neural related conditions, electrodiagnostic studies have been used to detect radiculopathies and peripheral neuropathies. Electromyographic testing can be helpful in medial and lateral plantar nerve compressions, but negative results do not rule out those conditions. Although 20–30% of patients with plantar fasciitis can exhibit the condition bilaterally, one should consider HLA-B27 test according to Gerster.

62.3.2.2 Management

Nonoperative Management

This is the mainstay of plantar fasciitis treatment. The American Orthopaedic Foot and Ankle Society (AOFAS) has previously recommended a minimum of 6–12 months of nonoperative treatment before surgical intervention and recently stated that 90% of patients will improve within 2 months of conservative treatment. Nonsurgical treatment includes plantar fascia-specific stretching exercises (Fig. 62.3), Achilles stretching exercises, night splints, orthotics, anti-inflammatory medications, steroid injections, extracorporeal shock wave therapy (ESWT), and botulinum toxin type A (Botox) injections.

Operative Treatment

A relatively recent study performed by DiGiovanni and colleagues attempted to identify the preferred method of surgical management of plantar fasciitis between a group of orthopedic foot and ankle surgeons. Available options were

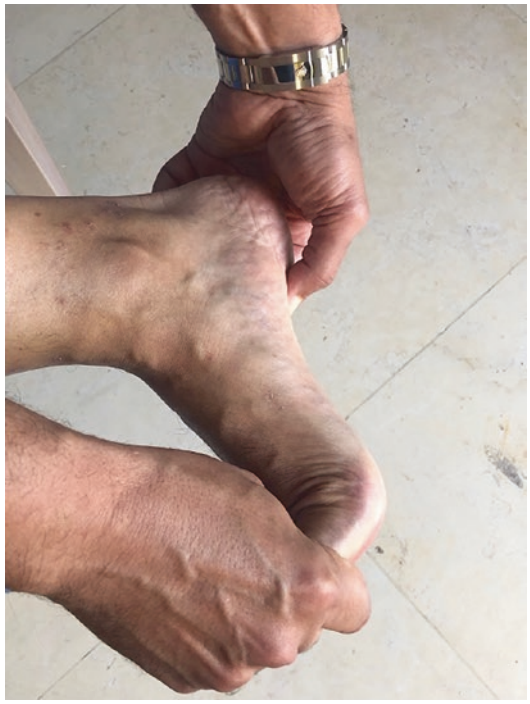

Fig. 62.3 Photograph showing a patient performing the plantar fascia-specific stretching exercises. Note the dorsiflexion of the ankle joint and dorsiflexion of toes while performing massage of the fascial bands

open partial plantar fascia release with decompression of first branch of lateral plantar nerve, endoscopic partial plantar fascia release, complete releases, and additional gastrocnemius recession for any one of the previous surgical options.

Open Partial Plantar Fascia Release and Decompression of First Branch of Lateral Plantar Nerve

The procedure was described by Baxter and Thigpen in 1984, and they reported very good results. However, later on, several other authors have not been able to reproduce these good results. Gould and DiGiovanni described a modification of the open release with the addition of proximal and distal tarsal tunnel releases.

Endoscopic Partial Plantar Release

The theoretical advantage behind this technique is the concept that this procedure is a minimally invasive one and will consequently have much lesser morbidity. Although studies have demonstrated that it is indeed associated with more rapid recovery, improved function, and improved pain, these studies are generally limited by small sample sizes and short follow-up periods. Also, some authors argue that this procedure does not address the neurogenic compression and does not provide as efficient visualization of the plantar fascia structures as the traditional open procedures.

Gastrocnemius Recession

This procedure has been gaining popularity among investigators especially recently. It has been used in isolation and also as an adjuvant with other procedures. The advocates of isolated recession for treatment of plantar fasciitis argue that there are good chances of complete relief of the condition, and the patients still have the ability to perform partial plantar fascia releases if the condition persists. Additionally, there is increased ankle dorsiflexion range. However, it is also associated with complications as probable weakness of push off, poor cosmesis, and sural nerve injuries.

Summary

Morton's neuroma is not really a true neuroma, but rather a fibrosis of the interdigital nerve, which might result from pressure or repetitive irritation resulting ultimately in increased nerve thickness. This condition more commonly occurs in the second or third intermetatarsal webspace, with the third intermetatarsal space being the most common location. Acquired adult flatfoot (AAFD) is one of the more common causes of collapse of the medial longitudinal arch. Several causes have been proposed for the etiology of AAFD ranging from inflammatory arthropathies to neuromuscular, genetic, and traumatic causes. Tibialis posterior tendon dysfunction still remains the more common pathology. The standard radiographic evaluations are weight-bearing anteroposterior, lateral, and hindfoot radiographs of both feet. The conservative management of AAFD includes immobilization, nonsteroidal anti-inflammatory drugs, bracing, and physical therapy. The surgical treatment is generally mer-

ited when conservative measures fail. One must take into consideration the general associated medical comorbidities, the functional demands of the patient, and other factors as smoking and alcohol consumption. Plantar fasciitis is considered the most common cause of inferior heel pain, and it is estimated that it is responsible for around 11–15% of all foot complaints. The American Orthopaedic Foot and Ankle Society (AOFAS) has previously recommended a minimum of 6–12 months of nonoperative treatment before surgical intervention and recently stated that 90% of patients will improve within 2 months of conservative treatment. Available options were open partial plantar fascia release with decompression of first branch of lateral plantar nerve, endoscopic partial plantar fascia release, complete releases, and additional gastrocnemius recession for any one of the previous surgical options.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. A patient presents with pain, intense burning, and a sensation of electric shock at the level of the plantar side of the forefoot, in correspondence of the III intermetatarsal space with irradiation to the third and fourth toes. This sensation increases on palpation and during walking. Which of the following pathologies is most likely to affect the patient?
   (a) Morton's neuroma
   (b) Tenosynovitis of the toe flexors
   (c) Chronic instability of the third metatarsophalangeal joint
   (d) Metatarsophalangeal arthrosis
   (e) Ledderhose disease
2. In the flatfoot, the medial arch is:
   (a) Reduced
   (b) Unmodified
   (c) Increased
   (d) Sometimes increased, sometimes decreased
3. The Mulder's click test is used for the diagnosis of:
   (a) Morton's neuroma
   (b) Plantar fasciitis
   (c) Flatfoot
   (d) Calcaneus fracture
4. The most common cause of adult acquired flatfoot deformity is:
   (a) Tibialis posterior tendon dysfunction
   (b) Talus fracture
   (c) Calcaneus fracture
   (d) Diabetic neuropathy
5. Surgical treatment for plantar fasciitis is:
   (a) Recommended only in case of conservative treatment failure
   (b) The gold standard
   (c) Performed after 1 week of conservative treatment
   (d) Only performed using endoscopic techniques

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Charcot Arthropathy of Foot and Ankle

63

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Overview

Charcot foot arthropathy is a progressive, noninfectious, destructive inflammatory process of the foot and ankle. As modern-day medical advances are made, and life span and longevity rise, consequently, it is expected that the number of diabetic-related complications shall also increase.

63.1 Introduction

Charcot foot arthropathy is a progressive, noninfectious, destructive inflammatory process of the foot and ankle. It was originally described by the French neurologist Jean-Martin Charcot, as a sequela of tertiary syphilis. However, in the vast majority of patients today, it is secondary to long-standing diabetes. In the USA alone, there are more than 29 million adults with diabetes, and these numbers are increasing worldwide.

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As modern-day medical advances are made, and life span and longevity rise, consequently, it is expected that the number of diabetic-related complications shall also increase. This will ultimately lead to financial and emotional burdens on the health-care system. Charcot arthropathy secondary to diabetes mellitus by itself severely reduces the overall quality of life and dramatically increases the morbidity and mortality of patients.

63.1.1 Pathophysiology

There is no single cause for Charcot arthropathy. However, two causative theories for the pathogenesis of Charcot arthropathy exist, which are the neurotraumatic and neurovascular theories.

The **neurovascular theory**, first described by Charcot, relates to an overactive vaso-autonomic neuropathy that causes hyperemia. Increased blood flow increases venous pressure and enhances fluid filtration through capillary leakage, consequently leading to increased compartmental pressure and deep tissue ischemia, ultimately compromising the foot and ankle leading to joint instability. Hyperemia might also result in greater delivery of osteoclasts and monocytes resulting in increased bone resorption.

The **neurotraumatic theory** on the other hand depicts any type of trauma in the context of

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decreased sensation as the initial cause. This then sparks an acute inflammatory process with the release of proinflammatory mediators and a decrease in inflammatory mediators.

If unprotected weight-bearing is continued in either of these situations, an increase in the release of proinflammatory mediators results that ultimately causes compromised bone form, integrity, and function. The imbalance between bone destruction and bone repair results in the appearance of simulated "hypertrophic bone nonunion" that is characteristic with radiographs associated with Charcot disease. However, more research is required to fully understand the actual pathogenesis process and mediators involved.

63.2 Clinical Presentation and Evaluation

63.2.1 History and Physical Examination

Charcot foot and ankle patients are usually between their fourth and sixth decades of life. They will present with an acute inflammatory-like episode of redness, hotness, and swelling. Pain is usually mild or absent. The patients will rarely recollect any significant traumatic incident, neither will any apparent constitutional manifestations be apparent. Naturally, the most common associated neuropathic disease will be diabetes. However, other less common disorders as syphilis, leprosy, parkinsonism, spinal and nerve root compromises, as well as sarcoidosis or rheumatoid disease should be searched for. Some authors have attributed the cause to be idiopathic in up to 40% of cases.

Upon examination, a direct skin temperature reading might show a difference of 10 °F or more between the normal and affected sides. Pulse presence and hyperemia will differentiate Charcot feet from diabetic feet with arterial compromise. Naturally, one must also look for cellulitis, ulcerations, secondary infection, purulent discharge, and abscess formations. Lastly, stability of the foot as a whole might give an idea about the stage of the disease as more advanced stages show higher degrees of instability or might even show fixed deformities with midfoot break and rocker-bottom deformity.

63.2.2 Imaging

Plain radiographs have a less than 50% specificity and sensitivity in the early stages of the disease and thus might be useful in ruling out other bony conditions. Radiographs might later show bone demineralization and polyarticular changes. Usually, the condition will start on the medial side with medial arch collapse especially of the cuboid height, which will progress and deform over time.

Although computerized tomography is more sensitive than plain radiographs, in detecting bone abnormalities and intra-articular fractures, due to its lack of ability to detect bony edema or microfractures that are associated with early stages of Charcot, it is therefore not routinely recommended as a diagnostic tool.

MRI on the other hand is equally effective at detecting both soft-tissue and bony changes and thus is of value in early stages of Charcot. Using the T2-weighted imaging sequences, high signal intensities might be apparent at the Lisfranc joints (early arch collapse) and subchondral bone changes of subtalar or cuboid bones. In addition, it can usefully rule out bone infection, abscess, sinus tracts, and fistulas.

Nuclear imaging techniques try to identify the biological activity in the tissues to differentiate Charcot arthropathy from osteomyelitis. Technetium 99m sulfur colloid bone marrow imaging combined with indium-111-labeled leucocyte scans may be used to improve the ability to distinguish between osteomyelitis and inflammation in the Charcot patient. Fluorodeoxyglucose proton emission tomography CT has recently been used as a diagnostic additive tool but still not as a screening test.

63.3 Charcot Arthropathy Classification

One of the most commonly used classification systems is the Eichenholtz classification (1966) which classifies Charcot stages according to the clinical, pathological, and radiological findings. A stage 0 was later added by Shibata in 1990 as the original classification lacked a stage for the early inflammatory changes without any radiological changes.

Other classification systems are based on anatomical/descriptive basis. Likewise, one of the most widely used anatomic classifications is that which was described by Brodsky. It is formed of five types. The most common is type 1: affecting tarsometatarsal and naviculocuneiform joints. Type 2 affects the talonavicular, calcaneocuboid, or subtalar joints. Ankle joint is affected in type 3A, while type 3B is the least common and affects the calcaneal tuberosity and might cause Achilles tendon avulsion. Type 4 is a combination of any of the above, while type 5 solely affects the forefoot. This classification system is simple and comprehensive and may even predict the severity and progression of the disease according to the degree of stability, deformity, and ulceration. However, it is also described as being too simple and lacks the ability to differentiate between the relatively benign midfoot arthropathy and the more complex perinavicular diseases.

Schon et al. also described an anatomical classification system to which the authors later added radiological measurements to distinguish between different types of Charcot arthropathy. However, this system is useful only for midfoot disease.

Perhaps, classification of Charcot arthropathy should include both temporal (Eichenholtz) and anatomic classifications (Brodsky), and clearly this is an area open for further research studies.

63.3.1 Charcot Arthropathy Ulcer Classification

There are numerous parameters that might need to be graded to ultimately create a comprehensive classification system for Charcot arthropathy ulcers. Among these are depth size location etiology and degree of arthropathy involved. In addition, other systemic factors need to be taken into consideration such as nutritional status, glycemic control, medical comorbidities, and secondary infection ischemia among others. All these factors if collectively considered might result in a cumbersome and impractical classification system. The most commonly used classification systems for diabetic ulcers are the Wagner-Meggitt classification, the depth ischemia, the University of Texas Classification, and the International Working Group on the Diabetic Foot (IWGDF) classification system. The latter is the most comprehensive of these classifications and is usually summarized by the acronym "PEDIS" (Perfusion, Extent and size of lesion, Depth and tissue loss, Infection severity, and Sensation). However, perhaps the most commonly used classification for ulcers is the Wagner-Meggitt classification.

63.3.2 General Guidelines in the Treatment of Charcot Arthropathy

Generally, when treating the diabetic foot and ankle, the goals are to obtain a stable plantigrade foot, to avoid abnormal plantar pressures in feet with bony deformities, to prevent ulcers from developing or reoccurring, and to allow the patient to ambulate using a combination of footwear and orthoses. However, this treatment strategy should not be limited to just that. The proper management should involve a whole team approach, ranging across an internal medicine specialist, that would be responsible for managing the diabetes itself and the associated comorbidities; a nursing specialist, that would usually provide instructions for adequate foot and antibiotic care; and in addition, routine visits to the pedorthist to maintain adequate shoe wear to allow the patient to have as normal a function and quality of life as possible.

As described previously, the condition usually progresses along Eichenholtz classification from physical signs of erythema, warmth, and swell-

ing, radiographically associated with periarticular bony debris, subchondral fragmentation, fractures, subluxations, and dislocations, which will continue on to new bone formation, sclerosis, and coalescence of large bony fragments. Eventually, this will show healed bone with some prominences that might predispose the patient to ulceration. However, this sequence might take months to years.

Diabetic foot and ankle care should begin with routine care of the skin and nails. This is an integral part of treatment and can help to avoid ulcerations and infections about the foot and ankle. Likewise, proper footwear is an important part of the overall treatment program, especially for those patients in the earliest stages of the disease, for those with a lack of sensation, or in patients with any kind of neuropathy. There are different types of shoe wear for Charcot arthropathy patients. These include healing shoes, in-depth shoes, external shoe modifications, and custom-made shoes. In addition, various orthoses or inserts can be added to the patient's footwear.

To date, total contact casting remains the gold standard for the treatment of diabetic foot ulceration. The minimal use of padding during its application allows for more precise contouring of the cast, which reduces the shear forces that might allow other conventional casting methods to slip within a very short time and cause further ulceration. The contraindications to total contact casting include infection, extensive drainage from the ulcer, poor surrounding skin, advanced arterial insufficiency, and poor patient compliance. Recurrent ulceration is known to be the most common complication of total contact casting, occurring in up to 40% of patients within the first 2 years of treatment. Generally, a total contact cast will allow for weight-bearing during the healing period of the ulcer, and it is usually changed every 1–2 weeks.

The Charcot Restraint Orthotic Walker (CROW), also known as a neuropathic walker, is a custom, bivalved, total contact ankle foot orthosis (AFO) that has full foot enclosure, a rocker-bottom sole modification, and a custom orthosis and is made from a cast model of the patient's foot and ankle (Fig. 63.1). This walker is usually recommended for patients with ulcers that are dry and less in size than 2 cm. On the other hand, if the ulcer is larger than 2 cm, moist, and particularly on plantar or malleolar surfaces, then surgical debridement, prominence removal, and reconstructive foot and ankle surgery might be considered initially.

Nonoperative interventions have been associated with a 49% risk of recurrent ulcerations, a 23% risk of bracing worn for greater than 18 months, and a 2.7% annual rate of amputation. To avoid these problems, surgical fixation and reconstruction are often necessary, especially in patients with recalcitrant wound issues and gross deformities or instability. One should apply appropriate fixation principles and constructs in order to avoid hardware failure, nonunion, and a recurrence of the deformity.

Surgical treatment for Charcot arthropathy of the foot and ankle is typically reserved for chronic, recurrent ulcerations or unstable unbraceable joints. These often present with an associated deformity or contracture of the

Fig. 63.1 Charcot Restraint Orthotic Walker (CROW)

extremity as well as with acute, displaced fractures often in patients who have adequate circulation. Usually, plates and screw bolts, long axial nails, and external fixators can be used in the management of these arthropathies.

The ankle is the least commonly affected area with CN. However, it is frequently managed surgically because the collapse that occurs is often so damaging that it frequently affects the anatomic foot alignment as a whole.

The goal is also to end with a stable well-aligned limb that can be safely and permanently braced to allow weight-bearing on a plantigrade foot. Like all Charcot neuropathies, successful fusion and stabilization of the Charcot ankle are only the first step in the long-term care of these patients. However, due to the large amount of stress seen at the ankle joint with normal walking, long-term care should also include permanent bracing, to help shield the fused joint from pressures that can create a new injury and start the process all over again.

On the other hand, even though relatively more common, Charcot midfoot deformity tends to be more difficult to treat efficiently. Most patients will be managed effectively with bracing, but there will remain a percentage of patients with significant deformity and instability who cannot be adequately managed without surgery. With the evolution of techniques and implants, a super-construct concept has been developed in an effort to increase stability and reduce the risk of fixation failure, especially in Charcot arthropathy of the midfoot. This super-construct concept is based on four main criteria:

(a) Fusion extending beyond the injury and including unaffected joints; (b) bone resection with shortening of the extremity allowing for deformity correction and reduction of soft-tissue tension; (c) use of the strongest devices tolerated; and (d) hardware application in a position that optimizes mechanical function.

External fixation for treatment of Charcot arthropathy has also been used and with good reported outcomes. However, due to numerous complications associated with its application and maintenance, it is not recommended for the inexperienced surgeon.

Take-Home Message

- Charcot foot arthropathy is a progressive, noninfectious, destructive inflammatory process of the foot and ankle.
- Charcot foot and ankle patients are usually between their fourth and sixth decades of life.
- Generally, when treating the diabetic foot and ankle, the goals are to obtain a stable plantigrade foot, to avoid abnormal plantar pressures in feet with bony deformities, to prevent ulcers from developing or reoccurring, and to allow the patient to ambulate using a combination of footwear and orthoses.
- To date, total contact casting remains the gold standard for the treatment of diabetic foot ulceration.

Summary

Charcot foot arthropathy is a progressive, noninfectious, destructive inflammatory process of the foot and ankle. It was originally described by the French neurologist Jean-Martin Charcot, as a sequela of tertiary syphilis. However, in the vast majority of patients today, it is secondary to long-standing diabetes. In the USA alone, there are more than 29 million adults with diabetes, and these numbers are increasing worldwide. There is no single cause for Charcot arthropathy. However, two causative theories for the pathogenesis of Charcot arthropathy exist, which are the neurotraumatic and neurovascular theories. Charcot foot and ankle patients are usually between their fourth and sixth decades of life. They will present with an acute inflammatory-like episode of redness, hotness, and swelling. Plain radiographs have a less than 50% specificity and sensitivity in the early stages of the disease and thus might be useful in ruling out other bony conditions. Although computerized tomography is more sensitive than plain radiographs, in detecting bone abnormalities and intra-articular fractures, due to its lack of ability to detect bony edema or microfractures that are associated with early stages of

Charcot, it is therefore not routinely recommended as a diagnostic tool.

One of the most commonly used classification systems is the Eichenholtz classification (1966), which classifies Charcot stages according to the clinical, pathological, and radiological findings.

Generally, when treating the diabetic foot and ankle, the goals are to obtain a stable plantigrade foot, to avoid abnormal plantar pressures in feet with bony deformities, to prevent ulcers from developing or reoccurring, and to allow the patient to ambulate using a combination of footwear and orthoses. The proper management should involve a whole team approach.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. The etiology of Charcot arthropathy has:
   (a) Two theories: neurotraumatic and neurovascular
   (b) Two theories: traumatic and infective
   (c) Three theories: endocrine, genetic, and traumatic
   (d) Hereditary characteristics
2. X-ray in Charcot arthropathy:
   (a) Has less than 50% of specificity
   (b) Has more than 80% of specificity
   (c) Is not routinely used
   (d) Is the gold standard
3. The most common classification system used is:
   (a) Eichenholtz classification
   (b) Denis classification
   (c) Ficat classification
   (d) Weber classification
4. The Charcot Restraint Orthotic Walker:
   (a) Is usually recommended for patients with ulcers that are dry and less in size than 2 cm
   (b) Is usually recommended for patients with ulcers that are dry and less in size than 5 cm
   (c) Is not usually recommended
   (d) Is used in all patients with Charcot arthropathy
5. Surgical treatment for Charcot arthropathy of the foot and ankle is reserved:
   (a) For chronic, recurrent ulcerations or unstable un-braceable joints
   (b) In each case
   (c) In the early stages of the disease
   (d) Only for non-ambulant patients

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Ankle and Foot Fractures

64

Kenneth J. Hunt, Marissa Jamieson, and Jason Koerner

Overview

This chapter gives an overview on the most commonly encountered foot and ankle fractures. Applied anatomy, epidemiology, common injury mechanisms, diagnosis, and appropriate management including surgical technique are covered.

64.1 Metatarsal Fractures

64.1.1 Definition/Anatomy

- The first metatarsal bears 30–50% of weight-bearing and has no intermetatarsal ligament support.
- Dense intermetatarsal ligaments between the second through fifth metatarsals help to maintain the length and stability of isolated fractures.
- Multiple metatarsal fractures lose the stability of the intermetatarsal ligaments.

64.1.2 Epidemiology

- Metatarsal fractures are among the most common injuries to the foot.
- The first metatarsal is the most common fracture in children less than 4 years old, and the fifth metatarsal is the most common in adults.

64.1.3 Pathogenesis

- Direct crush injury.
- Indirect mechanisms involve twisting the leg with the foot in a fixed position.
- Fifth metatarsal avulsion fractures result from an inversion twisting mechanism.

64.1.4 Classifications

- Based on the location (base, shaft, neck, head), description of the fracture pattern (non-displaced, comminuted, oblique, transverse, angulation), and articular involvement (head or base).

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Table 64.1 Fifth metatarsal fractures

Classification	Fracture location	Mechanism	Management
Zone 1 Pseudo-Jones	Base avulsion fracture	Twisting, inversion of the hindfoot	Protected weight-bearing in a stiff-soled shoe
Zone 2 Jones	Meta-diaphyseal junction, goes into the 4th–5th metatarsal articulation	Forced adduction of the foot	Non-weight-bearing × 6 weeks, surgical consideration in elite athletes
Zone 3	Metatarsal shaft	Repetitive stress, twisting of the foot	Protected weight-bearing in a stiff-soled shoe

- Fifth metatarsal fractures have their own unique classification system: zone 1 is an avulsion off the base by the peroneus brevis tendon and/or the lateral plantar aponeurosis, zone 2 is a fracture at the meta-diaphyseal junction (see section on Jones fractures), and zone 3 is a diaphyseal (shaft) fracture (see Table 64.1).

64.1.5 Diagnosis (Clinical and Imaging)

- Physical exam: ecchymosis and swelling to the forefoot with focal tenderness over the metatarsals.
- Standard weight-bearing foot radiographs including anterior-posterior (AP), oblique, and lateral images are sufficient for diagnosis.
- Computed tomography (CT) scans are rarely indicated for complex or intra-articular fracture patterns.

64.1.6 Treatment

- The majority of isolated fractures can be treated conservatively with protected weight-bearing in a hard-soled shoe or walking boot for 4–6 weeks.
- Surgical indications include multiple metatarsal fractures with significant shortening and/or displacement, open fractures, first metatarsal fractures with any displacement, significant sagittal plane deformity, and intra-articular involvement with displacement.
- Surgical techniques include open reduction and internal fixation with plate and screw constructs or retrograde/antegrade pinning with Kirschner wires.

64.2 Jones Fracture

64.2.1 Definition/Anatomy

- Fracture of the base of the fifth metatarsal at the meta-diaphyseal junction (zone 2).
- This area is predisposed to delayed healing due to the poor blood supply in this region.

64.2.2 Epidemiology

- Common in athletes.
- Cavus foot or varus hindfoot can exist as a predisposing condition.

64.2.3 Pathogenesis

- Common mechanism is forced forefoot adduction.

64.2.4 Classifications

- (See section on metatarsal fractures.)

64.2.5 Diagnosis (Clinical and Imaging)

- Physical exam: focal tenderness over the base of the fifth metatarsal, often pain with resisted eversion.
- Weight-bearing AP, oblique, and lateral foot radiographs are sufficient for diagnosis.
- MRI can be used for the diagnosis of a stress fracture.

64.2.6 Treatment

- Non-weight-bearing for 6 weeks in a short leg cast or walking boot.
- Intramedullary screw fixation should be considered for athletes to help prevent refracture and nonunion.
- Open reduction and internal fixation with a plate and screw construct in revision cases and cases of nonunion.

64.3 Lisfranc Injuries

64.3.1 Definition/Anatomy

- A disruption or dislocation of the tarsometatarsal (TMT) joint complex, which includes the metatarsal-cuneiform and metatarsal-cuboid articulations.
- The Lisfranc ligament runs from the medial cuneiform to the base of the second metatarsal and is critical in stabilizing the first and second TMT joint complex.
- The base of the second metatarsal is recessed between the medial and lateral cuneiforms acting as a "keystone" for the arch of the foot.

64.3.2 Epidemiology

- Relatively rare injuries and are missed in up to 20% of cases

64.3.3 Pathogenesis

- Direct crush injury to the foot
- Most commonly an indirect mechanism involving an axial load to a plantar-flexed foot or an abduction force to the foot (sports, missing a step off a curb, fall from a height, motor vehicle collision)

64.3.4 Classifications

- Purely ligamentous, which involves widening between the first and second metatarsals and/or medial and middle cuneiforms or is associated with fractures of the metatarsals and/or tarsals.
- Based on the direction of displacement/dislocation of the metatarsals: homolateral metatarsals displace in the same direction (typically lateral), and divergent metatarsals displace in opposite directions.

64.3.5 Diagnosis (Clinical and Imaging)

- Physical exam: Plantar ecchymosis is highly suggestive of a Lisfranc injury and focal midfoot tenderness.
- Weight-bearing AP, oblique, and lateral foot radiographs are critical for diagnosis because this tests the integrity and stability of the ligamentous complex.
- The uninjured side should always be included for comparison in order to detect subtle injuries (see Fig. 64.1).
- On the AP radiograph, the medial border of the second metatarsal should align with the medial border of the middle cuneiform, on the oblique view the medial border of the fourth metatarsal should align with the medial border of the cuboid, and on the lateral view the dorsal (superior) border of the first metatarsal should align with the dorsal border of the medial cuneiform.

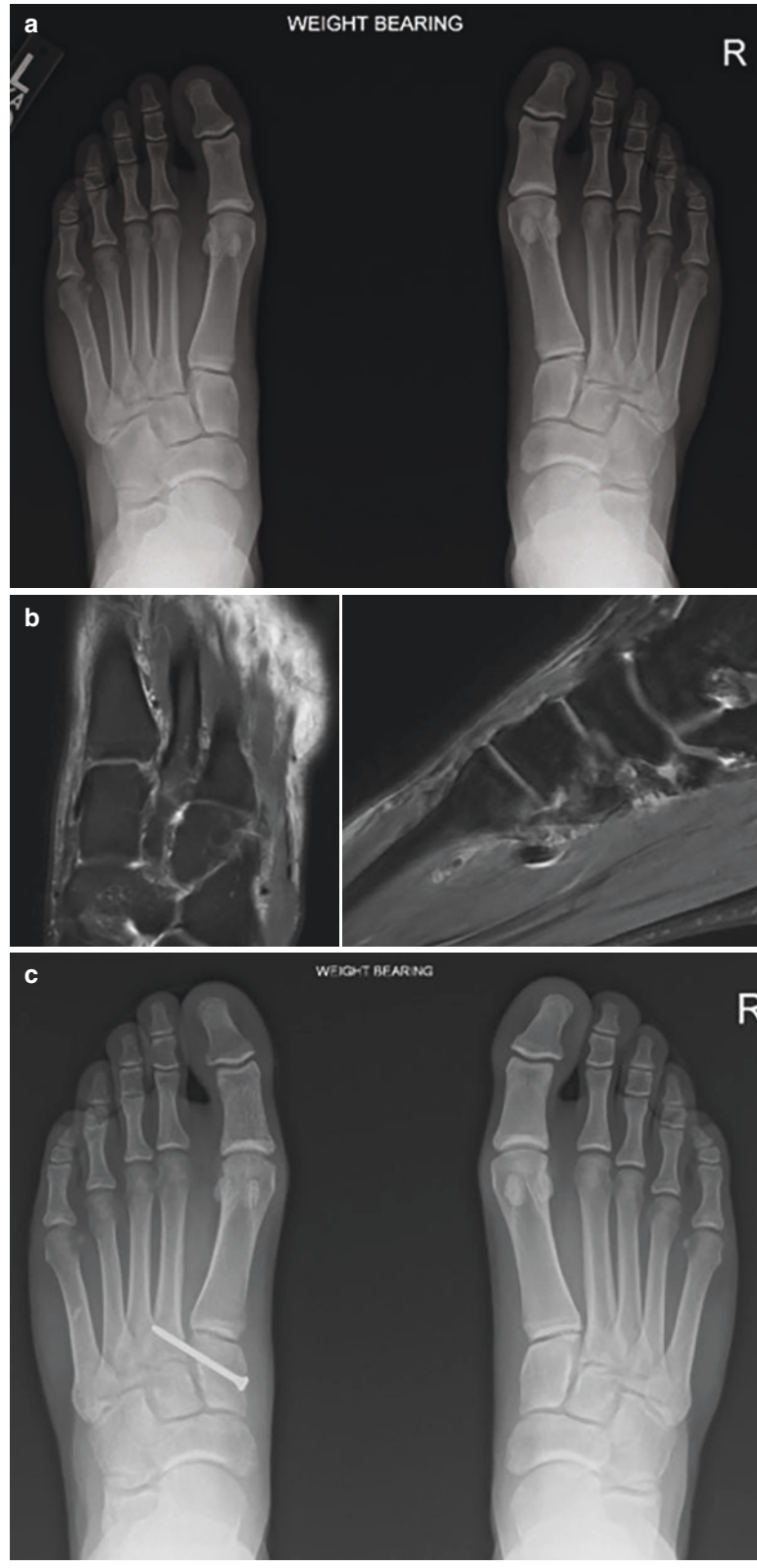

Fig. 64.1 (**a**) Weight-bearing bilateral AP radiograph showing a subtle ligamentous Lisfranc injury with widening between the first and second metatarsals; (**b**) axial and sagittal T2 MRI images showing a tear of the Lisfranc ligament; (**c**) status post-open reduction and screw fixation across the medial cuneiform and second metatarsal in line with the Lisfranc ligament

- A CT scan is often necessary if there are associated fractures in order to define the extent of fracture comminution and evaluate alignment.
- Weight-bearing CTs are helpful in identifying subtle widening of the TMT joints.
- MRI is typically not necessary unless there is a high index of suspicion for ligamentous injury and the patient is unable to tolerate weight-bearing.

64.3.6 Treatment

- Truly non-displaced injuries without displacement on weight-bearing or stress radiographs can be treated with protected weight-bearing in a short-leg cast or removable boot for 6 weeks followed by progressive weight-bearing.
- Unstable injuries need surgical intervention, which typically involves open reduction and internal fixation of the fractures with hardware (screws and/or plates) stabilizing the dislocated joints.
- Primary fusion of the TMT joints is sometimes used for highly comminuted fractures, significant instability, or delayed diagnosis/presentation.

64.4 Stress Fractures

64.4.1 Definition/Anatomy

- An overuse injury that occurs due to repetitive stress causing microfractures in the bone

64.4.2 Epidemiology

- Very common injuries, most common in the second and third metatarsals
- Common in runners, military recruits, ballerinas, and patients with the female athlete triad (eating disorders, amenorrhea, low bone mineral density)
- Associated with an increase or change in activity level

64.4.3 Pathogenesis

- Failure of the bone to respond to increased or repetitive stresses and/or abnormal loading
- Can also be caused by abnormal bone related to osteopenia or metabolic disease being unable to withstand physiologic stresses

64.4.4 Classifications

- Numerous classification systems
- Described based on location, MRI appearance (increased edema with or without fracture line), fracture line visible on radiographs, and displaced or non-displaced fracture

64.4.5 Diagnosis (Clinical and Imaging)

- Physical exam: Point tenderness over the bone and pain that is worse with weight-bearing is highly suggestive of a stress fracture, and swelling is variable.
- Standard foot/ankle radiographs are often normal initially and will show callus formation after 6–8 weeks.
- MRI is the most sensitive test for detecting a stress fracture or stress reaction; the earliest sign is increased T2-weighted signal indicating bone edema.
- CT can be used to better define the fracture line, particularly useful in navicular stress fractures.
- Nuclear imaging (bone scan) is highly sensitive but not specific and has largely been replaced with MRI.

64.4.6 Treatment

- The majority of foot and ankle.
- Stress fractures can be treated with protected weight-bearing in a stiff-soled shoe or a walking boot and gradual return to activities, typically 10–12 weeks until return to play.
- Painful activities including running or other weight-bearing exercises should be stopped until they are no longer painful.
- High-risk stress fractures including the base of the fifth metatarsal, navicular, and talus are prone to delayed healing and nonunion and should be treated with approximately 6 weeks of non-weight-bearing.
- Addressing any underlying vitamin D deficiency or osteopenia is imperative for appropriate healing.
- Any deformity in the foot that is contributing to abnormal loading (i.e., varus foot with fifth metatarsal fractures) should be corrected with orthotics or bracing.

64.5 Ankle Fractures

64.5.1 Definition/Anatomy

- Comprised of three parts: the medial malleolus, lateral malleolus, and posterior malleolus
- Mortise: Comprised of the medial and lateral malleoli with the tibial plafond over the talus
- Syndesmosis: Comprised of the anterior-inferior tibiofibular ligament (AITFL), posterior-inferior tibiofibular ligament (PITLF), interosseous membrane, interosseous ligament (IOL), and inferior transverse ligament (ITL), which maintains the tibiofibular joint distally

64.5.2 Epidemiology

- Most common fracture of the lower limb
- Accounts for 10% of all fractures

64.5.3 Pathogenesis

- A rotational injury about the ankle joint dependent on the position of the foot

64.5.4 Classifications

- Can describe as medial malleolar, lateral malleolar, or posterior malleolar fracture; bimalleolar fracture (combination of two of these fractures); or trimalleolar fracture (involves all three malleoli)
- Denis-Weber classification (classification of distal fibula fractures): A: below syndesmosis, B: at the level of syndesmosis, C: above syndesmosis
- Lauge-Hansen classification: supination-adduction (SA), supination-external rotation (SER), pronation-abduction (PA), and pronation-external rotation (PER)

64.5.5 Diagnosis (Clinical and Imaging) (Fig. 64.2)

- Plain radiographs: AP, lateral, mortise
- Must obtain full-length tibia to evaluate for Maisonneuve
- Evaluate syndesmosis (findings concerning for syndesmotic injury)
  - Decreased tibiofibular overlap: normal >6 mm on AP, >1 mm on mortise
  - Increased tibiofibular clear space: <6 mm overlap (AP or mortise) measured 1 cm above the joint
  - Increased medial clear space: normal <4 mm on mortise

64.5.6 Treatment

- Must first evaluate for dislocation; if dislocated, reduce immediately.
- Initial treatment for all ankle fractures consists of splinting in three-way splint or CAM

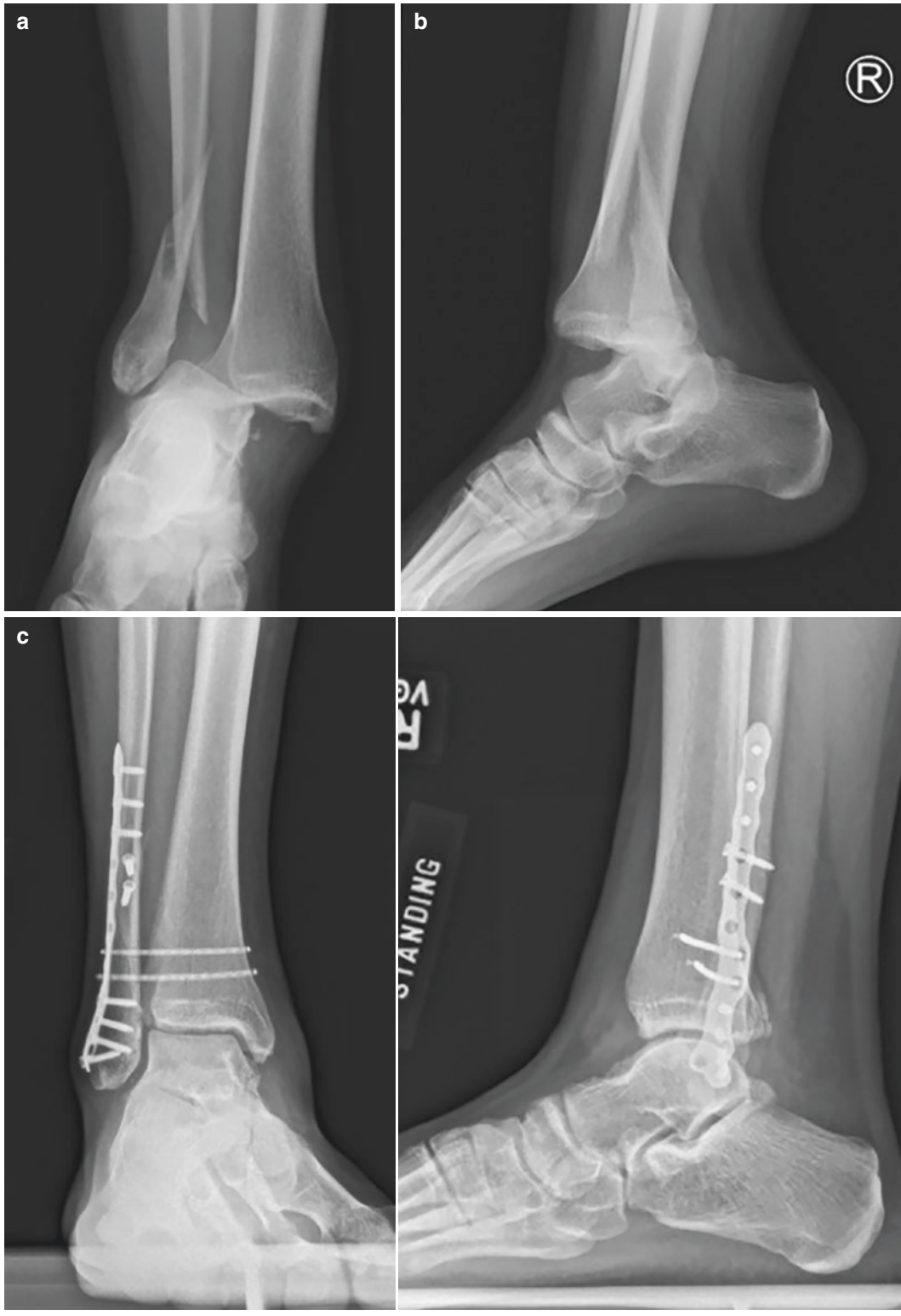

Fig. 64.2 AP (**a**) and lateral (**b**) radiograph showing a lateral malleolus fracture with tibiotalar subluxation, (**c**) mortise and lateral radiograph showing the injury status post-open reduction and internal fixation of the lateral malleolus fracture and syndesmosis

boot for stabilization. For operative fractures, allow 2 weeks for swelling to decrease prior to making incision.

- Nonoperative:
  - Medial malleolus: non-displaced
  - Lateral malleolus: <3 mm displacement with no talar shift, Weber A fractures
  - Posterior malleolus: <25% of joint surface
- Operative:
  - Parameters greater than those listed above, most ankle fractures will fall into this category
  - Intraoperatively must assess the syndesmosis with external rotation stress view or cotton test
  - If syndesmosis unstable, can fix with cortical screws or tight rope fixation
- The goal of operative treatment is to re-create an anatomic ankle joint along the articular surface to reduce the chance of post-traumatic arthritis.
- More stiff constructs are recommended for patients with diabetes mellitus.

64.6 Calcaneus Fractures

64.6.1 Definition/Anatomy

- Tarsal bone with multiple articulations and a major weight-bearing bone
- Articulates with the talus and cuboid
- Superolateral portion of talus comprised of anterior, middle, and posterior facet, which articulate with talus
- Sustentaculum tali: medial projection with multiple attachments, FHL passes beneath

64.6.2 Epidemiology

- Most commonly fracture tarsal bone
- Up to 75% involve the articular surface
- Associated injuries include vertebral body compression fractures and contralateral calcaneal fractures up to 10% of the time

64.6.3 Pathogenesis

- High-energy axial load
- Commonly seen with falls from height, floorboard intrusion during motor vehicle accidents

64.6.4 Classifications

- Described as extra-articular or intra-articular
- Intra-articular classifications:
  - Sanders classification: Uses CT to identify the number of fragments at the widest part of posterior facet
  - Essex-Lopresti: Identifies tongue-type versus joint depression

64.6.5 Diagnosis (Clinical and Imaging) (Fig. 64.3)

- Physical exam: Severe swelling, ecchymosis, and deformity of the heel. Evaluate for puckering skin, skin blanching, or open wounds especially in tongue-type pattern.
- Radiographs: AP, lateral, oblique of foot.
- Harris view (hindfoot alignment view): Obtained with maximum dorsiflexion and X-ray angle at 45°.
- Böhler's angle: Angle line between anterior and posterior facet with line tangential to superior tuberosity:
  - Normal 20–40°
- Angle of Gissane: Normal 120–145°.
- CT is the gold standard for evaluating these fractures.
- MRI can help identify stress fractures.

64.6.6 Treatment

- First step is to evaluate for tongue-type patterns and evaluate for skin compromise around the fracture site (surgical emergency)

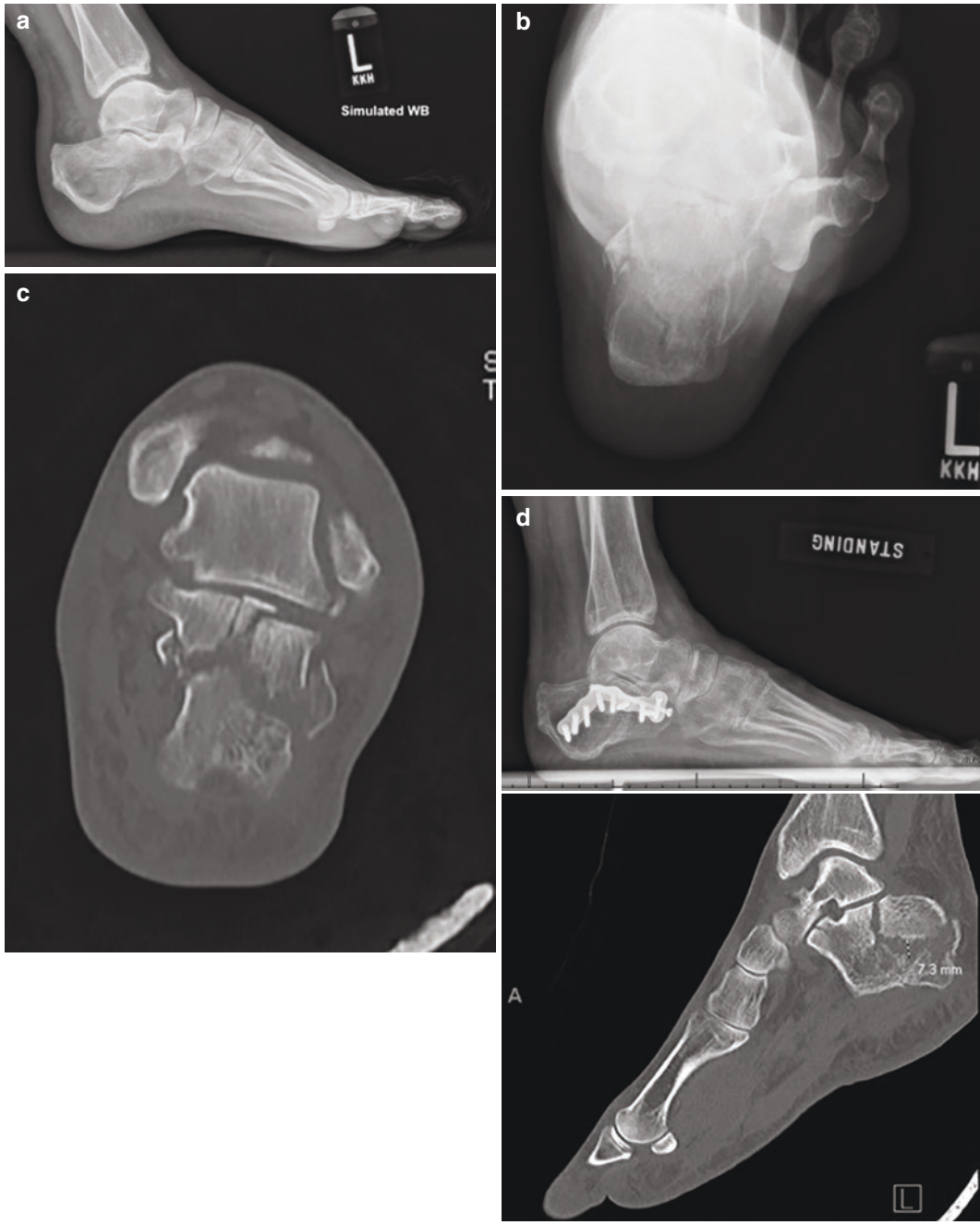

Fig. 64.3 (**a**) Lateral radiograph demonstrating an intra-articular calcaneus fracture, (**b**) axial Harris view demonstrating a comminuted calcaneus fracture, (**c**) coronal and sagittal CT image demonstrating fracture, (**d**) lateral radiograph status post-open reduction and internal fixation with a plate and screw construct

- Nonoperative management:
  - Stress fractures can be treated in cast for 6 weeks
  - Non-displaced fractures
- Operative management:
  - Tongue type, displaced fractures >2 mm, >25% of articular surface, flattening of Böhler's angle, varus alignment

  - Reduce all fractures to the "constant" fragment, which is the superomedial fragment:
    Contains the sustentaculum tali that has strong capsular and ligamentous attachments
- Skin assessment: Critical for surgical management, can perform wrinkle test to determine the ability to make incision

64.7 Talus Fractures

64.7.1 Definition/Anatomy

- Complex bone that has five articulating surfaces
- 70% of bone covered with articular cartilage
- No tendon or muscular attachments
- Has a direct blood supply to the bone but has tenuous blood supply, which puts this bone at high risk for avascular necrosis

64.7.2 Epidemiology

- Second most common fractured tarsal bone.
- <1% of all fractures.
- Talar neck fractures most common, followed by talar body and lateral process, respectively. Least common is talar head fracture.

64.7.3 Pathogenesis

- High-energy mechanism with ankle dorsiflexed with axial load.
- Different positions of the ankle will cause fractures on different parts of the talus.

64.7.4 Classifications

- Hawkins classification (classification for talar neck fractures): Type I: non-displaced, type II: subtalar dislocation, type III: subtalar and tibiotalar dislocation, and type IV: subtalar, tibiotalar, and talonavicular dislocation:
  - This classification also predicts the risk of avascular necrosis.

64.7.5 Diagnosis (Clinical and Imaging)

- Physical exam: Swelling, can have deformity depending on the type of fracture. Palpate lateral process of talus in patients with ankle sprains to not miss this fracture.
- Radiographs: AP, lateral, oblique of the foot:
  - Canale view: best view to visualize the talar neck:
    Obtained with beam 75° cephalad from horizontal. Foot position is maximum plantar flexion and 15° of pronation
  - Hawkins sign: Indicates revascularization of the talus:
    Lucency of the talar dome is good, if there is sclerosis at the talar dome; this means avascular necrosis is occurring.
- CT: Gold standard for evaluating articular surface, displacement, and comminution

64.7.6 Treatment

- Initial step is identifying if dislocation is present; if present, emergent closed reduction in emergency department
- Nonoperative management (short-leg cast):
  - Non-displaced talar neck fractures without articular step-off
  - All other talus fractures with <2 mm step-off
- Operative management:
  - Open reduction and internal fixation
    Displaced talar neck fractures
    All other talus fractures with >2 mm step-off
  - Fragment excision:
    Comminuted lateral or posterior process fractures
- Talar neck fractures will be treated with plates and screws, and other talus fractures can be treated with k-wires until union.
- Complications:

- 31% of talar neck fractures will develop avascular necrosis.
- Post-traumatic arthritis is most likely to occur in the subtalar joint (50%) and is the most common complication.

64.8 Navicular Fractures

64.8.1 Definition/Anatomy

- Named navicular because of its resemblance to a small boat.
- Navicular bone has multiple articulations with cuneiforms, cuboid, calcaneus, and talus.
- Plays an important role in the eversion and inversion of the foot.

64.8.2 Epidemiology

- Can be caused by acute trauma or repetitive bouts of trauma
- Stress fractures:
  - Usually seen in running athletes with repetitive trauma by running on hard surfaces
- Different positions of the foot at impact can cause navicular avulsion, tuberosity, or body fractures
- Mueller-Weiss syndrome
  - Spontaneous navicular avascular necrosis found in patients with chronic midfoot pain

64.8.3 Pathogenesis

- Navicular body: axial load
- Navicular tuberosity: contraction of PTT with eversion of the foot
- Navicular avulsion: forced plantar flexion, sometimes seen with forced eversion/inversion

64.8.4 Classifications

- Can describe by the location of fracture
- Sangeorzan classification: Navicular body fractures based on the degree of comminution and plane of fracture

64.8.5 Diagnosis (Clinical and Imaging)

- Physical exam: tenderness to palpation over the navicular bone with associated midfoot swelling
- Radiographs: standard AP, lateral, oblique views of foot
- If high suspicion for navicular fracture, can obtain CT scan or MRI to further evaluate

64.8.6 Treatment

- Stress fractures:
  - Most navicular stress fractures can be initially treated with cast immobilization and non-weight-bearing for 6–8 weeks.
  - Surgery is reserved for nonunions and high-level athletes.
- Traumatic fractures:
  - Most avulsion fractures and minimally displaced fractures can be treated with immobilization in short-leg cast.
  - Indications for surgery are fracture with >25% articular involvement and >5 mm displacement and can be treated with ORIF.
  - Symptomatic nonunions of avulsion fractures can be treated with fragment excision.

Take-Home Message

- Weight-bearing (if tolerated) radiographs should be the initial study performed in patients suspected of having a foot or ankle injury.
- Many fractures can be treated successfully conservatively with protected weight-bearing.
- High-risk fractures that often need surgical intervention include Lisfranc fracture/dislocations, ankle fractures, calcaneus and talus fractures, multiple metatarsal fractures, navicular fractures, and Jones fractures in athletes.
- Operative techniques typically involve open reduction and internal fixation with a variety of pins and/or plate and screw constructs.

Summary

Foot and ankle fractures are very common injuries. A patient with a history of trauma or twisting to the foot or ankle and localized swelling and tenderness should have standard weight-bearing radiographs of the injured extremity. This will be sufficient to make a diagnosis in the majority of cases. MRI and CT scans can be useful adjuncts in specific fracture patterns or when there is a high index of suspicion but negative radiographs. Prompt referral to an orthopedic surgeon is important for high-risk fractures.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Which fracture should be treated with ORIF?
   (a) Bimalleolar ankle fx
   (b) Isolated metatarsal fracture
   (c) Non-displaced cuboid
   (d) Zone 1 fifth metatarsal base avulsion
2. Which fracture should be treated with strict non-weight-bearing?
   (a) Jones fracture
   (b) Metatarsal neck fracture
   (c) Bad calc fracture
   (d) Navicular avulsion fracture
3. What is the next appropriate step in the evaluation of a patient with normal NWB foot radiographs who has tenderness along their dorsal midfoot and plantar ecchymosis?
   (a) Hard-soled shoe and WBAT
   (b) CT scan
   (c) MRI
   (d) Bilateral weight-bearing radiographs
4. What is a risk factor for the following injury? MRI of stress fx of foot:
   (a) Recent increase in activity level
   (b) Vitamin D deficiency
   (c) History of amenorrhea
   (d) Disordered eating
   (e) All of the above
5. For which of the following fractures is a CT scan appropriate?
   (a) Lateral malleolus fracture
   (b) Multiple cuneiform and metatarsal base fractures
   (c) Third and fourth metatarsal neck fractures
   (d) Lateral malleolus avulsion fracture
   (e) Comminuted calcaneus fracture
   (f) Both (b) and (e)

Further Reading

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Part XII

Shoulder

65 Acromioclavicular and Sternoclavicular Joint Injuries

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Overview

The acromioclavicular (AC) and sternoclavicular (SC) joints function to connect the upper extremity to the axial skeleton. Injury to these ligamentous-restrained joints often results from trauma. AC joint injuries are common and account for 9% of all shoulder injuries. SC joint injuries are rare, but potentially life-threatening secondary to their close proximity to important vascular structures and critical airways. Most injuries can be managed nonoperatively and reserve surgery for high-grade injuries or those that fail to heal nonoperatively. The anatomy, mechanisms of injury, physical examination, imaging, and treatment will be discussed for both AC and SC joint injuries.

65.1 Acromioclavicular Joint Injuries

65.1.1 Anatomy

The AC joint is a diarthrodial joint that helps suspend the upper extremity from the thorax. It contains an intra-articular meniscus-like structure to reduce pressure and enhance motion. The AC joint is surrounded by a thin capsule and circumferentially reinforced with ligaments, known as the AC ligamentous complex (ACLC). The ACLC is composed of anterior, posterior, superior, and inferior capsuloligamentous fibers. Nakazawa et al. described these fibers as a superoposterior bundle and anteroinferior bundle. On average, the ACLC inserts 2.8 mm onto the acromion and 3.5 mm onto the distal clavicle. The ACLC stabilizes the AC joint by reducing anterior-posterior translation and centering the AC joint during clavicular rotation. The superior and posterior segments of the ACLC provide the greatest restriction to posterior translation.

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UG. Longo, V. Denaro (eds.), *Textbook of Musculoskeletal Disorders*,
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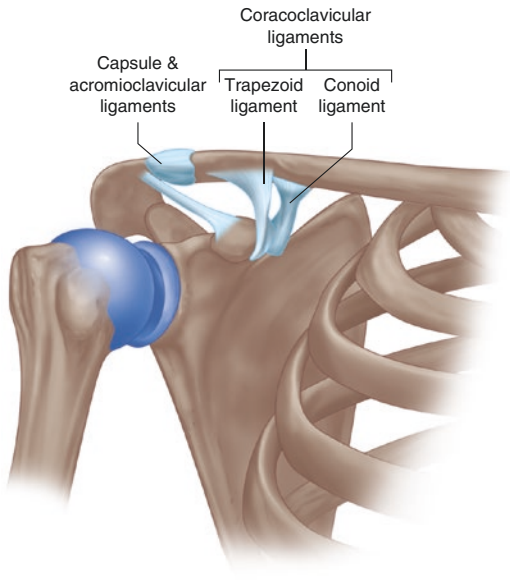

Fig. 65.1 Anatomy of the acromioclavicular (AC) joint. Static joint stability is provided by the AC ligamentous complex (ACLC) and the coracoclavicular (CC) ligaments. The conoid and trapezoid ligaments make up the CC ligaments. This image was originally published by Funk L, Imam MA. Acromioclavicular Joint Injuries. In: Funk L, Walton M, Watts A, Hayton M, Ng CY, eds. Sports Injuries of the Shoulder. Switzerland: Springer; 2020. p 153

The deltotrapezial fascia is confluent with the superior ACLC and provides additional static restraint. Moreover, the deltoid and trapezius muscles provide dynamic stabilization.

The coracoclavicular (CC) ligaments (conoid and trapezoid ligaments) provide additional stability to the AC joint (Fig. 65.1). The conoid is medial and runs superomedially from the posteromedial angle of the coracoid to the conoid tubercle of the clavicle. The trapezoid is lateral and runs superolaterally from the anterolateral angle of the coracoid to the clavicle. From the distal clavicle, the conoid and trapezoid attachments consistently attach at 17% and 31% of the total clavicular length, respectively. Knowledge of these attachments is important to replicate the native anatomy during anatomic reconstructions. Following injury to the ACLC, the trapezoid is the primary restraint to posterior translation and the conoid is the primary restraint to anterior and superior translation. The conoid ligament also resists anterior and superior clavicular rotation.

65.1.2 Mechanisms of Injury

AC joint injuries are caused by direct and indirect trauma. The AC joint is vulnerable to traumatic forces due to its superficial location with limited soft-tissue protection. Direct trauma transpires from a direct blow to the acromion with the arm adducted, often during contact sports (e.g., football, hockey). The noteworthy stability of the SC joint causes the forces to transfer to the AC joint, leading to injury. Biomechanical studies have shown a sequential disruption of the ACLC, CC ligaments, and ultimately deltoid and trapezial muscles. Conversely, indirect trauma ensues from a fall on an outstretched hand or elbow. Forces from the fall cause the humeral head to thrust into the acromion, resulting in superior acromial translation.

65.1.3 Physical Examination

Physical examination should begin with evaluation of the cervical spine to assess for radicular pain originating from a cervical nerve root (C5 provides afferent signals from the shoulder).

Inspect the shoulder with the patient upright and the arm unsupported. The weight of the arm provides traction to accentuate physical deformities (e.g., "step-off"). Tenderness to direct palpation of the AC joint is the most common examination finding (innervated by the lateral pectoral nerve). Pain disproportionate to the exam should raise suspicion for a coracoid fracture or type IV injury.

Special examination techniques used to evaluate the AC joint include the cross-arm adduction test, AC resisted extension test, and active compression (O'Brien) test. Each test has a low sensitivity performed individually (77%, 72%, 41%), but a high specificity performed together (97%). Pain with cross-arm adduction that is relieved following local anesthetic injection is highly suggestive of AC joint injury. AC joint pain secondary to resisted extension and active compression (in pronation, but not supination) is consistent with AC joint injury. Additional provocative tests include internal rotation behind the back and adduction with internal rotation (Hawkins-Kennedy test).

The shrug test differentiates between type III and type V injuries. It is performed by having the patient shrug both shoulders. If the clavicle reduces, the deltotrapezial fascia is intact (type III). If the clavicle fails to reduce, the deltotrapezial fascia is ruptured (type V).

65.1.4 Imaging

A standard set of radiographs to assess AC joint injuries include anteroposterior (AP), bilateral Zanca, and axillary views. AP views are nonspecific for AC joint injury but provide a broad understanding of the AC joint integrity. A bilateral Zanca view (AP orientation with 10–15° cephalic tilt) is the most important radiographic view for AC joint injury assessment. Differences in the AC joint and CC interspace are easily identifiable as the injured and non-injured shoulders can be evaluated simultaneously. An axillary view is particularly useful to evaluate posterior clavicular displacement (as seen in type IV injuries).

Less standard radiographic views include cross-arm adduction (Basamania), Stryker notch, and weighted stress views. The cross-arm adduction view detects an overriding clavicle during anteromedial scapular translation. The Stryker notch view (10° cephalic tilt with the patient supine and palm on his/her head) can detect a coracoid fracture. The weighted stress view (5 lbs weight tied to the wrists) provides humeral traction to distinguish between type II and type III injuries; however, it is rarely used due to patient discomfort and marginal clinical significance.

65.1.5 Classification

The Rockwood classification uses radiographic findings to stratify the severity of AC joint injury (types I–VI). This system provides information regarding the integrity of the ACLC, CC ligaments, and clavicular displacement; however, it is significantly limited given the lack of clinical findings. The International Society of Arthroscopy, Knee Surgery and Orthopaedic Sports Medicine (ISAKOS) Upper Extremity Committee expanded upon the Rockwood classification by considering scapular function and response to therapy (types IIIA and IIIB).

65.1.6 Management

There is consensus in treating low-grade AC joint injuries (types I and II) nonoperatively and high-grade injuries (types IV–VI) operatively. Controversy exists in managing type III injuries, although most opt for a trial of nonoperative rehabilitation and reserve surgery for those whose symptoms fail to improve. Recent evidence suggests that nonoperative management of high-grade injuries yields successful outcomes.

65.1.6.1 Nonoperative Management

The goal of nonoperative management is to achieve periscapular motor control to permit painless, effective function. Periscapular control is achieved by minimizing pain and increasing range of motion, muscle strengthening, and function-specific activities. Effective rehabilitation often progresses through brief immobilization, active-assisted range of motion, closed-chain exercises, open-chain exercises, scapular strengthening (e.g., isokinetic low rows), and sport/function-specific activities.

65.1.6.2 Operative Management

Numerous AC reconstruction techniques have been described and biomechanically evaluated. The techniques can be classified into four categories: ligament and tendon transfers, acromioclavicular joint stabilization, coracoclavicular interval stabilization, and anatomic ligament reconstruction. The operative techniques, biomechanical findings, and clinical outcomes will be discussed for each category.

65.1.6.3 Ligament and Tendon Transfers

Cadenet was the first to describe coracoacromial ligament (CAL) transfer for AC joint injuries. The Weaver-Dunn (W-D) technique was later described, which involved distal clavicle excision

and CAL transfer to the distal clavicle. Multiple modifications to the W-D procedure have been reported, including CC cerclage, transarticular K wire fixation, and transfer of a bone block to improve strength. AC joint stabilization has also been managed with transfer of the conjoined tendon or long head of biceps tendon to the distal clavicle.

CAL transfers have consistently demonstrated increased laxity and decreased strength compared to the native anatomy. Deshmukh et al. identified increased laxity in anterior, posterior, and superior directions with the W-D procedure. Furthermore, Harris et al. reported that the CAL ligament strength was 145 N, significantly less than the native CAL strength of 312 N identified by Soslowsky et al. Improvements in CAL transfer strength have been afforded by ACLC repair and augmented CC stabilization with suture anchors or bicortical screws.

Early reports of CAL transfers and modified W-D procedures showed favorable outcomes. However, a 2008 systematic review by Sood et al. concluded that only low-level evidence supports CAL transfer and supplemental fixation did not improve outcomes, questioning the utility of this technique.

65.1.6.4 Acromioclavicular Joint Stabilization

AC joint stabilization techniques include transarticular pins/wires and hook plates. Transarticular AC joint fixation using K wires was first described by Phemister in 1942. However, K wires have largely been abandoned secondary to complications with hardware failure and loss of reduction. Fialka et al. identified transarticular K wires as having the highest rate of implant loosening when compared to CC screws and synthetic AC ligament reconstruction. Bargren et al. identified a 40% failure rate at 6 weeks postoperatively, attributed to the inability to resist large bending moments.

Hook plates are contoured to the AC joint to provide rigid fixation. Overall, these devices have excellent stiffness, but fail to restore normal AC joint kinematics. McConnell et al. identified that hook plate stiffness was similar to the native AC joint (26 ± 17 N/m vs. 25 ± 8 N/m). Hook plates provide excellent superior loading stability but lack anterior-posterior restraint due to plate slippage underneath the acromion, resulting in reduction loss and potential acromial fracture. Today, hook plates are infrequently used secondary to increased infections, delayed rehabilitation to prevent implant failure, and controversy regarding implant removal.

65.1.6.5 Coracoclavicular Interval Stabilization

Copious devices have been produced to restore the CC interval, including CC (Bosworth) screws, suture loops, cortical buttons, suture anchors, and grafts (synthetic or natural). Various fixation techniques exist through bone tunnels or loops around the coracoid.

Bosworth first introduced CC screws in 1941, which provided rigid fixation across the CC interval. CC screws offer vertical and horizontal stability in addition to improved strength compared to native CC ligaments, hook plate fixation, and CC loop fixation using Mersilene tape. However, their application has grown out of favor as a result of injury to nearby neurovascular structures, screw breakage, and prevalence of secondary procedures for screw removal.

Further constructs have attempted to stabilize the CC interspace using sutures, grafts, and other synthetic materials anchored to the coracoid through bone tunnels or looped underneath it. Generally, these devices demonstrate excellent vertical stability; however, they fail to restore the stiffness, horizontal stability, and rotational stability of the native CC ligaments. Graft slippage along the coracoid can cause failed anterior-posterior restraint, resulting in anterior clavicular subluxation and loss of joint reduction. Jari et al. identified that CC suture slings resulted in increased anterior and posterior clavicular translation compared to native CC ligaments (110% and 330%, respectively). Kippe et al. identified that various CC suture slings provided insufficient rotational torque strength.

Newer, less invasive, techniques include cortical buttons drilled through the clavicle, coracoid, or both. These devices utilize suture, tape, or

natural grafts to stabilize the CC interval. Wellmann et al. demonstrated that subcoracoid flip buttons provide strength similar to the native CC ligaments and avoid excessive subcoracoid dissection. Early clinical success rates of nearly 90% have been reported for these devices.

65.1.6.6 Anatomic Ligament Reconstruction

Anatomic coracoclavicular ligament reconstruction (ACCR) utilizes auto- or allograft tendon to replicate the CC ligaments at their anatomic location. Multiple studies have demonstrated superior load-to-failure characteristics of this reconstruction technique compared to the modified W-D procedure, anatomic sutures, and graft-rope techniques. Furthermore, peak loads equivalent to the native CC ligaments can be reproduced, which help restore the biomechanical properties of the native ligaments. To improve horizontal and rotational stability, the remaining tendon can be used to augment, or reconstruct, the ACLC. Multiple studies have reported high patient satisfaction and favorable clinical outcomes, making the ACCR technique highly favorable. The authors' preferred technique is the ACCR.

65.2 Sternoclavicular Joint Injuries

65.2.1 Anatomy

The SC joint is formed at the junction between the medial clavicle and the saddle-like aspect of the sternum (Fig. 65.2). Between the articular surfaces is a fibrocartilaginous disc that provides shock absorption. The SC joint is limited in motion due to four major ligaments. The costoclavicular ligament provides stability to the medial clavicle and first rib. The SC capsular ligament is vital to prevent superior displacement of the medial clavicle. The interclavicular ligament is the greatest stabilizer for rotational motion. Lastly, the intra-articular disc ligament arises from the junction of the first rib and the sternum, passing through the SC joint and attaching to the superior/posterior aspects of the medial clavicle.

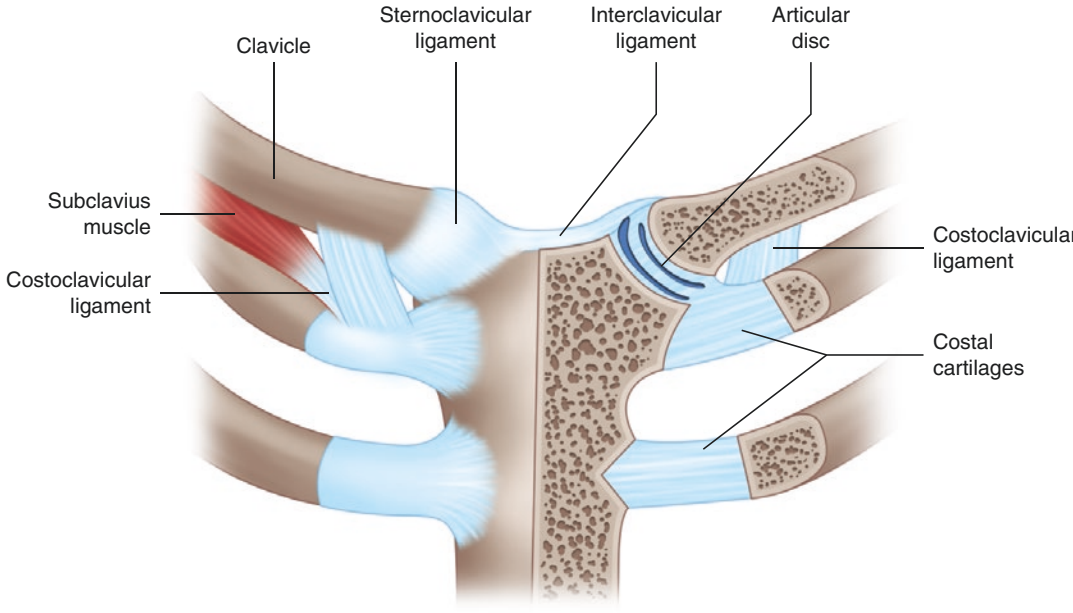

Fig. 65.2 Anatomy of the sternoclavicular (SC) joint. The bony anatomy depicts the clavicle, first and second ribs, and manubrium. Joint stability is maintained by the costoclavicular ligament, sternoclavicular ligament, interclavicular ligament, and articular disc. This image was originally published by O'Sullivan MB, Yang J, Barden B, Singh H, Divenere J, Mazzocca AD. Sternoclavicular Joint Anatomy. In: Bain GI, Itoi E, Di Giacomo G, Sugaya H, eds. Normal and Pathological Anatomy of the Shoulder. New York: Springer; 2015. p 186

65.2.2 Mechanisms of Injury

SC dislocations derive from significant trauma (e.g., motor vehicle collisions). Indirect force to the antero- or posterolateral shoulder results in either anterior or posterior dislocations. Contrarily, direct force to the anteromedial clavicle results in posterior dislocations.

65.2.3 Physical Examination

Importantly, SC dislocations can yield life-threatening, ipsilateral swelling, which may compress the trachea, esophagus, great vessels, or brachial plexus. Always assess for dyspnea, dysphagia, cyanosis, and paresthesia.

Inspection is likely to reveal gross deformity, especially with anterior SC dislocations. Patients often tilt their head towards the affected side to reduce joint stress. Direct palpation causes significant joint tenderness and typically reveals a step-off deformity. ROM is frequently limited secondary to substantial pain.

Thorough examination can help differentiate the different types of SC joint injuries. Pain with motion and moderate joint swelling suggest a type I injury (mild sprain). Palpable dislocation on manual stress testing suggests a type II injury (partial ligament disruption and subluxation). Severe pain, an arm supported in adduction, and the head tilted towards the affected side suggest type III injury (complete ligament disruption and dislocation).

65.2.4 Imaging

CT scans are the gold standard to evaluate SC joint injuries due to their speed, availability, and utility to detect dislocations, fractures, and soft-tissue injuries. The overlap of the bony anatomy surrounding the SC joint (e.g., medial clavicle, sternum, ribs, and vertebrae) makes routine radiographs difficult to interpret. The serendipity view (AP view angled 40° cephalad) is most useful for isolated SC joint injuries. MRI offers assessment of the trachea, esophagus, and vasculature; however, its main utility is delineating physeal injuries in children.

65.2.5 Management

Management of SC joint injuries depends on the severity and direction of the dislocation. Most injuries are managed nonoperatively with successful outcomes. Surgical reconstruction is reserved for recurrent dislocations or those that fail to reduce nonoperatively.

65.2.5.1 Nonoperative Management

Nearly all type I and II injuries are managed nonoperatively with ice, analgesics, and sling immobilization for 4–6 weeks. Type II injuries (moderate sprain or subluxation) may benefit from a clavicle strap or figure-of-eight bandage for 1 week prior to sling immobilization. If the SC joint is subluxated, reduction can be performed by applying a posteromedial force to the shoulders.

If the SC joint is dislocated (type III), reduction (closed or open) must be performed; however, the technique varies depending on the direction of dislocation. For anterior dislocations, closed reduction using sedation or anesthesia (local or general) is preferred. A 3–4 in. pad is placed between the scapulae of a supine patient, to retract the shoulders, and a posterior force applied to the anteromedial clavicle. A clavicle strap, sling, or figure-of-eight bandage is used to immobilize the arm for 6 weeks. Most anterior SC dislocations are unstable after closed reduction, but open reduction is discouraged due to potentially severe complications.

For posterior dislocations, closed reduction is the preferred treatment and yields low recurrence rates. It is recommended to have a thoracic surgeon available during reduction, as mediastinal involvement can result in life-threatening injury. With the patient supine, a 3–4 in. block is placed between the scapulae, and lateral traction is applied to the abducted arm as it is brought into extension. Grasping the medial clavicle with the fingers or a towel clip can aid with manipulation. A clavicle strap, sling, or figure-of-eight bandage is used to immobilize the arm for 6 weeks.

65.2.5.2 Operative Management

Open reduction is reserved for unsuccessful closed reduction and requires the presence of a thoracic surgeon if life-threatening complications arise.

Open reduction is achieved by exposing the SC joint, applying traction, and pulling the medial clavicle anteriorly. During exposure, it is critical to preserve as much of the anterior SC capsule as possible to enhance postreduction joint stability. The two methods of fixation are the medial clavicle excision and figure-of-eight reconstruction.

Medial clavicle excision is performed in conjunction with intramedullary ligament transfer. After medial clavicle excision, the medullary canal is curetted and two superior holes are drilled (1 cm lateral to the resection). The remnants of the intra-articular disc and capsular ligaments are then transferred through the medullary canal and secured to the clavicle.

Figure-of-eight reconstructions involve drilling two holes in the medial clavicle and manubrium. A semitendinosus graft is woven through the holes in a figure-of-eight and sutured into position. Biomechanical studies have shown that this technique provides greater stability than intramedullary ligament transfer and restores the native joint stability. The authors' preferred technique is the figure-of-eight reconstruction.

Take-Home Message

- The most common cause of injury to the acromioclavicular (AC) and sternoclavicular (SC) joints is trauma.
- AC joint injuries are more common than SC joint injuries, but SC joint injuries have the potential to be more severe and life-threatening.
- AC joint injuries are classified using the Rockwood and ISAKOS classification systems.
- Low-grade AC joint injuries are managed nonoperatively with brief immobilization and physical therapy. High-grade AC joint injuries are managed operatively.
- Operative management of AC joint injuries falls into four categories: ligament and tendon transfers, AC joint stabilization, CC interspace stabilization, or anatomic ligament reconstruction.
- SC joint injuries are often successfully managed nonoperatively.

Summary

The most common cause of injury to the acromioclavicular (AC) and sternoclavicular (SC) joints is trauma. AC joint injuries are more common than SC joint injuries, but SC joint injuries have the potential to be more severe and life-threatening. AC joint injuries are diagnosed using physical examination (pain at the AC joint on cross-arm adduction) and radiographs (AP, bilateral Zanca, and axillary views). SC joint injuries are diagnosed using physical examination (gross deformity) and CT scans. Posterior SC dislocations can cause ipsilateral swelling that compresses the trachea, esophagus, great vessels, or brachial plexus. Always assess for dyspnea, dysphagia, cyanosis, and paresthesia. AC joint injuries are classified using the Rockwood (types I–VI) and ISAKOS (types IIIA and IIIB) classification systems. Low-grade AC joint injuries (types I and II) are managed nonoperatively with brief immobilization and physical therapy. High-grade AC joint injuries (types IV–VI) are managed operatively. Type III injuries are typically managed nonoperatively, reserving surgery for those who fail to improve with conservative measures. Operative management of AC joint injuries falls into four categories: ligament and tendon transfers, AC joint stabilization, CC interspace stabilization, or anatomic ligament reconstruction. SC joint injuries are often successfully managed nonoperatively. Joint reduction (closed or open) is required for type III injuries (SC joint dislocation). Open reduction is reserved for unsuccessful closed reduction. Open reduction of the SC joint can be performed using medial clavicle excision or figure-of-eight reconstruction.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Which set of radiographic views are the most useful to diagnose and classify suspected AC joint injuries?
   (a) AP, scapular Y, bilateral Zanca
   (b) AP, scapular Y, Velpeau
   (c) AP, scapular Y, West point
   (d) AP, axillary, bilateral Zanca

2. How is the bilateral Zanca radiograph obtained to evaluate acromioclavicular joint injuries?
   (a) 10–15° cephalad tilt in the AP orientation with increased penetrance
   (b) 10–15° cephalad tilt in the AP orientation with decreased penetrance
   (c) 10–15° caudal tilt in the AP orientation with increased penetrance
   (d) 10–15° caudal tilt in the AP orientation with decreased penetrance
3. A 45-year-old male accountant fell off his bike and injured his nondominant shoulder while mountain biking over the weekend. The bilateral Zanca radiograph shows 25% increase in the coracoclavicular interspace. The axillary radiograph shows no anterior-posterior translation of the clavicle. What is the next step in management?
   (a) Sling for comfort and early physical therapy
   (b) Coracoclavicular screw
   (c) Weaver-Dunn reconstruction
   (d) Anatomic coracoclavicular ligament reconstruction
4. A 20-year-old male football player sustained a type V acromioclavicular joint injury after making a tackle. Which of the following ligaments has been disrupted?
   (a) Acromioclavicular
   (b) Coracoclavicular
   (c) Coracoacromial
   (d) Acromioclavicular and coracoclavicular
5. An 18-year-old male presents to the emergency department after a motor vehicle accident. Examination reveals unilateral jugular vein engorgement. Radiographic imaging reveals posterior dislocation of the sternoclavicular joint. What is the next step in management?
   (a) Closed reduction in the emergency department with local anesthetic
   (b) Closed reduction in the emergency department with sedation
   (c) Closed reduction in the operating room under general anesthesia with thoracic surgery on standby
   (d) Open reduction in the operating room under general anesthesia with thoracic surgery present

Further Reading

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Glenohumeral Instability

66

Patrick Garvin, Daniel P. Berthold, Colin Uyeki, and Augustus D. Mazzocca

Overview

Clinical cases of instability are classified by the degree of instability, the direction of instability, and the circumstances by which the instability event occurs. Instability can be a congenital issue that stems from dysplastic bony anatomy or systemic pathology.

66.1 Classification of Instability

Clinical cases of instability are classified by the degree of instability, the direction of instability, and the circumstances by which the instability event occurs. Instability can be a congenital issue that stems from dysplastic bony anatomy or systemic pathology. Collagen disorders such as Ehlers-Danlos syndrome can lead to instability due to ligamentous patholaxity. More commonly, instability events result from a traumatic injury to the upper extremity. Damage to the glenoid, humerus, labrum, rotator cuff muscle/tendon, or other ligamentous injury can destabilize the glenohumeral joint and lead to instability. In the absence of trauma, any compromise of the stabilizing mechanisms of the glenohumeral joint can also lead to instability. Repeated dislocation or subluxation of the glenohumeral joint is termed recurrent instability. Labral lesions are reported in Fig. 66.1.

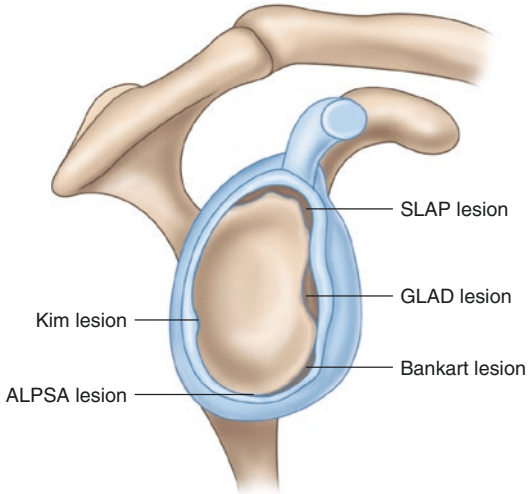

Fig. 66.1 Labral lesions

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https://doi.org/10.1007/978-3-031-20987-1_66

66.2 Traumatic vs. Atraumatic

Traditionally, recurrent instability has been classified into two distinct groups: TUBS and AMBRII. ***T****raumatic* instability events typically occur in a ***u****nidirectional* fashion and can have associated glenoid bone loss, known as a ***B****ankart* lesion. These injuries are commonly treated ***s****urgically*, and thus the TUBS acronym is a convenient way to categorize these patients. Conversely, ***a****traumatic* instability events typically occur in ***m****ultidirectional* fashion. These instability events commonly occur ***b****ilaterally*. ***R****ehabilitation* is the mainstay of treatment for these atraumatic episodes of instability. When conservative management fails, surgical intervention consisting of an ***i****nferior* capsular repair/imbrication can address patholaxity in the shoulder capsule to prevent instability. It is also important to consider the direction of instability, as determining the presence of anterior, posterior, or multidirectional instability has important implications when considering the associated injured structures that may require surgical repair.

66.3 Severity of Instability

The TUBS and AMBRII acronyms are helpful tools for classifying instability; however, it is important to classify instability events based on the degree of instability. Glenohumeral dislocation is defined as complete separation of the articular surfaces of the proximal humerus and glenoid. This may spontaneously relocate or may require manual reduction. A glenohumeral subluxation is defined as symptomatic translation of the humeral head on the glenoid without complete separation of the articular surfaces. Subluxations most commonly spontaneously return to a normal anatomical position within the glenohumeral joint.

66.4 Anatomy of the Shoulder

The stability of the glenohumeral joint is conferred by various static and dynamic constraints. Osseous anatomy consists of the articular surface of the proximal humerus, glenoid, coracoid process, acromion, and distal clavicle. The glenoid concavity permits stable articulation with the proximal humerus. This concavity is deepened by articular cartilage, which thickens at the periphery of the glenoid. A fibrocartilaginous labrum around the rim of the glenoid deepens this concavity. This concavity allows for compression of the humeral head within the glenoid and increases the surface area for glenohumeral contact. The labrum also provides attachments superiorly for the biceps tendon and inferiorly for the glenohumeral ligaments.

The capsuloligamentous structures contribute to stability based upon their insertion sites and the anatomic position of the shoulder. With the arm abducted to 90° and externally rotated, the anterior band of the inferior glenohumeral ligament is the primary constraint to anterior subluxation/dislocation. The middle glenohumeral ligament provides resistance to anterior/posterior/inferior translation with the humerus abducted at approximately 45°. The superior glenohumeral ligament limits inferior as well as anterior/posterior translation with the arm adducted. The rotator cuff muscles consist of the supraspinatus, infraspinatus, teres minor, and subscapularis and function to stabilize the humeral head into the glenoid socket. This applies a compressive load to the glenohumeral joint throughout the range of motion. An intact rotator cuff is of great importance for the concept of concavity compression, as tears of the rotator cuff may contribute to glenohumeral joint instability, especially in superior humeral head migration.

66.5 Acute Shoulder Dislocations: Pathogenesis, Clinical Findings, and Management

Glenohumeral dislocation occurs in approximately 2% of the general population. Most commonly, shoulder dislocations occur anteriorly. Classically, this occurs with the arm abducted, extended, and externally rotated. This combination of forces stresses the anterior and inferior capsule, glenoid, and rotator cuff musculature. Posterior dislocations are less common and can result from trauma or from forceful muscle contractions seen in convulsive seizures or electrical shocks. The strong internal rotators (latissimus dorsi, teres major, subscapularis, pectoralis major) overpower the weaker external rotators (infraspinatus, teres minor), causing the shoulder to dislocate posteriorly. Dislocations can also occur inferiorly, a condition known as luxatio erecta. While rare, this is caused most commonly by hyperabduction of the humeral shaft, which levers against the acromion superiorly and causes the humeral head to dislocate inferiorly.

The mechanism of injury, position of the arm, and direction of the force applied are important considerations. Injury with the arm held in abduction and external rotation is consistent with an anterior dislocation. Seizures, electric shock, or posterior directed force with the arm held in internal rotation favor posterior dislocation.

Physical examination should begin with inspection of the shoulder to look for atrophy, which can indicate denervation. In some cases of anterior dislocation, a sulcus sign can be observed and palpated lateral to the acromion. The humeral head may also be palpable anteriorly. Posterior dislocations can be subtler, mimicking the arm's normal position at rest. A critical component of the physical exam in an acutely dislocated shoulder is assessment of the neurovascular status of the upper extremity, both before and after reduction. Vascular and nerve injuries must be identified.

Radiographic evaluation is important to evaluate the direction of dislocation and associated fractures. X-rays consist of an AP view in the scapular plane, also known as a Grashey view, a lateral view in the plane of the scapula, and an axillary view. The axillary view is critical to evaluate the direction of humeral displacement and Hill-Sachs lesion, which can be difficult to visualize on AP radiographs (Fig. 66.2). The axillary view is obtained with the patient's arm abducted, and the X-ray beam is directed superiorly towards the axilla. If the patient cannot tolerate the abduction required to obtain an axillary view, a CT scan should be completed to ensure that the shoulder is located.

Acute shoulder dislocations should be reduced promptly to eliminate the risk of neurovascular compromise, minimize muscle spasm, and prevent progression of humeral impaction/bone loss. Reduction of an acute shoulder dislocation requires adequate analgesia and, importantly, muscle relaxation to facilitate reduction maneu-

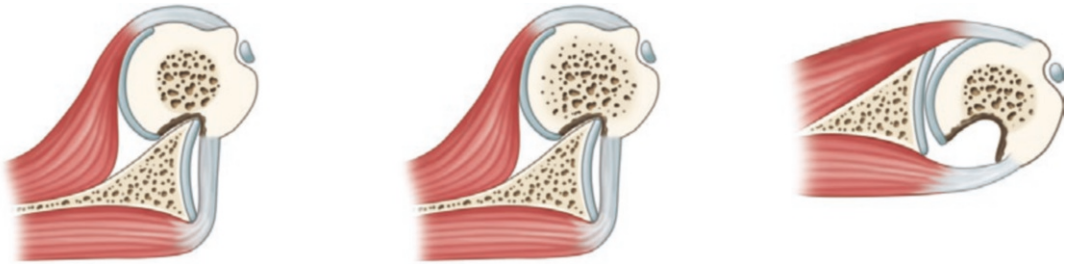


Fig. 66.2 Hill-Sachs lesion

vers. Providing traction/countertraction with gentle internal/external rotation of the shoulder is a reliable method.

66.6 Associated Bone, Soft-Tissue, and Neurovascular Injuries

An important feature of anterior dislocations is avulsion of the anteroinferior glenohumeral ligaments and labrum from the glenoid, known as a Bankart lesion. This can also involve fracture of the glenoid rim, known as a bony Bankart lesion. In anterior dislocations, the posterior aspect of the humeral head impacts the glenoid rim, leading to a compression fracture known as a Hill-Sachs lesion. In posterior dislocations, the anteromedial aspect of the humeral head can be fractured due to impaction on the posterior glenoid, or a "reverse Hill-Sachs" lesion. This has important implications for treatment, as large bony lesions of the humeral head can engage the glenoid with range of motion, a so-called off-track lesion. Evaluation of these associated bone defects is critical when evaluating surgical options.

Rotator cuff tears can also accompany traumatic shoulder dislocations, particularly in older patients. In patients over 40 years of age who suffer a shoulder dislocation, a thorough physical exam along with MRI imaging must be performed to rule out associated rotator cuff pathology.

The brachial plexus and axillary artery pass anteriorly, inferiorly, and medially to glenohumeral joint. As a result, it is critical to perform a thorough neurovascular exam during the initial evaluation. Assessment of distal pulses and perfusion is vital to detect vascular injury. A detailed neurological exam should test motor and sensory function of the upper extremity. The axillary nerve is the most commonly involved nerve as it lies in close proximity to the glenoid. Nerve injury can present as acute weakness or numbness after a dislocation, or present later with delayed recovery of shoulder motion due to persistent weakness in the deltoid.

66.7 Chronic or Recurrent Shoulder Dislocations: Pathogenesis, Clinical Findings, and Management

Historically, age is the most effective predictor of recurrence after a glenohumeral dislocation. Those involved with contact or overhead throwing sports are also more likely to have recurrent instability. Early surgical reconstruction for primary dislocation has been shown to decrease recurrence rates and improve function.

66.8 History and Physical Examination

Both age and activity level are important for clinical decision-making. It is also critical to obtain a thorough history of all previous instability events and prior treatments. In a patient with a history of instability, the following questions must be answered. Was a manual reduction required after the initial instability event? What position was the arm in? How many recurrent episodes of instability have occurred since the initial event? Were these dislocations or subluxations? What was the magnitude of trauma that caused the event? Have the recurrent episodes occurred with similar force, or have they occurred with less trauma? The answers to these questions grant significant insight into the direction and severity of instability. Evidence of worsening instability with a decreasing degree of trauma can indicate progressive bone loss.

Physical examination of the shoulder should begin with inspection of the shoulder musculature for any obvious atrophy. Particular attention should be paid to the deltoid, as atrophy, weakness, or diminished sensation can indicate an axillary nerve lesion. Range of motion of the shoulder should be examined both actively and passively, compared to the contralateral shoulder. Strength testing is performed to evaluate for focal weakness or pain and may indicate nerve injury or associated rotator cuff injury. Generalized ligamentous laxity should be evaluated by testing

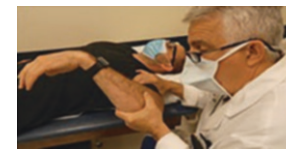
Test	Maneuver	Positive findings:	Demonstration:
Apprehension	• Shoulder held in 90 degrees of abduction and external rotation. shoulder stabilized at the elbow with one hand while the other externally rotates	• Patient develops apprehension or reproduces patients feelings of instability	
Relocation	• Examiner applies posterior force to humeral head	• Patient sense of apprehension or feeling of instability is relieved	
Load and Shift	• Patient lies supine with arm abducted and examiner stabilizes at the elbow and "loads" humerus anteriorly or posteriorly	• Humeral head translation/subluxation • Indicates anterior/posterior instability	
Jerk	• Shoulder is flexed to 90 degrees and internally rotated, then a compressive force is directed posteriorly through the elbow	• Humeral head translates/subluxes posteriorly over glenoid rim	

Fig. 66.3 Summary of various physical exam maneuvers useful in establishing diagnosis of instability

the hyperextension of the patient's thumb and metacarpal phalangeal joints and assessing for recurvatum of the elbows. The presence of symmetric, bilateral hyperlaxity suggests multidirectional instability. These patients may benefit from rehabilitation and physical therapy as opposed to surgical stabilization. It is also important to consider the presence of cervical pathology in any patient that presents with shoulder pain or weakness. A full shoulder evaluation should include a cervical spine examination to rule out cervical radiculopathy.

66.9 Special Tests

Special tests are important to evaluate for glenohumeral laxity. Provocative stability tests use maneuvers to reproduce instability symptoms and isolate pathology. These are highlighted in Fig. 66.3.

66.10 Diagnostic Imaging Studies

66.10.1 Radiographs

In addition to a complete shoulder series, additional views such as the West Point axillary and Stryker notch can be obtained. A West Point axillary view is obtained with the patient positioned prone and arm abducted. The X-ray beam is angled inferomedially to image the anteroinferior glenoid. This is useful for assessing Bankart lesions and glenoid rim bone loss in addition to the standard axillary view. Figure 66.4 demonstrates West Point axillary radiograph with avulsion of the anteroinferior glenoid.

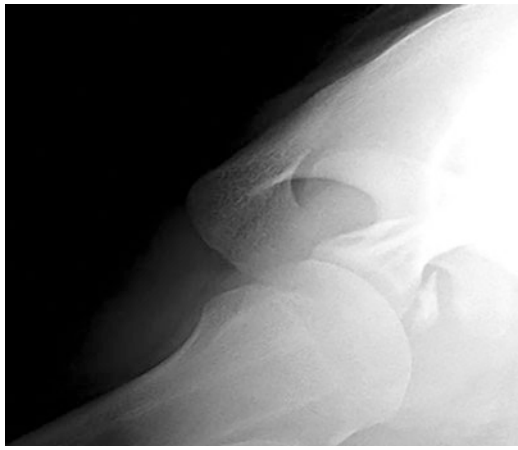


Fig. 66.4 West Point axillary radiograph showing avulsion of anteroinferior glenoid

66.10.2 Computed Tomography (CT)

CT scans can help quantify the magnitude of glenoid or humeral bone loss that can be difficult to assess on plain radiographs and is an important advanced imaging tool for surgical planning.

66.10.3 Magnetic Resonance Imaging (MRI)

MRI studies are useful to evaluate labral tears, rotator cuff tears, or other soft-tissue lesions. In particular, MRI can detect labral tears associated with instability (Bankart lesions) as well as avulsion of the glenohumeral ligament/capsule from the humerus (HAGL lesion).

66.10.4 Nonoperative Management

The majority of patients with recurrent instability will require surgery for stabilization. Patients with significant comorbidities, voluntary dislocators, or patients noncompliant with treatment/physical therapy may be appropriate for non-op management. Conservative management is also the mainstay of treatment for those with generalized hyperlaxity and multidirectional instability. Nonoperative management typically consists of a brief period of immobilization, followed by shoulder-strengthening exercises. Strengthening of periscapular musculature augments the humeral compression mechanism and stabilizes the humeral head in the glenoid concavity. Patients also undergo proprioceptive training to use the shoulder only in positions of stability and avoid positions of subluxation or apprehension.

66.11 Surgical Management of Recurrent Anterior Shoulder Instability

Surgical options for anterior instability consist of arthroscopic and open techniques. Surgical stabilization is indicated after a primary anterior shoulder dislocation in a young, high-demand patient, collision, or overhead athletes (especially throwing), and as patients with recurrent instability without bone loss. There has been a recent trend towards arthroscopic stabilization as opposed to open Bankart repair; however, both are effective.

Arthroscopic Bankart repair consists of suture anchors placed at the inferior aspect of the glenoid to anatomically repair the labrum. A capsular shift can also be performed for tensioning of the inferior glenohumeral ligament. Arthroscopic stabilization is effective when in the right scenario; however, preoperative investigation of glenoid and humeral bone loss is critical.

66.12 Treatment Considerations

66.12.1 Glenoid Bone Loss

Identifying glenoid bone loss is critical and can be addressed with various techniques, including the Bristow and Latarjet procedures, iliac crest bone grafting, and osteochondral distal tibial allograft grafting. These various techniques utilize bone block fixation to the glenoid and are effective at preventing recurrent instability. The Bristow procedure involves transfer of the tip of the coracoid process to the anterior glenoid; however, this was technically demanding and

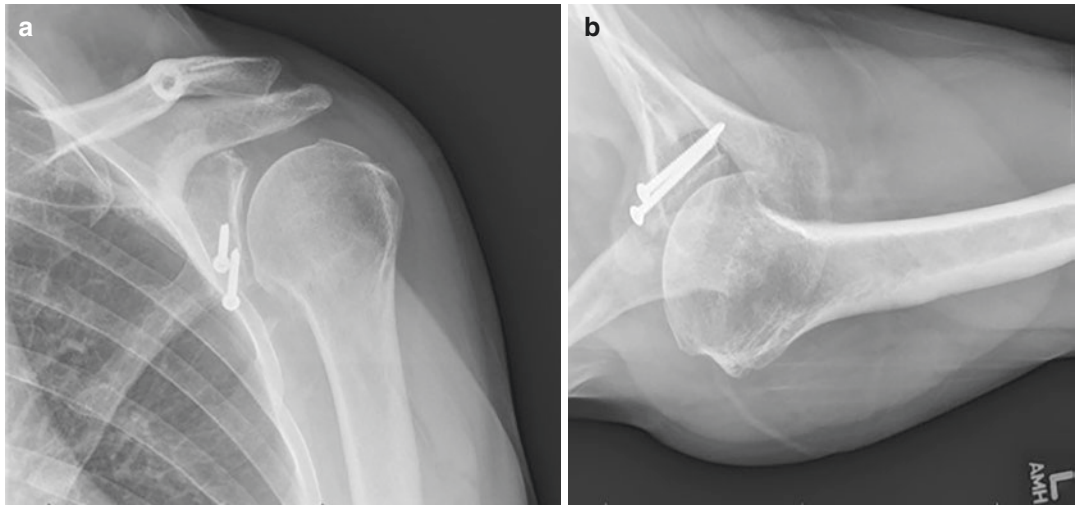


Fig. 66.5 Postoperative AP (**a**) and axillary (**b**) radiographs after Latarjet procedure

recurrence was problematic. More commonly, the Latarjet procedure is utilized, which can be performed as an open technique or arthroscopically assisted. In this procedure, a large portion of the coracoid process is harvested along with attached conjoint tendon of the biceps and coracobrachialis tendons, as well as the coracoacromial ligament. This is transferred to the anterior glenoid with screw fixation. An example of final construct after Latarjet can be seen in Fig. 66.5. This confers more stability than the Bristow procedure, in part due to a larger volume of bone. More importantly, the conjoint tendon and CA ligament provide a sling effect, adding an additional restraint to humeral subluxation. Great care is taken to avoid damaging the musculocutaneous nerve, which lies inferior to the conjoint tendon insertion. Nerve palsy, nonunion, hardware failure, and recurrent instability are some associated complications.

66.12.2 Humeral Head Lesions

Humeral head defects are seen in cases of anterior instability (Hill-Sachs lesion) as well as posterior instability (reverse Hill-Sachs lesion). In the presence of a large, "off-track" Hill-Sachs lesion, soft-tissue repair is likely to fail if bone loss is not corrected. Correction of the glenoid bone loss alone is sometimes sufficient to prevent Hill-Sachs engagement, but even small combined glenoid and Hill-Sachs lesions can lead to persistent instability. An emerging technique to address the Hill-Sachs defect is the arthroscopic remplissage. This involves placement of suture anchors into the Hill-Sachs defect and passage of the suture limbs through the posterior capsule and infraspinatus tendon. This may be performed in combination with arthroscopic Bankart repair and/or Latarjet to prevent further engagement of the Hill-Sachs defect on the glenoid. In the setting of engaging Hill-Sachs lesions that are too large to be addressed with remplissage, bone grafting techniques, arthroplasty, or rotational osteotomies can be performed.

66.13 Posterior Instability and Associated Lesions

While less common than recurrent anterior instability, posterior instability can result from posterior dislocations, labral tears, or repetitive trauma. This is classically seen in weight lifters and football lineman, as a result of strenuous pushing activities. In other rarer circumstances, glenoid retroversion can lead to posterior instability. Nonoperative and surgical treatment of posterior instability is conceptually similar to that of ante-

rior instability. Surgery involves open or arthroscopic posterior capsulolabral repair. Bone loss must be addressed, as failure to address an off-track reverse Hill-Sachs defect will lead to failure of soft-tissue repair. There are various options to address a large reverse Hill-Sachs lesion, including subscapularis tenodesis, bone grafting techniques, rotational osteotomy, and arthroplasty. One technique, known as the modified arthroscopic McLaughlin procedure, utilizes a subscapularis tenodesis. Suture anchors are placed into the reverse Hill-Sachs lesion, and limbs are passed in double-mattress fashion through the subscapularis tendon. Tensioning of the sutures pulls the lateral aspect of the subscapularis tendon into the defect, effectively filling the void and preventing engagement.

Take-Home Message

- Clinical cases of instability are classified by the degree of instability, the direction of instability, and the circumstances by which the instability event occurs.
- Instability can be a congenital issue that stems from dysplastic bony anatomy or systemic pathology.
- Traditionally, recurrent instability has been classified into two distinct groups: TUBS and AMBRII.
- Glenohumeral dislocation is defined as complete separation of the articular surfaces of the proximal humerus and glenoid.
- Rotator cuff tears can also accompany traumatic shoulder dislocations, particularly in older patients.
- Identifying glenoid bone loss is critical and can be addressed with various techniques, including the Bristow and Latarjet procedures, iliac crest bone grafting, and osteochondral distal tibial allograft grafting.

Summary

Clinical cases of instability are classified by the degree of instability, the direction of instability, and the circumstances by which the instability event occurs. Instability can be a congenital issue that stems from dysplastic bony anatomy or systemic pathology. Damage to the glenoid, humerus, labrum, rotator cuff muscle/tendon, or other ligamentous injury can destabilize the glenohumeral joint and lead to instability. Repeated dislocation or subluxation of the glenohumeral joint is termed recurrent instability. Traditionally, recurrent instability has been classified into two distinct groups: TUBS and AMBRII. Glenohumeral dislocation is defined as complete separation of the articular surfaces of the proximal humerus and glenoid. The stability of the glenohumeral joint is conferred by various static and dynamic constraints. Glenohumeral dislocation occurs in approximately 2% of the general population. An important feature of anterior dislocations is avulsion of the anteroinferior glenohumeral ligaments and labrum from the glenoid, known as a Bankart lesion. Rotator cuff tears can also accompany traumatic shoulder dislocations, particularly in older patients. Age is the most effective predictor of recurrence after a glenohumeral dislocation. The majority of patients with recurrent instability will require surgery for stabilization. Surgical options for anterior instability consist of arthroscopic and open techniques. Identifying glenoid bone loss is critical and can be addressed with various techniques, including the Bristow and Latarjet procedures, iliac crest bone grafting, and osteochondral distal tibial allograft grafting. Humeral head defects are seen in cases of anterior instability (Hill-Sachs lesion) as well as posterior instability (reverse Hill-Sachs lesion). While less common than recurrent anterior instability, posterior instability can result from posterior dislocations, labral tears, or repetitive trauma.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Which of these ligaments are injured in grade III dislocations of the acromioclavicular joint?
   (a) Acromioclavicular ligaments
   (b) Conoid ligament
   (c) Trapezoid ligament
   (d) A + B + C
2. The "piano key" deformity is typical of:
   (a) Colles fractures
   (b) Acromioclavicular dislocations
   (c) Glenohumeral dislocations
   (d) Fractures of the surgical neck of the humerus
3. Bankart lesion is characteristic of:
   (a) Full-thickness rupture of the rotator cuff
   (b) Fracture of the acromial end of the clavicle
   (c) Acromioclavicular dislocation
   (d) Recurrent glenohumeral dislocation
4. Which method is most useful to identify a Bankart lesion?
   (a) Computed tomography
   (b) Radiography in anteroposterior projection
   (c) Ultrasound
   (d) Magnetic resonance imaging
5. The Hill-Sachs lesion in shoulder dislocations involves:
   (a) The humeral head
   (b) The glenoid rim
   (c) The neck of the scapula
   (d) The surgical neck of the humerus

Further Reading

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The Rotator Cuff

67

Joseph P. DeAngelis

Overview
Rotator cuff disease represents a spectrum of pathology, ranging from tendinitis to a massive, irreparable rotator cuff tear. As a result, shoulder pain afflicts patients of all ages and can result from both overuse in a throwing athlete and inactivity associated with sedentary desk work.

67.1 Definition

The rotator cuff is the group of muscles and tendons that center the humeral head in the glenoid, providing the joint with dynamic stability throughout the mid-range of motion and activities of daily living. The coordinated action of the subscapularis, supraspinatus, infraspinatus, and teres minor establishes the fulcrum for arm motions away from the body and overhead, like lifting, reaching, pushing, pulling, carrying, and throwing. As a result, any injury to the rotator cuff will cause shoulder pain and dysfunction because the humeral head is not centered when the arm is moved away from the side of the body.

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67.2 Epidemiology

Rotator cuff pathology is the most common condition of the shoulder for which patients seek treatment. The prevalence of shoulder pain ranges from 16 to 34% in the general population. In the United States, rotator cuff problems account for more than 4.5 million office visits and approximately 40,000 surgeries, annually. With increasing physical demand, the incidence and prevalence of rotator cuff pathology are expected to increase as the world's population grows and industrialization expands.

67.3 Etiology/Pathogenesis

Rotator cuff pathology occurs on a continuum, ranging from inflammation in and around the tendons of the rotator cuff to a complete rupture, or detachment, of the tendon from its insertion on humerus. The cause of this progression is multifactorial and includes mechanical factors like overuse, degeneration, impingement, and overload, as well as genetic factors linked to inheritance.

In 1934, Codman proposed that rotator cuff tears developed from intrinsic tissue degeneration. His observation was supported by the recognition of how a zone of hypovascularity in the rotator cuff predisposed the tendon to injury. Subsequent work by Neer focused on the extrin-

UG. Longo, V. Denaro (eds.), *Textbook of Musculoskeletal Disorders*,
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sic, or mechanical, factors that contributed to tendon damage. He felt that bony impingement was central to rotator cuff tearing and described the acromion's effect on the supraspinatus.

In time, further analysis revealed that shoulder motion generates dynamic patterns of sheer and strain within the tendons of the rotator cuff. Overhead athletes develop patterns of rotator cuff injury related to tension overload, a phenomenon in which the ability of the rotator cuff to compress and stabilize humeral head is overwhelmed by the distractive forces of throwing. In moving from the extreme, end-range of motion, the shoulder experiences a complex mix of compressive and distractive loading, sheer stress, and tissue strain, as the arm rapidly accelerates towards ball release and decelerates during follow-through.

These various mechanisms combine with genetic factors that may predispose an individual to further degeneration. The result is a pattern of injury in which many rotator cuff lesions begin as partial tears on the articular surface of the supraspinatus and then progress to full-thickness tear that may involve the supraspinatus, infraspinatus, subscapularis, and biceps tendons.

67.4 Risk Factors

Age, above all else, is the primary risk factor for rotator cuff tearing. The prevalence of disease has a near-linear correlation with increasing age. Additionally, exposure to overhead activity, including both sports and occupations requiring lifting and throwing, increases the frequency of rotator cuff tearing and is associated with an earlier onset of disease. Both tobacco exposure and obesity have been associated with rotator cuff disease. In an observational cohort, there was an increased prevalence of asymptomatic tears in people with diabetes, obesity, hypercholesterolemia, and hypertension, as well as those who engaged in repetitive heavy labor and smoking. Several investigations have described the heritable nature of rotator cuff pathology and its genetic basis. Specific haplotypes have been identified, further underscoring the effect of inheritance. However, the direct effect of genetics on the disease's presentation remains unclear.

Most commonly, rotator cuff tears result from acute trauma. Sudden loading of the arm may occur when people attempt to arrest themselves during a fall. The deceleration, combined with the angular moment of rotation, results in a mechanical failure at, or near, the tendon's attachment to the bone. The portion of the rotator cuff affected and the extent of the injury correlate with the mechanism of injury and magnitude of force.

67.5 Classifications

Rotator cuff tears are classified in a variety of ways. Anatomic considerations include the extent of the tendon involved (partial vs. full thickness), the number of tendons involved, the degree of retraction of the torn tendon, and the presence of atrophy and/or fatty infiltration. Different authors have proposed a myriad of options for describing the patterns, severity, and location of rotator cuff pathology (Fig. 67.1).

Dr. Cofield's classification of rotator ruff tears groups full-thickness lesions based on the size of the tear and is commonly used. He described small tears involving less than 1 cm, medium tears between 1 and 3 cm, large tears between 3 and 5 cm, and massive tears greater than 5 cm.

The Bateman classification describes full-thickness tears after surgical debridement:

Grade 1 is less than 1 cm after debridement; grade 2 is 1–3 cm after debridement; grade 3 is less than 5 cm; and grade 4 is a complete, or global, tear in which there is no cuff left.

The Ellman and Gartsman classification focuses on the shape of the torn tissue when describing a full-thickness rotator cuff tear. Type 1 is a crescent, type 2 is a reverse "L," type 3 is "L" shaped, type 4 is trapezoidal, and type 5 is a massive tear.

To characterize the extent of retraction, the Patte classification describes the location of the tear in the coronal, or frontal, plane. Stage 1, the proximal tendon stump, is close to bony insertion; stage 2, the proximal stump, is at the level of

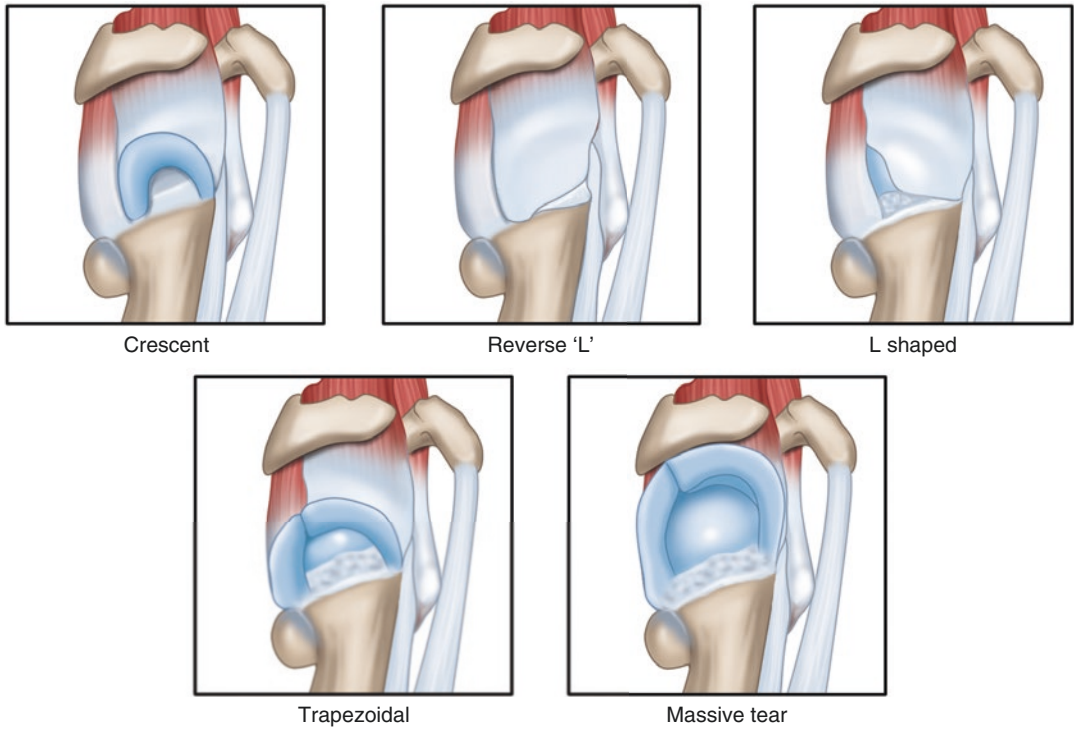

Fig. 67.1 Rotator cuff tear classification

humeral head; and stage 3, the proximal stump, is retracted to the glenoid.

When grouping partial-thickness rotator cuff tears, Ellman relied on an arthroscopic examination to assess the depth of the tear. Grade 1 is partial tear less than 3 mm deep; grade 2 is a partial tear 3–6 mm deep (not exceeding half of the tendon thickness); and grade 3 is greater than 6 mm deep.

Matsen's approach was intended to include a variety of clinical conditions that were associated with rotator cuff pathology. His system included the following: 1—asymptomatic cuff failure; 2—posterior capsular tightness; 3—subacromial abrasion without significant defect in the rotator cuff; 4—partial-thickness cuff lesion; 5—full-thickness cuff tear; 6—cuff tear arthropathy; 7—failed acromioplasty; and 8—failed cuff surgery.

To address the complicated presentation of rotator cuff disease, Snyder developed with Southern California Orthopedic Institute rotator cuff classification system to include the size, position, and quality of the tendon involved. The location of the tear was described as follows: A—articular surface; B—bursal surface; or C—complete tear. Partial-thickness tears were then grouped as 0—normal; 1—minimal superficial bursal or synovial irritation or slight capsular fraying over a small area; 2—fraying and failure of some rotator cuff fibers in addition to synovial bursal or capsular injury; 3—more severe rotator cuff injury fraying and fragmentation of tendon fibers often involving the whole of a cuff tendon (usually greater than 3 cm); and 4—very severe partial rotator cuff tear that contains a sizeable flap tear and more than one tendon.

Other authors have focused on the extent of injury in the sagittal plane. Patte described a separate classification where segment 1 is an isolated subscapularis tear (often traumatic and associated with a dislocation of the long head of the biceps); segment 2 is an isolated coracohumeral ligament tear; segment 3 is an isolated supraspinatus tear; segment 4 is a complete tear of the supraspinatus and leading one-half infraspinatus; segment 5 is a complete tear of the supraspinatus and the entire

infraspinatus; and segment 6 is a complete subscapularis, supraspinatus, and infraspinatus.

The quality of the rotator cuff tissue and the chronicity of the rotator cuff tear greatly influence the plan of care. To assess these important characteristics, Goutallier observed muscle bulk and content on the parasagittal reformats of a shoulder CT scan. Stage 1 represents normal muscle; stage 1 has some fatty streaks; stage 2 has less than 50% fatty muscle atrophy; stage 3 demonstrates 50% fatty muscle atrophy; and stage 4 finds greater than 50% fatty muscle atrophy.

67.6 Diagnosis

67.6.1 Clinical Presentation

While shoulder pain is the most frequent presenting symptom, pain is not specific and does not correlate with the presence of a rotator cuff tear. Many patients report lateral-sided arm pain at rest, with overhead activity, and at night. However, these complaints are not unique to rotator cuff injuries. The majority of rotator cuff tears are asymptomatic. While degenerative changes can be found in almost all people over the age of 50, the natural process of wear and tear does not necessarily impair function, and degenerative rotator cuff tears rarely require surgery.

Because the clinical presentation of rotator cuff disease is variable, it is difficult to correlate symptoms with tear size. The current literature contains evidence to support and refute the association of tear size/severity with symptoms. There is evidence that partial tears can be more painful than a full-thickness tear.

Because of the uncertain relationship between pain and rotator cuff disease, many clinicians rely on weakness as a sign of rotator cuff tearing. However, strength deficits can be identified in asymptomatic individuals, so weakness may or may not represent a presenting complaint. However, just as pain is not specific to rotator cuff pathology, so too weakness can confuse and confound a patient's presentation. Inflammation and trauma may result in muscle inhibition that presents with weakness.

67.6.2 Physical Examination

When examining a painful shoulder, the clinician aims to differentiate between an inflamed, painful tendon (as in outlet impingement) and an injured rotator cuff (partial- or full-thickness tear). This determination is difficult to achieve even in the most skilled hands.

Manual strength testing is believed to be the best method of assessing the rotator cuff's function and integrity, when accurately performed. A series of clinical maneuvers can improve the accuracy of the diagnosis, prior to imaging. The painful arc test, the drop arm test, and weakness in external rotation are the most effective series of tests in diagnosing for a torn rotator cuff. The classical test for the supraspinatus tendon is the Jobe test (Fig. 67.2).

67.6.3 Painful Arc Sign

When asked to actively abduct the arm, a positive painful arc occurs between 60 and 120° of abduction. Their pain should decrease above 120° of abduction.

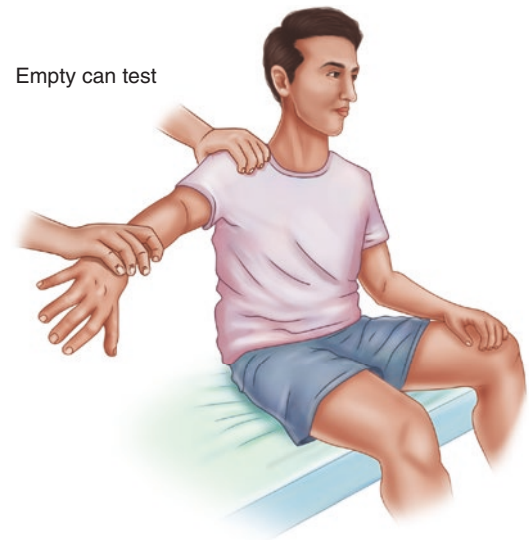

Fig. 67.2 Jobe test

67.6.4 Drop Arm Sign

A positive drop arm sign demonstrates the failure to smoothly control shoulder adduction. The examiner passively lifts the patient's shoulder to 90° of abduction. The patient is then asked to hold the arm abducted before slowly lowering it to their side. If the patient is unable to hold the arm abducted, it will drop to their sign as a manifestation of their rotator cuff dysfunction.

67.6.5 External Rotation Weakness

When tested with the elbow adducted at the patient's side, weakness in external rotation is concerning for posterior rotator cuff insufficiency. Both the infraspinatus and teres minor contribute to external rotator strength. Further interrogation of external rotation strength with the shoulder abducted to 90° intends to test the infraspinatus, while external rotation testing at 90° of abduction and 90 degrees of external rotation challenges the teres minor.

67.6.6 Subscapularis Test

Subscapularis tests are summarized in Fig. 67.3.

67.6.7 Radiographic Imaging

In patients with a clinical history and physical examination that are concerning for rotator cuff pathology, imaging of the affected shoulder can offer tremendous insight into their clinical diagnosis. Magnetic resonance imaging (MRI) and ultrasound are sensitive and specific when diagnosing rotator cuff tears, and MR arthrography demonstrates even greater sensitivity and specificity.

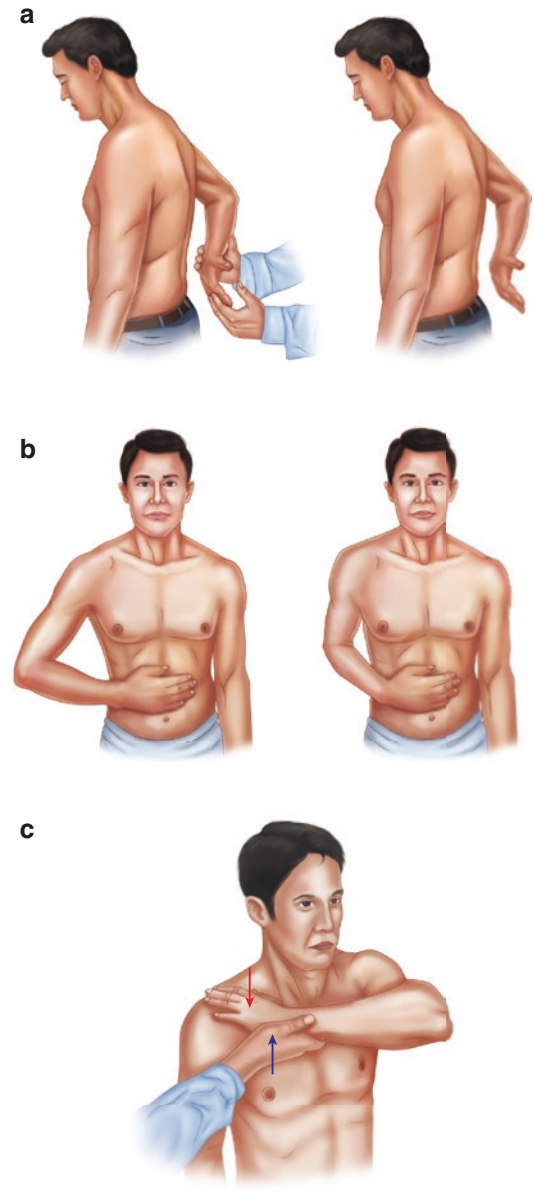

Fig. 67.3 Subscapularis tests. (**a**) Lift-Off Test (**b**) Belly Press Test (**c**) Bear Hug Test

67.7 Radiographs

Because of the cost and time associated with these modalities, plain radiographs remain the first imaging modality of choice in patients with a painful shoulder, regardless of their presenting complaints or examination findings. X-rays of the shoulder may reveal degenerative changes at the glenohumeral joint or calcific deposition in the rotator cuff. These findings help the clinician understand all of the factors that may contribute to the patient's complaint of shoulder pain.

Additionally, the acromiohumeral interval (AHI) quantifies the subacromial space as a proxy for the superior rotator cuff's function. When the supraspinatus is healthy, the humeral head remains centered in the glenoid and space beneath the acromion is maximized. When the supraspinatus is injured or its integrity is compromised, the humerus elevates and the AHI becomes smaller. When the rotator cuff is chronically deficient, the humerus articulates with the underside of the acromion and bony changes result. These end-stage changes manifest as acetabularization of the coracoacromial arch and the rounding of the humeral head, or femoralization.

67.8 Magnetic Resonance Imaging

The integrity of the rotator cuff is best assessed using a traditional MRI of the shoulder without contrast. It offers the most comprehensive view of the osseous and soft-tissue anatomy and can readily differentiate between the inflammatory changes associated with impingement and the structural abnormalities indicative of a rotator cuff tear. In addition to offering a critical view of the rotator cuff, MRI, as a three-dimensional imaging modality, highlights the shape of the acromion process, the humeral head, and the presence of degenerative changes at the acromioclavicular joint that may impact the rotator cuff's functions.

When considering the soft tissues about the shoulder, MRI will identify areas of tendon degeneration due to repeated chronic tendon injury (tendinosis), segments of the rotator cuff that have partial-thickness injuries, full-thickness tears, as well as abnormalities in the biceps tendon, the glenohumeral ligaments, and the labrum.

When the health and integrity of the rotator cuff are in question, the parasagittal reformats of the MRI offer a perspective on the muscle health and the chronicity of the injury. The presence of muscle atrophy, or a positive tangent sign, indicates a long-standing period of disuse or dysfunction and should direct clinical decision-making. Fatty infiltration, as described by Goutallier using CT, is also an important characteristic that is well seen on MRI. The presence of these degenerative changes also warrants consideration in patient management because it constitutes an irreversible change in the tissue conformity.

67.9 Magnetic Resonance Imaging with Arthrogram

An MR arthrogram of the shoulder is an enhanced version of the modality that benefits from the injection of contrast into the glenohumeral joint. Because of the process, an MR arthrogram requires an invasive procedure. However, it yields increased sensitivity and specificity because the intra-articular contrast improves image resolution as it mechanically tests the integrity of the rotator cuff. As a result, MR arthrograms are more accurate when detecting full-thickness and partial-thickness rotator cuff tears that involve the articular surface. Additionally, an MR arthrogram is more effective at diagnosing abnormalities of the biceps and labrum than a traditional MRI.

67.10 Ultrasound

In clinical settings with ultrasound readily available, this imaging modality can become the preferred initial evaluation of the rotator cuff. Ultrasonography has comparable accuracy to MRI when identifying and measuring the size of

a full-thickness or partial-thickness rotator cuff tear, if it is performed by an experienced examiner using high-quality equipment. The sensitivity and specificity for diagnosing a complete rotator cuff tear using ultrasound have been reported to be over 92%. For a partial-thickness rotator cuff tear, the same meta-analysis found the sensitivity and specificity to be 67 and 94%, respectively. Unfortunately, the diagnostic performance of a shoulder ultrasound varies with the operator, and the perspective offered by an ultrasound is limited, relative to MRI.

67.11 Computed Tomography with Arthrogram

In patients who are unable to undergo MRI, a CT arthrogram is a viable alternative if the scope of a shoulder ultrasound is too narrow. Because of the intra-articular contrast, CT arthrogram has sensitivity and specificity of greater than 90% when diagnosing supraspinatus and infraspinatus tears. Like MR, CT offers a wide field of view which includes the shoulder osseous anatomy, the extent of tendon retraction, if present, as well as a quantitative assessment of the rotator cuff muscles in cross section. This comprehensive perspective represents a considerable advantage over ultrasound for preoperative planning and surgical decision-making.

67.12 Treatment

Because the glenohumeral joint enjoys tremendous mobility, it is highly susceptible to injury. As a result, rotator cuff pathology falls along a spectrum of disease, ranging from inflammation to a complete, chronic rupture. The treatment of rotator cuff pathology, therefore, depends on an accurate diagnosis and aims to remedy the cause of the patient's pain and dysfunction. For many people with shoulder pain, conservative management is able to resolve their disability by improving the mechanical function of the healthy elements of the shoulder girdle. For other patients with structural changes to their anatomy, surgery intends to restore the normal anatomic attachments that center the humeral head in the glenoid as the shoulder moves.

67.13 Rotator Cuff Tendinitis/ Impingement

Shoulder pain is pervasive. Given the physical demands on modern, postindustrial workers, it is common for individuals to be seated at a computer or seated while driving a car for most of the workday. The result of his physical positioning is a protracted posture that predisposes the shoulder to impingement. With the head forward, the thoracic spine flexed, the shoulders rounded and internally rotated, and the scapula protracted, it is common for the muscles of anterior torso and upper body to become tight and strong, while the back and posterior shoulder are relatively weak. Recognizing this effect, the treatment of rotator cuff tendinitis that results from outlet impingement focuses on the restoration of normal scapular position by correcting this problematic posture.

Rehabilitation should decrease pain and inflammation, restore normal range of motion, improve individual muscle function/strength, restore functional capacity, and prevent reinjury. With shoulder impingement syndrome, the primary focus is improving posture by strengthening the periscapular muscles. Once corrected, the scapular mechanics allows for greater efficiency and endurance of the rotator cuff and improved overall shoulder function.

67.14 Rotator Cuff Tears

The treatment of a torn rotator cuff will depend upon the duration of symptoms, the patient's handedness, the type of tear (partial versus full thickness), and the patient's characteristics (age, comorbidities, and lifestyle).

All types of rotator cuff tears can be with and without surgery. While comparative trials have investigated both options, the benefits of surgery have not been clearly demonstrated.

67.15 Nonoperative Management

Currently, there are no universal indications for a rotator cuff repair, so most providers recommend surgery if/when conservative treatment fails. The goal of nonoperative management is to maximize shoulder function in spite of the injury while resolving the pain associated with the rotator cuff deficiency.

Rehabilitation efforts focus stretching of the shoulder to restore flexibility and motion, strengthening of the periscapular muscles and the remaining rotator cuff, and improving the shoulder's function. Patients who begin the course of physical therapy with low expectations regarding its effectiveness are more likely to undergo surgery.

Over time, most rotator cuff tears will increase in size, and significant increases in the tear's size may result in new pain or worsening weakness. With time and tear progression, there is an increased risk of muscle atrophy and fatty infiltration, so the conservative management of a rotator cuff tear may require surveillance, with or without repeat imaging.

Certain populations are most likely to benefit from serial observations—younger individuals represent the highest lifetime risk because of their age; active people who place high demand on their shoulder for professional or leisure activities; and anyone with a large, partial-thickness or incomplete rotator cuff tear.

To assist with discomfort, clinicians have employed corticosteroid injections to address the pain and inflammation associated with a rotator cuff injury, historically. Recent investigations have demonstrated that the benefit of the injection may be short-lived and the long-term effect may be significant. For this reason, the use of corticosteroids in the setting of a known rotator cuff tear should be limited to patients in whom a surgical repair is unlikely to happen.

Other interventions have been shown to be helpful at managing the symptoms associated with a rotator cuff injury. Ultrasound, electrical stimulation, iontophoresis, massage therapy, and acupuncture may provide relief, but do not change the nature or cause of the underlying problems.

67.16 Operative Management

When conservative treatment has been unsuccessful, a surgical procedure to restore the normal anatomy can address the pain associated with a rotator cuff tear while improving strength and function. However, the period of recovery following a rotator cuff repair is lengthy, so patients should be apprised of this requirement during the surgical decision-making process.

67.17 Acute, Full-Thickness Tears

In patients with an acute, full-thickness rotator cuff tear, a surgical repair aims to restore the anatomy of the musculotendinous cuff and prevent muscle atrophy, tendon degeneration, and retraction. Timely intervention may also prevent the degenerative process that culminates in irreversible fatty infiltration.

67.18 Partial-Thickness Tears

For patients with partial-thickness tears of the supraspinatus tendon, there is a risk of progression to a full-thickness tear. When indicated, partial-thickness rotator cuff tears can be managed with a primary repair or an arthroscopic debridement. Tears that involve less than 50% of the tendon's thickness can be treated with debridement. However, if more than 50% of the tendon's thickness is involved, repairing the tendon, not debridement, is likely to provide the best outcomes.

67.19 Chronic, Symptomatic Tears

After a structured course of conservative care, it is possible that a patient with a full-thickness rotator cuff tear may experience persistent weakness, pain, and/or disability. These individuals may benefit from a rotator cuff repair, depending on their age, the chronicity of the rotator cuff tear, its size, and the degree of atrophy/fatty infiltration.

67.20 Surgical Approach and Technique

A rotator cuff repair can be performed successfully with an arthroscopic or open approach. While the use of a standard delto-pectoral approach is rare when performing a rotator cuff repair, the "mini-open" technique is commonly performed with good results. However, the arthroscopic rotator cuff repair has gained popularity because it allows visualization of the entire glenohumeral joint, provides opportunity to address concomitant pathology, offers better postoperative motion, preserves deltoid function, decreases the infection rate, and reduces postoperative pain.

Regardless of the approach employed, the goal of a rotator cuff repair is to restore the normal anatomic relationship of the rotator cuff on the greater tuberosity. It can be achieved using a variety of surgical techniques and implants. A transosseous repair is performed by passing a large needle through the bone and the tendon and remains the gold standard because of its historical significance as the original method to repair a torn rotator cuff. With the advent of arthroscopic shoulder surgery, suture anchors have been developed that allow for rigid purchase in the bone using minimally invasive techniques. Once developed, suture anchors have been modified and manipulated to allow for an infinite number of configurations. In this way, a rotator cuff can be repaired using a single row of anchors, a double row of anchors, or two parallel rows of anchors to create a transosseous equivalent repair.

With all techniques, the clinical outcomes are good. Postoperatively, patient satisfaction and functional outcomes are improved. As an intervention, a rotator cuff reliably produces good results, even though the radiographic rates of healing are varied.

Over time, with any surgical technique, a re-rupture of the rotator cuff can occur. Current evidence suggests that the risk of a re-tear is related to the size of the tear, its chronicity, the age of the patient, the quality of the repair, the extent of the muscle atrophy and fatty infiltration, and nicotine use.

Take-Home Message

- Rotator cuff disease represents a spectrum of pathology, ranging from tendinitis to a massive, irreparable rotator cuff tear.
- When disease progression exceeds the body's ability to accommodate change, surgery may provide an avenue for decreased pain and improved function.
- In the setting of an acute, traumatic rotator cuff tear, restoration of the injured anatomy aims to normalize shoulder function by repairing the ruptured tendons to their normal insertion.
- When the severity or chronicity of the disease does not allow for a primary repair, other surgical approaches may be employed to minimize the patient's disability and improve their quality of life.

Summary

Rotator cuff pathology occurs on a spectrum, ranging from inflammation to massive, irreparable tears. In patients with rotator cuff disease, pain and weakness occur when the shoulder is used away from the body and overhead. The cause of this pathology is varied and can affect people in all walks of life and at any age. An accurate diagnosis is essential to the treatment and resolution of shoulder pain. A careful history and physical examination will guide the process. Radiographs offer clarity on the health and integrity of the shoulder osseous anatomy. When appropriate, MRI can be used to assess the health, integrity, and severity of any rotator cuff problem.

Most diagnoses can be improved with rehabilitation. Physical therapy and supportive modalities will maximize the function, strength, and alignment of the shoulder girdle. If this approach does not resolve the issues, surgery can help to restore torn or injured tissue to its normal alignment.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. What is the rotator cuff?
   (a) It is the group of muscles and tendons that center the humeral head in the glenoid.
   (b) It consists of a group of five muscles that center the humeral head in the glenoid.
   (c) It consists of two tendons that stabilize the humeral head.
   (d) It consists of a group of two muscles that stabilize the humeral head.
2. What is the function of the rotator cuff?
   (a) It provides dynamic stability throughout the mid-range of motion and activities of daily living. The coordinated action of the subscapularis, supraspinatus, infraspinatus, and teres minor establishes the fulcrum for arm motion.
   (b) It provides dynamic stability throughout the mid-range of motion and activities of daily living. The coordinated action of the infraspinatus and teres minor establishes the fulcrum for arm motion.
   (c) It provides dynamic stability throughout the mid-range of motion and activities of daily living. The coordinated action of the subscapularis and teres minor establishes the fulcrum for arm motion.
   (d) It provides static stability throughout the mid-range of motion and activities of daily living. The coordinated action of the subscapularis, supraspinatus, infraspinatus, and teres minor establishes the fulcrum for arm motion.
3. What is the most significant risk factor for rotator cuff tearing?
   (a) Age
   (b) BMI
   (c) Type of lesion
   (d) Sex
4. Name the most significant physical examination finding for a rotator cuff tear.
   (a) The painful arc test, the drop arm test, and weakness in external rotation
   (b) Sulcus signs and apprehension test
   (c) Sulcus sign and Napoleon test
   (d) Neer test and McMurray test
5. Which arthroscopic surgical technique for repairing a torn rotator cuff offers the best results?
   (a) b + c + d
   (b) Double-row repair
   (c) Single-row repair
   (d) Transosseous repair

Further Reading

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The Long Head of the Biceps Tendon

68

Lukas N. Muench, Daniel P. Berthold, and Knut Beitzel

Overview

Disorders of the long head of the biceps tendon (LHBT) include tendonitis, partial tearing, complete rupture, and lesions of the pulley reflection system along with subluxation and dislocation. Although being well documented, the underlying pathology is yet to be fully understood, which is probably due to the unique anatomic features of the LHBT.

68.1 Definition

The long head of the biceps tendon (LHBT) is of particular interest due to its intra-articular origin on the supraglenoid tubercle of the glenohumeral joint, with most of the fibers arising from the posterior aspect of the superior labrum and having an overall length of approximately 9 cm with a width of 5–6 mm. The intra-articular portion of the tendon is extrasynovial and has an average length of 34.5 ± 4.2 mm. Prior to entering the intertubercular groove, the tendon is guided by the structures of the pulley reflection system, comprising the superior glenohumeral ligament (SGHL) and the subscapularis tendon anteriorly as well as the coracohumeral ligament (CHL) and the anterior fibers of the supraspinatus tendon posterolaterally.

68.2 Epidemiology

The incidence of pathologies of the pulley reflection system during shoulder arthroscopy has been reported to be higher than 7% of cases and may be a result of acute trauma, repetitive microtrauma, or a degenerative process. Pulley lesions are frequently observed in patients with anterior shoulder pain and have been shown to be associated with rotator cuff tears, superior labrum anterior-posterior (SLAP) lesions, biceps instability, biceps tears, and internal anterosuperior impingement. Partial tearing (51%), instability (49%), and tenosynovitis (44%) have been found to be the most common indications for tenodesis of the LHBT, followed by SLAP tears (28%) and a positive clinical exam for LHBT-related pain (26%).

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68.3 Pathogenesis

The role of the biceps at the shoulder has been a matter of debate. Historically, the LHBT was believed to be an important humeral head depressor; however, recent research suggested the LHBT to rather be a passive centralizer of the humeral head on the glenoid. Despite the debate regarding its function, there is general agreement that the proximal LHBT is a considerable pain generator. The intra-articular portion of the LHBT is subject to compression, shearing, and friction forces, whereas the extra-articular distal segment is primarily exposed to tensional strain.

The vascular anatomy of the LHBT may also play a role in the pathogenesis of disorders. The proximal 3 cm of the intra-articular portion of the tendon comprises specific areas of hypovascularization along with an abundant neural innervation, being correlated with the areas of increased mechanical strain. This combination of specific multidirectional force distributions and the segmental blood supply is believed to be the primary contributor to the intra-articular degenerative process of the LHBT.

Pathologies of the pulley system have also been found to be associated with rotator cuff lesions. Anteromedial dislocations of the LHBT have been reported in the setting of subscapularis (SSC) tendon tears, whereas "hidden" lesions of the lateral CHL, SGHL, and SSC tendon have been described in association with supraspinatus (SSP) tendon tears.

68.4 Classifications

Multiple classifications for pathologies of the LHBT have been proposed, without an existing consensus. The classifications primarily focus on secondary lesions of the shoulder combined with LHBT instability. Disorders of the LHBT can be broadly classified into three groups: isolated tendonitis, tendonitis associated with an SLAP lesion or tendon instability, and tendon instability combined with a high-grade lesion of the stabilizing pulley system and a rotator cuff tear.

Habermeyer et al. classified lesions of the biceps reflection pulley into four types. Lesions of the SGHL resulting in anterior instability of the LHBT are considered as type 1. Type 2 describes a combined lesion of the SGHL with a partial rupture of the anterior portion of the SSP tendon. Type 3 is defined as a combination of an SGHL lesion and a partial rupture of the cranial portion of the SSC tendon. Type 4 combines a lesion of the anterior SSP tendon and a lesion of the cranial portion of the SSC tendon and is thought to be a result of anteroposterior instability.

Recently, the Habermeyer classification has been updated, as the former classification did not allow for correctly categorizing all pulley lesions due to the fact that in 36% of the pulley lesions, the SGHL fibers were not affected. Thus, an updated and simplified classification has been proposed comprising only three different types and allowing for a clear statement regarding biceps instability and its direction. Accordingly, type 1 describes a lesion of the medial pulley sling (SGHL and/or MCHL). Type 2 is defined as a lesion of the lateral pulley sling (LCHL), whereas type 3 is considered a combined lesion of the medial and lateral pulley sling (SGHL and CHL). Further, concomitant lesions of indirect pulley stabilizers (rotator cuff fibers) may be mentioned additionally according to existing classifications.

The arthroscopic classification according to Lafosse considers the direction and extent of LHBT instability, macroscopic lesions of the LHBT, and concomitant lesions of the SSC and/or SSP tendon.

68.5 Diagnosis

68.5.1 History

Patients with complaints involving the LHBT typically present with anterior shoulder pain, which may radiate distally following the course of the biceps muscle. Tendonitis is typically observed in patients over 50 years of age presenting with these complaints, along with pain

during resisted elbow flexion or forearm supination. In the setting of LHBT pathology, concomitant rotator cuff lesions have to be considered. In overhead athletes and those reporting traction injuries to the shoulder, lesions of the superior labrum (SLAP) should always be considered. Patients may feel a sensation of "snapping" in the anterior shoulder during abduction and external rotation of the arm in case of an instability of the LHBT. Rupture of the LHBT may present with an audible painful "pop" during sudden eccentric or concentric muscle contraction, followed by the typical "Popeye" sign, as the muscle retracts. In this setting, patients often describe an onset of pain prior to the injury, followed by relief following rupture of the tendon.

68.5.2 Clinical Exam

If the tendon is completely ruptured, physical examination may reveal a "Popeye" deformity. If the tendon is not ruptured, significant pain during motion, particularly during resisted elbow flexion or forearm supination, is a typical symptom. Tenderness is often observed during palpation of the bicipital groove. Occasionally, there may be ecchymosis in this area. In addition, it is important to examine for concomitant rotator cuff pathology, especially tears of the SSC and SSP tendon. Range of motion should also be assessed, in order to detect a possible incarceration of the LHBT stump in the glenohumeral joint (hourglass biceps). The O'Brien test can be used to identify an SLAP lesion. The Yergason and Speed tests can be used to evaluate pathologies occurring more distally to the tendon within the sulcus. However, clinical tests have to be considered as a set, as the specificity for diagnosis of each test alone is limited. Taylor et al. compared the diagnostic value of using a "three-pack" examination (bicipital groove palpation, throwing test, O'Brien sign) and found that together the test had a sensitivity of 73–98% with the specificity only ranging from 46% to 79%.

68.5.3 Imaging

In general, the diagnosis of an LHBT pathology is made clinically; however, it may be supplemented by imaging. Standard radiographs typically demonstrate non-pathologic findings. Subtle cystic erosions of the lesser tuberosity may be observed; however, this modality is usually unhelpful. Magnetic resonance imaging (MRI) with an intra-articular contrast agent may be the only reliable method to clearly distinguish between SLAP and pulley lesions (Fig. 68.1). Although being sufficient for detecting complete dislocation of the LHBT, preoperative imaging is limited for making a reliable diagnosis of LHBT pathology, especially in the setting of rotator cuff tears. Arthroscopic evaluation may be the only approach to objectively prove a diagnosed tendon instability, highlighting the importance of inspecting the entire intra-articular length of the LHBT prior to treatment. However, arthroscopy is unreliable in diagnosing extra-articular LHBT pathology, and macroscopic appearance of the tendon may not accurately reflect tendinopathy, complicating surgical decision-making.

Besides, ultrasound is a widely available, inexpensive imaging tool that has the advantage of a possible dynamic examination. Absence of the LHBT may be detected in the sulcus, and the tendons of the rotator cuff as well as the LHBT entering the pulley sling may be visualized. Tendinitis is often accompanied by fluid enhancement in the sulcus, although this remains an unspecific sign. The examiner needs to be skilled with the technique to achieve optimal results. If this is the case, especially lesions within the sulcus and medial subluxation of the biceps tendon towards the subscapularis muscle are reliable to detect; however, it is unreliable for intra-articular partial-thickness lesions of the tendon.

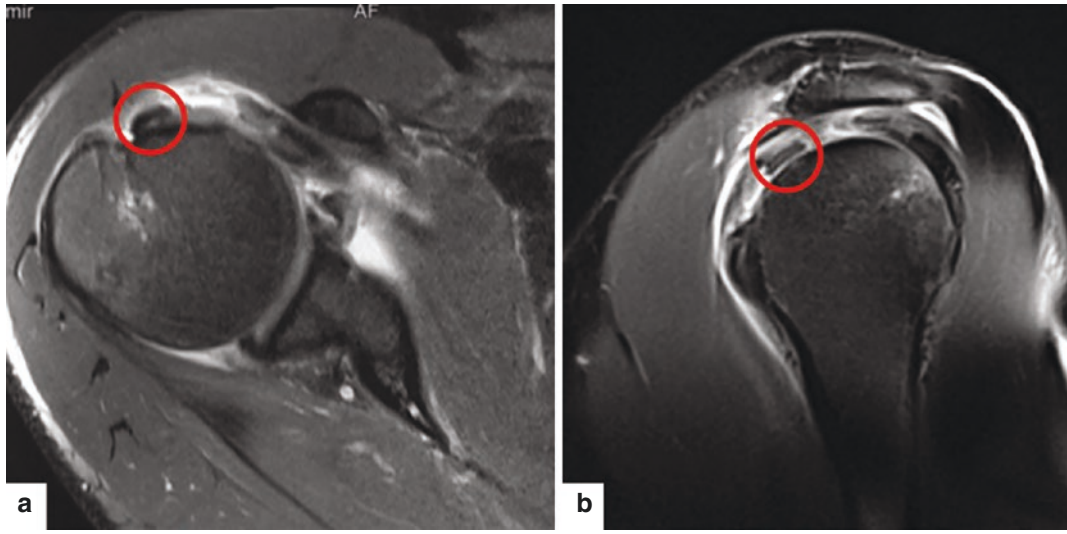

Fig. 68.1 MRI scan of a pulley lesion type IV according to Habermeyer with medial subluxation of the long head of the biceps tendon (red circle) as well as concomitant lesions of the cranial subscapularis and anterior supraspinatus. (**a**) Axial view, (**b**) parasagittal view

68.6 Treatment

Multiple surgical techniques have been proposed for the management of patients with LHBT-related pain, without an existing consensus regarding treatment. Both tenotomy and tenodesis have been reported to sufficiently decrease LHBT-related pain; however, clinical outcomes have been variable, with none of the techniques showing a clear functional superiority. Following tenotomy, cosmetic defects have been observed more frequently when compared to tenodesis. In the setting of biceps tenodesis, lower reoperation rates have been found following subpectoral fixation compared to suprapectoral fixation, as releasing the tendon from the bicipital groove may relieve the patient from most of the LHBT-related pain.

68.6.1 Tenotomy

Following induction of general anesthesia, the patient is placed in the beach-chair position. The surgical field is prepared and draped in a standard fashion with the arm being secured in a movable arm holder. Using a posterior standard portal, the glenohumeral joint is thoroughly evaluated for any concomitant intra-articular lesions due to the high incidence of combined lesions (e.g., rotator cuff, instability). The LHBT is inspected for its entire intra-articular length (Fig. 68.2). The origin of the tendon and the superior labrum are tested with a probe after establishing an additional anterosuperior portal under direct visualization to evaluate the biceps tendon anchor for SLAP lesions. The reflection pulley is evaluated for any lesions (hidden lesions of the SSC and/or SSP) or signs of instability. Finally, the tendon is pulled out of the sulcus to evaluate for possible fraying and/or synovitis, and the pulley structures are tested for stability.

A standard working portal is used to cut the biceps at its proximal insertion with either an arthroscopic scissor or a radiofrequency cutter, while preserving the integrity of the superior labrum. After detaching the tendon, it should be confirmed that the tendon slipped back into the sulcus. The proximal tendon stump is then debrided to a stable margin.

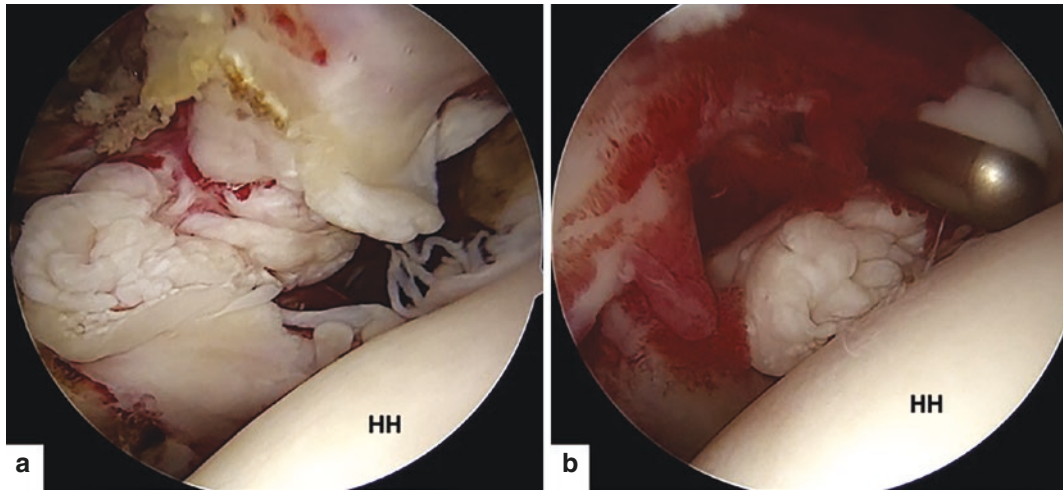

Fig. 68.2 Intraoperative view of an almost completely torn long head of the biceps tendon with extensive fraying and synovitis before (**a**) and after (**b**) tenotomy using a radiofrequency cutter. HH = humeral head

68.6.2 Mini-Open Subpectoral Biceps Tenodesis Using an All-Suture Anchor Onlay Fixation

Multiple methods are available for tenodesis of the LHBT, which can be distinguished according to the anatomic regions of fixation of the tendon stump. The author's preferred technique for a tenodesis of the LHBT is a mini-open subpectoral tenodesis using an all-suture anchor onlay fixation. As interference screw fixation has been reported to come along with various complications including humeral fractures at the drill hole, bioabsorbable screw reactions, and persistent pain, onlay fixation has been proposed to be a reliable alternative, in an attempt to reduce these risks.

Prior to tenodesis, arthroscopy is performed to identify any associated pathologies, to tenotomize the LHBT (Fig. 68.2), and to debride the proximal tendon stump. With the arm positioned in 90° of abduction and 90° of elbow flexion, the inferior border of the pectoralis major tendon is palpated. In the axillary crease, an incision is made from 1 cm above to 2 cm below the inferior border of the pectoralis tendon. A scalpel is used to cut down through the subcutaneous tissue while using an electrocautery to control bleeding. Once the inferior border of the pectoralis major has been identified, the fascia overlaying the coracobrachialis and short head of the biceps is incised in a proximal to distal manner. The pectoralis major tendon is retracted superiorly, leaving the LHBT palpable in the bicipital groove. Vigorous medial retraction should be avoided to prevent injury to the musculocutaneous nerve. With a clamp looped around the LHBT, it is then withdrawn from the bicipital groove.

Using an elevator, the periosteum in the bicipital groove is then stripped off, in order to prepare the cortical bed for fixation. A drill guide is positioned centrally in the bicipital groove and approximately 2 cm below the proximal border of the pectoralis major tendon. A 1.6 mm drill is used to create a unicortical bone tunnel for the placement of a single-loaded all-suture anchor (FiberTak, Arthrex Inc., Naples, FL). Beginning 1 cm proximal to the musculotendinous junction, one suture strand is used to whipstitch the LHBT with four passes at each side in a Krackow fashion until 3 cm distally. The excess proximal portion of the LHBT is sharply cut off, which is important to ensure appropriate tensioning. Subsequently, the free suture end is passed through the tendon, and with applying traction to the suture, the LHBT is tightened down to the anchor. Finally, the sutures are tied and the wound is closed.

68.6.3 Postoperative Rehabilitation

Following arthroscopic tenotomy without additional surgery, the patient should rest the arm for the first couple of days. Active and passive range of motion do not have to be restricted; however, active loading of the biceps should be avoided for the first 4–6 weeks following tenotomy.

In the setting of tenodesis, sling immobilization for 2 weeks with immediate active and passive range of motion is recommended. Resisted elbow flexion and forearm supination are restricted for 6 weeks following surgery. Overhead strengthening and heavy lifting are delayed for 3 months postoperatively.

Take-Home Message

- Pathologies of the LHBT include tendonitis, partial tearing, complete rupture, and lesions of the pulley reflection system along with subluxation and dislocation.
- The proximal LHBT is a considerable pain generator, as its intra-articular portion is subject to compression, shearing, and friction forces, while the extra-articular distal segment is primarily exposed to tensional strain.
- Both tenotomy and tenodesis have been reported to sufficiently decrease LHBT-related pain; however, clinical outcomes have been variable with none of the techniques showing a clear functional superiority.
- Following tenotomy, cosmetic defects have been observed more frequently when compared to tenodesis.
- The author's preferred technique for tenodesis of the LHBT is a mini-open subpectoral tenodesis using an all-suture anchor onlay fixation.

Summary

Pathologies of the LHBT include tendonitis, partial tearing, complete rupture, and lesions of the pulley reflection system along with subluxation and dislocation. The proximal LHBT is a considerable pain generator, as its intra-articular portion is subject to compression, shearing, and friction forces, while the extra-articular distal segment is primarily exposed to tensional strain. Both tenotomy and tenodesis have been reported to sufficiently decrease LHBT-related pain; however, clinical outcomes have been variable with none of the techniques showing a clear functional superiority. Following tenotomy, cosmetic defects have been observed more frequently when compared to tenodesis. The author's preferred technique for a tenodesis of the LHBT is a mini-open subpectoral tenodesis using an all-suture anchor onlay fixation, as interference screw fixation has been reported to come along with various complications including humeral fractures at the drill hole, bioabsorbable screw reactions, as well as persistent pain. Generally, surgical decision-making should be adjusted to patient-individual demands, using a technique the surgeon is most comfortable with.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. The biceps brachii muscle is innervated by:
   (a) Radial nerve
   (b) Suprascapular nerve
   (c) Circumflex nerve
   (d) Cutaneous muscle nerve
2. In voluntary shoulder dislocation, it is common to observe:
   (a) Rupture of the long head of the biceps
   (b) Rupture of the subscapularis tendon
   (c) Rupture of the supraspinatus
   (d) None of these injuries
3. Runs in the rotator interval:
   (a) Deltoid
   (b) Long head of the biceps
   (c) Subspinous
   (d) Short head of the biceps
4. The diagnosis of complete rupture of the long head of the biceps is:
   (a) Ultrasonographic

(b) Radiological in magnetic resonance imaging
(c) Predominantly clinical
(d) Clinical and ultrasonographic

5. In the scapulohumeral joint:
(a) The joint heads are separated by a fibrocartilaginous disc
(b) The tendon of the long head of the biceps runs
(c) The glenoid cavity is located on the vertebral margin of the scapula
(d) The joint capsule is extremely thick

Further Reading

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Adhesive Capsulitis

69

Miguel Angel Ruiz Ibán, Miguel García Navlet, Jose Ávila Lafuente, and Susana Alonso Güemes

Overview

Adhesive capsulitis of the shoulder, also known as frozen shoulder, is an idiopathic condition that presents with pain and stiffness of the shoulder. It is not related to previous injury to the shoulder (Fig. 69.1). It is characterized by initial inflammation followed by fibrotic alterations in the glenohumeral capsule (specially the rotator interval) and ligaments (in particular the coracohumeral ligament). The capsule is thickened and contracted, reducing the articular volume, and the synovial layer is initially increased with inflammatory reaction but is eventually lost.

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Its prevalence in the general population is around 3–5%. It typically affects adults between the fifth and sixth decades of life and is slightly more prevalent in females. Some comorbidities will be present in 85% of cases. The most important is diabetes mellitus with prevalences of 20% in type 1 diabetes and 10% in type 2 diabetes.

The natural course of the disease develops in three phases: The first phase (the freezing phase) lasts between 2 and 9 months and is characterized by intense shoulder pain and progressive development of stiffness. The patient complaint is most often isolated pain, especially severe during the night-time, and a minor trauma is sometimes recalled by the patient. In the second phase (the frozen phase), the pain often diminishes and disappears, and the stiffness is the main symptom and can last from 4 to 12 months. Progressively, in the third phase (the thawing phase), the stiffness solves progressively, and the patient regains function in 5–26 months.

The diagnosis of adhesive capsulitis is fundamentally clinical. Imaging techniques are not essential for the diagnosis but can help in differential diagnosis. The subject will present with pain and/or stiffness in varying degrees. On physical exam, there will be some degree of passive and active loss of range of motion, but strength is preserved. Other causes of pain and stiffness such as osteoarthritis, rotator cuff disease, or calcifying tendinitis should be excluded. Magnetic resonance imaging will show inflammatory changes initially followed by thickening of the capsule,

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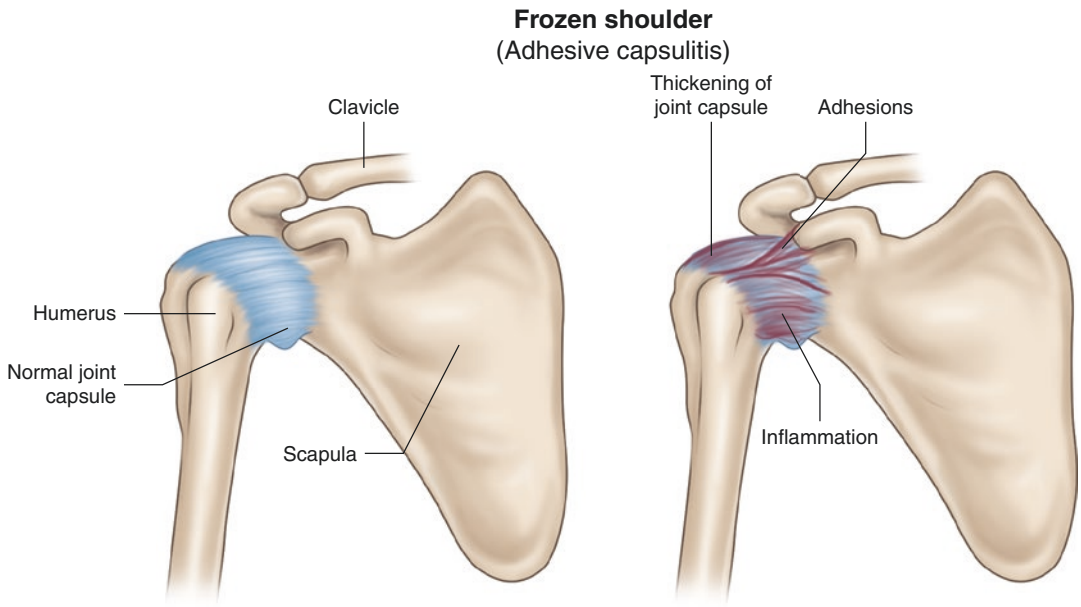

Fig. 69.1 Adhesive capsulitis

rotator interval and coracohumeral ligament, loss of the axillary pouch, and reduction of capsular volume.

The condition has always been considered to be self-limiting, with subjects evolving naturally to healing with time. Unfortunately, this is not the case as, even after 5 years, some degree of stiffness will be evident in 30% of cases and 5% of subjects will have persistent limiting symptoms. Thus, treatment should be offered to all subjects.

Initial treatment should focus on pain management. Oral nonsteroidal anti-inflammatory drugs or corticoids might provide short-term pain relief. Intra-articular steroid injections can be repeated and might be effective for up to 4 months. In cases with severe pain, suprascapular nerve blocks should be considered. For the management of stiffness, physical therapy is recommended, but intensive courses of assisted physical therapy have poorer outcomes than gentle self-administered, home-based, physical therapy. It is unclear if any of these treatments has any effect on the natural history of the disease. When symptoms persist for 3–6 months without improvement, surgical treatment should be considered: arthroscopic capsular release is a better option than mobilization under anaesthesia.

69.1 Definition

Adhesive capsulitis of the shoulder, also known as frozen shoulder, is a pathological condition on the shoulder that presents with pain and stiffness of the shoulder. It is caused by alterations of the glenohumeral capsule and ligaments that include inflammatory and fibrotic changes. It is an idiopathic condition, and that distinguishes it from secondary stiffness caused by trauma or surgery.

69.2 Epidemiology

The prevalence of adhesive capsulitis in the general population is estimated to be between 3% and 5% of the population. Five out of six subjects with the condition will be between 40 and 60 years old on onset. It seems to be slightly more prevalent in females (60% of cases), but males tend to have more severe symptoms and

have a worse outcome than females. It affects predominantly the non-dominant shoulder (60%), and in up to 50% of cases, the contralateral shoulder will be affected in a second episode. Simultaneous affection of both shoulders and relapse of the disease in the ipsilateral shoulder are rare occurrences.

There is a strong association with certain comorbidities, and comorbidities will be present in 85% of cases. The most important is diabetes mellitus with prevalences around 20% in type 1 diabetes and 10% in type 2 diabetes. Thyroid dysfunction also predisposes for adhesive capsulitis, and patients with hyperthyroidism have 1.22 risk of developing adhesive capsulitis. Other conditions such as obesity, myocardial infarction, hypoparathyroidism, Parkinson's disease, and emphysema are associated with increased risk of adhesive capsulitis. In particular, subarachnoid haemorrhage and Dupuytren's disease have a strong correlation with adhesive capsulitis.

69.3 Aetiology and Pathogenesis

Adhesive capsulitis is by definition idiopathic; thus, there will not be any findings in the physical exam, history, or imaging that justify the development of stiffness. Subjects who develop shoulder stiffness after trauma to the shoulder, immobilization, osteoarthritis, or a surgical procedure have shoulder stiffness secondary to these issues but not adhesive capsulitis.

The pathoanatomy of adhesive capsulitis is characterized by initial inflammation followed by fibrotic alterations in the glenohumeral joint that affect the capsule (specially the rotator interval, a triangular space in the anterior-superior part of the joint that is limited by the supraspinatus and subscapularis tendons) and the ligaments (in particular the coracohumeral ligament, located in the rotator interval). The capsule is thickened and contracted, reducing the articular volume, and the synovial layer is initially increased with inflammatory reaction but is eventually lost. The rotator interval doubles its thickness, and the coracohumeral ligament loses its elasticity and increases in size. Histologically, the capsule presents first inflammatory cell infiltration followed by fibrosis with a dense collagen matrix containing fibroblasts, some of them undergoing transformation into smooth muscle phenotypes (myofibroblasts), and there are increased levels of collagen type I and III.

From a biochemical point of view, there are numerous well-defined alterations that affect the cytokine environment, the extracellular matrix composition and the enzymes that regulate it (matrix metalloproteinases, MMPs), and the immune, vascular, and neural environment. There is an increase in proinflammatory cytokines, including interleukin-1α [IL-1α], IL-1β, IL-6, IL-8, cyclooxygenase-1 [COX-1], COX-2, and tumour necrosis factor-α. The matrix composition is altered with increased levels of vimentin, intercellular adhesion molecule-1, and fibronectin-1. These changes are attributed to a disbalance of MMP and tissue inhibitor of metalloprotease (TIMP) expression, both involved in the homeostasis of connective tissue remodelling. In brief, most MMPs (in particular MMP-14) are underexpressed and TIMPs are overexpressed. As inflammation has a clear role in the early pathogenesis of adhesive capsulitis, an immune origin has been suggested and increased populations of immune cells (T- and B-lymphocytes, macrophages, and mast cells) in the synovium of the rotator interval have been observed. Furthermore, there is an increase of nerve cells and neovascularization in the capsule.

69.4 Classification

When attempting to classify a subject with shoulder stiffness, the most important issue is to determine whether the problem is primary (that is, idiopathic or proper adhesive capsulitis) or secondary to trauma, osteoarthritis, immobilization, or surgery. Only subjects who do not have a clear antecedent that would justify the development of the stiffness can be properly diagnosed with adhesive capsulitis.

69.5 Natural History

The natural history of adhesive capsulitis has been well defined. The procedure has always been considered to be self-limiting, with subjects evolving naturally to healing with time, something that is nowadays disputed. Classically, the natural course has been divided into three phases (Fig. 69.2): a freezing phase, a frozen phase, and a thawing phase. The first phase (the freezing phase) lasts between 2 and 9 months and is characterized by intense shoulder pain and progressive development of stiffness. The patient complaint is most often isolated pain, especially severe during the nighttime, and a minor trauma is sometimes recalled by the patient. In the second phase (the frozen phase), the pain often diminishes and disappears, and the stiffness is the main symptom and can last from 4 to 12 months. Progressively, in the third phase (the thawing phase), the stiffness solves progressively, and the patient regains function in 5–26 months.

Another classification, that includes the anatomopathological and arthroscopic findings, distinguishes four stages: in the first stage, only pain is present and arthroscopic examination shows extended synovitis; in the second stage, pain is maintained and stiffness progressively develops with persistent synovitis and some loss of the axillary fold in the arthroscopic exam; in the third stage, stiffness is well established, synovitis disappears, and only loss of capsular space can be identified. To finish, in the fourth stage, the stiffness is gradually resolving, and arthroscopic evaluation shows persistent restriction of the capsular space.

One fundamental issue that determines the management is the final outcome expected, as classically adhesive capsulitis has been consid-

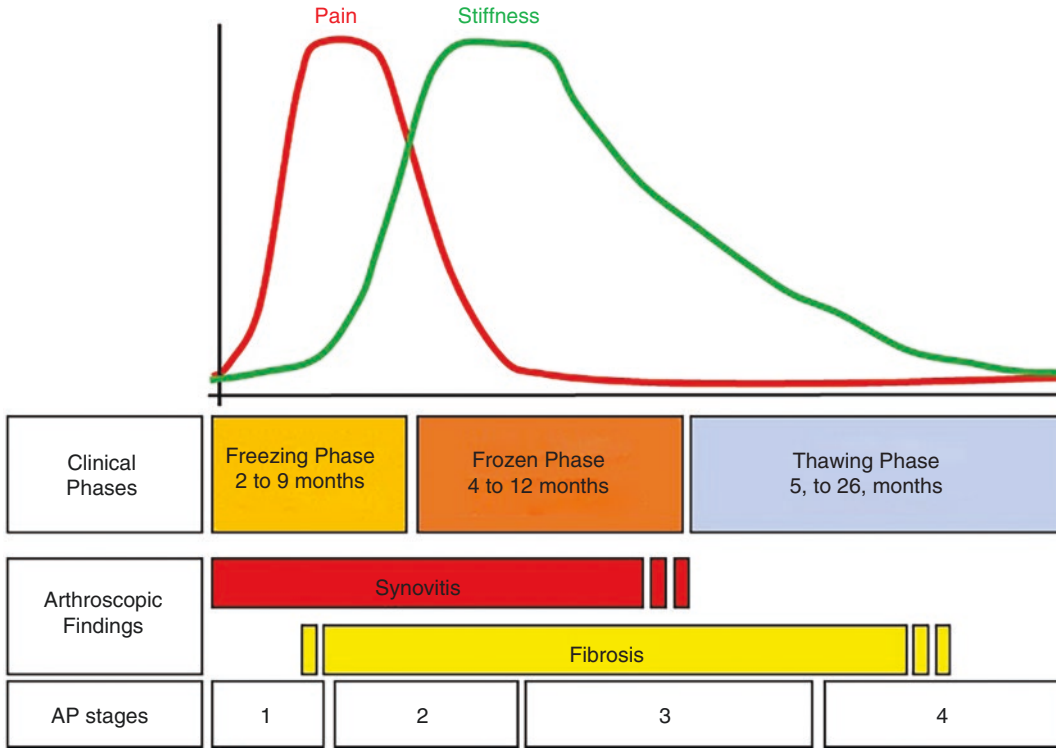

Fig. 69.2 The natural history of adhesive capsulitis. The three classical phases of the natural history are presented. Pain is pre-eminent in the first two phases, and stiffness gradually develops in the last two phases. During initial arthroscopic evaluation, synovitis is the main finding; but when the disease progresses, the reduction of the capsular volume, fibrosis of the capsule, and disappearance of the inferior capsular fold are more prominent

ered self-limiting. If complete healing could be expected, the case for continued conservative treatment should be very strong, despite the prolonged suffering sustained by the subject (adhesive capsulitis can last for many months or even 3–4 years). Unfortunately, this is not the case: even after 5 years, some degree of stiffness will be evident in 30% of cases and 5% of subjects will have persistent limiting symptoms.

69.6 Clinical and Radiological Diagnosis

The diagnosis is fundamentally clinical. The typical complaints of subjects with adhesive capsulitis are shoulder pain and stiffness. At the onset of the disease, pain is more prevalent, and progressively stiffness develops and becomes the main issue for the patient.

In any shoulder condition, the three main symptoms can be pain, loss of strength, stiffness, and/or instability. There should be a high suspicion for the diagnosis of adhesive capsulitis in any middle-aged subject that presents to the clinic with shoulder pain that has no clear origin (a traumatic onset or surgical history), specially if the subject has diabetes or has some degree of stiffness. When the main complaint is stiffness and pain is not that relevant on interrogation, once a traumatic origin has been excluded, the subject should be questioned about a previous episode of pain. Sometimes, the subject might relate the onset of pain with a minor injury or sprain, but to contemplate the diagnosis of adhesive capsulitis, major trauma should be excluded.

Physical exam should focus on the proper evaluation of the range of motion in abduction, flexion, and internal or external rotation. If loss of active range of motion is present, passive range of motion should be assessed. The subject with adhesive capsulitis and stiffness typically presents loss of passive range of motion and, during examination, usually a clear end point in the movement arc can be appreciated without significant pain. In patients with loss of active range of motion and normal passive range of motion, a muscular (rotator cuff tears) or neurologic (suprascapular nerve injury or brachial plexus palsy) problem should be suspected. If passive range of motion is especially painful and no clear end point can be observed, other problems such as rotator cuff disease, osteoarthritis, biceps tendinitis, or calcifying tendinitis should be excluded. Strength assessment of patients with adhesive capsulitis is usually normal.

A careful evaluation of the scapulothoracic rhythm (the way the scapula moves along the humerus in abduction, contributing to glenohumeral abduction with scapular external rotation and retraction) during active abduction should be performed, comparing the affected side with the normal side from posterior. The subject with adhesive capsulitis has stiffness related primarily to glenohumeral loss of range of motion, and this is compensated in part with scapulothoracic joint hypermobility (Fig. 69.3).

Imaging techniques are not essential for the diagnosis of adhesive capsulitis but can help in differential diagnosis. Plain radiographs of the shoulder can help in the diagnosis of conditions such as calcifying tendinitis or shoulder osteoarthritis that classically can present to the clinic with pain and stiffness without traumatic origin as the main symptoms. Most subjects with adhesive capsulitis will have completely normal plain radiographs, but in some cases, some disuse osteopenia can be observed after many months of symptoms.

Magnetic resonance imaging can help in the diagnosis of adhesive capsulitis. The typical findings include 1. thickening of the capsule at the axillary fold, the rotator interval, and the coracohumeral ligament; 2. hyperintensity on T2 signal of these structures; and 3. loss of the inferior capsular fold and, when associated with direct arthrography, loss of general capsular volume. Magnetic resonance imaging can help in the differential diagnosis with other causes of shoulder pain and stiffness such as osteoarthritis, rotator cuff disease, or neurological problems with muscle atrophy, which can be readily observed in the imaging evaluation.

Ultrasound imaging is increasingly popular as often can be performed in office by the orthopae-

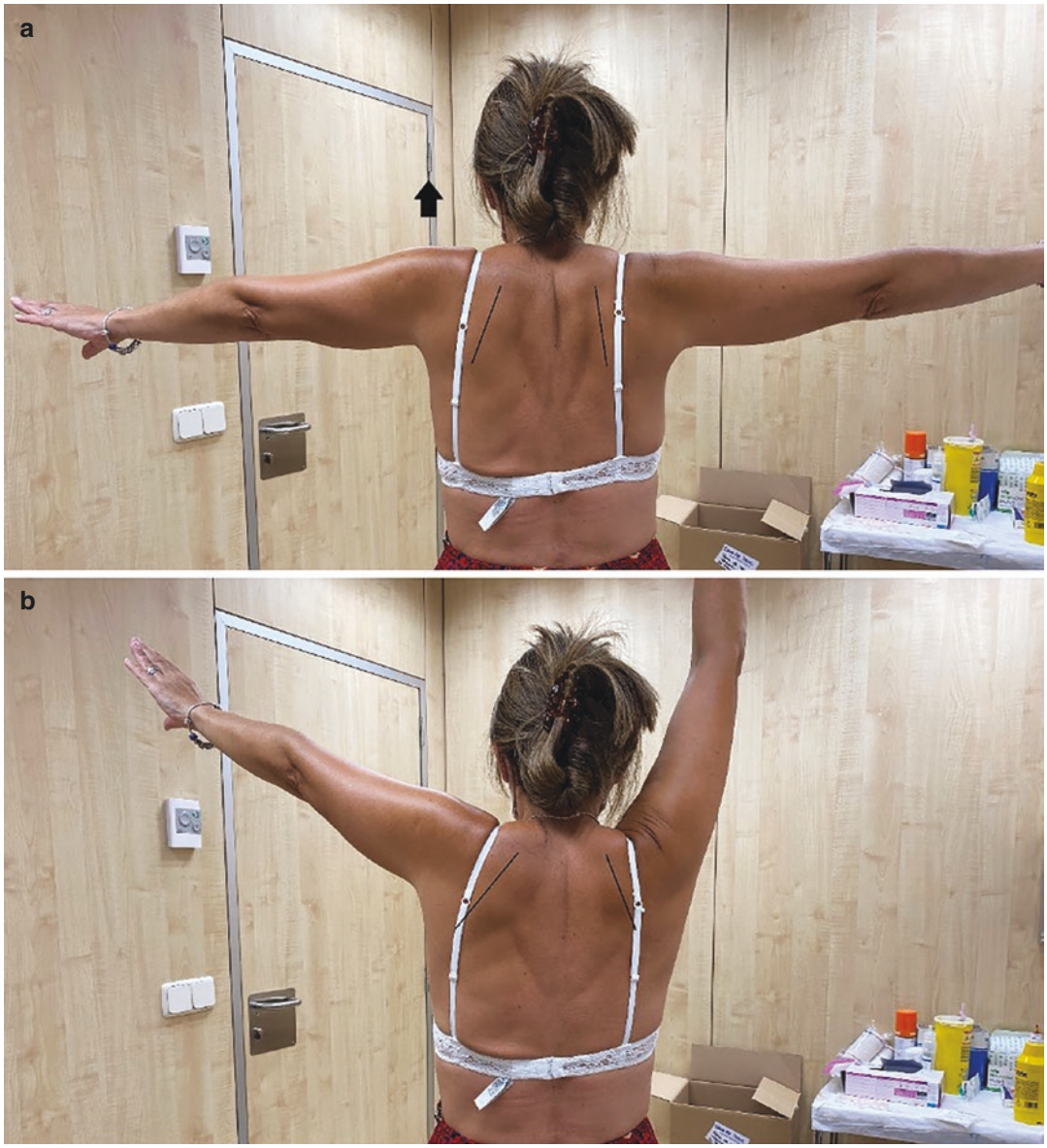

Fig. 69.3 Clinical presentation of adhesive capsulitis. This 51-year-old female has left shoulder adhesive capsulitis at the frozen phase. A. At 90° of abduction, there is some upward displacement of the affected shoulder (arrow), and excessive tilting of the medial scapular edge (black lines) can be appreciated due to loss of glenohumeral ROM and compensatory scapulothoracic hypermobility. B. When asked to reach 180° of abduction, the affected shoulder does not reach full abduction, and scapular disfunction is more evident

dic surgeon or general practitioner. The magnetic resonance findings of thickening of the rotator interval and coracohumeral ligament can also be appreciated during ultrasound evaluation but can be difficult to assess by less skilled evaluators. Anyway, there is clear room for ultrasound to help in differential diagnosis with rotator cuff disease and calcifying tendinitis.

69.7 Treatment

As previously mentioned, adhesive capsulitis is not a self-limited condition and can cause persistent symptoms and long-term disability in a relevant percentage of subjects. Thus, even if many of the therapeutic options available have efficacies that are limited in time or strength, some kind of treatment should be offered to all our patients with the condition (Table 69.1). Pain management is the most important issue in the initial phases; later, the management of stiffness is more relevant. A combination of treatment modalities is probably the best alternative for most patients (Fig. 69.4).

69.7.1 Conservative Treatment

Oral non-steroidal anti-inflammatory drugs (NSAIDs) in combination with physical therapy have been classically considered the mainstay of conservative management of adhesive capsulitis. Their use does not alter the course of the disease, but NSAIDs have shown to be a good alternative in the management of acute shoulder pain of many different origins. **Oral corticosteroids** can be a better alternative for the management of severe pain in the short term, but their effect is limited after 6 weeks of treatment and long courses of oral corticosteroids have significant associated risks. Both treatment alternatives are poor options for adhesive capsulitis management in diabetic subjects (due to increased cardiovascular risk and worse glycaemic control); in those, better control of the glycaemic situation is essential.

Physical therapy is one of the most used interventions for adhesive capsulitis, but there is strong controversy on its efficacy. There are many different physical therapy techniques available: manual therapy, mobilization techniques, and stretching. Different studies have shown that supervised aggressive exercises and stretching are inferior to home-based self-administered physical therapy. The focus of the therapy should be on avoiding pain, working in the pain-free range of motion, and scapular stabilization and scapulothoracic rhythm restoration.

Table 69.1 Treatment options in adhesive capsulitis

Treatment alternatives	Advantages	Disadvantages
Oral non-steroidal anti-inflammatory drugs	Readily accessible Safe and effective in the short term	Only efficacious in the short term Does not affect the natural history of the disease Contraindicated in diabetic subjects
Oral corticosteroids	Safe and effective in the short term	Should only be used for 2–3 weeks Does not affect the natural history of the disease Unsafe in diabetic subjects
Physical therapy	Readily accessible The patient expects this kind of treatment Can be self-administered and home-based	Might not affect the natural history of the disease Aggressive protocols are deleterious
Intra-articular steroid injections	Immediate pain relief Can help during physical therapy Can be repeated for up to 16 weeks	Might not affect the natural history of the disease Should be placed intra-articularly
Nerve blocks	Immediate pain relief Can help during physical therapy Can be supplemented with radiofrequency	Does not affect the natural history of the disease Invasive
Arthrodistension	Can improve range of motion quickly	No clear advantages to steroid injections Requires specific devices
Mobilization under anaesthesia	Might be as effective as capsular release in the short term	Surgical procedure Significant risk of complications
Arthroscopic capsular release	Effective in the long term Limited complications	Surgical procedure Expensive Technically demanding

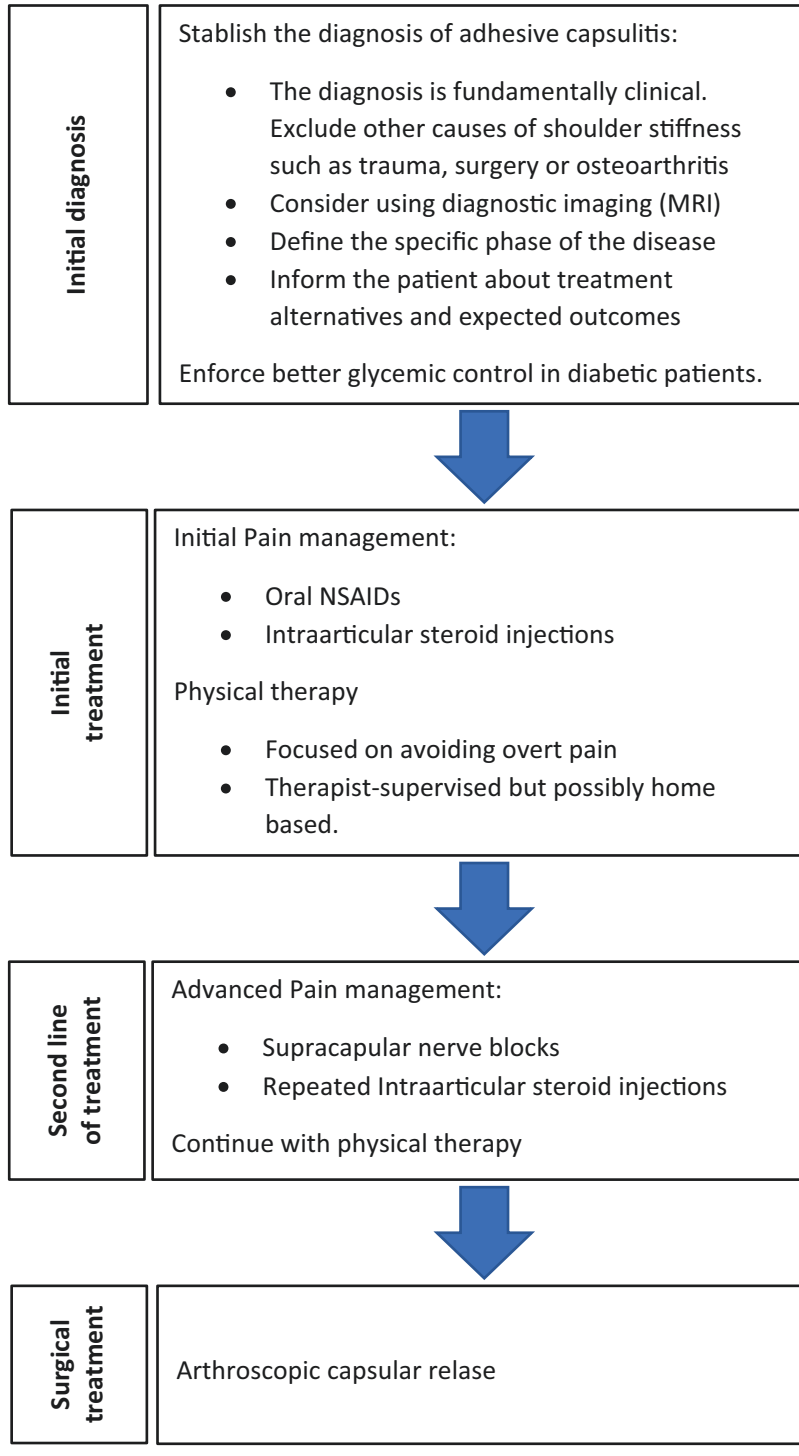

Fig. 69.4 A proposed management algorithm for adhesive capsulitis

Intra-articular steroid injections are a good alternative for the acute management of pain in adhesive capsulitis. Although they do not seem to affect the long-term outcome and the natural history of the disease, they are effective, reducing pain and improving function, for 3 to 6 weeks, and can be repeated, being effective for as long as 16 weeks after the first injection. Both low doses and high doses of corticosteroid (40 mg or 20 mg of triamcinolone acetonide) seem to be effective, but the injection should be placed inside the joint as its efficacy is superior to those administered in the subacromial space. Thus, ultrasound guidance, if available, should be recommended. Intra-articular hyaluronic acid has also been suggested to be effective, but the evidence available is limited.

In patients with severe pain that does not respond to other conservative alternatives, **suprascapular nerve blocks** are an interventional alternative that has been shown to be as effective and safe as intra-articular steroid injections. The addition of pulsed radiofrequency or continuous nerve blocks are also alternatives for more lasting pain relief.

Hydrodilatation of the shoulder capsule consists of injecting (using fluoroscopic or ultrasound guidance) of a large volume of sterile fluid (usually saline with local anaesthetic) into the joint. That causes an increase of the intra-articular pressure, distension of the capsule, and its eventual rupture. Again, there is strong controversy on its efficacy as it does not seem to be overtly superior to intra-articular steroid injections.

69.7.2 Surgical Treatment

Many patients with adhesive capsulitis will improve after a course of conservative treatment using one or more of the alternatives presented before. But a small subset of subjects will present persistent pain and shoulder dysfunction despite 3–6 months of conservative management. In these, two surgical alternatives are available: mobilization under anaesthesia and arthroscopic capsular release.

Mobilization under anaesthesia consists of obtaining complete anaesthesia of the shoulder (either with general anaesthesia or a complete interscalene nerve block) followed by gentle progressive mobilization of the shoulder joint with the aim of producing capsular tears that will allow the subject to regain full range of motion. Different studies have shown that its efficacy is comparable to arthroscopic capsular release in the short term, the main disadvantage of mobilization being that the capsular lesion is not obtained in a controlled fashion and that other structures such as the labrum or the tendons can be damaged; furthermore, serious complications such as humeral or glenoid fractures and neurovascular lesions have been described; full restoration of external rotation is not always achieved due to concerns of extracapsular damage.

Arthroscopic capsular release is considered the best surgical alternative for the treatment of refractory adhesive capsulitis. The surgeon will perform an arthroscopic evaluation of the glenohumeral joint, confirming the extent of the disease; this is followed by a controlled release of the rotator interval, the coracohumeral ligament, and the capsule, usually starting from anterior and progressing as needed to inferior and posterior. This procedure is safe with a small risk of neural damage (related to the relative closeness of the axillary nerve to the inferior capsule) and has shown to be effective in the long term. Patients with long-standing stiffness and diabetes will invariably fare worse.

Take-Home Message

- Adhesive capsulitis of the shoulder, also known as frozen shoulder, is an idiopathic condition that presents with pain and stiffness of the shoulder. It is not related to previous injury to the shoulder.
- It is characterized by initial inflammation and pain followed by fibrotic alterations that cause stiffness in the glenohumeral capsule.
- Prevalence is around 3–5%. It affects adults between the fifth and sixth decades of life and is slightly more prevalent in females. There is strong association with diabetes mellitus.
- The natural course of the disease develops in three phases: The freezing phase lasts between 2 and 9 months and is characterized by intense shoulder pain. In the frozen phase, pain often diminishes and disappears, and stiffness is the main symptom and can last from 4 to 12 months. Progressively, in the thawing phase, the stiffness slowly solves but 30% of subjects will have persistent stiffness.
- The diagnosis is fundamentally clinical.
- On physical exam, there will be some degree of passive and active loss of range of motion, but strength is preserved.
- Magnetic resonance imaging will show inflammatory changes initially followed by thickening of the capsule pain and stiffness. It can also help to exclude osteoarthritis, rotator cuff disease, or calcifying tendinitis.
- Initial treatment should focus on pain management with oral non-steroidal anti-inflammatory drugs and intra-articular steroid injections. In cases with severe pain, suprascapular nerve blocks should be considered.
- Physical therapy is recommended, but intensive courses of assisted physical therapy have poorer outcomes than gentle self-administered, home-based, physical therapy.
- When symptoms persist for 3 to 6 months without improvement, surgical treatment should be considered: arthroscopic capsular release is a better option than mobilization under anaesthesia.

Summary

Adhesive capsulitis of the shoulder, also known as frozen shoulder, is an idiopathic condition that presents with pain and stiffness of the shoulder. It is not related to previous injury to the shoulder. It is characterized by initial inflammation followed by fibrotic alterations in the glenohumeral capsule (specially the rotator interval) and ligaments (in particular the coracohumeral ligament). The capsule is thickened and contracted, reducing the articular volume, and the synovial layer is initially increased with inflammatory reaction but is eventually lost. Its prevalence in the general population is around 3–5%. It typically affects adults between the fourth and sixth decades of life and is slightly more prevalent in females. Some comorbidities will be present in 85% of cases. The most important is diabetes mellitus with prevalences of 20% in type 1 diabetes and 10% in type 2 diabetes. The natural course of the disease develops in three phases: The first phase (the freezing phase) lasts between 2 and 9 months and is characterized by intense shoulder pain and progressive development of stiffness. The patient complaint is most often isolated pain, especially severe during the night-time, and a minor trauma is sometimes recalled by the patient. In the second phase (the frozen phase), the pain often diminishes and disappears, and the stiffness is the main symptom and can last from 4 to 12 months. Progressively, in the third phase (the thawing phase), the stiffness solves progressively, and the patient regains function in 5 to 26 months. The diagnosis of adhesive capsulitis is fundamentally clinical. Imaging techniques are not essential for

the diagnosis but can help in differential diagnosis. The subject will present with pain and/or stiffness in varying degrees. On physical exam, there will be some degree of passive and active loss of range of motion, but strength is preserved. Other causes of pain and stiffness such as osteoarthritis, rotator cuff disease, or calcifying tendinitis should be excluded. Magnetic resonance imaging will show inflammatory changes initially followed by thickening of the capsule, rotator interval and coracohumeral ligament, loss of the axillary pouch, and reduction of capsular volume. The condition has always been considered to be self-limiting, with subjects evolving naturally to healing with time. Unfortunately, this is not the case as, even after 5 years, some degree of stiffness will be evident in 30% of cases, and 5% of subjects will have persistent limiting symptoms. Thus, treatment should be offered to all subjects. Initial treatment should focus on pain management. Oral non-steroidal anti-inflammatory drugs or corticoids might provide short-term pain relief. Intra-articular steroid injections can be repeated and might be effective for up to 4 months. In cases with severe pain, suprascapular nerve blocks should be considered. For the management of stiffness, physical therapy is recommended, but intensive courses of assisted physical therapy have poorer outcomes than gentle self-administered, home-based, physical therapy. It is unclear if any of these treatments has any effect in the natural history of the disease. When symptoms persist for 3–6 months without improvement, surgical treatment should be considered: arthroscopic capsular release is a better option than mobilization under anaesthesia.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Regarding adhesive capsulitis, which one of the following sentences is FALSE:
   (a) Adhesive capsulitis of the shoulder is also known as frozen shoulder
   (b) It is an idiopathic condition that presents with pain and stiffness of the shoulder
   (c) It is often related to previous injury or surgical insult to the shoulder
   (d) It is characterized by initial inflammation
2. Regarding the epidemiology of adhesive capsulitis, which one of the following sentences is FALSE:
   (a) Its prevalence in the general population is around 3–5%
   (b) It typically affects adults between the fourth and sixth decades of life
   (c) It is more prevalent in males
   (d) The prevalence in type 1 diabetes mellitus is 20%
3. Regarding the natural history of adhesive capsulitis, which one of the following sentences is FALSE:
   (a) The natural course of the disease develops in three phases:
   (b) The first phase (the freezing phase) lasts between 2 and 9 months and is characterized by intense shoulder pain and progressive development of stiffness
   (c) In the second phase (the frozen phase), the pain often diminishes and disappears, and the stiffness is the main symptom and can last from 4 to 12 months
   (d) It is a self-limited condition that heals always completely
4. Regarding the diagnosis of adhesive capsulitis, which one of the following sentences is FALSE:
   (a) Plain radiographs of the shoulder can help in the diagnosis of the thickening of the capsule
   (b) The diagnosis is fundamentally clinical
   (c) The typical magnetic resonance findings include 1. thickening of the capsule at the axillary fold, the rotator interval, and the coracohumeral ligament; 2. hyperintensity on T2 signal of these structures; and 3. loss of the inferior capsular fold
   (d) Ultrasound imaging can also appreciate the thickening of the rotator interval

5. Regarding the treatment of adhesive capsulitis, which one of the following sentences is FALSE:
   (a) Oral NSAIDs and intra-articular steroid injections can provide short-term pain relief
   (b) Physical therapy is recommended
   (c) Intensive therapy has worse outcomes than self-administered, home-based, exercises
   (d) Hydrodistension and mobilization under aesthesia are the preferred forms of treatment

Further Reading

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Glenohumeral Arthritis

70

Alfonso Ricardo Barnechea Rey

Overview

Glenohumeral arthritis, albeit uncommon, can be very debilitating and painful. It can be primary or can be caused by many conditions. Mild cases can be managed conservatively, whereas prosthetic replacement can improve more advanced cases.

70.1 Introduction

Arthritis at the glenohumeral joint is not as frequent as in the case of other joints such as the hip or knee. It is also not as frequent as a cause of shoulder pain. Nonetheless, when untreated or in advanced cases, it is a very debilitating and painful condition, so early recognition and management are of importance. It is more frequent in women, and its incidence increases after the age of 50.

70.2 Etiology

Glenohumeral arthritis is caused by numerous conditions, and it can be divided into several categories. Primary osteoarthritis is believed to occur in the same manner as in other primary arthritis, due to a degeneration of articular cartilage. Main risk factors include shoulder overuse, overhead sports, history of trauma, obesity, and genetic/hereditary background. Secondary causes include post-traumatic, instability, infection (both acute and chronic), inflammatory, chondrolysis, rotator cuff disorders, and avascular necrosis, among others (Table 70.1).

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Table 70.1 Causes of glenohumeral arthritis

Degenerative
• Osteoarthritis (primary, idiopathic)
Chondrolysis
Post-traumatic
• Post-dislocation arthritis
• Generic post-traumatic
Avascular necrosis
Connective tissue disorders
• Rheumatoid arthritis
• Spondyloarthropathies
• Systemic lupus erythematosus
Crystal deposition arthropathy
• Gout
• Pseudogout (calcium pyrophosphate)
• Apatite[II]
• Amyloid deposition
Postinfectious
• Post-acute septic arthritis
• Tuberculous arthritis
Rotator cuff tear arthropathy
Idiopathic destructive arthritis of the shoulder
Dialysis arthropathy
Neuropathic arthropathy
Endocrine/metabolic
• Hemochromatosis
• Alkaptonuria
• Acromegaly
• Hyperparathyroidism
Synovial causes
• Pigmented villonodular synovitis
• Primary synovial osteochondromatosis
Other
• Sarcoidosis
• Polymyalgia rheumatica

70.3 Pathophysiology and Pathoanatomy

In the setting of primary glenohumeral osteoarthritis, involvement usually starts at the intra-articular synovial membrane. As the condition progresses, articular cartilage erosion starts, usually in the posteroinferior aspects of both the glenoid and the humeral head. This leads to pain and stiffness, especially on external rotation. Continuous erosion and subsequent pain lead to greater stiffness, capsular retraction, and instability due to abnormal glenohumeral motion (the soft tissues fail to stabilize the head in the center of the joint), leading to greater erosion and pain.

In the case of inflammatory diseases, direct synovial and articular damage is caused by antibodies and inflammatory mediators. In post-traumatic arthritis, it is believed that necrosis due to trauma to the microvasculature is the direct cause, and in instability-related arthritis, abnormal, eccentric humeral head motion relative to the glenoid is the cause of chondral erosion.

Rotator cuff arthropathy (RCA) is defined as glenohumeral arthritis produced by superior migration of the humeral head, this in turn due to the inability of the rotator cuff to adequately depress it; this occurs when there is a massive rotator cuff tear, involving the supraspinatus in conjunction with the subscapularis and/or the infraspinatus. The superior migration of the head causes eccentric loading of the joint, causing superior erosion; depending on the tendons involved, erosion can be more anterior or posterior.

70.4 Clinical Presentation

Initially, glenohumeral osteoarthritis presents with progressive shoulder pain and inability to perform tasks that involve greater range of motion such as overhead or external rotation activities. Progressively, stiffness and weakness appear as pain increases with motion; the pain starts to wake the patient at night. In advanced stages, pain at rest and severe disability occur.

On clinical examination, one must inspect the shoulder carefully. Neck examination allows to find signs of cervical spine arthritis or other causes of referred shoulder pain. Inspection and palpation of the scapula allow to look for cuff muscle atrophy (if localized, evidence of massive tears or neurological involvement), and active scapular motion is many times abnormal; usually, loss of the scapulohumeral rhythm (the normal coordinated movement between scapula and humerus) reflects scapular muscular weakness.

Pain and tenderness at the posterior joint line may orient to primary, degenerative causes, whereas tenderness in the anterior joint line may orient to inflammatory causes. Passive and active range of motion must be evaluated and compared

to the healthy side (if unilateral); important differences between passive and active range of motion indicate secondary causes such as rotator cuff disorders or instability.

70.5 Diagnosis and Imaging

Usually, a complete history and detailed clinical examination can orient the diagnosis, but one must rule out differential diagnoses (Table 70.2). One must complement evaluation with an adequate set of images, always starting with X-ray imaging. A true anteroposterior (beam must be oriented 30–40° laterally, so it is tangential to the joint line), lateral (or Y-view) scapular, and axillary views are necessary. One can evaluate the articular space (mostly for narrowing), presence of osteophytes (especially so-called goat-beard osteophytes at the inferior head border), both glenoid and head deformities, and subchondral cysts (Fig. 70.1).

CT imaging is helpful in determining the direction of both humeral and glenoid erosion and in assessing bone stock, especially in the glenoid; this is important in advanced cases to see if the glenoid can support a prosthetic implant or if bone grafting or augmentation will be needed.

MRI imaging is helpful in evaluating soft tissues; in early stages, it allows to detect articular cartilage erosion, synovial thickening, early-stage osteonecrosis, and capsulolabral or cuff tendon damage.

Other imaging tests such as radionuclide imaging tests may help detect monoarticular inflammatory, infectious, or tumoral causes.

Table 70.2 Differential diagnoses for glenohumeral arthritis

Stiff shoulder
• Primary (frozen shoulder)
• Secondary
Chondrolysis
Pseudoparesis/pseudoparalysis from massive tears
Neurologic
• Cervical radiculopathy
• Axillary nerve injury

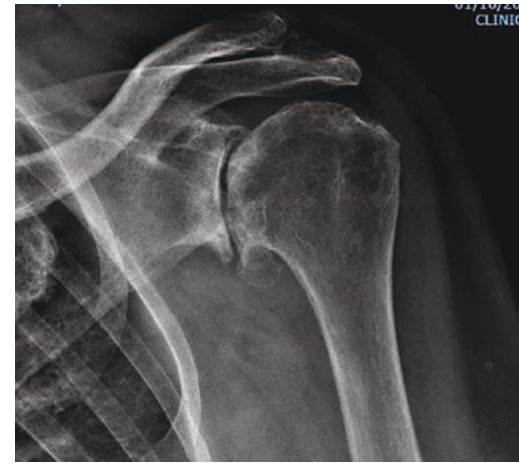

Fig. 70.1 X-ray true anteroposterior (AP) view of a right shoulder with primary osteoarthritis. Note the loss of articular space, humeral head, and glenoid deformity, and the "goat-beard" osteophyte at the inferior edge of the head

Laboratory tests such as CRP and ESR may orient to an inflammatory cause, but their very low specificity relegates them to a follow-up tool rather than diagnostic. Other specific tests may help to rule out other causes, such as connective tissue disorders.

70.6 Management

In early cases or in patients with other medical comorbidities that preclude surgical treatment, conservative treatment is advocated. The goal is to reduce pain and discomfort and, if possible, improve range of motion and overall functionality. The patient must be instructed on avoiding pain-generating activities and positions, home exercises, and general pain-relieving measures. Anti-inflammatory medication can help with pain, especially in inflammatory diseases; one must instruct the patient on their side effects and risks.

Physical therapy in this setting is paramount; its goal is to help restore normal scapulohumeral rhythm (the correct scapular-to-humeral relationships when glenohumeral motion occurs), release contractures, help with capsular stretching, and

Table 70.3 Surgical alternatives for glenohumeral arthritis

Arthroscopic debridement and release
• Mild-to-moderate cases
• Limited effectiveness
Replacement arthroplasty
• Surface arthroplasty
– Young patients
– Localized cartilage involvement
• Anatomic total shoulder arthroplasty (ATSA)
– Patients with intact or repairable rotator cuff
• Reverse total shoulder arthroplasty (RTSA)
– Requires good deltoid function

strengthen dynamic stabilizers. One must not expect to have a dramatic increase in range of motion, sometimes not at all; pain relief and better daily activity improvement are the main goals.

When conservative management fails, or when arthritis has advanced, surgical treatment offers pain relief and moderate functional improvement. Strategies are summarized in Table 70.3. In moderate cases, arthroscopic articular debridement along with synovectomy can help with pain relief, although the effect is limited.

The mainstay of surgical treatment for moderate-to-severe glenohumeral arthritis is prosthetic replacement. The goal is to relieve pain, as well as to restore glenohumeral motion to a more functional level when possible.

Surface arthroplasty consists of a cap that replaces only the articular cartilage and a portion of the subchondral bone at the humeral head; this may or may not include a component at the glenoid side. It is indicated in mild-to-moderate, more localized cartilage involvement with slight or no soft-tissue damage in younger, more active patients.

Total anatomic shoulder arthroplasty consists of replacing the entire humeral head with a metallic, spherical-section head, fixed to a metallic stem that goes into the medullary humeral canal, and the glenoid surface is replaced by a polyethylene component, directly fixed to either the bone or a metallic base that anchors into the glenoid bone. It is indicated in cases of moderate-to-severe arthritis with a functional or repairable rotator cuff (Fig. 70.2).

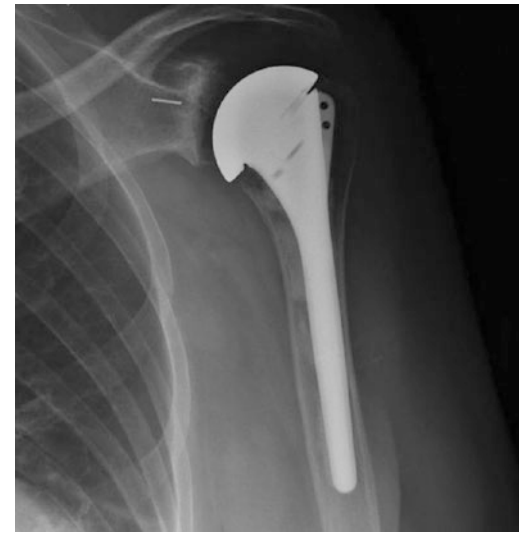

Fig. 70.2 Anatomic total shoulder arthroplasty at 1 month after surgery. The case was presented in Fig. 70.1. The glenoid component is made of cross-linked polyethylene, hence its radiolucent aspect (it has the radio-opaque line in the glenoid neck as a marker)

In cases of rotator cuff arthropathy or important involvement of the rotator cuff and/or soft-tissue static stabilizers (bone, labrum, ligaments), reverse shoulder arthroplasty provides excellent pain relief while maintaining stability due to its configuration. By putting a hemispherical component on the glenoid side, medialization of the glenohumeral center of rotation allows more deltoid muscle fibers to become abductors; also, tensioning of the deltoid results in a better lever arm and increased joint reaction forces that increase prosthetic stability. These conditions make the necessity of an intact cuff unnecessary (Figs. 70.3 and 70.4). However, there is a sine qua non condition for the patency of both the deltoid muscle and the axillary nerve, because this muscle becomes the main, if only, joint dynamic stabilizer. The other consideration is that if there is lack of external and/or internal rotation due to involvement of the anterior and/or posterior cuff, the reverse prosthesis will not correct it and additional gestures (such as tendon transfers) may be needed.

Results of prosthetic shoulder replacement are generally good for pain relief and acceptable for basic daily living activities, but rarely does it

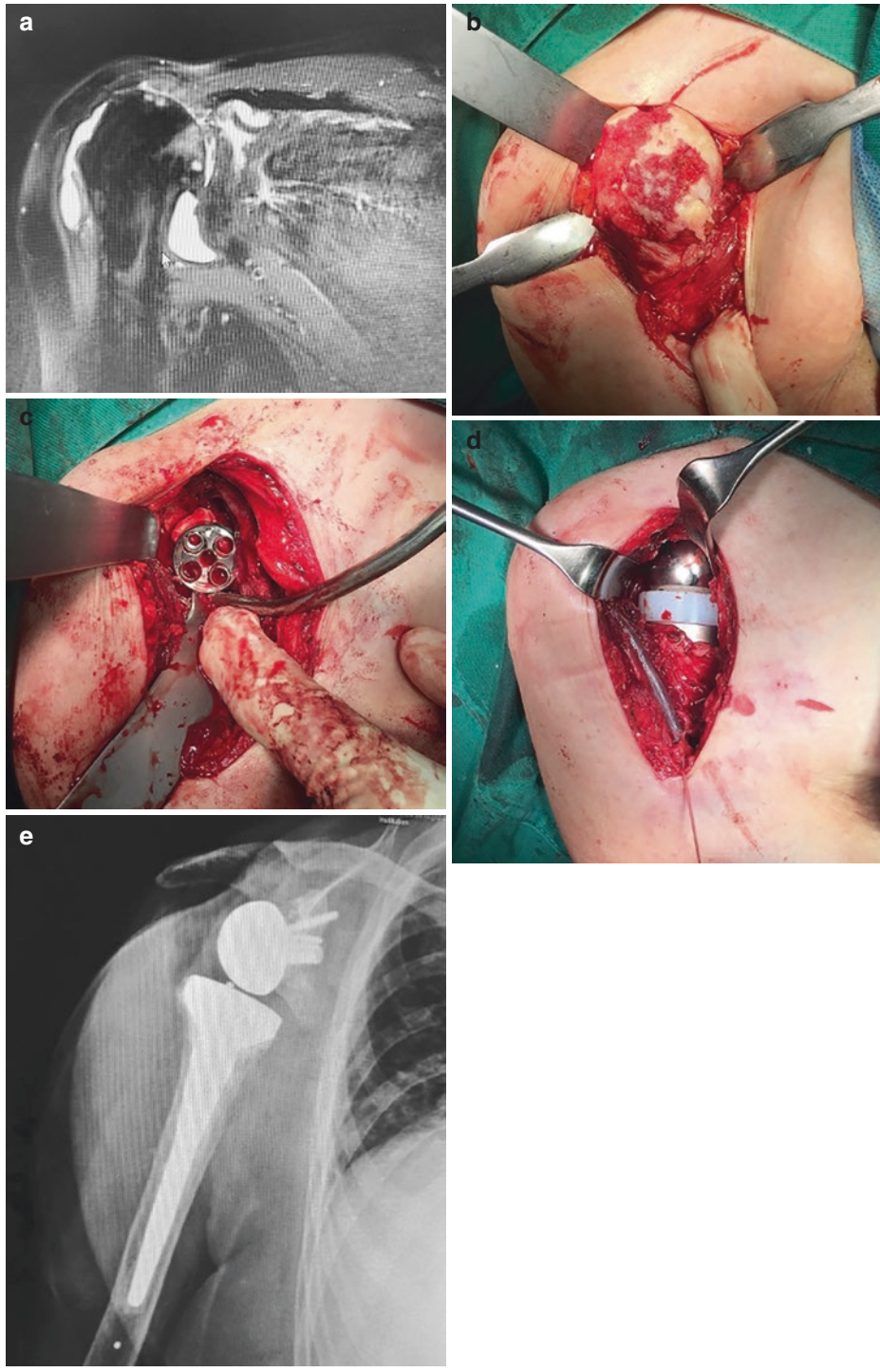

Fig. 70.3 Reverse total shoulder arthroplasty for treatment of arthritis due to rotator cuff arthropathy. Preoperative MRI shows superior migration of the humeral head due to a massive cuff tear (Fig. 70.3a); no supraspinatus tendon can be seen on the articular space. On surgery, damage to the humeral head can be appreciated (Fig. 70.3b). The base for the glenoid component is shown in Fig. 70.3c, as well as an autograft from the resected head. After placement of all components, the prosthesis is reduced (Fig. 70.3d). Postoperative X-rays show the reverse prosthesis in place (Fig. 70.3e)

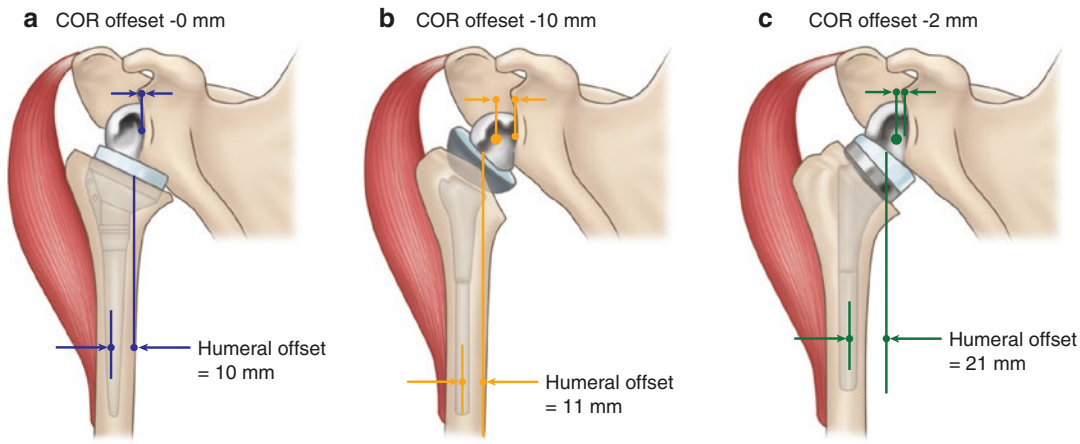

Fig. 70.4 Reverse shoulder arthroplasty

achieve normal or near-normal ranges of glenohumeral motion. After replacement surgery, a comprehensive, staged physical therapy program is needed in order to restore muscle strength and basic glenohumeral motion. Complications can occur, such as prosthetic loosening, nerve (especially the axillary nerve) damage, infection, and fractures around the implant.

Take-Home Message

- Main risk factors include shoulder overuse, overhead sports, history of trauma, obesity, and genetic/hereditary background.
- Secondary causes include post-traumatic, instability, infection, inflammatory, chondrolysis, rotator cuff disorders, and avascular necrosis, among others.
- Rotator cuff arthropathy (RCA) is defined as glenohumeral arthritis produced by superior migration of the humeral head.
- Initially, glenohumeral osteoarthritis presents with progressive shoulder pain and inability to perform tasks that involve greater range of motion such as overhead or external rotation activities.
- One must complement evaluation with an adequate set of images, always starting with X-ray imaging and continuing with CT and MRI.
- In early cases or in patients with other medical comorbidities that preclude surgical treatment, conservative treatment is advocated.
- When conservative management fails, or when arthritis has advanced, surgical treatment offers pain relief and moderate functional improvement.

Summary

Arthritis at the glenohumeral joint is not as frequent as in the case of other joints such as the hip or knee. It is also not as frequent as a cause of shoulder pain. Nonetheless, when untreated or in advanced cases, it is a very debilitating and painful condition, so early recognition and management are of importance. Primary osteoarthritis is believed to occur in the same manner as in other primary arthritides, due to a degeneration of articular cartilage. Main risk factors include shoulder overuse, overhead sports, history of trauma, obesity, and genetic/hereditary background. Secondary causes include post-traumatic, instability, infection, inflammatory, chondrolysis, rotator cuff disorders, and avascular necrosis, among others. In the setting of primary glenohumeral osteoarthritis, involvement usually starts at

the intra-articular synovial membrane. This leads to pain and stiffness, especially on external rotation. Rotator cuff arthropathy (RCA) is defined as glenohumeral arthritis produced by superior migration of the humeral head, this in turn due to the inability of the rotator cuff to adequately depress it; this occurs when there is a massive rotator cuff tear, involving the supraspinatus in conjunction with the subscapularis and/or the infraspinatus. Initially, glenohumeral osteoarthritis presents with progressive shoulder pain and inability to perform tasks that involve greater range of motion such as overhead or external rotation activities. One must complement evaluation with an adequate set of images, always starting with X-ray imaging. CT imaging is helpful in determining the direction of both humeral and glenoid erosion and in assessing bone stock, especially in the glenoid. MRI imaging is helpful in evaluating soft tissues. In early cases or in patients with other medical comorbidities that preclude surgical treatment, conservative treatment is advocated. Physical therapy in this setting is paramount. When conservative management fails, or when arthritis has advanced, surgical treatment offers pain relief and moderate functional improvement.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. They are indications for shoulder replacement except one:
   (a) Primary glenohumeral arthrosis
   (b) Recurrent shoulder dislocation
   (c) Dislocation fracture of the humeral head
   (d) Eccentric arthropathy as a result of rotator cuff rupture
2. Rupture of the rotator cuff is often the final phase of one of these conditions:
   (a) Glenohumeral arthrosis
   (b) Recurrent shoulder dislocation
   (c) Humeral acromion friction syndrome
3. Radiographic examination in cases of rotator cuff rupture may reveal:
   (a) Mushroom-shaped deformity of the humeral head
   (b) Juvenile bone cyst
   (c) Rise of the humeral head
   (d) Reduction of the glenohumeral joint rhyme
4. One of these muscles is not part of the rotator cuff of the shoulder:
   (a) Subspinatus
   (b) Subscapularis
   (c) Deltoid
   (d) Supraspinatus
5. The most important limitation for the reverse shoulder arthroplasty is:
   (a) Deltoid insufficiency
   (b) Massive rotator cuff tears
   (c) Supraspinatus tears
   (d) Poor bone stock

Further Reading

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Berton A, Gulotta LV, Petrillo S, Florio P, Longo UG, Denaro V, Kontaxis A. The effect of humeral version on teres minor muscle moment arm, length, and impingement in reverse shoulder arthroplasty during activities of daily living. J Shoulder Elbow Surg. 2015;24(4):578–86.

Shoulder Fractures

71

Miguel Angel Ruiz Ibán, Ignacio de Rus Aznar, Raquel Ruiz Díaz, Isabel Delgado Martinez, and Jorge Díaz Heredia

Overview

Fractures of the shoulder affect both young, active individuals and older adults. In younger individuals, these fractures appear in the context of sport- or work-related injuries and sometimes due to high-energy trauma.

71.1 Introduction

Fractures of the shoulder affect both young, active individuals and older adults. In younger individuals, these fractures appear in the context of sport- or work-related injuries and sometimes due to high-energy trauma. In the elderly, there is a strong association between these fractures and osteoporosis.

This chapter focuses on the following traumatic injuries of the shoulder: clavicle fractures, scapular fractures and proximal humerus fractures, as instability of the glenohumeral joint and acromioclavicular injuries are addressed in other chapters. To finish, the concepts of scapulothoracic dissociation and floating shoulder are also succinctly addressed.

71.2 Clavicle Fractures

71.2.1 Definition

The clavicle is the only long bone that ossifies through intramembranous ossification. Its subcutaneous position and the fact that it is the only bony connection between the upper extremity and the trunk make it especially prone to trauma, and clavicle fractures are very common.

Most clavicle fractures will occur in the midshaft portion, but distal clavicle fractures are not uncommon. These distal injuries can also affect

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UG. Longo, V. Denaro (eds.), *Textbook of Musculoskeletal Disorders*,
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the complex ligamentous structures that connect the scapula and the clavicle. Medial clavicle fractures are extremely uncommon and generally associated with high-energy trauma.

71.2.2 Epidemiology

The clavicle is the most common broken bone in the body, representing by itself up to 4% of all fractures in the adult population. The estimated annual incidence of clavicle fractures is between 30 and 60 per 100,000 and seems to be steadily rising, at least in developed countries. Midshaft fractures account for approximately 70–80% of fractures and distal third fractures for 20–30%, with proximal end fractures being extremely uncommon, accounting only for 3% of all clavicle fractures.

All age groups are prone to sustain clavicle fractures as these develop both in the newborn (clavicle fractures can develop during labour) and in the elderly and frail. In adults, its age distribution is bimodal with peaks in the younger population under 30 years of age and in the elderly and frail. In the younger population, midshaft fractures tend to be more common, but in the older population, distal clavicle fractures are more frequent.

71.2.3 Aetiology and Pathogenesis

There are three typical injury mechanisms that account for most clavicle fractures. Simple falls on the shoulder are the most common mechanism (around 31% of cases) and account for a disproportionate number of fractures in the older population, with the lateral end of the clavicle being the most commonly injured zone. Motor vehicle accidents (including bicycle injures) are the second most common aetiological mechanism (in 27% of cases) and with sporting activity injuries (in 23% of cases) are typical in younger subjects, affecting mainly the middle third of the clavicle.

The role that osteoporosis has in clavicle fractures in the elderly should not be overlooked. Although clavicle fractures are not considered typical fragility fractures (as opposed to hip, spine or wrist fractures), its increased incidence in the older population is a clear sign that the association is relevant, and the diagnosis of osteoporosis should be considered in this type of patients.

71.2.4 Classification

Allman classified clavicle fractures according to their situation along the bone in three types (Fig. 71.1a): type I: midshaft; type II: lateral; and type III: medial. Each of these fracture types has vastly different management methods and is further classified into subgroups.

Lateral, or distal, clavicle fractures are classified according to the degree of involvement of the coracoclavicular complex. Neer's classification divides these fractures into six different types (Fig. 71.1b): type I are nondisplaced fractures with intact CC ligaments, with the rest being displaced fractures with different patterns of coracoclavicular ligament involvement.

Midshaft clavicle fractures are usually classified with the AO classification into simple, wedge and complex fractures (Fig. 71.1c).

71.2.5 Clinical and Radiological Diagnosis

The subject with a clavicle fracture is often easily diagnosed as swelling, haematoma and local crepitation, with variable degrees of deformity alerting the clinician of the general outline of the injury. A complete neurological and vascular evaluation of the affected upper limb and succinct chest trauma is necessary as, even if neurovascular injury is relatively rare, brachial plexus or axillary vessel injury and pneumothorax have been described in association with clavicle fractures.

Simple radiological imaging is often enough to ascertain the type of fracture, and two AP clavicle conventional X-rays (one with craniocaudal tilt and another with caudocranial tilt) will define the injury pattern for most midshaft and distal clavicle fractures. CT can be useful in identifying medial third clavicle fractures, which can be

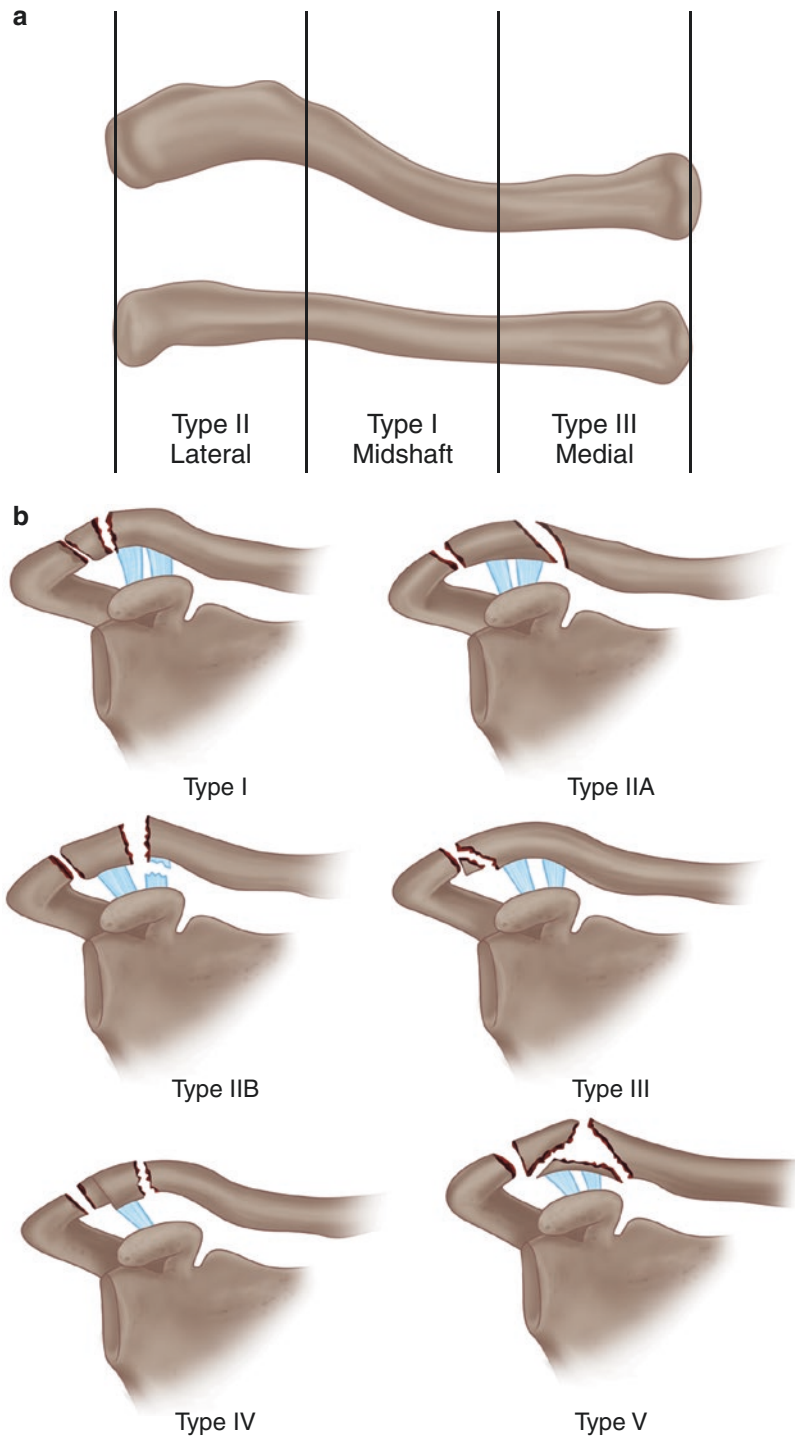

Fig. 71.1 Classification of clavicle fractures: (**a**) Allman classification distinguishes three types: type I: midshaft; type II: lateral; and type III: medial. (**b**) Neer's classification divides distal clavicle fractures into six different types according to the different patterns of coracoclavicular ligament (CCL) involvement: type I are nondisplaced fractures with intact CC ligaments, with the rest being displaced fractures; in type IIA, the fracture lies medial to the CCL; type IIB fractures occur between the CCL and lateral to them with injury to these ligaments; type III fractures are intra-articular lesions without CCL involvement; type IV are physeal fractures in immature individuals; and type V fractures are comminuted injuries with significant displacement and intact CCL attached to a third fragment. (**c**) Midshaft clavicle fractures are classified with the AO classification into simple, wedge and complex fractures with further subtypes that take into account the severity of the injury

overlooked in conventional X-rays. Sometimes, CT scans are also helpful in complex fracture patterns to assess the need for surgical management and help in preoperative planning.

71.2.6 Treatment

Nondisplaced fractures of the clavicula, independently of their position and pattern, do not require surgical treatment and should be managed conservatively. The arm should be placed in a sling for 2–3 weeks, and the subject should be actively discouraged to use their affected arm. A combination of oral analgesics and NSAIDs with local measures such as ice packs can help in the management of early inflammation and pain. Once this initial period has passed, radiological confirmation of the absence of secondary displacement is needed and the sling can be discontinued progressively during the next 3 weeks. Also, physical therapy can start with the initial focus on passive range-of-motion restoration, avoiding strengthening exercises and abduction over 90° for the first 6 weeks, when a second radiological follow-up is recommended. Return to contact sports should be avoided up to the fifth month.

The management of displaced clavicle fractures is very controversial, depending on the characteristics of both the fracture and the patient, and is best approached differentiating midshaft, lateral and medial fractures. Despite these, there are some absolute indications for surgical treatment: open fractures of any degree, severe soft-tissue damage with risk of skin perforation, acute neurovascular injury and floating shoulder. The management of non-union of clavicle fractures is presented at the end of this section.

71.2.6.1 Treatment of Displaced Midshaft Clavicle Fractures

Classically, displaced midshaft clavicle fractures have been treated conservatively as non-union is considered to be rare and symptomatic malunion rarely develops. But there is growing evidence from different randomized controlled trials that, although conservative management might be adequate for most subjects, there is an increased risk of non-union (10–20%) and symptomatic malunion when compared with surgical treatment. These complications are increased in the presence of comminution, severe displacement and smoking habit. On the other hand, surgical treatment, although it reduces the non-union rate (to 1–2%, possibly requiring further surgery) and shortens the recuperation time, is associated with the risk of infection (0–4%) and implant-related complications, such as local irritation requiring implant removal (that is needed for 15–30% of cases). As a general rule, surgical treatment should be offered to younger active patients, especially in fractures with displacement or shortening over 2 cm, shortening or comminution, but the surgeon should carefully present the advantages and disadvantages of each option.

Conservative management of displaced fractures follows that described for nondisplaced fractures. The use of a figure-of-eight bandage has been proposed as an alternative to a simple sling. This would have the advantage of free movement of the injured arm and possible reduction of displacement. Different studies have compared the outcomes of these two alternatives, and, in general, sling treatment seems to result in lower short-term pain, less shortening and increased satisfaction of the patient and thus is recommended for most patients.

When surgical treatment is indicated, there are two surgical options: intramedullary nailing and plating. Intramedullary nails are placed from the lateral end of the clavicle and are usually reserved for non-comminuted fractures; despite some biomechanical advantages, the need of more or less systematic implant removal due to lateral clavicle irritation makes it relatively unpopular in Europe. Open reduction and internal fixation with plates and screws is considered the standard of care for displaced comminuted clavicle fractures. There is no consensus on ideal plate position (either superior or anterior), but contemporary precontoured titanium plates with locked screws allow for non-union rates below 1%. Despite this, there is a very limited neurovascular risk that should be taken into account when obtaining informed consent and during surgery.

71.2.6.2 Treatment of Displaced Lateral Clavicle Fractures

The management of these fractures is guided by the Neer classification. Type I and III fractures can be managed conservatively if there is no gross displacement, but type II and V injuries cause disruption of the coracoclavicular ligaments (CCL) and will benefit from surgical treatment, as, for example, displaced type II fractures managed conservatively have a high rate of complications (up to 60%). There are many alternatives for the surgical management of these fractures: hook plates, standard distal clavicle plating, K-wire/tension band fixation, bone suturing and CCL suspension devices. When the distal fragment allows for secure screw fixation, traditional precontoured distal clavicle plates are probably the best option, but comminution and fracture pattern can force the surgeon to use other alternatives.

71.2.6.3 Treatment of Displaced Medial Clavicle Fractures

When displacement of more than one diameter of the clavicle is present or one of the absolute indications for surgery are present (see above), surgical treatment of medial clavicle fractures is required as the non-union rate can be 15–20%. Open reduction and internal fixation with plate and screws from an anterior approach is the best alternative although there are no specific plates designed for this anatomical site.

71.2.6.4 Management of Non-union of Clavicle Fractures

When, after 3–4 months of conservative treatment, symptomatic non-union develops in a clavicle fracture, surgical treatment should be considered. The patients will usually complain of local pain, arm weakness, and neurological or vascular symptoms. A CT scan will allow for confirmation of the non-union and for surgical planning. Resection of the non-union, followed by plate and screw fixation with the addition of autologous bone graft, is considered the best surgical alternative.

71.3 Scapula Fractures

71.3.1 Definition

The scapula is a flat triangular bone surrounded by substantial muscular masses that sits over the thorax and connects the trunk and the upper limb. Scapular fractures are uncommon fractures that often develop in the context of high-energy trauma. The complex anatomy of the scapula dictates that many different fracture patterns can develop. Correct identification of the fracture type is necessary for proper management.

71.3.2 Epidemiology

Scapula fractures account for less than 1% of all fractures and between 3 and 5% of the fractures of the shoulder girdle. They are most commonly seen in young and middle-aged patient as these group of patients are most frequently involved in the high-energy trauma that often causes scapular fractures. Scapula fractures are rarely isolated injuries; 90% of patients will present with another relevant injury associated with them, the most common being the thorax (80%), ipsilateral extremity (50%), head (50%) or spine (25%).

71.3.3 Aetiology and Pathogenesis

The scapula is well protected by a thick soft-tissue envelope that protects it from trauma. Furthermore, the high mobility of the glenohumeral joint, the most important articular link to the upper extremity, further protects it from the forces that usually cause fractures. These two factors make scapular injuries relatively uncommon. The most common injury mechanism is a direct trauma to the lateral or posterosuperior aspect of the upper trunk produced in the course of a high-energy trauma or during sporting activities. Uncommonly, fractures of the coracoid, acromion or spine can develop due to repetitive

stress; these develop usually in subjects with other significant shoulder problems such as rheumatoid arthritis, reverse shoulder arthroplasty or cuff tear arthropathy.

71.3.4 Classification

Scapula fractures are classified according to the international classification for scapula fractures (Fig. 71.2). This system divides the scapula into three sections: the articular segment, which includes the glenoid fossa and the articular rim; the processes, which include the coracoid and the acromion; and the remaining body of the scapula. A scapula fracture can affect one or more of these segments, and the pattern of the body and articular segment are further subclassified. For intra-articular fractures of the glenoid, the Ideberg classification is used (Fig. 71.3).

71.3.5 Clinical and Radiological Diagnosis

The patient with a scapula fracture typically presents with local pain with the arm held in an adducted position. Some local swelling is present, but important ecchymosis is rare. Deformity of the contour of the shoulder develops in fracture that affects the acromion or scapular spine. A complete neurovascular evaluation of the affected upper limb is necessary to rule out more severe injuries.

If the fracture has been produced in the course of a high-energy trauma, explicit attention should be given to look for associated injuries; specifically, the thorax, ipsilateral upper extremity, head and spine should be evaluated to discard potentially missed injuries. Conversely, scapular fractures are sometimes overlooked in patients with high-energy trauma and secondary diagnosis, days or weeks after initial management is not uncommon.

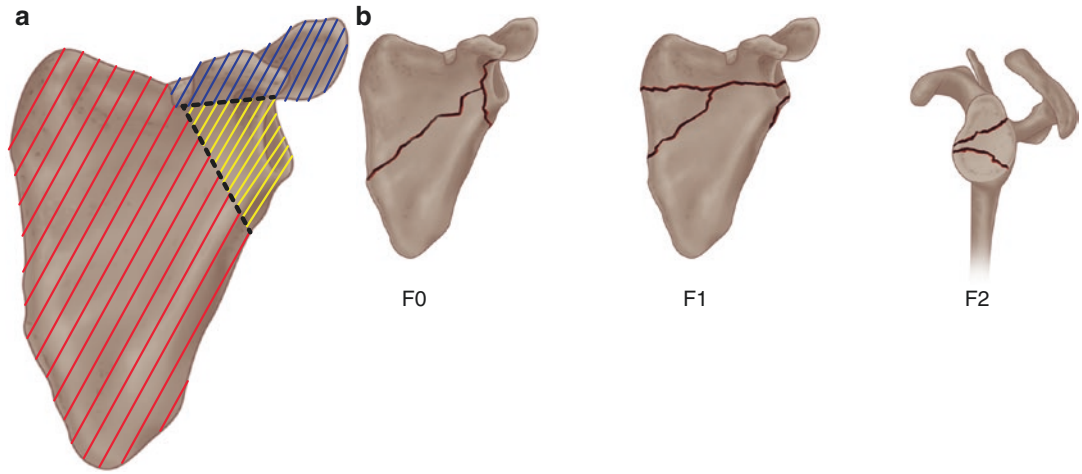

Fig. 71.2 The international classification for scapula fractures. (**a**) The scapula is divided into three-part articular segment (involving the glenoid fossa, F), processes (P) and body (**b**). Further subdivision of each allows for detailed characterization of the fracture pattern. The process fracture can affect the coracoid (P1), the acromion (P2) or both (P3). Body involvement is classified as a simple pattern (B1) with fracture lines that exit at 2 or less points or complex (B2) with fracture lines exiting at 3 or more points. B: The articular fractures (F) are divided into fractures of the articular segment, without glenoid fossa involvement (F0); simple patterns that involve the glenoid fossa (F1) including rim, transverse and oblique fractures; and multifragmentary joint fractures that involve the glenoid fossa with 3 or more articular fragments (F3)

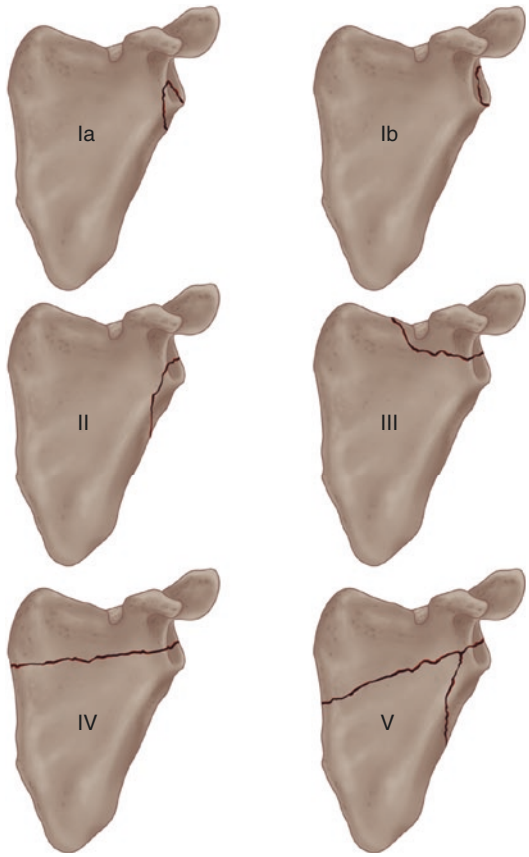

Fig. 71.3 The Ideberg classification for intra-articular scapula fractures. Type Ia: anterior rim fracture. Type Ib: posterior rim fracture. Type II: fracture line through glenoid fossa exiting scapula laterally. Type III: fracture line through glenoid fossa exiting scapula superiorly. Type IV: fracture line through glenoid fossa exiting scapula medially. Type V: combination of types IV and any of the others

Simple radiological studies of the shoulder or thorax usually allow for initial diagnosis, but a scapula CT scan is the preferred diagnostic tool; in particular, 3D reconstructions will allow for precise fracture pattern recognition and orient the clinician in treatment decisions.

71.3.6 Treatment

The management of scapula fractures is guided by the degree of displacement and the fracture pattern. As a general rule, nondisplaced fractures can all be managed conservatively with pain management (oral NSAIDs and local cold application) and a protective sling worn for 1–2 weeks followed by radiological confirmation of the absence of secondary displacement and gentle physical therapy to restore range of motion and strength. For displaced fractures, the management is dictated by the fracture pattern.

71.3.6.1 Displaced Process Fractures

Acute isolated displaced fractures of the acromion, coracoid and spine are rare, once stress fractures are excluded. Mild displaced fractures can be managed conservatively, but those with significant displacement (>1 cm) or that present in the context or other severe injuries to the scapular girdle (scapulothoracic dissociation or floating shoulder, see below) should be managed with open reduction and internal fixation and good outcomes can be expected.

71.3.6.2 Displaced Body Fractures

Classically, all these injuries were managed conservatively regardless of the displacement observed as consolidation is almost always successful and the shoulder girdle has a large potential to compensate for malunion. Despite this, poorer outcomes associated with residual snapping of the scapula and scapular dyskinesis are sometimes present. Thus, an attempt to single out fractures that could benefit from open reduction and internal fixation with plates has been made: nowadays, surgery is indicated when medial displacement of the glenoid is greater than 25 mm, when there is body angular deformity of 45° or more and when the glenopolar angle (measured between two lines: one parallel to the glenoid surface and another drawn between the upper edge of the glenoid and the distal tip of the scapula) is less than 20–30°.

71.3.6.3 Displaced Glenoid Fractures

These fractures, being true intra-articular fractures of a large joint, are always considered for open reduction and rigid internal fixation, but the displacement and percentage of articular involvement that merit surgery are in discussion, as smaller mildly displaced lesions might

be managed conservatively. Surgery is clearly indicated in fractures that associate with recurrent shoulder instability or those in which the humeral head follows the displaced fracture (static subluxation). For the rest of the cases, displacement over 4 mm and affection of more than 20% of the articular surface are often considered for surgery, but patient characteristics (comorbidity or high-demand subjects) can modify these limits. Internal fixation can be securely obtained with interfragmentary compression screws with arthroscopic assistance in subjects with smaller fragments, but large fragments need more extensive procedures with plating through a posterior Judet approach to the scapula.

71.4 Proximal Humerus Fractures

71.4.1 Definition

Fractures of the proximal humerus are common injuries that affect the metaphysis and epiphysis of the humerus. They present in both young active individuals and elderly osteoporotic patients. Most of them can be managed conservatively, but some might require surgery and the decision to operate or not is not always straightforward.

The peculiar characteristics of this zone, with a large articular surface with possibly compromised vascularization and extensive tendinous attachment at the tuberosities that can cause displacement and function impairment, make these fractures specially challenging.

71.4.2 Epidemiology

Proximal humerus fractures represent 4–6% of all fractures, being the third most common non-vertebral osteoporotic fractures in older adults after hip and distal radius fractures. They occur twice as often in women as in men. Their incidence seems to be increasing slightly in the developed world and is around 100/100,000 cases per year.

71.4.3 Aetiology and Pathogenesis

The aetiology of these fractures is varied. In the elderly, these often are low-energy fractures, occurring by an indirect mechanism when the subject falls from her own height over her hyperextended hand. In the younger population, high-energy trauma (traffic accidents, convulsions, sport-related trauma …) is the usual mechanism, usually resulting in more complex fracture patterns that can be associated with severe soft-tissue damage. As in other locations, pathological fractures must be considered.

71.4.4 Classification

Classically, proximal humerus fractures have been classified using the Neer classification (Fig. 71.4). The proximal humerus is divided into four segments: the greater tuberosity (GT), the lesser tuberosity (LT), the articular surface and the shaft. A segment is considered displaced if there is separation larger than 1 cm or angulation larger than 45° from the adjacent segment. If no segment is displaced, the fracture is classified as a one-part fracture; if both tuberosities and the articular segment are displaced, it is considered a four-part fracture. This classification is routinely used and frequently guides the treatment algorithm, but it was never designed to help in surgical decision-making and the displacement criteria were arbitrarily chosen. It has also very limited reproducibility between different observers.

The AO/OTA classification organizes the fractures in three main groups: A, unifocal and extra-articular fracture, with intact bloody supply; B, bifocal and extra-articular fracture, with possible injury to bloody supply; and C, articular fracture involving the anatomic neck, with high likelihood of necrosis. The fracture-dislocation can be included in group B (if tuberosities are affected) or C (if the articular surface is affected).

More recently, Foruria et al. described the most frequent patterns of fractures in proximal humerus and their predictable outcomes after

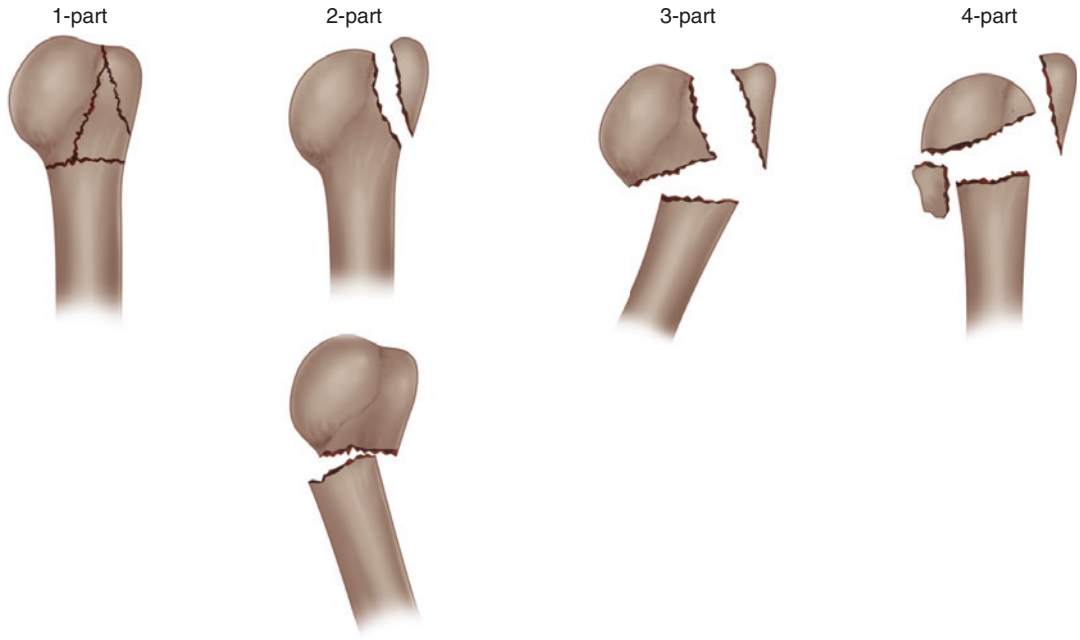

Fig. 71.4 The Neer classification of proximal humerus fractures. The proximal humerus is divided into four segments: the greater tuberosity (GT), the lesser tuberosity (LT), the articular surface and the shaft, divided at the surgical neck (SN). A segment is considered displaced if there is separation larger than 1 cm or angulation larger than 45° from the adjacent segment. A fracture pattern is classified as a one-part fracture if all segments are nondisplaced. If one segment is displaced form the rest, it is considered a two-part fracture; if two segments are displaced, then it is considered a three-part fracture. When all segments are displaced from each other, it is a four-part fracture

conservative treatment. Most fractures can be classified into four distinct fracture patterns: posteromedial (varus) impaction, lateral (valgus) impaction, isolated greater tuberosity or anteromedial impaction. This classification attempts to guide treatment decision-making as the valgus impaction pattern (that associates a fracture of the greater tuberosity and sometimes lesser tuberosity involvement) causes the worst outcome in terms of pain if conservative treatment is indicated and the posteromedial impaction pattern presents with poorer outcomes when range of motion is taken into account.

A specific group of fractures that require special attention are fracture-dislocations. When the surgical or anatomical neck of the humerus is damaged and the humeral head dislocates, there is a high risk of damage to the vascularization of the humeral head.

71.4.5 Clinical and Radiological Diagnosis

Patients frequently present to the emergency area with pain in the shoulder and the joint held in adduction and internal rotation, the elbow in flexion and the forearm held by the other hand. Clinical examination reveals fractures' cardinal symptoms: pain, swelling and decreased range of motion. Later on, inspection shows the Hennequin ecchymosis, a pathognomonic sign of proximal humerus fracture that extends from the affected arm to the forearm and chest. Anamnesis should include information on the dominance, functional requirements, occupation and comorbidities in order to guide the therapeutic management.

It is mandatory to assess the indemnity of the brachial plexus, especially the axillary nerve (evaluating the lateral shoulder sensibility and

the possibility to extend the shoulder) and the axillary artery. In addition, as in other locations, it is important to explore the nearest structures that can be injured during the trauma (for example, the scapula, the cervical spine, the ribs or the elbow).

Once a proximal humerus fracture is suspected, a scapular AP and Y view must be required to the X-ray department. The Velpeau's axillary view is often helpful. Except in the most simple, nondisplaced patterns, in which conservative treatment can be prescribed right away after X-ray examination, a CT scan is required almost always. This allows the clinician to precisely define the fracture's pattern (evaluating the fragments, especially the intra-articular ones, and its displacement), helping the surgeon in decision-making and, eventually, in preoperative planning.

71.4.6 Treatment

Proximal humerus fracture treatment is one of the most controversial topics in shoulder surgery. Often, the literature shows treatment options based on the Neer classification, but even Dr. Neer noted that his classification was developed to describe fractures, not to guide treatment.

For nondisplaced or minimally displaced (<5 mm) fracture patterns, conservative management should be the norm, even in younger individuals. On the other side, fracture-dislocations almost always require surgical treatment. For the rest of the fracture patterns, there is considerable discussion on which is the best treatment option, a decision that should be based not only on the fracture pattern but, most importantly, on the age, activity level and comorbidities of the person suffering from the injury. Older age by itself is the worst predictor of poor outcomes regardless of the type of treatment selected.

In general, surgical treatment should be considered for surgical neck fractures that are angulated >45° or present complete translation, tuberosity fractures with more than 1 cm displacement and more than 45° of valgus or varus malalignment.

Conservative treatment consists of the use of a simple sling for a period of 7–14 days. After 2 weeks, patients are encouraged to start pendulum and active motion exercises. After 6 weeks, progressive strength exercises are allowed. This treatment is recommended for nondisplaced fractures and patients with low functional demands irrespective of the fracture pattern.

Closed reduction and percutaneous pinning are indicated for surgical neck fractures after closed reduction. This is a technically demanding option that requires good bone quality, minimal metaphyseal comminution and intact medial calcar. The surgeon will place 3–4 K-wires (frequently, 2 from the lateral distal cortex and 1 from the superior greater tuberosity) that are left in place for 3 weeks; meanwhile, the patient has to use a sling. This procedure presents potential complications during the pinning, so caution has to be taken in order to avoid the axillary nerve or, less frequently, the cephalic vein, the musculocutaneous nerve and the biceps tendon.

Open reduction and internal fixation (ORIF) with a precontoured titanium plate with screws is the preferred method for 2-, 3- or 4-part fractures in young population. It is mandatory to reach an anatomical reduction before plate implantation besides controlling the head varus collapse by introducing an inferomedial screw and, if necessary, add an allogeneic fibula strut to maintain the medial support. Complication rates can be relatively high, and they include osteonecrosis, infection, screw protrusion or stiffness.

Intramedullary nailing (IMN) is an alternative option for surgical neck fractures, 3-part fractures in younger patients or combined proximal humerus and humeral shaft fractures.

Compared to ORIF, IMN presents inferior biomechanically outcomes, but favourable rates of fracture healing and range of movement with lower implant-related complications.

Shoulder arthroplasty is considered for complex fractures and fracture-dislocations. In elderly patients with low functional requirements who suffer from a fracture-dislocation or a fracture with non-reconstructible tuberosities and/or poor bone stock, reverse shoulder arthroplasty is a good indication. However, clinical outcomes are unpredictable but are improved if tuberosity healing is achieved.

In younger patients (<65 years) with head splitting components or fracture-dislocations with anatomical neck involvement and metaphyseal comminution, ORIF is expected to fail due to humeral head necrosis, and hemiarthroplasty must be considered. This is, probably, one of the most demanding shoulder surgeries because outcomes are subject to tuberosity reduction and healing and restoration of humeral height and version.

71.5 Scapulothoracic Dissociation

A scapulothoracic dissociation is a traumatic injury to the shoulder girdle that causes a disruption of the scapulothoracic joint. It is caused by high-energy trauma, is frequently associated with other serious injuries and can be life-threatening. There is damage to the acromioclavicular joint or clavicle (the osteoligamentous attachments of the arm to the body), the muscles of the shoulder girdle (trapezius, deltoid, both pectoralis, rhomboid …) and the axillary artery and brachial plexus. The physical exam shows swelling of the shoulder girdle and weakness, numbness and pulselessness in the upper extremity. Simple chest X-rays will characteristically show the osseous lesions and lateral displacement of the medial edge of the scapula. Once diagnosed, CT, magnetic resonance, angiography and electrodiagnostic testing might be necessary. Surgery in the emergency setting is reserved for bleeding of vascular injuries, but urgent stabilization of the osseous lesions is necessary, followed by management of the neurologic injury as needed, as neurologic injury will usually define the outcome.

Take-Home Message

- Fractures of the shoulder affect both young, active individuals and older adults.
- The clavicle is the most common broken bone in the body.
- Clavicle fractures can be classified according to their situation along the bone in midshaft, lateral and medial.
- Simple radiological imaging is often enough to ascertain the type of clavicle fracture.
- Nondisplaced fractures should be managed conservatively, but the management of displaced clavicle fractures depends on the characteristics of both the fracture and the patient.
- Scapula fractures are rare and are most commonly seen in young and middle-aged patients involved in high-energy trauma.
- 90% of patients with scapular fractures will present with another associated relevant injury.
- The management of scapula fractures is guided by the degree of displacement and the fracture pattern: nondisplaced fractures can all be managed conservatively, but displaced fractures might require open reduction and internal fixation.
- Fractures of the proximal humerus are common injuries that affect both young active individuals and elderly osteoporotic patients.
- A CT scan will precisely define a proximal humerus fracture's pattern helping the surgeon in decision-making.
- For nondisplaced or minimally displaced proximal humerus fractures, conservative management should be the norm.
- Surgical alternatives for proximal humerus fracture are varied: closed reduction and percutaneous pinning, open reduction and fixation with plates and screws, intramedullary nailing and arthroplasty.

Summary

Fractures of the shoulder affect both young, active individuals and older adults. In younger individuals, these fractures appear in the context of sport- or work-related injuries and sometimes due to high-energy trauma. In the elderly, there is a strong association between these fractures and osteoporosis.

The clavicle is the most common broken bone in the body, representing by itself up to 4% of all fractures in the adult population. Clavicle fractures can be classified according to their situation along the bone in midshaft, lateral and medial. The subject with a clavicle fracture is often easily diagnosed with swelling, haematoma and local crepitation, with variable degrees of deformity alerting the clinician of the general outline of the injury. Simple radiological imaging is often enough to ascertain the type of fracture. Nondisplaced fractures should be managed conservatively with the arm in a sling for 2–3 weeks. The management of displaced clavicle fractures is very controversial, depending on the characteristics of both the fracture and the patient. Despite these, there are some absolute indications for surgical treatment: open fractures, severe soft-tissue damage with risk of skin perforation, acute neurovascular injury and floating shoulder.

Scapula fractures are rare and are most commonly seen in young and middle-aged patients involved in high-energy trauma. 90% of patients will present with another associated relevant injury. The patient with a scapula fracture typically presents with local pain with the arm held in an adducted position. Some local swelling is present, but important ecchymosis is rare. Deformity of the contour of the shoulder develops in fracture that affects the acromion or scapular spine. If the fracture has been produced during a high-energy trauma, explicit attention should be given to look for associated injuries. A scapula CT scan is the preferred diagnostic tool. The management of scapula fractures is guided by the degree of displacement and the fracture pattern: nondisplaced fractures can all be managed conservatively with a protective sling worn for 1–2 weeks. For displaced fractures, the management is dictated by the fracture pattern and might require open reduction and internal fixation.

Fractures of the proximal humerus are common injuries that affect the metaphysis and epiphysis of the humerus. They present in both young active individuals and elderly osteoporotic patients. Patients frequently present to the emergency area with pain in the shoulder and the joint held in adduction and internal rotation, the elbow in flexion and the forearm held by the other hand. Clinical examination reveals fractures' cardinal symptoms: pain, swelling and decreased range of motion. Simple X-rays will allow for initial diagnosis, but a CT scan allows the clinician to precisely define the fracture's pattern helping the surgeon in decision-making. Proximal humerus fracture treatment is one of the most controversial topics in shoulder surgery. For nondisplaced or minimally displaced (<5–10 mm) fracture patterns, conservative management should be the norm, even in younger individuals. On the other side, fracture-dislocations almost always require surgical treatment. For the rest of the fracture patterns, there is considerable discussion on which is the best treatment option, a decision that should be based not only on the fracture pattern but, most importantly, on the age, activity level and comorbidities of the person suffering from the injury. Older age by itself is the worst predictor of poor outcomes regardless of the type of treatment selected. Surgical alternatives are varied: closed reduction and percutaneous pinning, open reduction and fixation with plates and screws, intramedullary nailing and arthroplasty are alternatives that should be considered.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Which one of these is NOT a clear indication for surgical management of a midshaft clavicle fracture?
   (a) Open fractures
   (b) Simple oblique fracture with 3 mm displacement
   (c) Severe soft-tissue damage with risk of skin perforation

(d) Acute neurovascular injury
(e) Associated displaced glenoid fracture (floating shoulder)

2. With respect to the management of midshaft clavicle fractures, which one of the following is FALSE?
   (a) Nondisplaced fractures of the clavicula should be managed conservatively
   (b) The management of displaced clavicle fractures depends on the characteristics of both the fracture and the patient
   (c) Most, if not all, displaced fractures of the midshaft clavicle should be treated with intramedullary nailing
   (d) The complications of conservative management are increased in the presence of comminution, severe displacement and smoking habit
   (e) Surgical treatment reduces the non-union rate (to 1–2%, possibly requiring further surgery) and shortens the recuperation time
3. Regarding the management of scapula fractures, which of the following sentences is FALSE?
   (a) Nondisplaced fractures can all be managed conservatively
   (b) Acute isolated displaced fractures of the acromion, coracoid and spine with significant displacement (>1 cm) should be managed with open reduction and internal fixation
   (c) All body fractures should be managed conservatively regardless of the displacement observed
   (d) Displaced glenoid fractures are considered always for open reduction and rigid internal fixation
   (e) Some displaced glenoid fractures can be managed with interfragmentary compression screws with arthroscopic assistance in subjects with smaller fragments
4. Regarding proximal humerus fractures, which of the following sentences is FALSE?
   (a) They represent 4–6% of all fractures
   (b) They are the third most common non-vertebral osteoporotic fractures in older adults after hip and distal radius fractures
   (c) They occur twice as often in men as in women
   (d) In the elderly, these are often low-energy fractures, occurring by an indirect mechanism when the subject falls from her own height
   (e) In the younger population, high-energy trauma (traffic accidents, convulsions, sport-related trauma …) is the usual mechanism
5. Regarding the management of proximal humerus fractures, which of the following sentences is FALSE?
   (a) Classically, proximal humerus fractures have been classified using the Neer classification
   (b) The valgus impaction pattern (that associates a fracture of the greater tuberosity and sometimes lesser tuberosity involvement) causes the worst outcome in terms of pain
   (c) When the surgical or anatomical neck of the humerus is damaged and the humeral head dislocates, there is a high risk of damage to the vascularization of the humeral head
   (d) It is mandatory to assess the indemnity of the brachial plexus, especially the axillary nerve
   (e) A CT scan is rarely needed for most displaced fractures

Further Reading

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Part XIII

Elbow and Wrist

72 Osteoarthritis, Tendinitis, Bursitis of the Olecranon, Biceps Detachment

Sheraz Malik and Chris Peach

Overview
This chapter provides a general overview of the most common elbow problems encountered in orthopaedic practice. Each section covers the significant points of the underlying pathology, important clinical features and management options. The chapter also highlights indications for surgical interventions.

72.1 Elbow Arthritis

The two main causes of elbow arthritis are rheumatoid arthritis and osteoarthritis.

72.1.1 Rheumatoid Arthritis

This is the most common cause of elbow arthritis. The prevalence of rheumatoid arthritis is approximately 2% in the population with female-to-male ratio of 3:1. 50% of patients with rheumatoid arthritis develop elbow arthritis, with the vast majority affected bilaterally. As with all joints affected by rheumatoid arthritis, the clinical course involves symptom flares followed by periods of remission.

Patients can develop rheumatoid nodules over the olecranon, which are painful hard lumps, as well as more generalised symptoms of synovitis and swelling. The main problem in the rheumatoid elbow is pain, but movements may also become restricted, causing profound functional limitation, and in severe cases, the elbow may become unstable. Often due to the associated functional disturbance in the hands, this elbow stiffness significantly affects the patient's upper limb function even though the patient might have previously adapted well to their hand and finger deformities.

On the lateral aspect of the elbow, inflamed synovium produces tenderness over the radial head, and on the medial aspect, synovial swelling may cause ulnar nerve compression. Involvement of the ulnohumeral joint leads to restriction in flexion and extension, whereas involvement of the proximal radioulnar joint may affect supination and pronation. In long-standing disease, laxity of soft tissues and destruction of articular surfaces result in joint instability.

Radiographs show periarticular osteopenia and bone erosions, often affecting the greater sigmoid notch of the ulna. The radial head may be deformed.

The mainstay of treatment is medical management with disease-modifying anti-rheumatic

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drugs (DMARDs) and non-steroidal anti-inflammatory medications (NSAIDs). Intra-articular steroid injections can provide temporary local pain relief, and custom-made splints may be helpful in acute flare-ups. Injections are administered into the soft spot of the elbow at a point between the lateral epicondyle, radial head and olecranon process. Physiotherapy is important to encourage joint movement and maintain muscle power. The advancements in medical management have reduced the need for surgical interventions. Surgical management involves procedures such as arthroscopic synovectomy, radial head excision and total elbow replacement. Total elbow replacement is suitable for older, low-demand patients who have exhausted other management options.

72.1.2 Osteoarthritis

Symptomatic osteoarthritis of the elbow is much less common than rheumatoid arthritis. Secondary osteoarthritis is more prevalent than primary osteoarthritis. It is seen in younger patients and develops due to an underlying joint pathology, such as previous fractures or dislocations (i.e. post-traumatic arthritis), septic arthritis, osteochondral defects, inflammatory arthritis or gout. Primary degenerative osteoarthritis is seen in middle-aged patients and is more common in males than females. It typically affects the dominant limb of individuals in occupational groups exposed to repetitive mechanical loading, e.g. farmers, builders and professional sportspersons.

The main problem in elbow osteoarthritis is loss of range of movement, especially extension. This mechanical block results from contracture of joint capsule and presence of osteophytes. Pain is present at extremes of motion. Ulnar nerve neuropathy may result from encroaching osteophytes.

Classical radiological findings in osteoarthritis are loss of joint space, periarticular osteophytes and subchondral cysts and sclerosis. Loose bodies may also be detected on radiographs, but these are usually better demonstrated on a CT scan. In secondary osteoarthritis, signs of the underlying pathology (e.g. prior fracture implants) may be evident as well.

Treatment is aimed at symptom control and consists of analgesia according to the analgesic ladder and activity modification. The clinical course consists of periods of exacerbation and remission. Surgery is indicated when medical management fails to control symptoms and quality of life is impaired. Surgical options are the following:

- Arthroscopic osteocapsular release, debridement of osteophytes and release of capsular contractures and removal of loose bodies.
- Outerbridge-Kashiwagi procedure (OK procedure), open debridement of osteophytes and capsular release.
- Ulnar nerve decompression +/− transposition.
- Total elbow replacement is considered for carefully selected patients with end-stage arthritis. After total elbow replacement, patients need to limit the use of arm to light activities only. It is contraindicated in young and high-demand patients due to high risk of loosening, the most common cause of revision surgery. Other complications include infection, nerve injury and periprosthetic fracture.

72.2 Elbow Tendinopathy

The most common cause of elbow pain is tendinopathy, namely lateral and medial epicondyloses. Elbow movements are typically unaffected in both conditions. Patients with long-standing symptoms may however have reduced grip strength.

72.2.1 Lateral Epicondylosis

Lateral epicondylosis, or tennis elbow, is chronic symptomatic degeneration, i.e. tendinosis, of the common extensor origin at the lateral epicondyle. This overuse condition usually affects patients between the ages of 35 and 55 years with an equal gender distribution and is seen more commonly in the general population than in tennis

players. Typically, the extensor carpi radialis brevis (ECRB) tendon is most affected.

Lateral epicondylosis was previously thought to arise from an acute inflammation of the tendon, i.e. tendinitis; however, the findings of histopathological studies demonstrate that actually relatively few inflammatory cells such as macrophages and neutrophils are present in the affected tendon tissue. The current consensus is that repetitive wrist extension and forearm rotation lead to microtears in the ECRB tendon, and the subsequent proliferation of fibroblasts and myofibroblasts and formation of vascular granulation tissue disrupt the ordinary arrangement of tendon. This degeneration and repair process is termed angiofibroblastic hyperplasia [1].

Patients complain of pain directly over or just distal to the lateral epicondyle. The pain is often exacerbated on activities such as typing, handling tools and playing racquet sports. On examination, patients have point tenderness 1–2 cm distal to the lateral epicondyle. Symptoms are reproduced on resisted wrist extension with the elbow extended and forearm pronated. Plain xray films are usually normal, but in some cases may show calcification at the common extensor tendon origin. Ultrasound or MRI scans can be used to confirm the diagnosis if there is history of trauma or associated mechanical symptoms, such as locking. Other causes of lateral elbow pain include lateral ulnar collateral ligament injury, radial tunnel syndrome, arthritis and osteochondritis dissecans. Tennis elbow rarely occurs in young or old patients, and differential diagnoses such as osteochondritis dissecans or osteoarthritis should be considered, respectively.

Lateral epicondylosis is generally self-limiting and spontaneously resolves within 18 months in 90% of cases. Symptoms are usually adequately managed with activity modification, NSAIDs and physiotherapy with eccentric loading exercises of extensor muscle mass. Steroid injections are used with caution as they can adversely affect the natural history of the condition. Platelet-rich plasma injections (PRP) are becoming a popular treatment, although evidence for their routine use is lacking. Surgery is considered if the symptoms persist despite adequate conservative treatment for 12 months. The surgical options include excision of the degenerative ECRB tissue, common extensor origin release or arthroscopic ECRB release [2].

72.2.2 Medial Epicondylosis

Medial epicondylosis, or golfer's elbow, is tendinosis of the flexor-pronator origin at the medial epicondyle. The pathophysiology is that repetitive stress loading causes degenerative changes (i.e. microtears, which fail to heal normally, resulting in granulation tissue formation) in the pronator teres tendon, leading to pain. It affects the same age group of patients as tennis elbow, and also with equal gender distribution, but the incidence of medial epicondylosis is five times less than lateral epicondylosis.

Although popularly associated with playing golf, it is more common in sportspersons who repetitively throw, e.g. baseball pitchers, bowlers and javelin throwers, a motion in which usually the wrist is flexed and forearm pronated, resulting in overuse injury of the flexor-pronator muscle group. Medial epicondylosis is also seen in athletes participating in racquet sports, weightlifting and archery, and workers lifting heavy loads or exposed to constant vibration at the elbow, e.g. construction workers, plumbers and carpenters. The dominant limb is affected in most of the cases.

Patients complain of pain at or around the bony prominence of the medial epicondyle, exacerbated on activities such as golf swings, throwing and gripping tools. Tenderness is elicited on palpation of the pronator teres tendon, just anterior and distal to the medial epicondyle. The most effective test is with resisted elbow and wrist flexion and resisted forearm pronation. This can be examined by getting the patient to bring their hand towards their face as if washing and the examiner providing resistance on the radial border of the hand, causing medial elbow pain. Some patients may also have features of associated ulnar neuropathy, which is caused by compression from flexor-pronator origin swelling from tendinopathy. Plain films are usually normal, and if there is history of a preceding injury, ultrasound and MRI scan can be useful to exclude damage to underlying structures, e.g.

ulnar collateral ligament. Beware of medial collateral ligament pathology, however, in throwing athletes who present with symptoms mimicking medial epicondylosis.

Most patients with medial epicondylosis respond to conservative management with activity modification, NSAIDs and physiotherapy. A wrist splint to limit wrist flexion may also prove helpful. Surgery is rarely required and involves debridement, repair and, if needed, reattachment of flexor-pronator tendon origin.

72.3 Olecranon Bursitis

The olecranon bursa is a smooth sac between the skin and the underlying olecranon process at the posterior aspect of the elbow (Fig. 72.1). It is an extra-articular and a relatively superficial structure, which allows the skin to move without restraint over the bony prominence. The normal bursa is small, but it may become inflamed and accumulate fluid. The olecranon bursa is in fact the most common cause for an elbow swelling. The commonest trigger for olecranon bursitis is repetitive microtrauma—this has been given various pseudonyms, e.g. 'student's elbow', 'miner's elbow' and 'plumber's elbow'. Other causes include acute trauma, gout, calcium pyrophosphate deposition and rheumatoid arthritis. The olecranon bursa is also prone to infection via a transcutaneous route, with *Staphylococcus aureus* being the most common causative organism. This can be due to a skin abrasion, puncture wound or iatrogenic causes including injections. In the context of infection, the disorder is referred to as an infected olecranon bursa.

The diagnosis of olecranon bursitis is clinical, and treatment is conservative. Patients usually present with a notable fluctuant swelling at the back of the elbow. A careful enquiry may reveal clues as to the underlying cause. The condition usually responds to a brief period of activity modification, elbow protection and NSAID treatment. Recurrent olecranon bursitis may be treated with surgical excision, but wound healing can be a problem and is a considerable surgical risk. Differential diagnoses include acute injuries such as triceps tendon rupture and olecranon fracture. The elbow may also become markedly swollen and bruised in the days after a fracture of the humerus, due to the fracture haematoma tracking distally under the influence of gravity.

A history of fever, marked tenderness and overlying cellulitis suggest infected olecranon bursa. Elbow movements are typically preserved, as the bursa does not communicate with the elbow joint. This is treated with antibiotics, but if the patient is systemically unwell, incision, drainage and debridement of bursa are performed, and

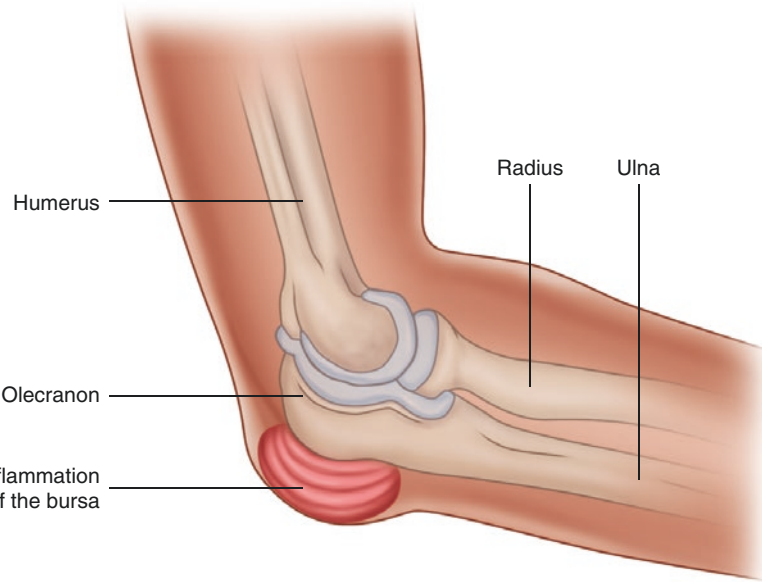

Fig. 72.1 Olecranon bursitis

the tissues are allowed to heal by secondary intention. A universally hot and painful joint, loss of elbow movements and a septic patient are more indicative of septic arthritis of the elbow. This requires urgent washout in theatre [3]. Olecranon bursitis and infected olecranon bursa are uncommon in children, whereas septic arthritis of the elbow is a more likely differential diagnosis in this age group.

72.4 Biceps Rupture

The distal biceps tendon inserts onto the radial tuberosity. The biceps is an important functional muscle of the upper limb and is a strong forearm supinator and weak elbow flexor. It can be affected by a broad spectrum of pathology, ranging from bicipitoradial bursitis and tendinopathy through to partial and complete tendon rupture.

Rupture of distal biceps tendon is seen almost exclusively in males, usually between the ages of 30 and 60 years, and mostly in the dominant arm. The main mechanism of injury is eccentric loading of biceps when elbow at 90 degrees of flexion during a lifting activity experiences a sudden extension force. In other words, the biceps is contracting fully under load and then is lengthened by elbow extension, thus exceeding the strength of the tendon insertion, resulting in rupture of the tendon from the bone. Aetiological factors that contribute to tendon rupture include hypovascularity of tendon, intrinsic tendon degeneration, mechanical impingement between the radial tuberosity and ulna and bicipitoradial bursitis. Manual work, weight training and anabolic steroids are known risk factors.

Patients report a painful snap at the front of the elbow during a strenuous activity. Weakness in supination is more noticeable, e.g. when using a screwdriver or turning door handles, whereas flexion power is less affected, since the intact brachialis is the main flexor of the elbow. There may be a change in the contour of biceps muscle, and ecchymosis over the medial aspect of the elbow and forearm. The Hook test is diagnostic and performed by asking the patient to look at the palm of the affected side, which produces shoulder and elbow flexion and forearm supination. The examiner then tries to hook the index finger around the biceps tendon from the lateral side of the elbow, and the test is abnormal if this is not possible. Hook test can be normal in partial tears. If there is any doubt, prompt ultrasound scan or MRI can be used to confirm the diagnosis, although this is a clinical diagnosis in the majority of patients.

Partial tears are usually treated conservatively, whereas most complete ruptures are fixed to restore strength and function. The operation is best performed within a few weeks from injury. Conservative treatment for complete ruptures leads to long-term weakness in supination and pain but can be appropriate for older, low-demand individuals.

Bicipitoradial bursitis and distal biceps tendinopathy are less common and have less male predilection than distal biceps tendon rupture. Patients present with deep-seated pain at the front of the elbow and forearm. Fluid accumulation around the course of the biceps tendon may result in swelling in the antecubital fossa. Pain may increase with resisted forearm supination or elbow flexion, leading to weakness. The diagnosis may be confirmed with ultrasound or MRI. If adequate conservative treatment with NSAIDs and physiotherapy for 6 months fails to improve symptoms, low-dose steroid injection into the bursa or around the tendon is effective. More resistant cases can be treated with endoscopic debridement of area of tendinosis or primary repair of the tendon usually providing good outcomes.

Take-Home Message

- Elbow tendinopathies, including tennis and golfer's elbow, are self-limiting and rarely require surgical intervention. Biceps tendon ruptures are best treated in the first 3 weeks following injury and therefore require prompt referral.

Summary

The elbow is an important structure with a significant contribution to the function of the upper limb by positioning the hand in space. The knowledge of anatomy, common symptoms and signs is normally enough to formulate a considered list of differential diagnoses. Clinical examination can be complex; however, if this is carried

out bearing in mind the anatomical location of the pathological processes, this can aid the clinical decision-making often without further specialised imaging being necessary.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. What are the main clinical signs of the rheumatoid elbow?
   (a) There will often be clinical manifestations of the disease elsewhere, including hand deformities, rheumatoid nodules over the ulna/olecranon area and history of joint replacement in the lower limbs
   (b) There will often be specific clinical manifestations of the disease in some locations, including feet, knee and cervical spine
   (c) The presence of systemic symptoms such as fatigue and weight loss are usually not present
   (d) Patients might experience morning stiffness and joint swelling, especially in the late stages of the disease
2. What are the most common operations for elbow arthritis prior to joint replacement?
   (a) Operative intervention prior to joint replacement aims to address symptoms caused by factors other than the severe joint degeneration
   (b) Surgical intervention with arthroscopy is always required in the early stages of the disease to prevent progression
   (c) Synovectomy, a surgical procedure to remove inflamed synovial tissue, is ever performed to alleviate symptoms
   (d) Osteotomy, a surgical procedure to lenghtening the bones, can be considered to redistribute joint forces
3. What are the red-flag signs to look out for in tennis elbow?
   (a) The red-flag signs to look out for are patients presenting with symptoms of tennis elbow when they are either young or old. These patients rarely have tennis elbow and for young patients they often have osteochondritis dissecans and in the more elderly population they have arthritis
   (b) The red-flag signs are inflammation signs in older patients and paraesthesia in young patients
   (c) Fever
   (d) Osteoporosis
4. Can a distal biceps rupture be treated conservatively?
   (a) Although patients are not immediately disabled after the injury, if the patient relies on supination as part of their work or sport, they will have significant ongoing symptoms
   (b) Distal biceps rupture requires urgent surgical repair
   (c) Never
   (d) Conservative treatment is the only management possible
5. Should a swollen olecranon bursa be aspirated or injected with steroid?
   (a) Most olecranon bursal swellings are due to aseptic causes, and the natural history is that it resolves spontaneously. Infection of a swollen olecranon bursa is frequently iatrogenic and can be caused by passage of a needle into it. Therefore, in this naturally resolving condition for the majority, it is essential to reassure the patient that time will heal the swelling rather than resort to interventions that can make the situation worse
   (b) Most olecranon bursal swellings are due to septic causes, and the natural history is that they do not resolve spontaneously. Therefore, it is essential to perform a surgical treatment
   (c) Aspiration is avoided
   (d) Injection with a steroid can worsen the symptoms

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Adult Elbow: Fractures

73

Marc B Gibson and Philip M Ahrens

Overview
The management and diagnosis of adult elbow fractures is complex and deserves careful attention to detail. Meticulous clinical examination and appropriate radiological investigations are critical to an accurate delineation of the fracture pattern and damage to any neurological structures and associated soft-tissue stabilizers of the elbow.

73.1 Fractures of the Distal Humerus

73.1.1 Definition + Epidemiology

- Distal humerus fractures account for 2% of all fractures and represent one-third of all elbow fractures.
- Occur predominantly in young males, and females in their sixth decade of life associated with osteoporosis.

73.1.2 Pathogenesis

- In the younger population, fractures usually result of a high-energy impact. In elderly, often from low-energy falls.
- Elbow flexed <90° at the time of injury leads to a transcolumnar fracture pattern due to axial loading.
- Elbow flexed >90° tends to result in an intercondylar fracture.

73.1.3 Classifications

- Classified according to location and extent of the fracture:
  - Supracondylar fractures
  - Single-column fractures *(Milch classification)*
  - Both column fractures
- *Milch classification* of single-column fractures (NB. lateral condyle fractures are more common than medial condyle fractures):
  - Type I: the lateral trochlear ridge remains intact.
  - Type II: involves a fracture through the lateral trochlear ridge; the elbow may dislocate!
- *AO classification* of distal humerus fractures. The importance with intra-articular fractures is the requirement for good fracture reduction and stable fixation.

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73.1.4 Diagnosis (Clinical Features and Imaging)

- Clinically present with pain, swelling, and deformity. Compartment syndrome may occur as part of the clinical presentation. Avoid range-of-motion examination due to the risk of neurovascular damage.
- Check radial, ulnar, and median nerve function. Distal pulses must also be checked due to possible damage of the brachial artery.
- Imaging must include plain radiographs with anterior-posterior (AP) and lateral views.
- Computerized tomography (CT) is often obtained for accurate fracture delineation and surgical planning.

73.1.5 Treatment

- *Nonsurgical*. Nondisplaced fractures should be immobilized for 2 weeks in a cast and then early gentle elbow mobilization to prevent stiffness. Immobilize nondisplaced lateral condyle fractures in forearm supination and medial condyle fractures in pronation. Elderly low-demand patients where risks of surgery remain high and nonoperative treatment should be considered.
- *Closed reduction and percutaneous pinning (CRPP)*: Displaced condyle fractures (Milch type I) may be treated with closed reduction and percutaneous pinning. *NB. Milch type II fractures require open reduction and internal fixation.*
- *Open reduction and internal fixation (ORIF)*: Displaced supracondylar and both column fractures require open reduction and internal fixation, often with double plating (lateral plate for the medial column and a posterior plate for the lateral column). Access the elbow via a posterior approach, then perform either an olecranon osteotomy, triceps split (*Campbell*), or a triceps-sparing exposure with elevation of the triceps to access the fracture.
- Extra-articular fractures require reduction and fixation of both columns to the epiphysis. When treating intra-articular fractures, it is necessary to anatomically reduce and fix the distal articulating epiphysis first. The reconstructed epiphysis is then fixed to both columns.
- *Arthroplasty*: In highly comminuted or both column fractures in elderly osteoporotic, poorly compliant, or low-demand patients, it is recommended to perform a cemented total elbow arthroplasty.
- *External fixation*: Comminuted open fractures with significant soft-tissue damage may require an external fixator prior to definitive internal fixation.

73.2 Fractures of the Capitellum

73.2.1 Definition + Epidemiology

- A coronal fracture (purely intra-articular) of the distal humerus at the capitellum
- Rare; accounts for only 1% of all elbow fractures
- Often missed due to subtle radiographic changes

73.2.2 Pathogenesis

- Typically result from a low-energy fall onto an outstretched arm. With the elbow in a semi-flexed position, the axial compressive force through the radial head creates a shear force at the capitellum resulting in a fracture.
- Due to the lack of soft-tissue attachments, avulsion fractures do not occur here.
- Occasionally associated with injury to the radial head and/or lateral collateral ligament.

73.2.3 Classification

- *Bryan and Morrey classification* with McKee modification (Fig. 73.1):
  - Type I: large bony fragment
  - Type II: shear fracture of articular cartilage

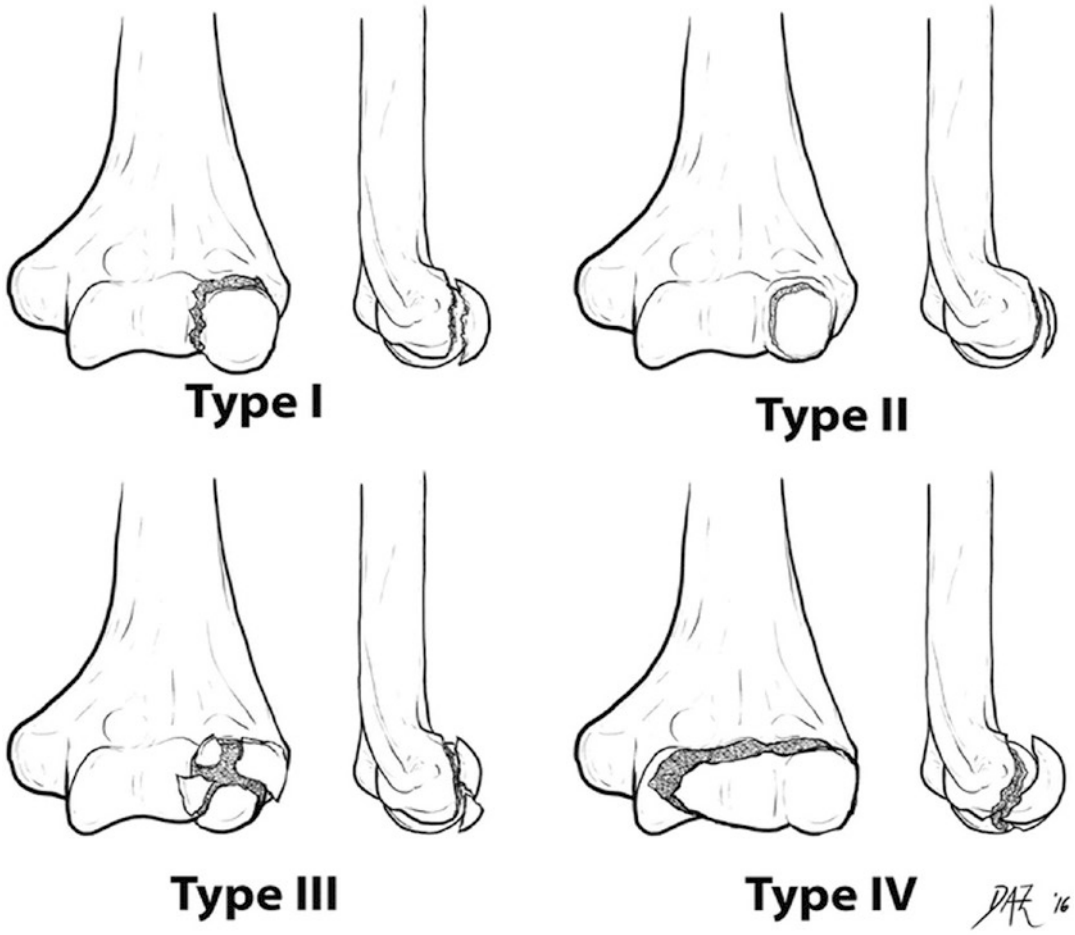

Fig. 73.1 Modified Bryan and Morrey classification

- Type III: comminuted fracture
- Type IV: fracture involves trochlea

73.2.4 Diagnosis

- Present with elbow pain in addition to lateral tenderness, swelling, and ecchymosis. Patients may have a mechanical block to flexion/extension and, less often, rotation. Elbow joint instability may also be apparent when an associated medial collateral ligament injury (medial tenderness) has occurred.
- Neurovascular exam must be performed in all cases.
- AP and lateral radiographs are recommended, but as the fracture fragments consist mainly of cartilage, X-rays often do not reveal the true size of the fracture fragment. The lateral radiograph is key to diagnosis showing the "double-arc" sign. Careful assessment of the radial head must also be performed during radiography.
- CT may provide more accurate information on the size and displacement of the fracture, therefore assisting the treatment plan.

73.2.5 Treatment

- *Nonsurgical:* Nondisplaced fractures (<2 mm displacement) should be splinted for 2–3 weeks and then gentle range of motion to prevent elbow stiffness.
- *ORIF:* Displaced fractures >2 mm require fixation. Access the capitellum via a lateral

elbow approach. Reduce and fix the fragment with two posterior-to-anterior headless screws to avoid penetration of the articular surface. A posterior approach may be indicated in cases associated with medial elbow injuries or distal humeral fractures.
- *Excision:* If the fracture fragment(s) is comminuted and unsalvageable, then fragment excision is recommended followed by early joint motion.
- *Arthroplasty:* For unreconstructable fractures in elderly patients with associated medial column instability, total elbow replacement is recommended.

73.3 Fractures of the Radial Head and Neck

73.3.1 Definition + Epidemiology

- Account for 2% of all fractures. Among the most common (33%) of all elbow fractures.

73.3.2 Pathogenesis

- Commonly result from a fall on an outstretched elbow with the hand in supination.
- Majority are minimally displaced isolated injuries with a very good prognosis.
- If part of an elbow dislocation injury, may also involve a fracture of the coronoid; then termed *Terrible Triad Injury*.
- When associated with disruption of the interosseous membrane and the distal radioulnar joint (DRUJ), this is termed an *Essex-Lopresti injury* or *Longitudinal radioulnar dissociation (LRUD).*
- When association with a fracture of the proximal ulna, this is termed a *Monteggia injury.*

73.3.3 Classifications

- *Johnston modified Mason classification* (Fig. 73.2):
  - Type I: nondisplaced fractures; no block
  - Type II: displaced fractures; limited motion
  - Type III: comminuted fractures; motion block
  - Type IV: associated with an elbow dislocation

73.3.4 Diagnosis

- Clinical features include immediate pain and swelling along the lateral elbow. Localized tenderness over the radial head, just distal to the lateral epicondyle. Forearm rotation is painful or limited (blocked) and associated with crepitus.
- If forearm pronation and supination are full after intra-articular injection of local anesthesia, nonoperative treatment is indicated.
- Physical exam should include assessment of elbow stability and DRUJ integrity.
- AP and lateral radiograph views are recommended. For subtle fractures, perform a radiocapitellar view which allows clearer visualization of the radial head without coronoid overlap.
- Wrist radiographs should also be performed if wrist pain is present acutely.

73.3.5 Treatment

- *Nonsurgical.* Type I nondisplaced fractures or minimally displaced fractures (less than 2 mm) with no block to forearm rotation only require a soft supporting arm sling for no more than 7 days, followed by early mobilization. After 2–3 months, one should expect good to excellent function.
- *ORIF:* For fractures with a mechanical block that are displaced (greater than 2 mm) and reconstructable, ORIF of fracture fragments is required with small interfragmentary compression screws. Access the radial head via a lateral (Kocher or Kaplan) approach.
- *Fragment excision*: When a fracture fragment(s) lies inside the joint and blocks motion, then isolated fragment excision is required, usually via a mini-open lateral approach.

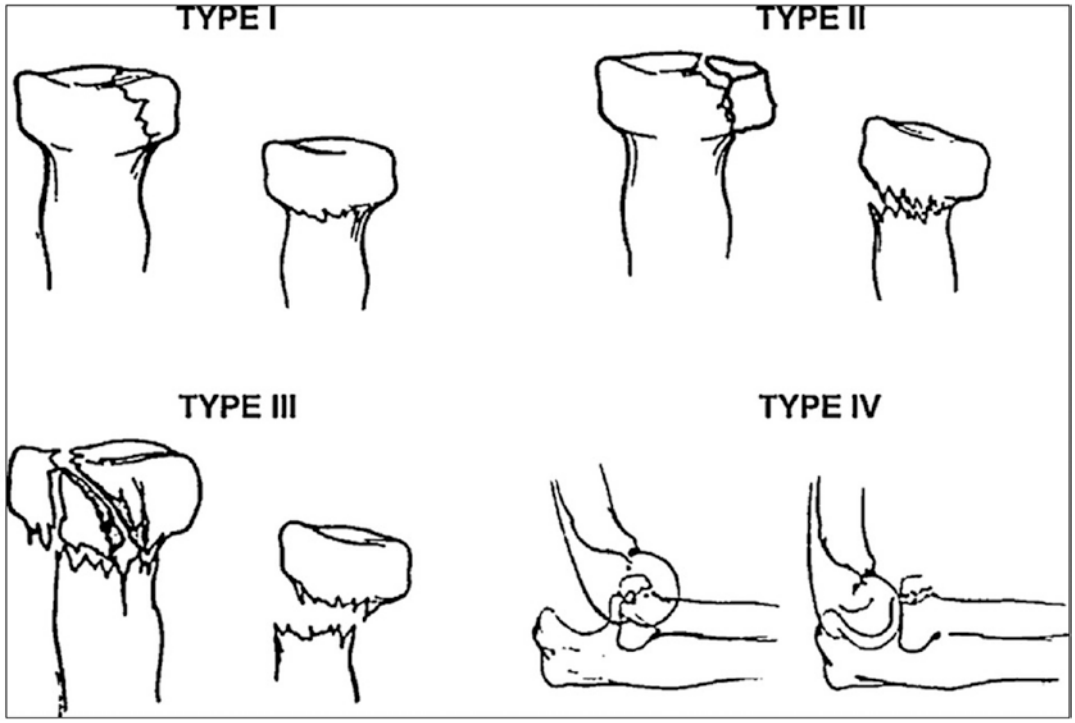

Fig. 73.2 Modified Mason classification

- *Head excision*: Unreconstructable painful comminuted radial head fractures in elderly low functional demand patients that exhibit a block to forearm rotation are best treated with excision of the radial head. Removal of the head is performed at the level of the annual ligament via a lateral elbow incision. Intraoperative elbow stability must be assessed, and collateral ligament reconstruction performed if necessary.
- *Arthroplasty*: Radial head replacement should be considered in younger patients if the head is unreconstructable (Type III fractures) and elbow stability remains a concern; this is often the case in fracture-dislocations (Type IV fractures) where the lateral collateral ligaments or interosseous membrane of the forearm is also damaged. The accepted choice of implant is an uncemented metallic radial head.

73.4 Fractures of the Olecranon

73.4.1 Definition + Epidemiology

- Account for 10% of all adult elbow fractures.
- All fractures have an intra-articular component.
- Bimodal distribution; high-energy injuries in young patients, and low-energy falls in the elderly.
- Due to the subcutaneous position of the proximal ulna, the olecranon is vulnerable to direct trauma.

73.4.2 Pathogenesis

- Commonly result from a fall directly onto the posterior aspect of the elbow. Impaction of the distal humerus splits the olecranon at its narrowest point.
- Nondisplaced fractures are rare due to the strong pull of the triceps muscle.

73.4.3 Classifications

- *Mayo classification*
  - Type I: nondisplaced fracture (<2 mm)
  - Type II: displaced but stable elbow (A: noncomminuted, B: comminuted)
  - Type III: displaced with elbow instability (A: noncomminuted, B: comminuted)

73.4.4 Diagnosis

- Present with a palpable gap at the proximal ulna (fracture site) and an inability to actively extend the elbow indicating discontinuity of the triceps mechanism.
- Careful neurological evaluation is required, as ulnar nerve injures may accompany these fractures.
- AP and true lateral radiographs are essential for determining the fracture pattern.

73.4.5 Treatment

- *Nonsurgical:* Nondisplaced fractures may be treated in a cast for 2 weeks at 80° of elbow flexion and then through protected range-of-motion exercises avoiding flexion past 90°. In case of fractures in elderly low-demand patients in whom surgery is associated with high risks, nonsurgical management often achieves acceptable clinical outcomes.
- *Tension band wiring*: Simple transverse fractures may be treated using this method. Many patients require subsequent hardware removal due to protrusion and pain, if using metalwork (Kirschner wires).
- *ORIF:* Dorsal hook plate and screw fixation is preferred in comminuted or distal oblique fractures.

73.5 Fractures of the Coronoid Process

73.5.1 Definition + Epidemiology

- Fracture almost exclusively associated with elbow dislocations/instability (*see Sect. 73.6*)
- Isolated fractures very uncommon

73.5.2 Pathogenesis

- During elbow dislocation, the distal humerus slides anterior impacting the coronoid process causing it to fracture.
- Associated conditions include posteromedial rotatory instability, posterolateral rotatory instability, and *Terrible Triad* injuries (coronoid fracture plus radial head fracture plus elbow dislocation).

73.5.3 Classification

- *Regan and Morrey* classification
  - Type I: tip of coronoid fracture
  - Type II: <50% of coronoid height fracture
  - Type III: >50% of coronoid height fracture

73.5.4 Diagnosis

- Present almost exclusively following elbow dislocation.
- Plain radiography interpretation can be difficult due to overlapping structures.
- CT is best for classifying injuries and guiding treatment.

73.5.5 Treatment

- *Nonoperative.* Nondisplaced fractures of the coronoid with a stable elbow may be managed with a brief period of immobilization then early range of motion.
- *ORIF (suture fixation):* Small lesions (Types I and II) with elbow instability fixed with transolecranon sutures are adequate.
- *ORIF:* Some Type II and III fractures require fixation with retrograde screws or a plate to restore elbow stability.
- *External fixator:* In patients with poor bone quality or very unstable elbow joints, a hinged external fixator will provide stability while allowing early motion of the elbow.

73.6 Dislocation of the Elbow

73.6.1 Definition + Epidemiology

- Second most common major joint dislocation (after shoulder).
- Most common dislocated joint in children.
- Posterior most common type of acute dislocation (80%) involves both the radius and ulna, which move posterior in relation to the humerus.
- Predominantly affects patients aged 10–20 years.

73.6.2 Pathogenesis

- Usually results from a fall on the outstretched arm (axial load) with the elbow in almost full extension and forearm supinated.
- As the elbow continues to hyperextend and exert a valgus stress, a resultant anterior and medial force is generated putting the anterior capsule and collateral ligaments (soft-tissue stabilizers) under tension which then fail. The key bony stabilizers of the elbow joint (radial head and coronoid process) are also put under excessive force as the humerus slides anterior; therefore these structures commonly fracture in elbow dislocations.
- When elbow dislocation presents with a fracture of the coronoid and radial neck, this is termed a *Terrible Triad injury.*

73.6.3 Classification

- Anatomic description; position of the ulna relative to the humerus:
  - Posterior (NB. posterolateral most common)
  - Anterior
  - Lateral
  - Medial
  - Divergent (ulnar and radius move apart)
- Simple vs. complex
  - Simple: dislocation with no associated fracture
  - Complex: dislocation with associated fracture *(e.g. Terrible Triad injury)*

73.6.4 Diagnosis

- Severe pain, swelling, deformity, and inability to move the elbow joint.
- Olecranon process unduly prominent due to the posterior position of the forearm.
- Careful assessment of neurovascular status as nerve injuries frequently occur.
- Plain radiographs remain a very sensitive diagnostic tool, which also illustrate associated fractures.
- CT scaning is performed to assess complex injury patterns and guide treatment.

73.6.5 Treatment

- Closed reduction of the posterior dislocation, under sedation or anesthesia, can be achieved with longitudinal traction with the elbow in slight flexion. In rare circumstances, surgical exploration might be required if interposed fragments or soft tissues within the joint prevent joint relocation.
- Following reduction, immobilize the elbow in 90° of flexion with the forearm supinated. Aim not to immobilize the elbow longer than 7 days to avoid severe stiffness.
- If an associated radial head and/or coronoid fracture is present (e.g. *Terrible Triad injury*), appropriate fixation, as described earlier in this chapter, is required to stabilize the elbow acutely.
- During open bony fixation (ORIF), assessment and repair of the posterolateral/collateral ligament complex is also paramount to maintain elbow stability.
- If patients' elbow remains unstable despite fixation, then an external hinged fixator may be required for a period of time until healing of the soft-tissue stabilizers is achieved.

Take-Home Message

- Limb neurologic status must be assessed in all cases at the time of initial review.
- CT scaning is recommended in complex injuries as plain radiography often under reports fracture displacement and comminution.
- Capitellum fractures are often missed on plain radiographs.
- 35% of radial head fractures are associated with a concomitant soft-tissue or skeletal injury (e.g. elbow dislocation, Essex-Lopresti injury, Monteggia injury).
- With elbow dislocations, there is always an associated soft-tissue or bony injury.
- Fracture of the coronoid is almost always associated with an elbow dislocation.
- Displaced articular fractures require accurate reduction and internal fixation.
- Arthroplasty surgery is reserved more for elderly low-demand patients.
- Poorly managed radial head or neck fractures may result in painful restricted forearm rotation.
- Early mobilization is a key factor in preserving range of movement.

Summary

The management and diagnosis of adult elbow fractures is complex and deserves careful attention to detail. Meticulous clinical examination and appropriate radiological investigations are critical to an accurate delineation of the fracture pattern and damage to any neurological structures and associated soft-tissue stabilizers of the elbow. Regardless of the treatment strategy, elbow fractures are particularly vulnerable to post-injury complications, especially stiffness. Early mobilization following conservatively or surgically treated elbow injuries is a key factor.

Radial head injuries present most commonly and fortunately have a very good prognosis with good results seen in 90% of cases after early range-of-motion rehabilitation is performed. In the more severe cases (Type III fractures), arthroplasty patients do better with regards to stability, satisfaction, and complications when compared to radial head ORIF. Other complications commonly seen following elbow trauma include post-traumatic arthritis, heterotopic ossification, valgus deformity, or laxity resulting in potential ulnar nerve palsy (tardy palsy) and joint instability.

Recurrent instability, especially posterolateral rotatory instability, after elbow dislocation can be a major problem if not identified early and treated appropriately, however, recurrent dislocations of the elbow are very rare without a bony fracture or ligament injury.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. What is the most common elbow fracture?
   (a) Radial head fracture
   (b) Capitulum humeri
   (c) Proximal ulna
   (d) Distal humerus
2. What are the most common bony injuries associated with elbow dislocation?
   (a) Radial head and coronoid fractures
   (b) Distal ulna
   (c) Distal radius
   (d) Capitulum humeri
3. What is the most common mode of elbow instability?
   (a) Posterolateral rotatory instability
   (b) Anterolateral instability
   (c) Posteromedial instability
   (d) Anteroposterior instability
4. Which elbow fractures commonly require ORIF?
   (a) Bi-column distal humerus and olecranon fractures
   (b) Capitulum humeri
   (c) Radial diaphysis
   (d) Ulnar diaphysis
5. In which patient groups is total elbow replacement surgery suitable?
   (a) Elderly low-demand patients
   (b) Young patients

(c) Athletes
(d) Patients with low BMI and comorbidities

6. What is the most common clinical complication of elbow fracture?
(a) Joint stiffness
(b) Infection
(c) Plate breakage
(d) Neurological injuries

Further Reading

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Fractures of the Forearm and the Wrist

74

Enrico Cautero and Alessandro Mazzola

Overview

The radius, the ulna, the proximal, and the distal radioulnar joints form a virtual ring in the forearm. Like all ring structures, their breakage generally occurs in two different points, with potential damage to the surrounding soft tissues.

74.1 Fractures of the Radial Head

Fractures of the radial head are the most frequent fractures of the elbow: they occur in about 20% of all acute elbow injuries. The dynamics of the trauma generally consists of a fall onto an outstretched hand (FOOSH) with hyperextended and supine elbow. More than 30% of these fractures are associated with other lesions, both capsuloligamentous (lateral collateral ligament (LCL), distal radial ulnar joint (DRUJ)) and bony (fracture of the olecranon or coronoid). Therefore, a careful assessment of the entire forearm must be carried out in these patients. The Mason classification of radial head fractures is based on the location of the fracture and the amount of comminution and displacement involved. Since the radial head guarantees lateral stability of the elbow by limiting its valgus and allows rotation of the forearm, any alteration that compromises its function affects the entire elbow function.

In case of undisplaced or minimally displaced (<2 mm) fractures, immobilization in a cast with the forearm in supination for 2 weeks followed by active ROM is sufficient for a complete healing. An ongoing radiographic evaluation is strongly recommended.

In case of displaced radial head fractures, the goal is the anatomical reduction and osteosynthesis of fracture fragments or, when it is not feasible, its replacement with prosthesis that acts as a joint spacer.

The excision of the radial head, especially when the interosseous membrane has also been injured, exposes the elbow to the risk of chronic instability. It may potentially evolve into valgus deformity of the elbow, loss of range of motion (ROM), and instability of the DRUJ. A more detailed analysis of radial head fractures is discussed in the chapter on elbow fractures.

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74.2 Coronoid Fractures

Coronoid fractures are traumatic elbow fractures that are generally pathognomonic for an episode of elbow instability. They are relatively uncommon and often occur in association with elbow dislocations. Repair of coronoid fractures may be necessary for restoring elbow joint stability. Coronoid fractures are discussed in the chapter on elbow fracture.

74.3 Fractures of Both Forearm Bones

Fractures of the radius and the ulna are the most common fractures of the upper extremity. The causative mechanism is generally a direct trauma (FOOSH). They can occur proximally (near the elbow), in the middle of the forearm, or distally (near the wrist). The evaluation of the DRUJ and elbow integrity is crucial in order to exclude the presence of Monteggia or Galeazzi fracture-dislocations. In the presence of fractures of the forearm, a compartment syndrome must always be excluded: bleeding from the major blood vessels of this area may lead to increased pressure levels in the anterior compartment of the forearm, causing acute ischemia and necrosis of the flexor muscles. Due to ischemic muscle fiber shortening, a permanent flexion deformity at the wrist and fingers resulting in a clawlike presentation of the hand occurs. This clinical pattern is known as Volkmann's contracture. The radiographic evaluation of forearm fractures requires at least two projections of the forearm including the elbow and the wrist; dislocations or subluxations of the radial capitellum must be excluded by checking that the line passing through the diaphysis, neck, and radial head crosses the center of the capitulum humeri. To ensure that the forearm and surrounding joints are able to function correctly again, the reduction and synthesis of these fractures must be anatomical. Therefore, in adults, the best choice is to treat them with open reduction and fixation in the shortest time possible. It also prevents further complications by quickly removing the compression of the hematoma on the anatomical compartment. In cases of undisplaced fractures of the radius and the ulna, immobilization in a rigid elbow is the treatment of choice. In case of open fractures with loss of substance and serious damage to the soft tissues, temporary external fixation and damage control are the treatment preferred by the authors. The final synthesis is performed using angular stability plates once the anatomical reduction is obtained; the option to firstly reduce the radius and the ulna depends on the surgeon's preferences. The surgical approach to the radius can be volar (following the Henry's approach) or dorsal (following the Thompson's approach). As regards the ulna, the incision follows the posterior subcutaneous edge, and the cleavage plane is between the flexor and extensor tendons. The plate can be positioned either volar or dorsal. Once the fracture is reduced, the pronosupination of the forearm is tested: if not complete, it is necessary to try again the fracture reduction.

These fractures generally heal after 4 months. Postoperative management involves a 2-week period of immobilization followed by cautious mobilization of the wrist and elbow. Pronosupination is recovered after a 4-week radiographic check in order to assess a good healing phase.

74.4 Wrist Fractures

Wrist fractures are one of the most common causes of orthopedic urgency. They account for 17% of the fractures in adults and are more frequent in men. In the elderly, they are due to low-energy traumas, resulting from falls on osteoporotic bones. In young people, they are more frequently caused by high-energy impacts. They can be associated with lesions of the proximal radial ulnar joint (PRUJ), ulnar styloid, triangular fibrocartilage complex (TFCC), and intercarpal ligaments. The elbow involvement should always be excluded. The diagnosis of wrist fractures is radiographic with anteroposterior (AP), lateral, and oblique projections. In more complex cases, a CT scan investigation is necessary. The MRI is useful for the study of the capsuloligamentous structures.

The classification of wrist fractures distinguishes undisplaced or displaced, extra- or intra-articular, multi-fragmented, and stable or unstable fractures. The most frequent fractures are named with eponyms (Table 74.1).

Table 74.1 Eponyms of wrist fractures

Die-punch fracture	Depressed fracture of the lunate fossa in the articular surface of the distal radius
Barton's fracture	Fracture-dislocation of the radiocarpal joint with intra-articular fracture involving the volar or dorsal lip (volar or dorsal Barton's fracture)
Chauffeur's fracture	Radial styloid fracture
Colles' fracture (Fig. 74.1)	Low-energy, dorsally displaced, extra-articular fracture
Smith's fracture	Low-energy, volarly displaced, extra-articular fracture

Undisplaced extra-articular fractures are generally immobilized with a rigid brace for 3 weeks. For fractures with distal epiphyseal involvement of the radius, a 5-week period of immobilization is preferred. For displaced fractures, the goal of treatment is the anatomical restoration of the wrist length, volar tilt, and radial inclination. In some patients, this result can be achieved through closed reduction with external maneuvers under local anesthesia. The wrist is then immobilized in a rigid brace. Postreduction radiographic check determines the degree of reduction, stability, and treatment strategies (Table 74.2).

Factors that contribute to making the fracture unstable are coexisting fracture of the base of the ulnar styloid, fractures involving the sigmoid notch, enlargement of the DRUJ, dorsal subluxation of the ulna, and injury of the TFCC. Surgical

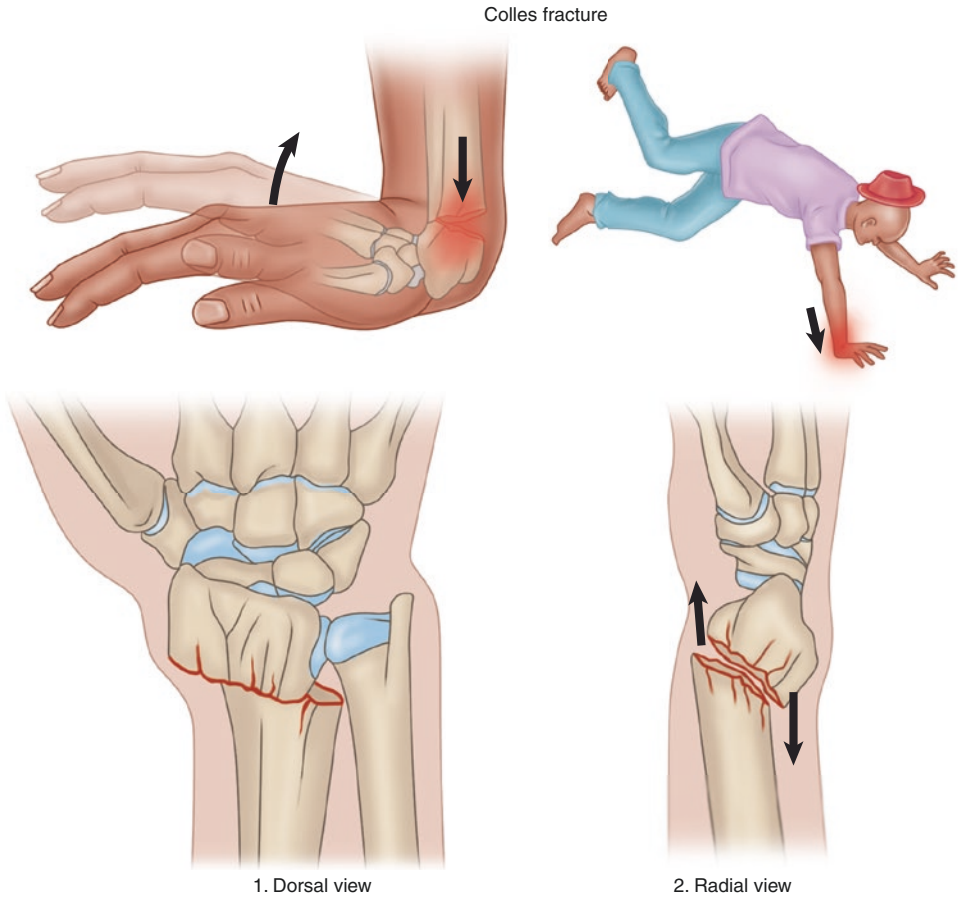

Fig. 74.1 Colles' fracture

Table 74.2 Criteria for an acceptable wrist reduction

<5 mm shortening compared to opposite wrist
No dorsal angulation
>15° of inclination
Articular step-off <2 mm
Congruent DRUJ

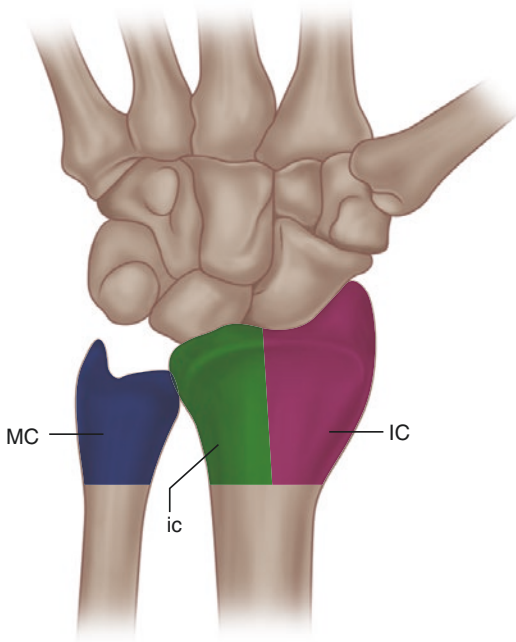

Fig. 74.2 The proximal wrist joint, showing the medial (mc), intermediate (ic), and lateral (lc) columns

options include percutaneous pinning and external fixation or rigid fixation with plate and screws. An external fixation is indicated in cases of polytraumatized patients that need temporary fixation, during initial treatment of severe open fractures, and to supplement suboptimal internal fixation. The most important surgical concept in distal radius fractures (DRFs) is to divide the wrist into three columns: radial, intermediate, and ulnar (Fig. 74.2). The radial column includes the scaphoid fossa and the radial styloid process: it is a platform that supports the carpus and prevents radial translation when the wrist is loaded in ulnar deviation. It also works as a support for ligaments, limiting the ulnar translation of the carpus. Furthermore, it is the insertion site of the brachioradialis muscle which produces the main deforming force of this column in cases of DRFs. The intermediate column incorporates the lunate and the sigmoid fossae: the volar rim of the lunate facet gives origin to the short radio-lunate ligaments. They prevent volar subluxation or dislocation of the carpus in addition to the volar distal radioulnar ligament. The volar ridge of the intermediate column extends distally more than the radial one. It should be taken into account while applying any hardware to the volar distal radius and in plate manufacturing. Dorsally, the intermediate column is the insertion site of the dorsal radiocarpal ligaments: it serves as a support for the carpus preventing dorsal subluxation or dislocation when the radiocarpal joint is loaded. Finally, the ulnar column contains the ulnar head and the TFCC which stabilizes the DRUJ and allows forearm rotation.

Current indications for volar plate are unstable fractures in younger patients, volar shear fractures, die-punch fractures, intra-articular step-off >2 mm that cannot be controlled with closed reduction, radial shortening >3 mm, and dorsal tilt >10° from neutral.

74.5 Isolated Ulnar Fractures

Fractures of the ulnar shaft not affecting the PRUJ are often caused by a direct trauma to the forearm (nightstick fracture). They are frequently undisplaced or minimally displaced fractures (<10° in any axis and with translation <50%), and conservative treatment in sling or brace is generally preferred.

Displaced fractures, fractures involving the proximal third of the ulna, or those associated with vascular nerve damage must be treated surgically with open reduction and internal fixation. Traditional AO plating of the bones of the forearm has been shown to be very reliable, with union rates of around 95%.

74.6 Monteggia Fracture

A Monteggia fracture-dislocation is defined as a fracture of the proximal third of the ulnar shaft with an associated dislocation of the PRUJ.

Typically, the Monteggia fracture-dislocation occurs as a result of a FOOSH: Giovanni Battista

Monteggia was the Italian surgeon who firstly described this type of injury in 1814.

A prompt diagnosis is essential to avoid the onset of dangerous complications; according to the Bado classification of Monteggia fracture-dislocations, type 1 is the most common form, and it corresponds to the fracture of the proximal or middle third of the ulna with anterior dislocation of the radial head.

In contrast to what happens in children, where reduction and closed treatment are frequently adopted, in adults, the fracture is generally treated surgically by open reduction and internal fixation of the ulna. In most cases, the reduction of the radial head is obtained with the anatomical ulnar alignment. Sometimes, especially in pediatric patients, the interposition of the annular ligament can make the reduction of the radium unstable.

74.7 Galeazzi Fracture

A radial shaft fracture associated with DRUJ dislocation is also known as a Galeazzi fracture-dislocation. It was firstly described by Riccardo Galeazzi in 1934.

Nowadays, internal fixation of the radial shaft fracture is the gold standard of treatment, but there are different options for the treatment of the DRUJ: simple splinting, pinning, or excision of the distal ulna. It seems that the distance between the radial fracture and the DRUJ is important for the residual instability. Evaluation of the DRUJ stability in supination after radial fixation and its treatment with pinning and TFCC reconstruction are the key points.

Long arm cast in supination is placed for 4–6 weeks. Then, passive and active exercises of the elbow can start.

74.8 Essex-Lopresti Fracture-Dislocation

An Essex-Lopresti injury is a severe injury of the upper limb. It consists of a typical triad: comminuted radial head fracture, disruption of the DRUJ, and tearing of the interosseous membrane (IOM). It generally occurs when a high-energy load is axially applied to the forearm, usually as a result of a FOOSH. Wrist radiogram is mandatory in order to evaluate the dorsal subluxation of the ulna.

Early surgical treatment is required to manage the radial head: it should be fixed adequately or, if impossible, replaced. The excision of the radial head should be avoided, because it results in proximal migration of the radius with severe wrist and elbow pain. Treatment of the DRUJ can be obtained with a long arm cast in full supination with or without pinning for 4–6 weeks.

Take-Home Message

- Whenever possible, the patient should be asked for the dynamics of the reported trauma.
- The forearm can be considered as a ring; therefore, its breakage in a single point is rare.
- Careful evaluation of all structures, both bony and ligamentous, proximal and distal to the fracture is mandatory.
- In case of displaced fractures with possible soft tissues suffering or vascular nerve injury, if a definitive fixation is not feasible, a temporary reduction and stabilization must be carried out promptly.

Summary

Fractures of the forearm are a common cause of orthopedic emergency room access: among all, fractures of the wrist are the most frequent.

The dynamics of the causative trauma, the bone quality, and the site of injury are the factors influencing the fracture pattern. A prompt diagnosis, based on clinical and radiographic evaluations, both downstream and upstream of the fracture, is necessary to choose the correct treatment and to prevent complications.

The innovation of internal fixation devices has improved the treatment options available to the surgeon for restoring the correct anatomy and achieving a complete functional recovery.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. What are the coronoid fracture signals?
   (a) An episode of elbow instability and they often occur in association with elbow dislocations
   (b) An episode of elbow instability and they often occur in association with dislocation or subluxation of the proximal humerus
   (c) An episode of glenohumeral instability
   (d) An episode of elbow instability and they occur after an exposed fracture of the distal humerus
2. Which is the most common cause of orthopedic urgency?
   (a) Wrist fractures
   (b) Pelvic fractures
   (c) Spine fractures
   (d) Hand fractures
3. What should be carefully evaluated in a patient presenting with a fracture of the radial head due to falling from height?
   (a) The wrist should be carefully assessed with a lateral view in supine position to exclude lesions of the DRUJ indicative of an Essex-Lopresti fracture-dislocation
   (b) The wrist should be carefully assessed with an anteroposterior view in supine position to exclude lesions of the DRUJ indicative of an Essex-Lopresti fracture-dislocation
   (c) The elbow should be carefully assessed with a lateral view in supine position to exclude lesions of the DRUJ indicative of an Essex-Lopresti fracture-dislocation
   (d) The elbow should be carefully assessed with an anteroposterior view in supine position to exclude lesions of the DRUJ indicative of an Essex-Lopresti fracture-dislocation
4. Monteggia fracture consists of:
   (a) Fracture of the proximal third of the ulnar shaft and dislocated PRUJ
   (b) Fracture of the distal third of the ulnar shaft and dislocated PRUJ
   (c) Fracture of the proximal third of the radial shaft and dislocated ulna
   (d) Fracture of the distal third of the radial shaft and dislocated ulna
5. Colles' fracture is the eponym for which kind of wrist injury?
   (a) Distal radial extra-articular fracture, dorsally displaced
   (b) Distal ulnar extra-articular fracture, dorsally displaced
   (c) Distal radial extra-articular fracture, volarly displaced
   (d) Distal ulnar extra-articular fracture, volarly displaced

Further Reading

Sandeep JS. A historical report on Riccardo Galeazzi and the management of Galeazzi fractures. J Hand Surg Am. 2010;35(11):1870–7.

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Longo UG, De Salvatore S, Mazzola A, Salvatore G, Mera BJ, Piergentili I, Denaro V. Colles' fracture: an epidemiological nationwide study in Italy from 2001 to 2016. Int J Environ Res Public Health. 2023;20(5):3956.

Part XIV

Hand

Hand Pathologies

75

Luke Wojdyla, Tudor Hughes,
and Giovanna Stelitano

Overview
Hand and wrist injuries involve a broad spectrum of traumatic and degenerative conditions. Extensor and flexors tendons could be affected by inflammatory and stenotic process that causes various diseases.

75.1 Tendon Injuries and Overuse Syndromes

75.1.1 Extensor Tendon Injury

In most cases, the extensor tendon injuries affect long fingers, and partial disruptions (less than 50%) do not require surgical treatment if the patient can extend the finger against resistance. On the contrary, complete (or partial >50%) lacerations need to be treated with direct suture repair, but the postoperative rehabilitation depends on the zone of injury. The eight zones of injury, respectively, correspond to terminal extensor tendon at the DIP joint (I), middle phalanx (II), PIP joint (III), proximal phalanx (IV), MCP joint (V), metacarpal zone (VI), wrist joint (VII), and distal forearm (VIII).

A laceration at zone I injury causes the typical deformity called mallet finger, which is described as "bony" in the presence of an avulsed fragment of bone. The DIP joint extension splinting can be used for acute injuries (<12 weeks) or nondisplaced bony mallet injury, whereas for the displaced condition, there are two options: CRPP or extension block pinning. The chronic mallet injury can be treated with nonoperative or surgical options (tenodermodesis and the Fowler tenotomy); furthermore, the association with the swan neck deformity requires the Fowler central slip tenotomy or the spiral oblique retinaculum ligament SORL reconstruction.

A zone II injury occurs because of a dorsal laceration or crush. The treatment is essentially the same as that of the zone IV and VI injuries, and it depends on the entity of the laceration: if it is <50%, it needs only early mobilization, and if not, direct surgical suture.

The disruption of the extensor at zone III results in an extensor lag and DIP joint hyperextension (boutonnière deformity) (Fig. 75.1). When an acute damage occurs, the Elson test is performed to evaluate the presence of central slip injury. The treatment of an acute boutonnière deformity is represented by splinting for closed injuries and direct repair for open injuries. On the

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https://doi.org/10.1007/978-3-031-20987-1_75

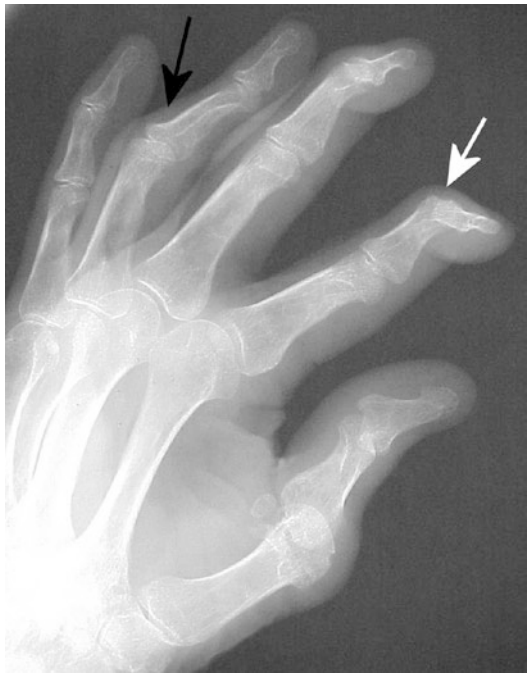

Fig. 75.1 Boutonniere and swan neck deformities. When the extensor tendon is disrupted distally, a swan neck deformity with flexion at the DIPJ may ensue (white arrow), or if the extensor hood that holds the tendon centrally over the phalanges is disrupted more proximally, the extensor tendons may pass to the volar side of the PIPJ causing a boutonniere deformity (French for buttonhole) with flexion at the PIPJ and extension at the DIPJ (black arrow)

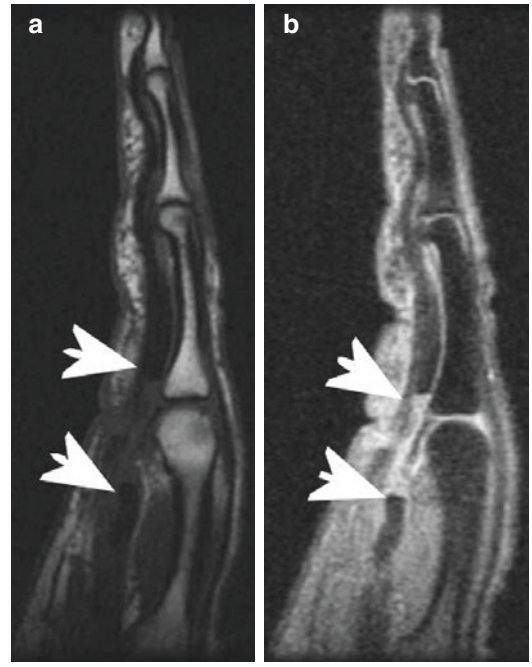

Fig. 75.2 Complete laceration of the flexor tendons. Sagittal T1 (**a**) and T2FS (**b**) MRI images of the ring finger showing laceration of the FDP and FDS between the two white arrows

contrary, chronic cases may need a dynamic splinting, terminal extensor tenotomy, PIP volar plate release, and central slip reconstruction.

A zone V injury is treated with early mobilization and dynamic splinting, but in the case of a sagittal band rupture, repair or reconstruction of this band can be used if the nonoperative option fails. About zone VII and VIII injuries, they are often associated with an extensor retinaculum laceration, and for this reason, the treatment provides its repair and static immobilization with the wrist in extension and MCP partially flexed.

75.1.2 Flexor Tendon Injury

The flexor tendon injuries have several common points regardless of the specific area of interest: they are commonly associated with neurovascular damage, and the examiner should test each digit for active DIP and PIP flexion. Imaging is best with ultrasound (US) or MRI. The treatment is essentially operative (in fact, the nonoperative management of partial disruption often leads to trigger finger and gap formation); furthermore, the lacerations affecting more than 60% of the tendon (Fig. 75.2) need simultaneous core and epitendinous repair within 3 weeks. To obtain an optimal suture strength, the core sutures should have 6–8 strands, high caliber, and a purchase of approximately 1.2 cm; for a good outcome, the surgeon should preserve A2 and A4 pulleys, testing the integrity of the repair during the performance, and use a postoperative active flexion protocol. In the case of primary repair failure or chronic injury, the finger requires a flexor tendon reconstruction that is generally performed using a two-stage technique. Furthermore, there are descriptions and specific treatments based on the Verdan zones, whereas for the FPL injury as described Kleinert (dynamic splinting) and Duran (passive flexion exercises) rehabilitation protocols.

75.1.3 Stenosing Tenosynovitis (Trigger Finger)

The stenosing tenosynovitis affects more frequently middle and ring finger of women over 50 and is often associated with diabetes and inflammatory arthropathy. It is characterized by a progressive mechanical locking, which is staged by Green classification. US is useful in the diagnosis. The treatment provides an initial approach with corticosteroid injection and, with failure of this treatment, surgical release of A1 pulley with additional A3 release or resection of ulnar FDS slip in case of non-recovery.

75.1.4 De Quervain Tenosynovitis

De Quervain tenosynovitis involves the first compartment of the extenders, which includes the abductor pollicis longus and extensor pollicis brevis tendons of the thumb. The patient reports swelling and tenderness at the dorsoradial zone of the wrist, and the examiner finds positive Finkelstein and Eichhoff tests. The diagnosis can be suggested by radiographs and confirmed with US or MRI (Fig. 75.3). The nonoperative treatment is represented by rest, thumb spica splinting, NSAIDs, and local corticosteroid injections, and, if it fails, release of the first extensor compartment should be used.

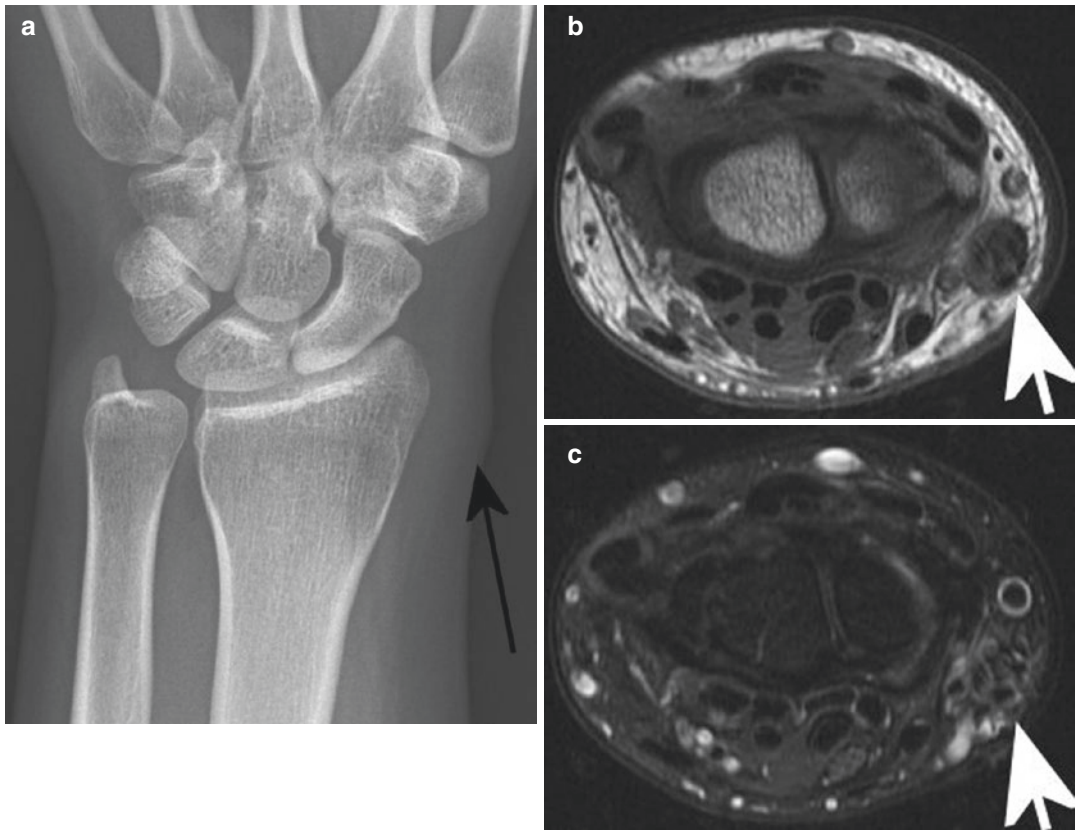

Fig. 75.3 De Quervain tenosynovitis. The first dorsal compartment of the wrist (abductor pollicis longus and extensor pollicis brevis) is prone to chronic repetitive microtrauma and subsequent tenosynovitis (particularly nursing mothers in the nondominant wrist, from holding the baby). The PA view of the left wrist (**a**) shows focal soft-tissue swelling at the level of the radial styloid. The axial T1 MRI (**b**) at the level of the wrist shows the markedly enlarged and heterogenous tendons (white arrow) and the axial T2 fat-saturated image at the same level; (**c**) the fluid signal surrounding the markedly abnormal tendon shreds caused by tenosynovial fluid and synovial hypertrophy (white arrow)

75.1.5 Intersection Syndrome and Acute Calcific Tendinitis

Intersection syndrome at the wrist is divided into proximal (where APL and EPB pass dorsally over ECRB and ECRL) and distal (where EPL passes dorsally over ECRL and ECRB) and is a tenosynovitis and/or bursitis secondary to chronic friction. The symptoms are tenderness, swelling, and crepitus 4–5 cm proximally to the radiocarpal joint (Fig. 75.4). Acute calcific tendinitis is an overuse syndrome due to repetitive resisted wrist flexion, which especially affects FCU. It is characterized by acute pain, discoloration, and swelling at the wrist (Fig. 75.5). In both cases, if ice, splinting, NSAIDs, and local (oral for the tendinitis) corticosteroid fail, the treatment may need to be release of the extensor compartment and debridement of inflamed bursa.

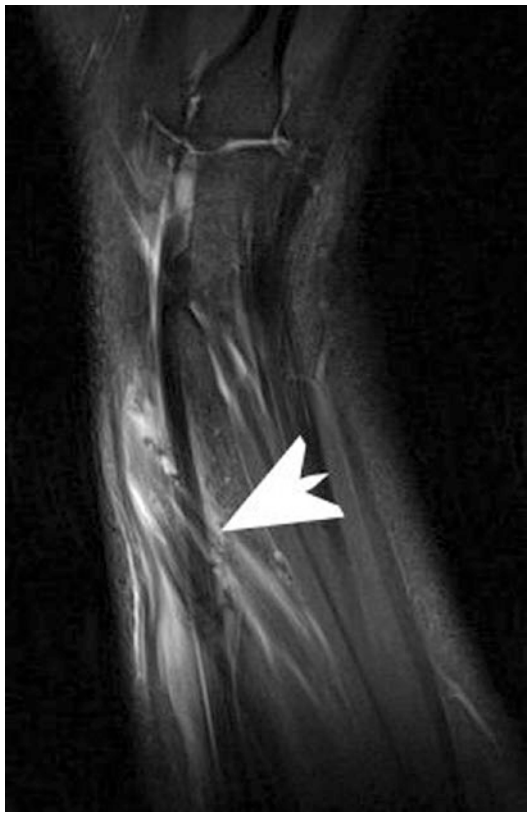

Fig. 75.4 Proximal intersection syndrome. This is caused by friction from chronic repetitive microtrauma as the first dorsal tendon compartment passes superficially over the second. The coronal T2FS MRI image of the dorsal wrist in an 18-year-old man shows high T2 signal from fluid due to this inflammation (white arrow)

75.1.6 ECU Tendinitis and Subluxation

Repetitive supination of the forearm, flexion of the wrist, and ulnar deviation can cause an inflammation of the ECU tendon at the distal ulna, which can be treated with a nonoperative management. A traumatic origin requires Muenster splint if acute and repair or reconstruction of the overlying extensor retinaculum in chronic subluxation.

75.1.7 Distal Radioulnar Joint, Triangular Fibrocartilage Complex

The distal radioulnar joint (DRUJ) includes several structures (among which triangular fibrocartilage complex should be mentioned) important for its stability that may be affected by some alterations. These pathologies are often reported as a wrist pain, which must be investigated with wrist arthroscopy.

Palmer classification divides pathologies of TFCC in traumatic injuries (class 1) and degenerative TFCC tears (class 2) providing important information for treatment. Regarding diagnosis, MRI (especially following dilute gadolinium arthrogram) is highly accurate (Fig. 75.6) and arthroscopy is the gold standard for detection of TFCC tears thanks to trampoline, hook, and suction test. In the case of traumatic injuries, the initial management always provides a nonoperative treatment, and, if it fails, the surgical choice depends on the characteristics of the subclass: a central lesion requires a debridement, the peripheral one and the distal avulsion need to be repaired, whereas the radial avulsion often benefits from the reduction of radius. The degenerative TFCC tears are often associated with a positive ulnocarpal variance and increased loading, and the main symptom is a chronic wrist pain located at the ulnar area exacerbated by forearm rotation and grip; more-

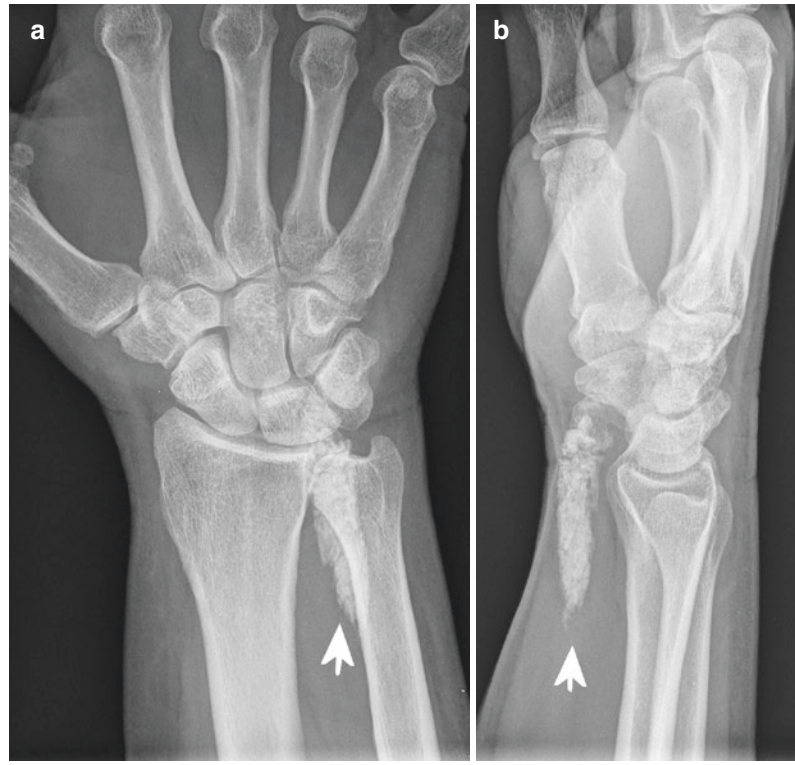

Fig. 75.5 FCU calcific tendonitis. This is due to calcium hydroxyapatite deposited within the tendon. The PA (**a**) and lateral (**b**) radiographs show the characteristic location of the calcium (white arrows), although it is usually much smaller than this

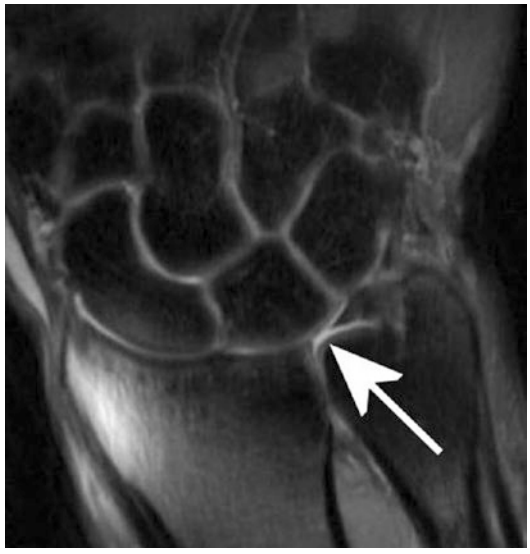

Fig. 75.6 Triangular fibrocartilage perforation. The Cor T1FS images following intra-articular injection of dilute gadolinium into the radiocarpal joint space show a palmer 1a tear of the central triangular fibrocartilage (white arrow) with continuity of the radiocarpal joint space with the distal radioulnar joint (bright signal) through the perforated triangular fibrocartilage

over, it can be determined by loading of wrist in extension and ulnar deviation. Surgical management is used if the nonoperative treatment fails, and it is represented by the reduction of ulnocarpal loading with ulnar shortening osteotomy, wafer resection, or debridement.

The DRUJ can be affected by two pathologic conditions: instability and post-traumatic osteoarthritis. The acute dislocation should be treated with closed reduction and immobilization if it occurs alone, with an open or arthroscopic approach if it is associated with distal ulna fracture and TFCC tears, and with ORIF of the radial shaft in the case of Galeazzi injury. A chronic DRUJ instability can be assessed using CT scans (Fig. 75.7), and it can be resolved with TFCC repair or ligament reconstruction. A post-traumatic DRUJ OA should be initially treated with a nonoperative management, eventually followed by several surgical possibilities: distal ulna resection (Darrach procedure), hemiresection or interposition arthroplasty, Sauvé-Kapandji procedure, ulnar head or total joint implant arthroplasty (Fig. 75.8), and one-bone forearm.

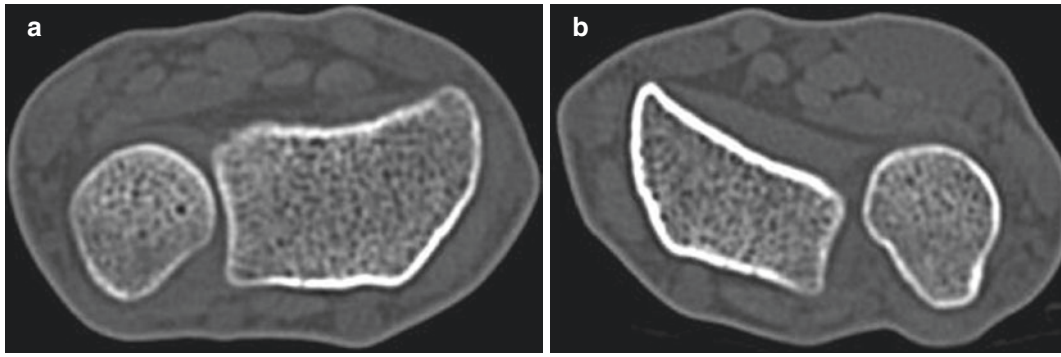

Fig. 75.7 Distal radioulnar joint instability. The axial CT images of the left (**a**) and right (**b**) wrists in full supination show instability of the right DRUJ in full supination with the radius dorsally subluxed on the ulna

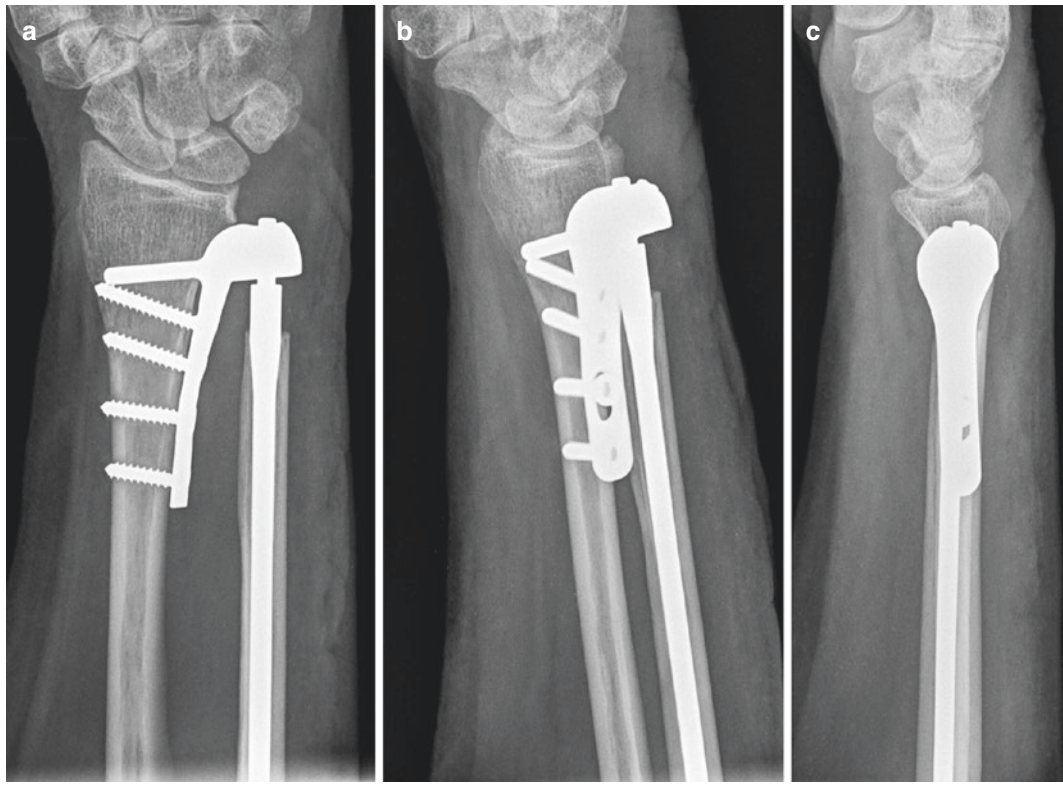

Fig. 75.8 Distal radioulnar joint arthroplasty. The PA (**a**), oblique (**b**) and lateral (**c**) radiographs of a 68-year-old female following DRUJ arthroplasty

75.1.8 Compression Neuropathies and Peripheral Nerve Injuries

The compression syndromes are a set of painful neuropathies due to the compression of a nerve. Those of the upper limb concern the median, ulnar, and radial nerve and the thoracic outlet, and they share some sensory and/or motor symptoms and signs, such as initially loss of light touch followed by pain and temperature sensitivity, paresthesia, weakness, dropping of objects, and clumsiness. Furthermore, they can lead to irreversible changes: demyelination, fibrosis, and axonal loss. The diagnosis is essentially clinical but also instrumental; the physical examination includes strength (of single muscles, grip, pinch)

and neurosensory testing (Semmes-Weinstein monofilaments and two-point discrimination). The electrodiagnostic techniques are represented by sensory and motor nerve function tested by electromyography, which detects the electrical activity of muscle during voluntary contraction, and nerve conduction study, used to obtain measures of nerve conduction velocity, distal latency, and amplitude. Imaging with US and MRI is very useful to find the exact level of compression. The choice of treatment depends on the physical and electrodiagnostic testing and the possible presence of multiple compression sites (double-crush phenomenon).

The suffering from median nerve entrapment can present with different syndromes according to the compression zone: carpal tunnel syndrome (carpal tunnel), pronator syndrome (arm or forearm), and anterior interosseous syndrome (AIN) (bicipital bursa and accessory head of the FPL). CTS occurs with paresthesia and pain in the volar aspect of radial digits, thenar weakness followed by atrophy, positive Tinel sign and Phalen test, loss of two-point discrimination, and night-time symptoms. Clinical analysis is sufficient for the diagnosis, and the electrodiagnostic tests are not always necessary. Also, imaging is rarely needed to diagnose CTS, but when performed will show the characteristic appearance of a swollen T2 bright nerve proximal to the carpal tunnel (Fig. 75.9). Regarding the management, nonoperative treatment includes activity modification, night splints, NSAIDs, and local corticosteroid injections, whereas surgery offers several options: open, mini-open, or endoscopic release of the transverse carpal ligament.

The pronator syndrome is characterized by the same symptoms of CTS, and in addition, the patient also reports proximal volar forearm pain and sensory disturbances over the thenar region. In the AIN syndrome, the patient refers to inability to grasp, but sensitivity and opposition are preserved. Tinel sign will be positive around the pronator teres muscle, and pronation against resistance leads to symptoms. Surgical treatment is necessary only if nonoperative treatment fails, and it consists of the release of all potential sites of compression.

The ulnar nerve can be compressed at the cubital tunnel and Guyon canal (ulnar tunnel syndrome). A patient affected by the cubital tunnel syndrome reports paresthesia of fifth and ulnar half of fourth finger and dorsal ulnar hand, as well as an important motor weakness, detected with Froment, Jeanne, Wartenberg, and Masse sign; interosseous atrophy (Fig. 75.10); and last two fingers clawing. The management at the cubital tunnel includes both nonoperative and operative treatment (in situ decompression, anterior transposition, and medial epicondylectomy). The ulnar tunnel syndrome is generally due to a ganglion cyst, and success of treatment depends on the correct identification of the cause.

Signs and symptoms due to the radial nerve compression neuropathy depend on the involved zone of the nerve, and they all require a surgical decompression or exploration after 6 months of symptoms. The affection of the radial nerve proper is generally caused by a distal humerus shaft fracture and produces motor symptoms of triceps, brachioradialis, ECRL, and distal muscles, innervated by the posterior interosseous nerve (PIN). The PIN compression syndrome, in addition to the weakness of the muscles innervated (Fig. 75.11), is characterized by radial deviation during active wrist extension and lateral elbow pain. Finally, radial tunnel syndrome occurs only with lateral elbow and radial forearm pain without motor or sensory dysfunction, whereas the patient affected by cheiralgia paresthetica (Wartenberg syndrome) reports typical pain, numbness, and paresthesia over the dorsoradial hand.

The peripheral nerve function may be compromised by compression, stretch, blast, crush, avulsion, transection, or tumor invasion. These modifications determinate different kinds of damage: neurapraxia (simple conduction block due to mild nerve stretch), axonotmesis, and neurotmesis are, respectively, characterized by incomplete and complete nerve injury and lead to Wallerian degeneration distal to injury.

The surgical repair should be performed within 2 weeks and, in case of larger gaps, grafting is required. The most important peripheral nerve injuries for the upper limb are traumatic

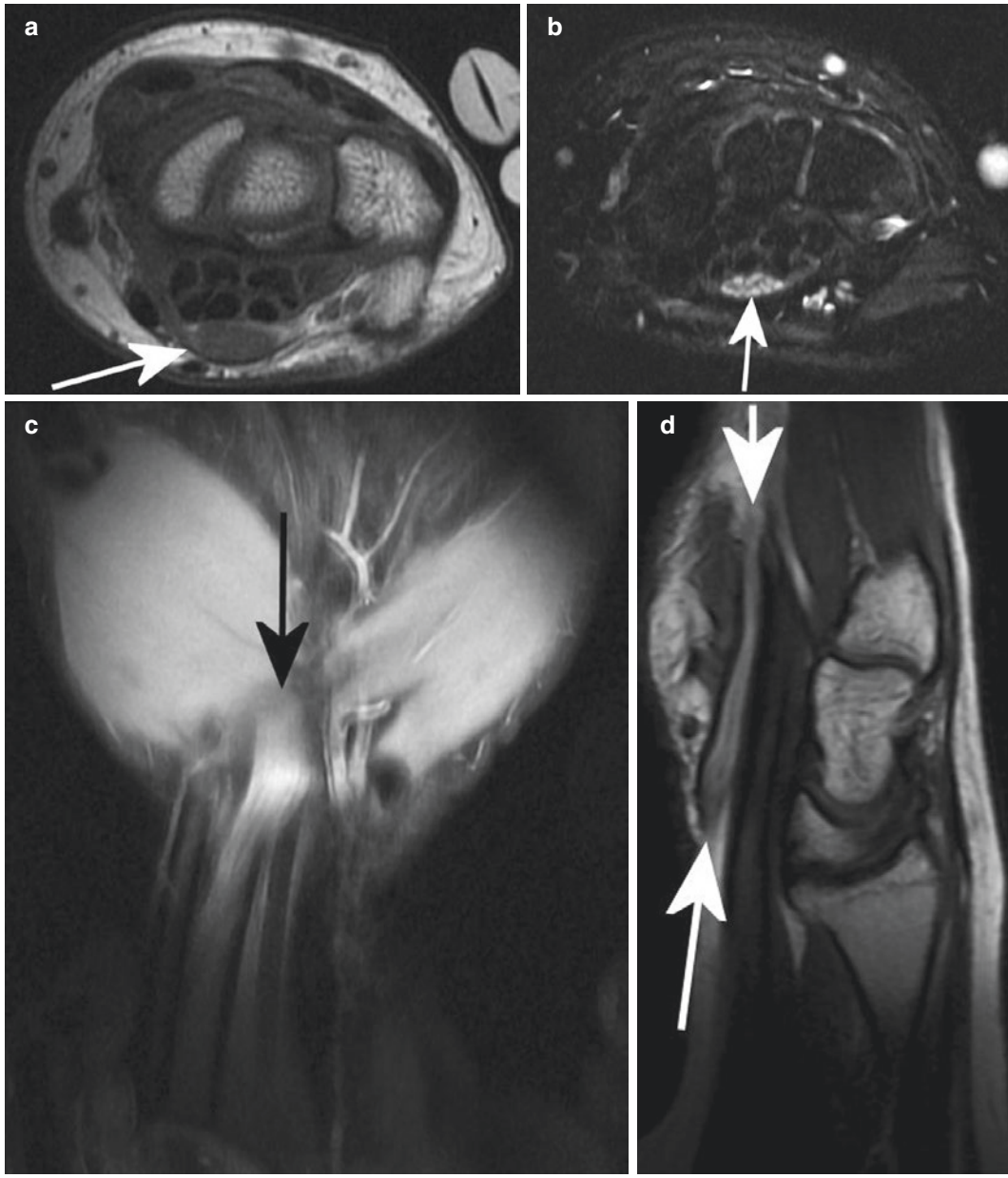

Fig. 75.9 Carpal tunnel syndrome. This is not routinely imaged, relying more on clinical examination, EMG, and nerve conduction studies. However, in more complex cases, there is a role for imaging. The axial T1 MRI at the level of the midcarpal (**a**) shows a thickened median nerve (white arrow) and the corresponding T2-weighted fat saturation image, and (**b**) increased fluid signal within the nerve (white arrow). In longitudinal images ((**c**) coronal, black arrow, and (**d**) sagittal, white arrows), the affected median nerve can be seen to taper as it passes deep to the carpal tunnel retinaculum

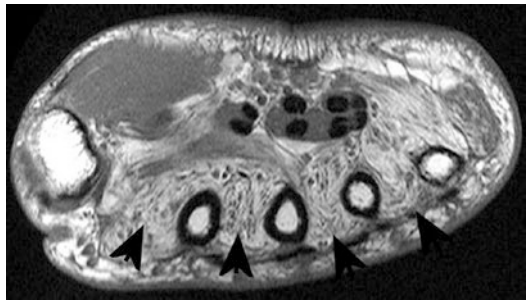

Fig. 75.10 Ulnar neuropathy muscle atrophy. The axial T1 MRI at the level of the metacarpals shows fatty atrophy of the interossei muscles (black arrows) and also the ulnar sided lumbricals and hypothenar eminence muscles due to chronic compression of the ulnar nerve

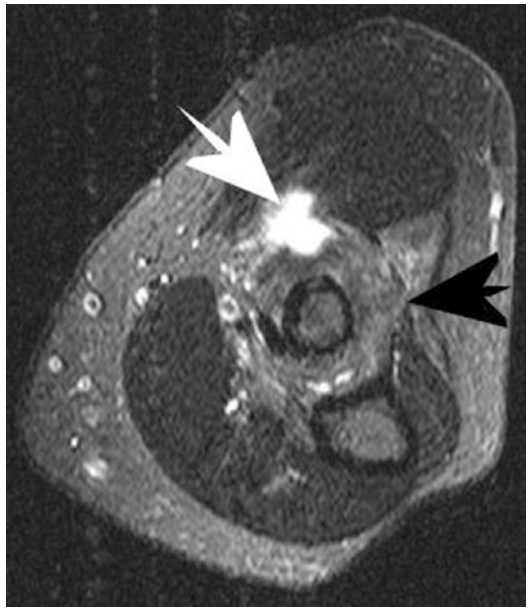

Fig. 75.11 Posterior interosseous nerve syndrome. In this 51-year-old female, the PIN is compressed by a ganglion cyst (white arrow) as it passes beneath the arcade of Frohse to enter between the two heads of supinator muscle. Consequently, the muscles of the posterior compartment including supinator, extensor digitorum, extensor carpi ulnaris, and extensor carpi radialis brevis show muscle edema (black arrow)

brachial plexus injury, obstetric brachial plexopathy, and cerebral palsy. The first one has a surgical treatment (early or delayed for incomplete lesions) and may need an Oberlin transfer in case of insufficient number of proximal axons available. On the contrary, the last one can be initially managed with a nonoperative treatment.

75.2 Arthritis

Osteoarthritis commonly affects DIP joints and trapeziometacarpal of the thumb and is diagnosed by clinical features (pain, swelling, and decreased motion) and classic radiographic findings (joint space narrowing, subchondral cysts and sclerosis, and osteophytes) (Fig. 75.12). The choice of the surgical technique depends on the specific joint affected: arthrodesis for DIP joint, arthroplasty for PIP joint (except for that of the index), and many options that involve partial excision of trapezium (arthrodesis, arthroscopic hemitrapeziectomy, ligament reconstruction-tendon interposition with trapezium excision (Fig. 75.13), simple trapeziectomy).

Rheumatoid arthritis is a systemic autoimmune inflammatory disease that primarily affects hand and wrist, especially MCP joints. The patient reports local inflammatory symptoms and radiological features (osteopenia, erosion, joint space narrowing, and subluxation) (Fig. 75.14). This pathology may cause many alterations at the hand such as rheumatoid nodules (may need surgical excision), tenosynovitis eventually followed by tendon rupture (respectively, treated with tenosynovectomy and tendon transfer), caput ulnae syndrome, Z deformity, rheumatoid thumb, and rheumatoid wrist. In this case, the arthritis weakens the structures of stability of the wrist, and, for advanced radiocarpal destruction, total wrist arthrodesis should be performed.

There are many other inflammatory diseases that involve the hand, such as juvenile rheumatoid arthritis (characterized by radial deviation of the MCP joints and ulnar deviation of the wrist in JRA), psoriatic arthritis (DIP joint pencil-in-cup deformity), SLE, scleroderma (Fig. 75.15), gout, and pseudogout (Fig. 75.16).

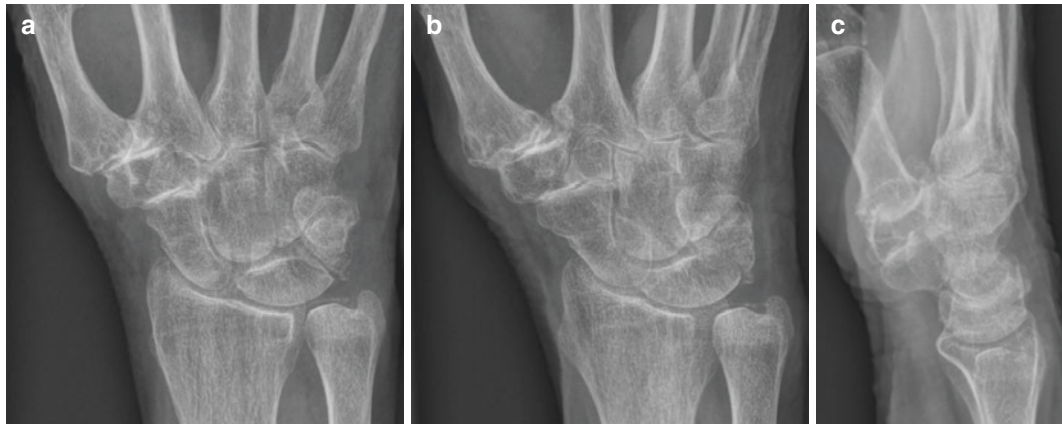

Fig. 75.12 Thumb carpometacarpal joint and triscaphe joint osteoarthritis. In this 81-year-old female, the AP (**a**), oblique (**b**), and lateral (**c**) radiographs show the characteristic findings of osteoarthritis, with joint space narrowing, subchondral cysts and sclerosis, and marginal osteophytes

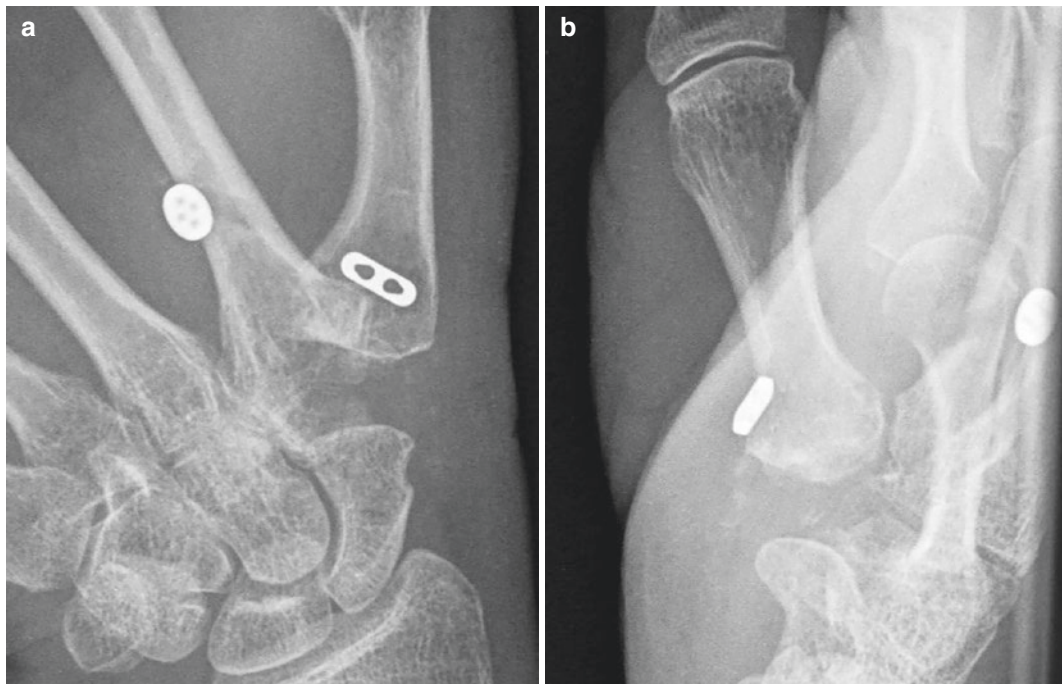

Fig. 75.13 Thumb carpometacarpal joint arthroplasty with trapeziectomy, ligament reconstruction, and tendon interposition (anchovy procedure) utilizing endobuttons. The PA (**a**) and lateral (**b**) radiographs show the trapeziumectomy and the metacarpal tunnel with endobuttons supporting a tightrope between the first and second metacarpals

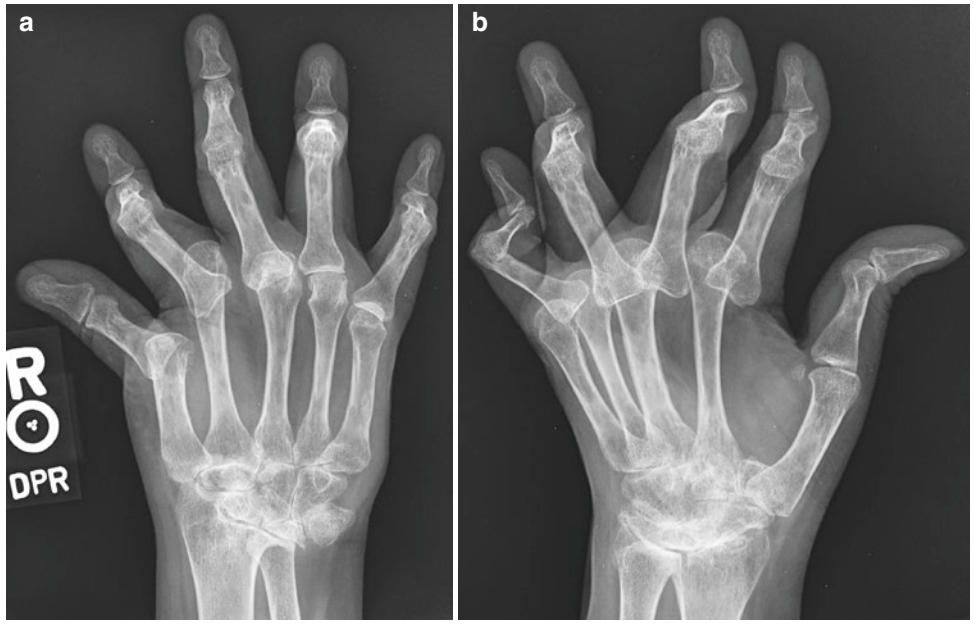

Fig. 75.14 Advanced rheumatoid arthritis. The PA radiographs of the right hand (**a**) and left hand (**b**) in a 34-year-old female show the characteristic findings of advanced rheumatoid arthritis, with intercarpal joint space narrowing, carpal erosions, and severe MCPJ subluxations. The symmetry between the hands is another feature of rheumatoid arthritis

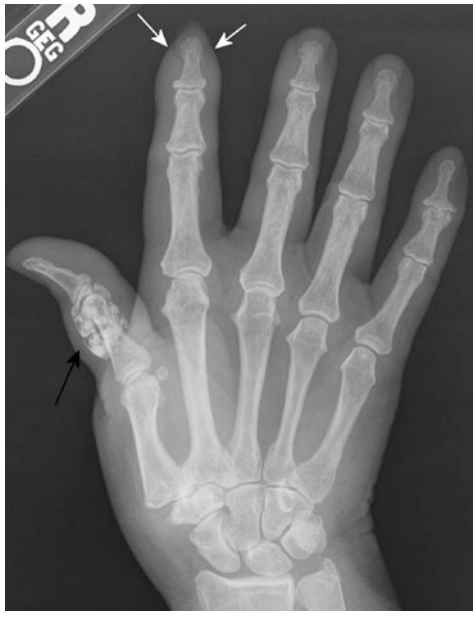

Fig. 75.15 CREST syndrome. This is the association of calcinosis, Raynaud's syndrome, esophagus, scleroderma, and telangiectasia. The PA view of the right hand shows tapering of the terminal soft tissues of the index finger due to vascular insufficiency from Raynaud's (white arrows) and soft-tissue calcifications due to scleroderma (black arrow)

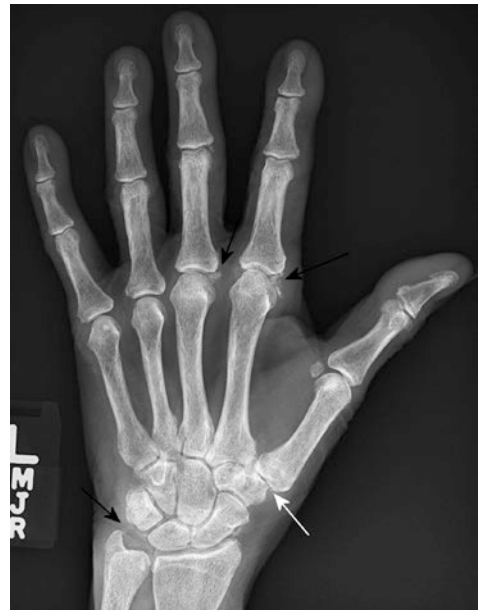

Fig. 75.16 Calcium pyrophosphate dihydrate deposition and arthropathy. The PA view of the left hand shows the classical distribution of chondrocalcinosis due to CPPD deposition at the triangular fibrocartilage, intercarpal ligaments, and second and third MCPJs (black arrows). In addition, there is early arthropathy at the thumb CMCJ (white arrow)

75.3 Idiopathic Osteonecrosis of the Carpus

The Kienböck disease is an idiopathic osteonecrosis of the lunate characterized by fragmentation and collapse of this bone that affects men aged 20–40 years. It has a multifactorial etiology, and the diagnosis is based on the observation of dorsal wrist pain, mild swelling, limited motion, weakness, and radiographic findings typical of the advanced stages (lunate sclerosis and lunate collapse) (Fig. 75.17) and MRI signs in initial stages (Fig. 75.18). The treatment depends on Lichtman stage and ulnar variance: in stages I–IIIA, the usual management is to save the lunate, whereas surgery in stages IIIB–IV performs salvage procedures because lunate pathology prevents its revascularization.

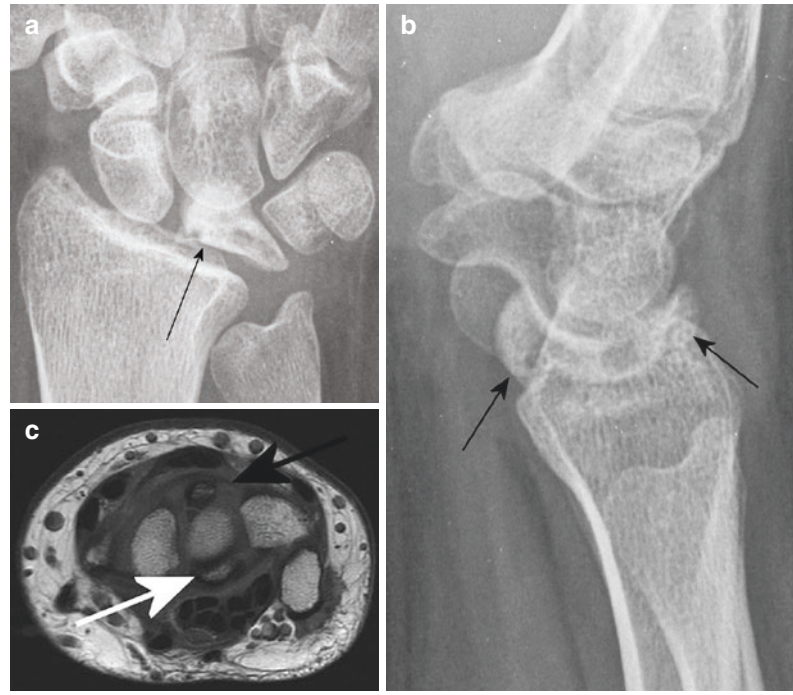

Fig. 75.17 Kienböck disease. Spontaneous avascular necrosis of the lunate is partly related to having a short ulna, as can be seen in (**a**). The necrosed lunate bone has become sclerotic and collapsed (black arrow). On the lateral view (**b**), the lunate can be seen to have broken into volar and dorsal fragments (black arrows) known as the broken canoe sign. (**c**) The axial T1-weighted MRI image again shows the volar (white) and dorsal (black arrow) fragments

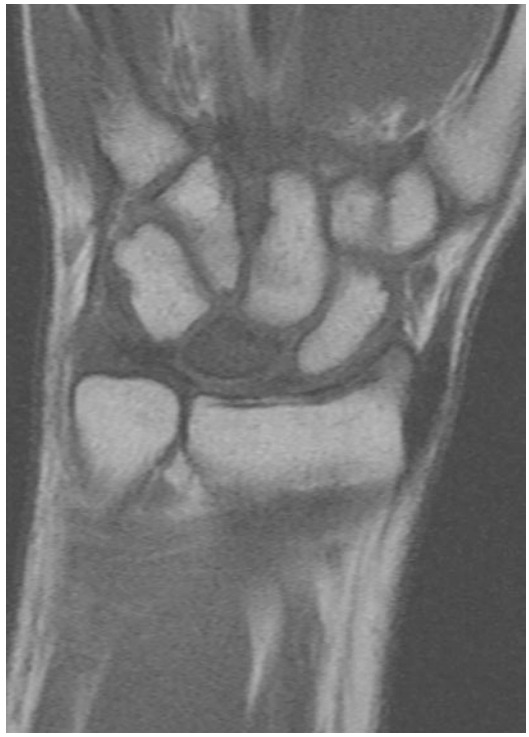

Fig. 75.18 Kienböck disease. Coronal T1 MRI at an earlier stage of the osteonecrosis shows the uniform low T1 signal throughout the lunate that is characteristic

75.4 Dupuytren Disease

Dupuytren disease is a benign fibroproliferative disorder characterized by the progression of a nodule in the palmar fascia to pathological cords and MCP and/or PIP joint flexion contractures (Figs. 75.19 and 75.20). The etiology is unknown, but it is caused by cytokine-mediated transformation of normal fibroblasts into myofibroblasts. The Dupuytren disease evolves in three stages: proliferative, involutional, and residual. The diagnosis is based on clinical features, and the treatment may be nonoperative or surgical. The first one consists of collagenase injection at the cord affected or needle aponeurotomy (NA) followed by manipulation and night splint; the surgical management is represented by open fasciectomy, which can be performed with different techniques and with a postoperative protocol that consists of active ROM and static night-time splinting to maintain extension correction.

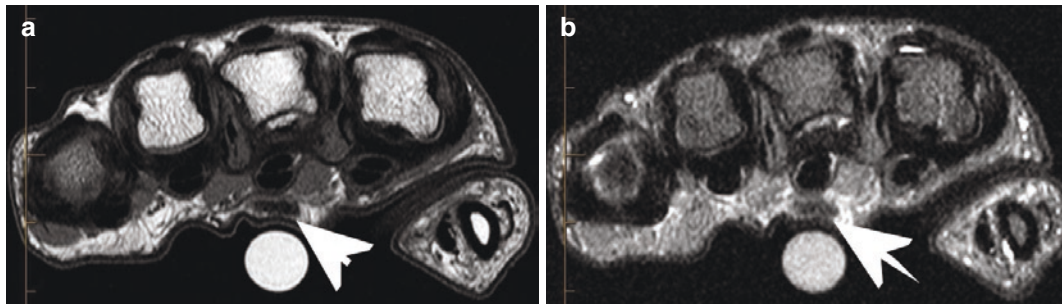

Fig. 75.19 Dupuytren disease. Axial T1 images (**a**) and T2 images with fat saturation (**b**) show the band of low-signal fibrous tissue within the subcutaneous tissues beneath the marker (white arrows)

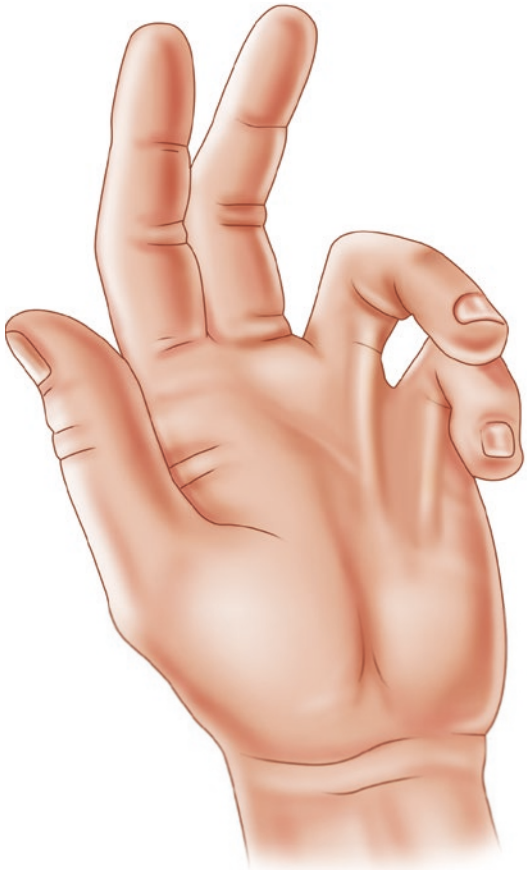

Fig. 75.20 Dupuytren disease

Take-Home Message

- Complete (or partial >50%) extensor tendon lacerations need direct suture repair.
- The treatment for flexor tendon injuries is essentially operative; furthermore, the lacerations affecting more than 60% of the tendon need simultaneous core and epitendinous repair within 3 weeks.
- The stenosing tenosynovitis affects more frequently middle and ring finger of women over 50 and is often associated with diabetes and inflammatory arthropathy.
- De Quervain tenosynovitis involves the first compartment of the extenders.
- Osteoarthritis commonly affects DIP joints and trapeziometacarpal of the thumb and is diagnosed by clinical features and classic radiographic findings.
- The Kienböck disease is an idiopathic osteonecrosis of the lunate characterized by fragmentation and collapse of this bone that affects men aged 20–40 years.
- Dupuytren disease is a benign fibroproliferative disorder characterized by the progression of a nodule in the palmar fascia to pathological cords and MCP and/or PIP joint flexion contractures.

Summary

In most cases, the extensor tendon injuries affect long fingers, and partial disruptions (less than 50%) do not require surgical treatment if the patient can extend the finger against resistance. On the contrary, complete (or partial >50%) lacerations need to be treated with direct suture repair, but the postoperative rehabilitation depends on the zone of injury.

The flexor tendon injuries have several common points regardless of the specific area of interest: they are commonly associated with neurovascular damage, and the examiner should test each digit for active DIP and PIP flexion. Imaging is best with ultrasound (US) or MRI. The treatment is essentially operative (in fact, the nonoperative management of partial disruption often leads to trigger finger and gap formation); furthermore, the lacerations affecting more than 60% of the tendon need simultaneous core and epitendinous repair within 3 weeks.

The stenosing tenosynovitis affects more frequently middle and ring finger of women over 50 and is often associated with diabetes and inflammatory arthropathy.

De Quervain tenosynovitis involves the first compartment of the extenders, which includes the abductor pollicis longus and extensor pollicis brevis tendons of the thumb.

The intersection syndrome at the wrist is divided into proximal and is a tenosynovitis and/or bursitis secondary to chronic friction.

The distal radioulnar joint (DRUJ) includes several structures important for its stability that may be affected by some alterations. These pathologies are often reported as a wrist pain, which must be investigated with wrist arthroscopy.

The compression syndromes are a set of painful neuropathies due to the compression of a nerve. Those of the upper limb concern the median, ulnar, and radial nerve and the thoracic outlet, and they share some sensory and/or motor symptoms and signs, such as initially loss of light touch followed by pain and temperature sensitivity, paresthesia, weakness, dropping of objects, and clumsiness.

Osteoarthritis commonly affects DIP joints and trapeziometacarpal of the thumb and is diagnosed by clinical features and classic radiographic findings. The choice of the surgical technique depends on the specific joint affected: arthrodesis for DIP joint, arthroplasty for PIP joint, and many options that involve partial excision of trapezium.

The Kienböck disease is an idiopathic osteonecrosis of the lunate characterized by fragmentation and collapse of this bone that affects men aged 20–40 years.

Dupuytren disease is a benign fibroproliferative disorder characterized by the progression of a nodule in the palmar fascia to pathological cords and MCP and/or PIP joint flexion contractures.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Dupuytren's syndrome most commonly affects:
   (a) I and II index finger
   (b) III and IV finger
   (c) IV and V finger
   (d) IV finger
2. Dupuytren's disease presents:
   (a) Pain
   (b) Cyanosis of the fingers
   (c) Hypersensitivity to cold
   (d) None of the above
3. A 50-year-old worker complains of strength deficit in his right hand and radiating pain from the wrist to the forearm, especially at night. He reports tingling paresthesia I, II, and III and radial side of the IV finger. This symptomatology is reminiscent of upper limb compression neuropathy. Which nerve is probably involved?
   (a) Median nerve
   (b) Ulnar nerve
   (c) Radial nerve
   (d) Posterior interosseous nerve
4. A 65-year-old man presents to the outpatient clinic complaining of progressive and disabling rigidity in flexion of the fourth and fifth fingers of the right hand. On physical examination, this flexion attitude is not reducible and is accompanied by the presence of a sclerotic chord like thickening in the palmar at the palmar level, in correspondence of the fourth and fifth fingers. A possible diagnosis is:
   (a) Dupuytren's disease
   (b) Lesion of the median nerve
   (c) Radial nerve injury
   (d) Ulnar nerve injury
5. A 55-year-old man complains of intense right elbow pain radiating to the forearm with paresthesia radiating to the forearm, the fifth finger, and the ulnar side of the fourth finger. There is a modest ipsilateral strength deficit. The clinical picture suggests:
   (a) Cubital tunnel syndrome
   (b) Round pronator syndrome
   (c) Carpal tunnel syndrome
   (d) Radial tunnel syndrome

Further Reading

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Longo UG, Loppini M, Del Vescovo R, Cazzato L, Zobel BB, Maffulli N, Denaro V. Safety of dorsal wrist arthroscopy portals: A magnetic resonance study. Surgeon. 2018;16(2):101–6.

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Fractures of the Hand

76

Matthew Carr and Tudor Hughes

Overview
The hands and fingers are common sites of trauma and fractures. These fractures are almost never life-threatening, but the hands are essential for many occupations and daily tasks; thus, hand trauma is often associated with significant disability and costs. Due to these factors, swift identification and management of any hand fracture or dislocation are critical to persevere function and ensure the best possible outcome.

76.1 Introduction

The bones of the fingers and hands are relatively fragile structures and very common sites of musculoskeletal trauma. Given the importance of the hands and fingers to the patient's livelihoods, prompt identification and correct management of finger and hand fractures are vital to avoid disability and retain function.

When there is a suspicion for hand fracture from history and physical exam, the radiograph is almost always the first-line modality for identification. Trauma radiographs should always be performed with at least two different views (generally PA and lateral) as one view is seldom optimal to determine the presence or the displacement of the fracture and preferably three views. Furthermore, it may also be beneficial to obtain radiographs for the ipsilateral wrist and forearm as the mechanism of injury for many hand fractures is concomitant with fractures in the proximal upper extremity. While radiographs are normally sufficient, there are some scenarios where obtaining a CT or MRI is the correct course of action. CT is often useful in the evaluation of fractures of the carpal bones as the multiple overlying structures in that area can make identifying fractures difficult by radiograph. Another common reason to obtain CT exams in hand trauma is for operative planning. MRI is also becoming increasingly relevant in the evaluation for fractures. Fractures can be radiographically occult, and MRI is the most sensitive modality for identification of fractures; however, its high cost prevents routine use.

Once radiographs are obtained, the first thing one should evaluate is the adequate positioning/quality and if the correct structures are profiled; for example, a common scenario is two fingers being buddy taped together, which severely limits evaluation of the clinical area of interest on the lateral radiograph. Another consideration is ensuring that the correct views are obtained, such as obtaining scaphoid views if there is a concern for scaphoid pathology. Once the quality of the

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scan is evaluated, many like to ascertain an overall sense of the pathology: Are there obvious fractures and diffuse soft-tissue swelling? Is the bone mineralization appropriate? After that, look at the mechanism of injury if available.

On frontal and oblique views, trace each of the bone cortices and check for small bone fragments. Then evaluate for the contour of the carpal bones and specifically the scaphoid, which should be smooth on these projections. On lateral radiographs, look for osseous fragments at the dorsal aspect of the hand, which may be seen in triquetral fractures and small osseous fragments about the digits, which should increase suspicion for avulsion injury. When evaluating for fractures, it is also important to note that nondisplaced fractures are easily overlooked and to be very suspicious of any abnormal lucencies or irregularities when trauma is involved. One common pitfall is that the most commonly missed fracture is the second one. If a fracture is identified, do not fall victim to "satisfaction of search" and be particularly vigilant for additional fractures or dislocations.

After assessing for fractures, assess for dislocations. On PA views, trace the three carpal arcs of Gilula (Fig. 76.1e). Assess for any abnormal overlap, such as those seen in carpometacarpal dislocations, and for loss of the clear joint space and disruption of the double-M-shaped pattern between the carpals and metacarpals. Assess the carpal bones to ensure that there are no abnormalities in shape. Look at the scapholunate interval, which should be less than 3 mm in adults. On lateral views, evaluate the shape of the scaphoid again, as these are commonly missed fractures, and ensure that there are no lunate or perilunate dislocations. Assess for dorsal or volar dislocations or abnormal angulations. Aside from assessing for fractures and dislocation, one should also evaluate for secondary signs of injury, such as soft-tissue swelling, subcutaneous gas, or deformities.

Scaphoid: Of the carpal bones, the scaphoid is the most common significant fracture in the setting of trauma. Its elongated structure bridges both the proximal and distal rows, which makes it particularly vulnerable to mechanical forces. In particular, the scaphoid bone is classically injured after a "fall onto outstretched hand" (FOOSH) where the wrist is hyperextended and deviated radially. It has an unusual blood supply where the perforating arteries enter the bone distally and flow proximally, which makes the proximal pole particularly vulnerable to osteonecrosis. Thus, it is critical to properly identify fractures in this location. Unfortunately, its oblique orientation relative to the remainder of the carpus causes fractures to be difficult to evaluate on standard PA and lateral radiographs (Fig. 76.1a–c). Even with dedicated scaphoid views, these fractures are often occult and require CT or MRI to identify (Fig. 76.1d).

Lunate and perilunate dislocation: Another commonly missed but potentially devastating injury of the carpal bones is a lunate dislocation. This happens commonly after high-energy trauma resulting in dorsiflexion of the hand. The lunate is dislocated volarly and primarily identified on the lateral radiograph (Fig. 76.2). These injuries require prompt management and surgical repair as the lunate can compress the median nerve when dislocated, causing neuropathy. Perilunate dislocation and perilunate fracture-dislocations (not to be confused with lunate dislocation) occur when there is a volar dislocation of the other carpal bones, usually the capitate, relative to the lunate, which remains in normal alignment with the radius (Fig. 76.3). These also happen in high-energy traumas with hyperextension of the hand with ulnar deviation (as opposed to radial deviation, which is associated with lunate dislocations).

Hook of hamate and trapezial ridge: Fractures of the hook of the hamate (Fig. 76.4) can be seen with direct blows to the hamate, which is most commonly seen in athletes such as golf or baseball players. These fractures are important to identify due to the adjacent Guyon's canal, which contains the fibers of the ulnar nerve and can result in ulnar neuropathy if damaged. Similar to the hook of the hamate, the ridge of the trapezium also extends volarly from the body and can also be injured from direct blows to the volar surface (Fig. 76.5). Both the hook of the hamate and trapezial ridge fractures are commonly over-

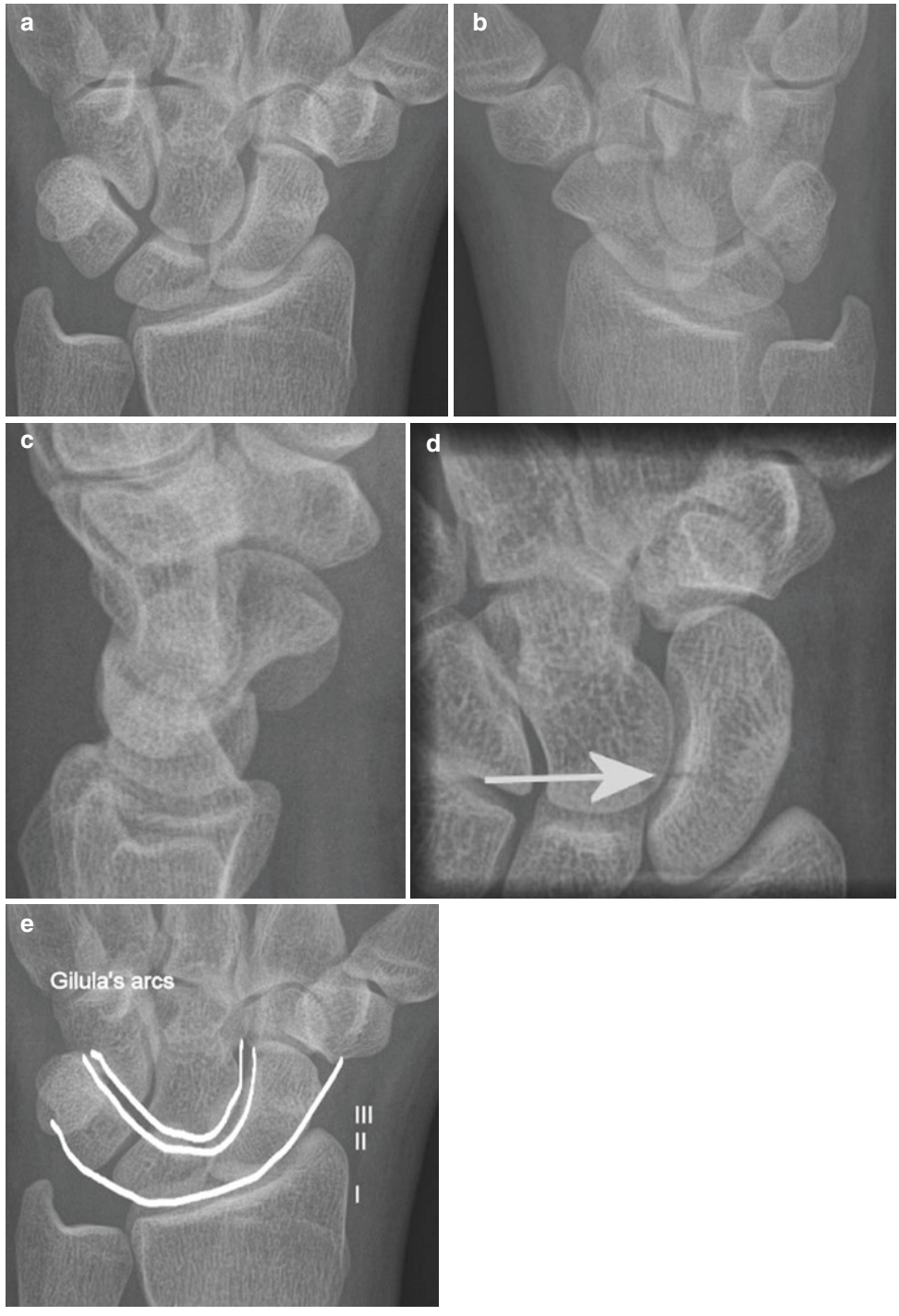

Fig. 76.1 Scaphoid fracture in a 19-year-old female. Traditionally, there are three views of the wrist, PA, oblique, and lateral (**a–c**). This example well demonstrates the need for a coned-down ulnar deviation view (**d**), which is the only view in which the proximal waist of scaphoid fracture is well demonstrated (white arrow). On PA view, one can follow the three carpal arcs of Gilula (**e**)

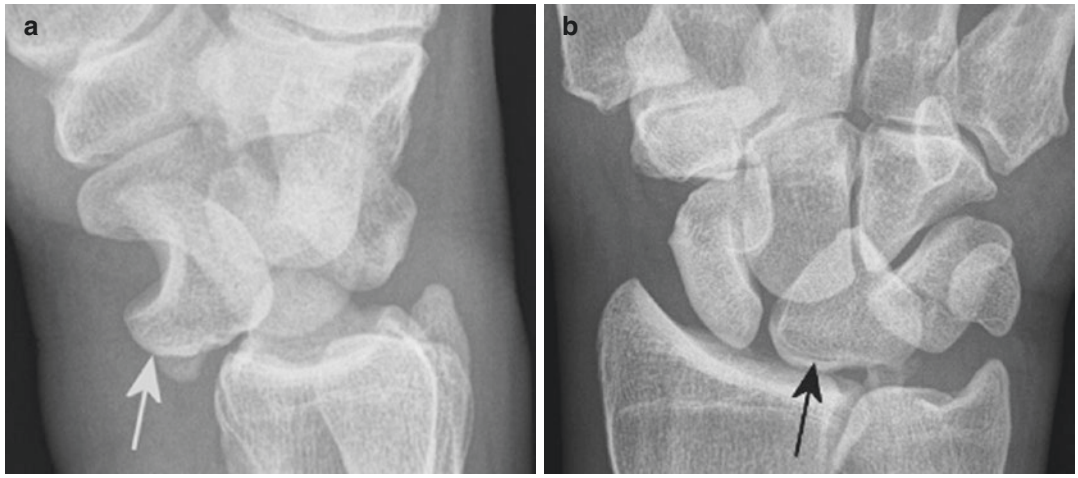

Fig. 76.2 Lunate dislocation. This is sometimes overlooked, because no fracture is seen, and the usually volar displaced lunate (white arrow) is mistaken for another carpal bone such as the pisiform (**a**). On the PA view (**b**), there is loss of the normal carpal rows and the usually lentiform shape of the lunate takes on a triangular shape (piece-of-pie sign, black arrow)

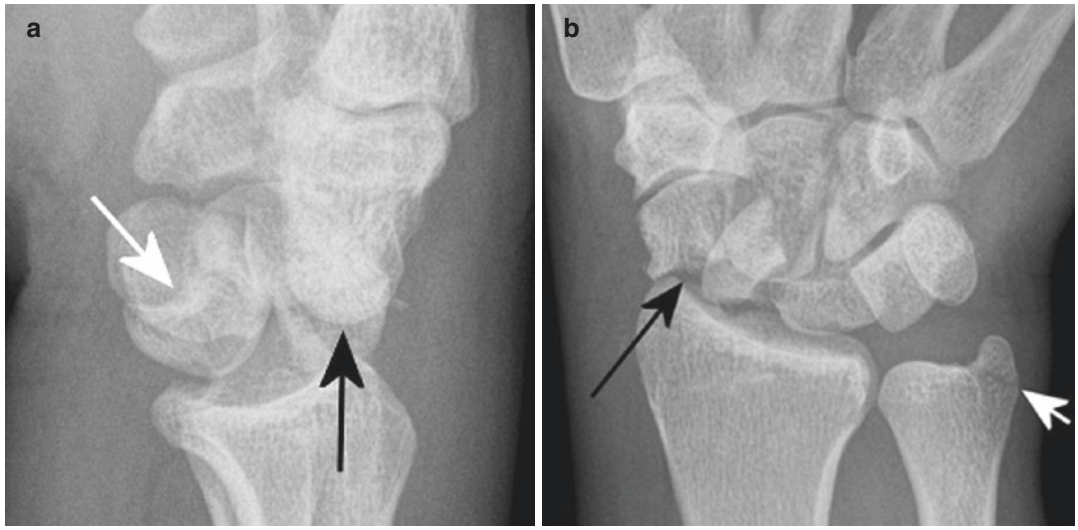

Fig. 76.3 Trans-scaphoid perilunate fracture dislocation in a 32-year-old man. The lateral view (**a**) shows the capitate (black arrow) dislocated posteriorly from the lunate (white arrow). On the PA view (**b**), the additional fractures of the ulnar styloid (white arrow) and waist of scaphoid (black arrow) are demonstrated, making this a greater arc injury

looked on routine radiographs. If there is clinical concern, a dedicated carpal tunnel radiograph should be obtained, which involves dorsiflexion of the wrist, so these structures are better profiled as the metacarpals and carpal bones will not overlap them on this view. Even with dedicated radiographs, these fractures can be difficult to see and a CT scan may be required.

Dorsal triquetral avulsion: After the scaphoid, the triquetral bone is the second most commonly injured bone in the carpus. The usual mechanism of injury is again fall onto out-

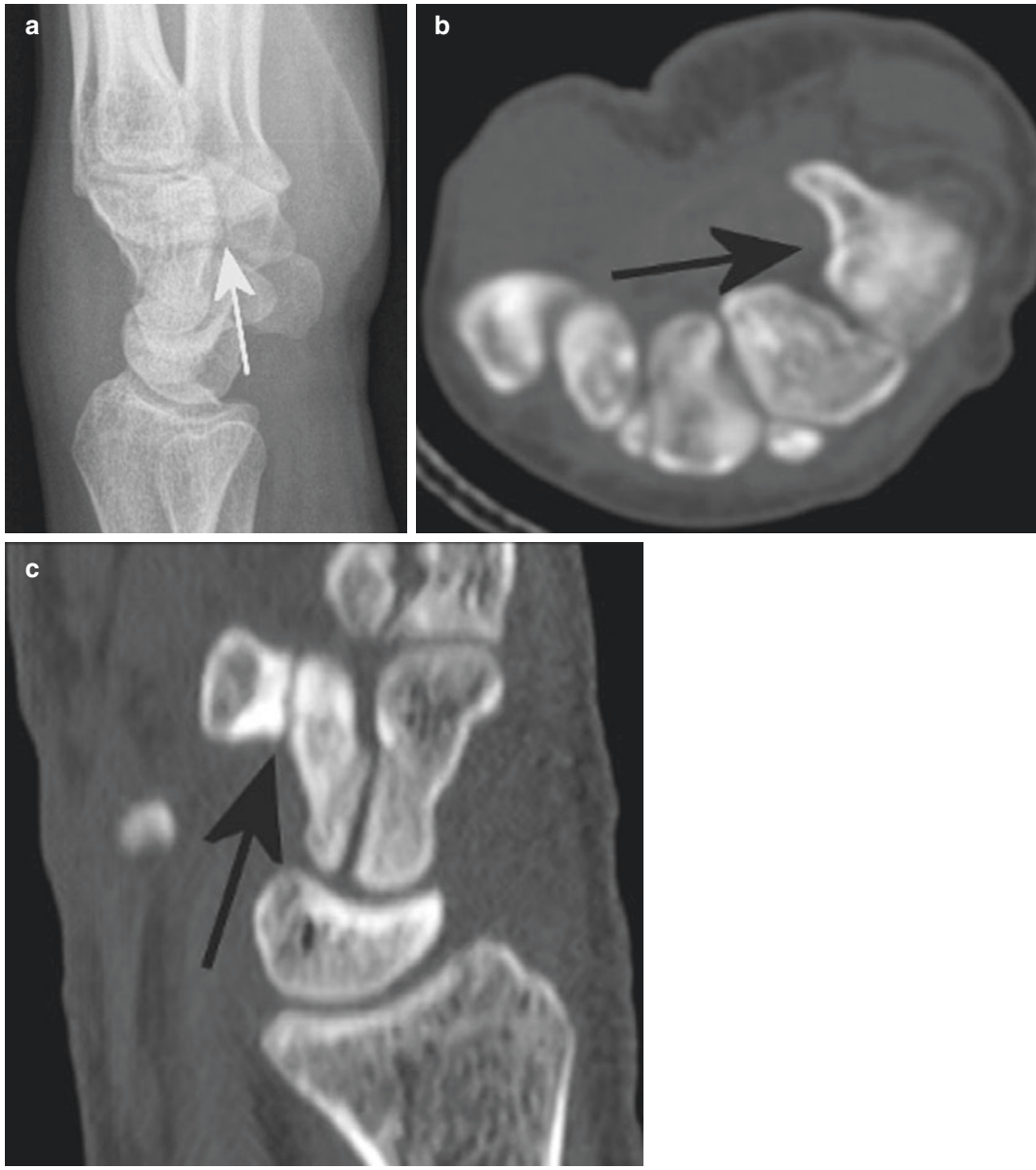

Fig. 76.4 Hook of hamate fracture. These fractures can be very difficult to see on conventional radiographs (**a**, white arrow) including carpal tunnel views, which are technically difficult. CT more clearly shows these on the axial (**b**, black arrow) or sagittal (**c**, black arrow) planes

stretched hand with hyperextension of the hand or less commonly a direct blow to the dorsum of the hand. Lateral radiographs are used to identify this avulsion fracture, where an avulsed flake of bone is seen lying posteriorly to the triquetral bone (Fig. 76.6). While surgical intervention is rarely required, these avulsion fractures can cause persistent (and painful) symptoms in some patients.

Base of thumb, Bennett, Rolando, and beak ligament avulsion: Given that opposable thumbs are one of the defining features of our species, it is not a surprise that the thumb is a common source of musculoskeletal injury. The majority of

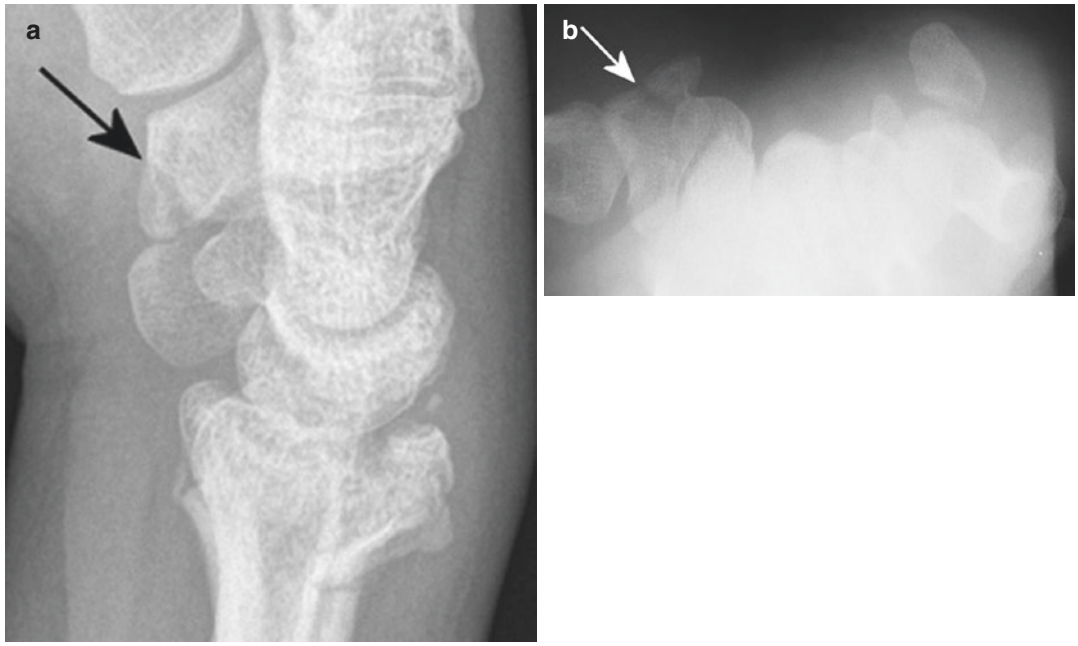

Fig. 76.5 Trapezial ridge fracture. Another fracture that may be difficult to identify. The lateral view (**a**) shows the fracture (black arrow), confirmed on the carpal tunnel view (**b**, white arow). Also note the distal radius fracture

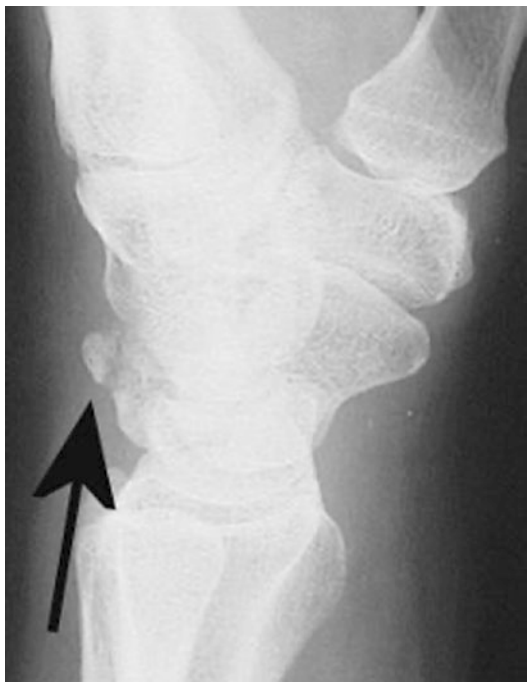

Fig. 76.6 Dorsal triquetral avulsion fracture. A small fracture fragment is seen on the dorsum of the midcarpal region (black arrow)

thumb fractures have involvement of the metacarpal base. For example, a Bennett fracture is a fracture of the base of the first metacarpal after forced abduction. It is a two-piece fracture with intra-articular involvement where the small fragment of the first metacarpal remains attached to the anterior oblique ligament (beak ligament) and articulates with the trapezium while the shaft of the first metacarpal becomes retracted laterally by the now unopposed abductor pollicis longus tendon (Fig. 76.7). While usually the treatment consists of spica splinting, a large or unstable fracture may require surgical fixation. A Rolando fracture is a three-part fracture that involves a volar fragment remaining in place and articulated with the joint, while the main dorsal fragment subluxes dorsally and radially due to the unopposed pulling of abductor longus tendon. It is often thought of as a comminuted Bennett fracture where the fracture lines form a T or Y (Fig. 76.8). The mechanism of injury is typically an axial blow to the flexed first metacarpal, classically from a fistfight, which explains the significant young male predominance who present with

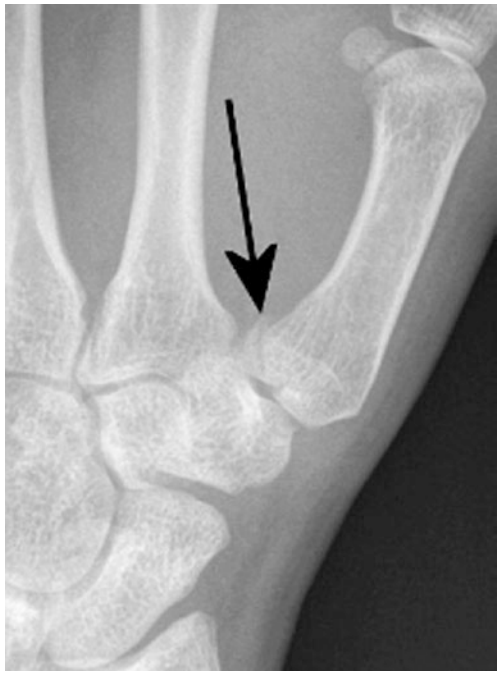

Fig. 76.7 An uncomminuted intra-articular Bennett fracture at the metacarpal base of the thumb (black arrow)

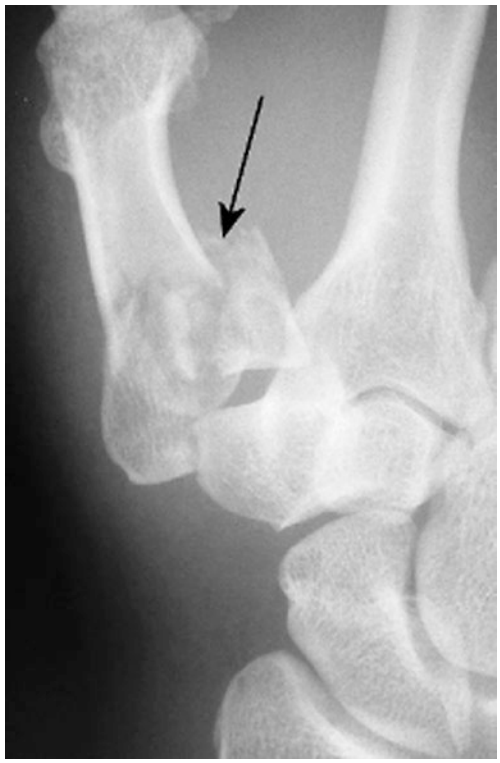

Fig. 76.8 Comminuted intra-articular Rolando fracture at the metacarpal base of the thumb (black arrow)

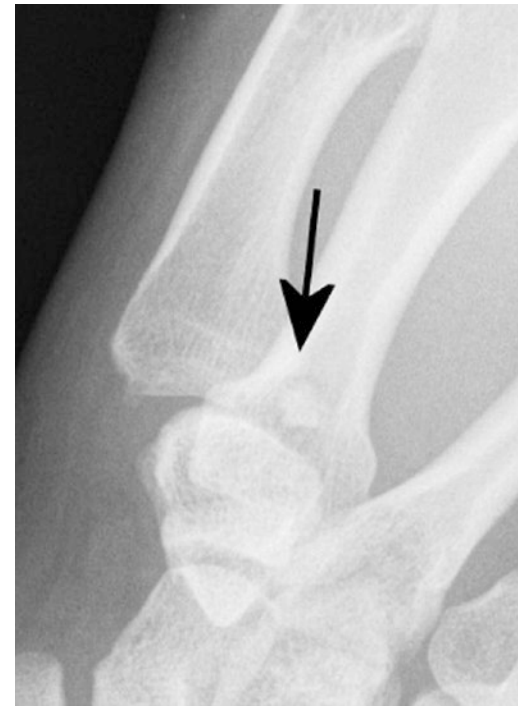

Fig. 76.9 Beak ligament avulsion. Similar to a very small Bennett fracture only avulsed by the deep anterior oblique ligament. These can be hard to see (black arrow)

this fracture. The anterior oblique ligament (beak ligament) can also be involved in a small isolated avulsion fracture, which can be hard to identify (Fig. 76.9).

Fourth and fifth carpometacarpal joint fracture dislocations: Carpometacarpal joint (CMCJ) fracture-dislocations are uncommon but very important factures to identify. They are usually associated with hand pain and swelling after throwing a punch, so they are commonly found in the young male demographic; however, they can be associated with falls as well. Loss of the clear joint space, disruption of the M-shaped joint space pattern between the carpals and metacarpals, and soft-tissue swelling on PA radiographs should increase suspicion for this pathology, which can then be confirmed on oblique or lateral radiographs, which should show abnormal positioning of the carpal metacarpal bones (Fig. 76.10). They are commonly associated with the base of the metacarpal fractures and less commonly hamate fractures. Once a dislocation is identified, it requires reduction and usually inter-

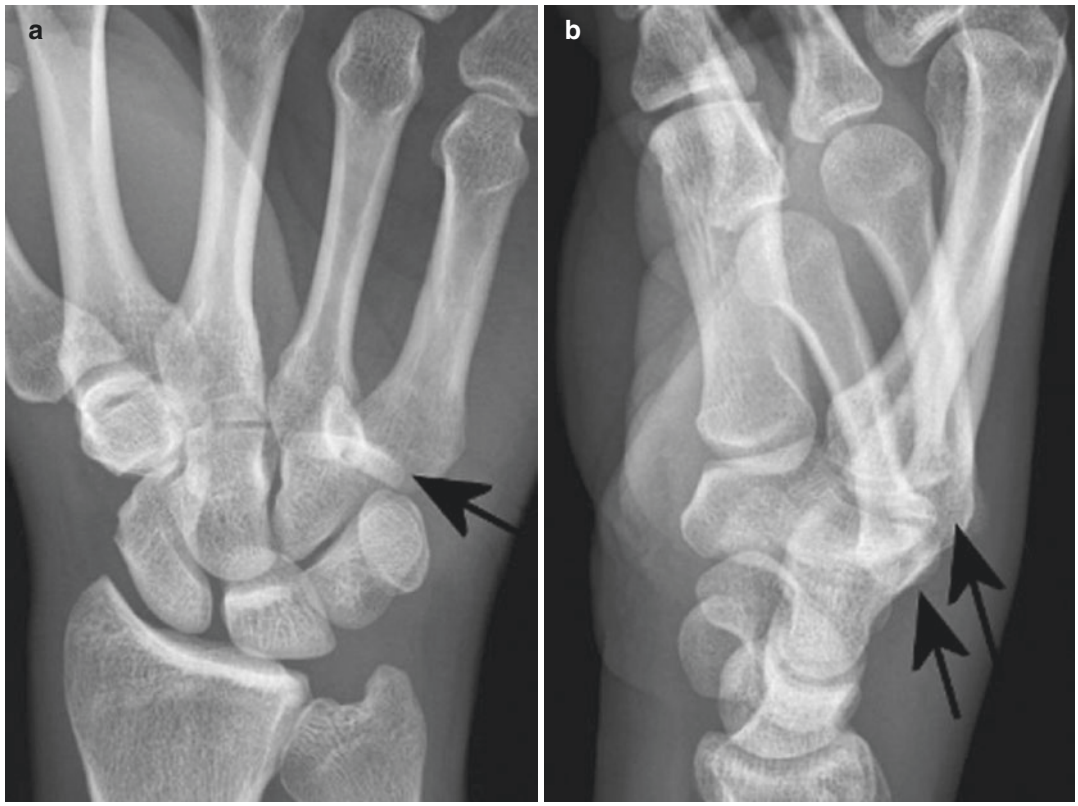

Fig. 76.10 Fourth and fifth carpometacarpal dislocations. Sometimes difficult to see on the PA (**a**) view, unless the overlap of the hamate and fourth and fifth metacarpal bases is appreciated (black arrow). On the lateral view (**b**), the loss of parallelism of the fourth and fifth metacarpals (black arrows) with the second and third metacarpals is a big clue that the fourth and fifth are dislocated. Look for the commonly accompanying dorsal hamate fracture

nal fixation as they are often unstable even after reduction. If it is associated with a metacarpal base fracture, the fracture will require reduction and possibly internal fixation.

Boxer's fracture: Among fractures of the metacarpals, the boxer's fracture, a minimally comminuted transverse fracture of the fifth metacarpal neck, is the most common. It is an impaction fracture that occurs after a direct blow to a solid surface (usually a face or wall) with a clenched fist, which causes axial loading and fracture of the fifth metacarpal, hence the fracture's namesake. It unsurprisingly has a predominance with the young male population. The fracture is usually easily identified on standard radiographs, but careful attention is given to other associated fractures, the degree of displacement, the amount of angulation of the fracture fragment, and the fracture planes (with transverse being the most common) (Fig. 76.11). These factors will determine if surgical management is required.

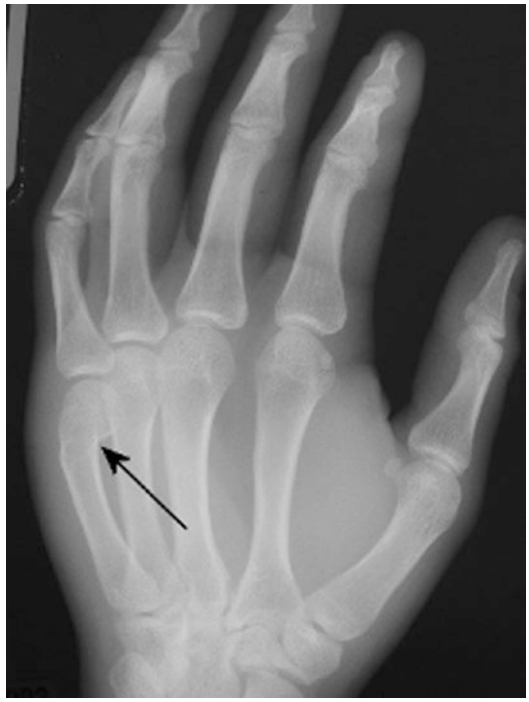

Fig. 76.11 The boxer's fracture (better described as a brawler's fracture due to poor technique) is a transverse fracture of the fifth metacarpal neck (black arrow), usually with volar and radial deviation of the head on the shaft, from a glancing blow contacting a hard surface, be it victim or wall

76.2 Gamekeeper's Thumb and Skier's Thumb

Damage to the ulnar collateral ligament (sometimes with a bony avulsion (Fig. 76.12)) of the metacarpophalangeal joint of the thumb has different names depending on the etiology of the injury. If the injury is from chronic/repetitive overuse, the injury is referred to as "gamekeeper's thumb" as gamekeepers would repeatedly break the neck of game using their thumb and index finger, eventually causing overuse and eventual tearing of the ligament. As its popularity waned, break dancers and then finally skiers became the most common population to have this injury, where skiers would fall with the pole straps in hand and accidentally cause hyperabduction of the thumb. These injuries can involve either partial- or full-thickness tears of the ulnar collateral ligament with or without an accompanying avulsion fracture. If no avulsion fracture is present, the patient may only have widening of the ulnar portion of the metacarpophalangeal joint on radiographs. If there is significant instability, displacement, or a Stener's lesion (interposition of the adductor pollicis muscle aponeurosis between the ruptured ulnar collateral ligament and its site of insertion on the base of the proximal phalanx, diagnosed with MRI or ultrasound), surgery may be required.

Phalangeal shaft, intra- and extra-articular: Fractures of the phalangeal shaft are an extremely common fracture type, with the distal phalanges and fifth digit being the most common. Everyone reading this book has had experience where they have had their finger jammed in a car door or had something dropped on their fingers. Notably, these fractures are common in workplace accidents and can result in significant disability. Once a fracture is identified, one should pay careful attention to the degree of displacement, the amount of angulation, and whether there is intra-articular involvement (Fig. 76.13). If there is significant displacement and intra-articular involvement, these fractures often require operative intervention.

Volar plate, without and with finger dislocations: The volar plate, particularly the volar plate attachment of the proximal interphalangeal joint (PIP), includes areas that are vulnerable to fracture with hyperextension of the distal finger. This hyperextension causes avulsion by the volar plate at the base of the middle phalanx (Fig. 76.14). Usually, there is instability and risk of dorsal dislocation due to much of the stabilizing collateral ligaments of the interphalangeal joint being attached to the avulsed fragment. When characterizing these fractures, the Eaton classification is often used, which evaluates the size of the avulsed fragment (specifically the portion of articular surface), degree of impaction, and direction of dislocation. Depending on these factors, surgical management may be required.

Mallet fractures: In contrast to volar plate fracture, mallet fractures are avulsion fractures

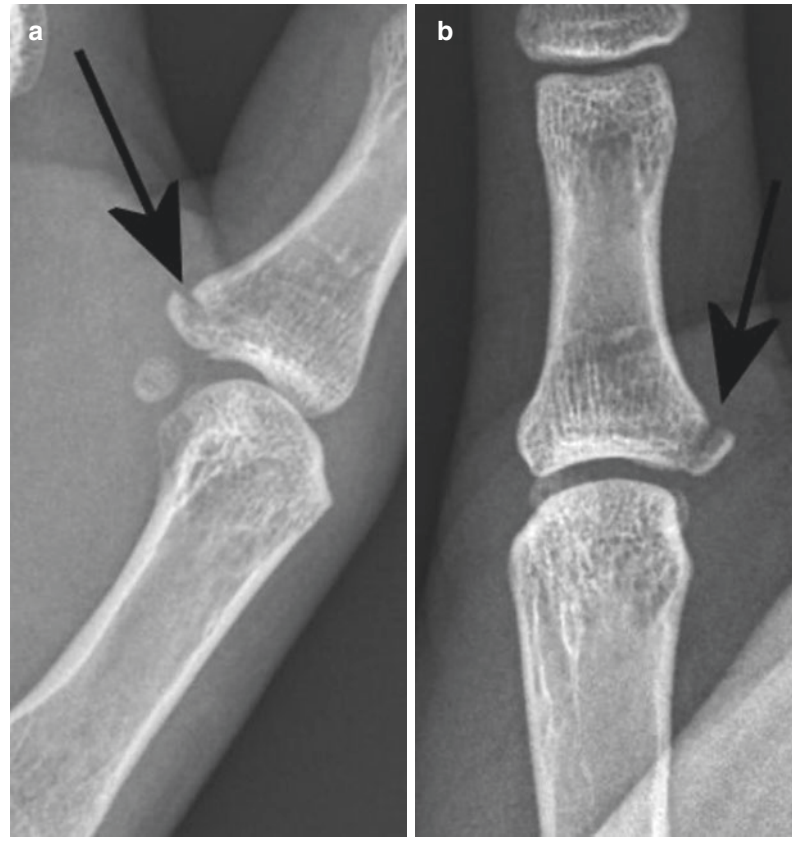

Fig. 76.12 Ulnar collateral ligament avulsion of the thumb metacarpophalangeal joint. The ulnar/medial collateral ligament is prone to injury with repetition (gamekeeper's thumb) or acutely (skier's thumb). 40% of the time, a bony avulsion will occur, usually from the proximal phalangeal base (a Lat, b PA). When undisplaced, this will heal well. When proximally displaced to the medial side of the adductor aponeurosis, surgical reduction is necessary

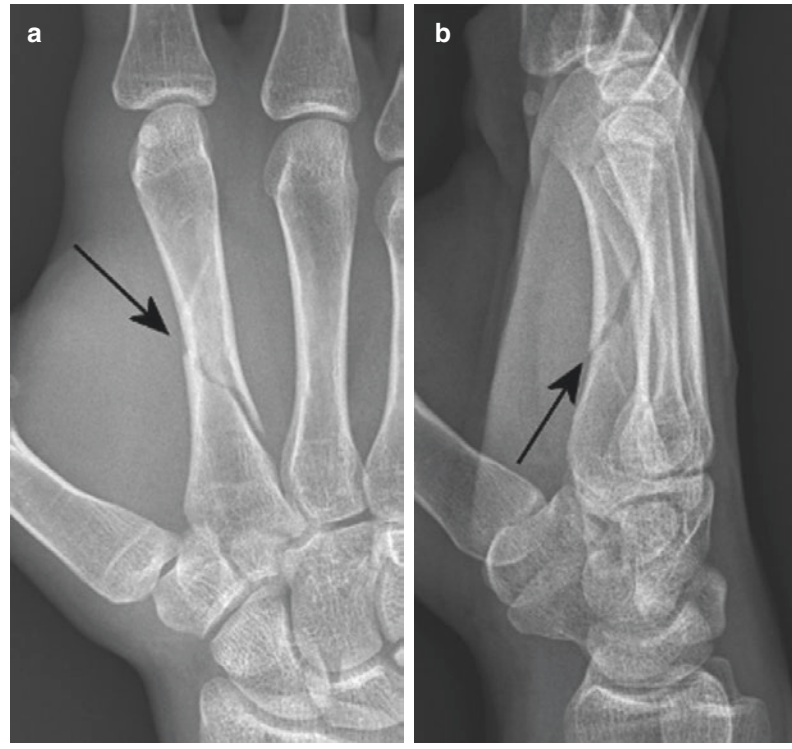

Fig. 76.13 Spiral fracture of the metacarpal shaft. A spiral fracture is one which is oblique on two views at right angles to one another (a PA, b lateral). The main thing to look for is extension into an adjacent joint, which may require surgical anatomic reduction

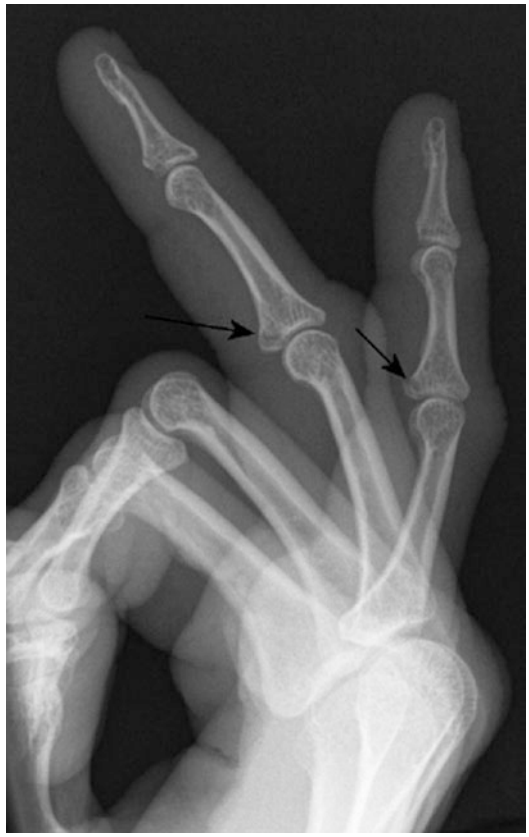

Fig. 76.14 Volar plate avulsion fractures from the bases of the fourth and fifth middle phalanges (black arrows). These occur when forced extension happens during clenching of the fist. They can be multiple as in this case, so adjacent fingers should be carefully interrogated for additional fractures

involving the extensor mechanism, where the dorsal portion of the base of the distal phalanges is avulsed off. These commonly occur with direct axial or flexion injury to the associated digit and are commonly associated with sports injuries. This injury is characterized by inability to extend the finger and slight resting flexion, which results in the finger resembling a mallet, hence its namesake. These fractures are most often identified on lateral radiograph (Fig. 76.15). Of note, patients with this clinical presentation will sometimes have isolated tendinous injuries without any associated fracture. These injuries are called "mallet finger" and can usually be diagnosed via ultrasound. Mallet injuries usually do not require

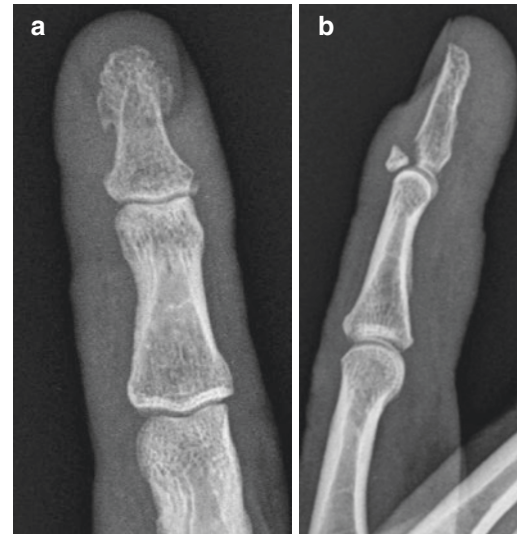

Fig. 76.15 Mallet fracture at the base of distal phalanx. These occur due to sudden forced flexion, often when a ball strikes the tip of the finger. They can be difficult to see on the PA view (**a**) but are usually obvious on the lateral (**b**). Look for secondary volar subluxation of the major fragment

operative management unless there is significant displacement, or in the management of chronic injuries.

76.3 Terminal Tuft Fractures

Fractures of the distal phalanges (Fig. 76.16) are the very common hand fractures, accounting for nearly half of all hand fractures. Notably, these fractures are common in workplace accidents, such as direct blunt trauma, for example striking the tip of the finger with a hammer that can result in significant disability. These are often associated with damage to the associated nail bed. Patients commonly have formation of a subungual hematoma, which is often more painful than the fracture itself and requires drainage, often with electrocautery, and usually provides immediate relief to the patient. Findings such as soft-tissue destruction and subcutaneous gas can suggest an open fracture, which usually has significant disruption of the underlying nail bed and pulp and can progress to osteomyelitis. Luckily,

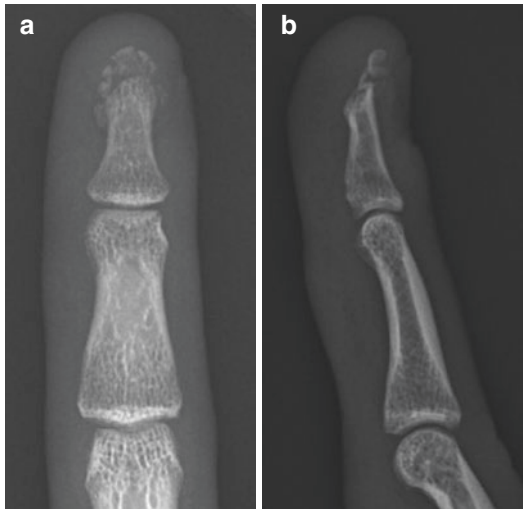

Fig. 76.16 Terminal tuft fracture. These are usually caused by direct blunt trauma such as striking the tip of the finger with a hammer (**a** PA and **b** lateral). Look for loss of the nail or gas under the nail, which could indicate an open fracture

surgical fixation is rarely required for these injuries as re-opposition of the tissues is normally adequate.

Take-Home Message

- When obtaining radiographs, one should obtain at least two different views (PA and lateral) and ideally a third view (oblique).
- In certain instances, such as when a scaphoid fracture is suspected, special views such as the scaphoid view can be obtained.
- Or, in the case of a hook of hamate or trapezial ridge fracture, carpal tunnel radiographs can also be obtained, which profiles the hook of the hamate and trapezial ridge. Despite getting additional radiograph views, certain fractures like a scaphoid fracture can still be occult on radiograph and can only be seen on cross-sectional imaging such as a CT or the more sensitive MRI. In addition to fractures, dislocations are also a serious concern. Lunate and perilunate dislocations can be seen on lateral radiographs of the wrist and are critical to identify as they can cause compression of the median nerve and potentially permanent nerve damage if treatment is delayed.
- The thumb is also a common source of fracture and ligamentous damage, as seen with the ulnar collateral ligament damage seen with gamekeeper's thumb or base of the first metacarpal fractures, such as the Bennett and the more severe Rolando fracture. Sports and fistfights are also a common source of hand injuries, particularly with fracture-dislocations of the fourth and fifth carpometacarpal, hence the apt name boxer's fracture (minimally comminuted transverse fracture of the fifth metacarpal neck). Careful attention should be given to the lateral radiographs when examining the more distal portions of the fingers, where mallet fractures (avulsion fractures involving the extensor mechanism at the base of the distal phalanges) and volar plate fractures (particularly the attachment of the proximal interphalangeal joint) with or without dislocations are most easily identified. Work-related injuries (especially with hammers) or simple everyday life injuries, such as having a car door closed on one's finger, commonly cause fractures to the terminal tuft (with or without possible damage to the nail bed). Given the commonality of hand fractures, the variety of fractures and dislocations, and the difficulties identifying these fractures, clinicians must have an in-depth understanding of where to look for fractures, what imaging to obtain when there is a suspicion, and prompt management when a fracture is identified.

Summary

The hands are vital for everyday life, and their ubiquitous use and versatile function make them a particularly common location of fractures and pathology. This same complexity also makes them vulnerable to complications and associated disability if pathology in this area is not identified correctly and in a timely manner. In the setting of trauma, one should always keep a high suspicion for fractures in the hand. In order to identify these fractures, a thorough physical exam and radiographs in at least two views (PA and lateral), and ideally oblique (as many fractures are only visible on one view), should be obtained first. Any irregularity or lucency on radiograph should also be thoroughly interrogated as non-displaced fractures are easy to be overlooked. If there is suspicion for a fracture, particularly in the carpal bones, have a low threshold for obtaining dedicated/specialized radiographs such as carpal tunnel view/scaphoid view, or obtaining a CT scan or MRI, which is more sensitive for detecting fractures. Once a fracture is identified, one should evaluate the amount of displacement, fracture plane (i.e., transverse vs. spiral vs. comminuted), degree/direction of angulation of fracture fragments, and whether there is any intra-articular involvement. Most importantly, if a fracture is identified, one should be careful not to fall victim to the "satisfaction of search." If a patient has one fracture, there is a good chance that they have additional fractures, which are commonly missed. Proper identification and description will then affect the proper management (conservative versus surgery), which will result in the greatest chance of recovery with persevered function.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. A patient presents the day after a fall on an outstretched hand and is having parethesia of the volar first, second, and third digits. What single radiographic view would be the most important to obtain?
   (a) PA radiographs
   (b) Oblique radiographs
   (c) Lateral radiographs
   (d) Scaphoid view
   The clinical concern is for median nerve compression, which can be caused by a lunate dislocation, best seen on a lateral view.
2. There is concern for fracture of one of the carpal bones after a fall. What is the most sensitive way of identifying that facture?
   (a) MRI
   (b) Standard radiographs
   (c) Scaphoid view radiographs
   (d) CT scan
   Although CT has good specificity, MRI is more sensitive for fractures.
3. What ligament is responsible for the radial/lateral retraction of bone fragments in a Bennett fracture?
   (a) Abductor pollicis longus
   (b) Ulnar collateral ligament
   (c) Anterior oblique ligament (beak ligament)
   (d) Abductor pollicis muscle
4. Which two main fracture types are best identified on carpal tunnel view?
   (a) Triquetral avulsion fracture and scaphoid fracture
   (b) Hook of hamate and triquetral avulsion fracture
   (c) Trapezial ridge and triquetral avulsion fracture
   (d) Hook of hamate and trapezial ridge
5. After a skiing accident, a patient presents with pain and swelling of the ulnar side of the first MCP joint and laxity on valgus stress. Standard trauma radiographs are obtained (PA, oblique, and lateral) and are negative. What is the most appropriate next step in management?
   (a) Repeat radiographs with a carpal tunnel view
   (b) MRI or ultrasound
   (c) CT scan
   (d) Immobilization for 4–6 weeks

Further Reading

Christian Fox J. Clinical emergency radiology, 2nd Ed. Cambridge: Cambridge University Press; 2017;ISBN-13:978–1107065796. ISBN-10: 1107065798

Rogers LF, West OC. Imaging skeletal trauma. 4th ed. Philadelphia, PA: Saunders; 2015. ISBN: 9780323278195

Longo UG, De Salvatore S, Mazzola A, Salvatore G, Mera BJ, Piergentili I, Denaro V. Colles' fracture: an epidemiological nationwide study in Italy from 2001 to 2016. Int J Environ Res Public Health. 2023;20(5):3956.

Part XV

Neurological Pathologies

Traumatic Nerve Injuries

77

Mark Henry

Overview

Traumatic peripheral nerve injuries threaten permanent loss of function. The risk of functional loss is proportionate to the type and extent of nerve injury and the distance from target receptors. Many closed nerve injuries will recover spontaneously without intervention. Lacerations should be repaired or reconstructed promptly to permit regenerating axons to reach their receptors as soon as possible. Sensory receptors are more tolerant of delayed reinnervation. Motor end plates become permanently depleted over time, precluding functional recovery if regenerating axons arrive later than 12–18 months after injury. The mechanism of injury, clinical examination, and ancillary tests guide treatment decisions that include various surgical techniques to reinnervate distal receptors.

77.1 Definition

Traumatic injury to a peripheral nerve includes any acute process that compromises nerve function. Closed mechanisms of injury include blunt force trauma involving sudden compression or traction forces. Open mechanisms of injury range from pure sharp lacerations to combinations of cutting, crushing, and tearing.

77.2 Epidemiology

Unlike some other medical conditions, traumatic nerve injury is not limited to any select population. From the very young to the very old, any patient can encounter a set of circumstances leading to nerve injury. The greatest incidence occurs during the working years of adult life when patients are most exposed to conditions likely for nerve injury.

77.3 Etiology/Pathogenesis

Sharp laceration accounts for the majority of nerve injuries. Common environments include the kitchen where knives and broken glass easily slip and cut nerves in the hand and wrist. Punching or falling through glass panes can lacerate nerves in the more proximal upper extremity. Laceration by sharp edges of metal occurs in

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UG. Longo, V. Denaro (eds.), *Textbook of Musculoskeletal Disorders*,
https://doi.org/10.1007/978-3-031-20987-1_77

manufacturing workplaces. Industrial machinery or vehicular trauma can impart heavy crush or avulsion injury to nerve. Even sleeping with the body weight on a nerve for many hours is enough to wake with a nerve palsy.

77.4 Classification

The cross-sectional architecture of a peripheral nerve consists of numerous individual nerve cells (axons) traveling together, longitudinally connecting the proximal cell bodies in the spinal column to the distal sensory and motor receptor. Each axon is enveloped in a conductive sheath of myelin protein manufactured by Schwann cells and housed in its own structural conduit, the endoneurial tube. A Sunderland grade I injury disrupts the myelin sheath or endoneurial environment enough to cease nerve function, but the axon itself survives. Sunderland grade II signifies injury to the nerve cell resulting in axon death, but the endoneurial tube is not disrupted. Multiple axons in their endoneurial tubes are grouped together into fascicles encased by the next structural layer called perineurium, an extension of the blood-brain barrier. Sunderland grade III injuries disrupt the axons and their endoneurial tubes, but the perineurium remains intact. Sunderland grade IV injuries further disrupt the protective perineurium, leaving intact only the most peripheral encasement, epineurium. With disruption of the epineurium, Sunderland grade V injuries render the nerve fully separated with an intervening gap. The original five-part Sunderland classification was subsequently modified to include grade VI, representing a pattern of mixed grades occurring in adjacent fascicles. Different types of nerve injury are reported in Fig. 77.1.

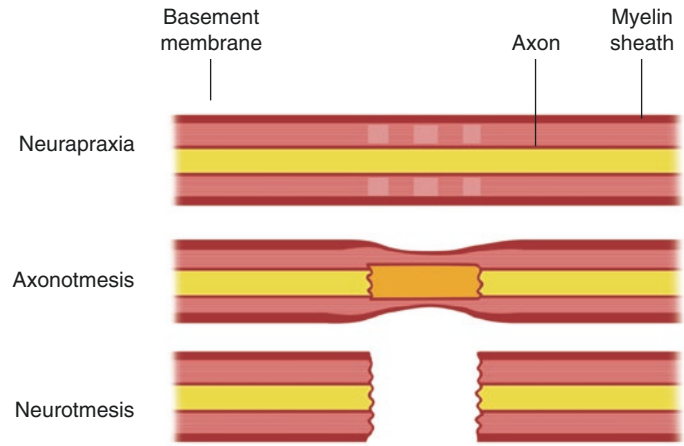

Fig. 77.1 Different types of nerve injuries

77.5 Diagnosis

Peripheral nerves may be pure motor, pure sensory, or mixed. Diagnosis begins with the history of injury, focusing on the mechanism and anatomic location. This establishes the likely type and grade of injury and the specific nerve(s) involved. Assessment continues with the physical examination, mapping the exact distribution of functional loss, sensory and motor. In most cases, this information is sufficient to establish the treatment plan. More complex scenarios may leave uncertainty regarding multilevel injuries that overlap to produce the observed deficits. With closed mechanisms of injury, the Sunderland grade cannot be known with certainty just based on the clinical deficit alone.

The most frequently employed ancillary study is electrical nerve testing. There are two components: nerve conduction study (NCS) and electromyography (EMG). During NCS, abnormalities of conduction are tested with a proximally applied electrical impulse recorded from a distal surface electrode. During EMG, thin needles inserted into specific muscles produce characteristic waveforms when stimulated, providing information about the condition of the nerve supplying that muscle. Similar techniques are also used during surgery with electrodes applied directly to the surface of the nerve to guide intraoperative treatment decisions. Less utilized are imaging studies: magnetic resonance imaging and ultrasound. Imaging studies have varying degrees of accuracy to reveal physical continuity of the nerve, constriction vs. enlargement, or signal intensity changes indicating response to injury.

77.6 Treatment

Optimal treatment is based on the contributing variables: mechanism of injury, age of the patient, location and time since injury, and specific motor/sensory deficits.

77.6.1 Nonoperative Treatment

The majority of closed mechanism of nerve injury traumas will ultimately recover spontaneously over the appropriate time period based on the grade of injury and distance from target receptors. For that reason, most closed injuries are initially observed for anticipated recovery. A baseline NCS/EMG is obtained at 3 weeks from injury. The relevant findings that distinguish a Sunderland I from higher grade injury will not be evident if obtained too early. In select rare cases, unique circumstances may indicate a higher grade injury and prompt early surgical evaluation including intraoperative electrical nerve testing. There are no current medical therapies that enhance nerve regeneration. Patients are taught therapy exercises while awaiting regeneration: desensitization, passive joint and tendon range of motion, and splinting to prevent contracture. If there are no clinical signs of functional recovery, a repeat NCS/EMG may demonstrate electrical signs of reinnervation that precede clinical signs. Distal motor receptors, motor end plates, are irreversibly lost over time. Potential to regain function declines at a rate of approximately 1% per week. Axons regenerating at 1 mm/day must reach the motor end plates by 12–18 months after initial injury for any meaningful recovery. The presence or absence of clinical/electrical signs of reinnervation guides appropriately timed next interventions.

77.6.2 Neurolysis

The decision to pursue neurolysis is typically made by 3–4 months following injury, certainly no later than 6 months. The surgical plan must include a treatment strategy to account for each possible variation of injury pattern. Neurolysis alone, as opposed to nerve graft or transfer, is indicated by demonstration of axon regeneration across the site of injury. Regenerating axons will have traveled a sufficient distance after 3–4 months to place the stimulating and recording electrodes far enough apart to accurately measure nerve action potentials (NAP). While there is no universally agreed-upon criterion, the amplitude of the NAP across the zone of injury should be at least 50% that of an NAP obtained proximal to the zone of injury. All dense investing scar tissue is removed from around the nerve at the site of injury along with decompression

performed at known sites of anatomic narrowing such as the carpal or cubital tunnel passages.

77.6.3 Direct Repair

Unlike closed nerve injuries, direct nerve lacerations should be taken emergently to the operating room for direct repair. Within only a matter of days, tension-free approximation may become impossible because of retraction of the nerve stumps, local tissue edema, and fibrosis. Nerve suture is performed at high magnification under the operating microscope. Specialized microsurgical instruments permit delicate handling of the tissues and precise manipulation. Each nerve ending is trimmed perpendicular to its longitudinal axis, creating fresh margins of undamaged tissue without protruding fascicles. Using 9-0 or 10-0 nylon, the least number of sutures are placed only through epineurium to avoid damaging fascicles. Each fascicle should be aligned with itself across the junction. Multiple clues guide correct alignment including epineurial vessels and pattern of different-sized fascicle groups. The edges of the epineurium should just meet each other circumferentially and create a perfect cylinder without gaps. Fascicles should neither buckle nor escape. Forcing the two ends together too tightly distorts the repair, misaligns fascicles, and can hinder regeneration. The repair should remain intact without gaps when the adjacent joints are positioned in neutral extension or the range that permits limb function. If a reasonable limb position cannot be achieved without undue tension, consideration must be given to interposing a nerve graft. Axon regeneration is impaired across a junction under tension. The outcomes of nerve repair are influenced by multiple variables, the most powerful of which is patient age. Single-digit-aged children can achieve recovery indistinguishable from normal in many circumstances. The quality of regeneration and recovery then diminishes into the teens and adulthood. The second most important variable is distance to target receptors combined with time delay between injury and repair. Sensory receptors retain the capacity for reinnervation longer than the 1% per week attrition of motor recovery potential. Mixed nerves require proper alignment of motor and sensory fascicles; any misdirected axon regeneration will not result in functional recovery. Certain nerves with specific functional roles demonstrate relatively consistent outcomes such as the well-established hierarchy of recovery for radial nerve over median nerve over ulnar nerve [1]. Clean, sharp lacerations are more favorable as opposed to avulsion mechanisms that impart traction injury over substantial distance from the point of disruption.

77.6.4 Nerve Grafting

Nerve grafting is indicated for direct lacerations that cannot be repaired without excessive tension or Sunderland grade III–V crush/avulsion injuries that show no evidence of regeneration by intraoperative NAP. The key to successful grafting is elimination of damaged tissue on both the proximal and distal ends. Any fibrosis obstructs the endoneurial tubes and blocks regeneration. Standard teaching called for starting with the damaged end and trimming sequentially under the microscope until the cut end appeared normal. This often results in a repair margin that is still compromised. The better method is to begin the microscope examination in undamaged nerve tissue and progress towards the zone of injury. As soon as tissue change is encountered, that represents the correct margin for resection. After resecting damaged nerve tissue from both ends, the limb is positioned at full length to determine the tension-free size of the defect. A structural pathway for axon regeneration is then placed between the two ends in the form of nerve autograft or allograft [2]. The same surgical techniques employed in direct nerve repair apply to nerve grafting. Autograft refers to sections of nerve harvested from expendable sensory branches. The most common donor is the sural nerve that supplies sensation over the lateral ankle. Autograft provides a structural path for regeneration, nerve growth factors, and viable Schwann cells that are critical to promote regeneration over greater distances. To avoid the additional surgery and donor-site morbidity of autograft, hollow conduits were studied for decades. Conduits ultimately proved inferior to the other means of avoiding autograft, cadaveric nerve allograft. To avoid antigenicity, allograft must be laboratory processed to remove cellular

material as well as tissue debris that obstructs the endoneurial tubes. Allografts still contain structural proteins and nerve growth factors but lack Schwann cells. Outcomes of nerve grafting are influenced by the same variables as direct repair, in addition to the length and type of graft matter. At shorter distances, the absence of Schwann cells has not created a discernible difference in outcomes. But with increasing defect size, especially for critical motor targets, autografts prove superior to allografts. The longest allografts available are 5–7 cm.

77.6.5 Nerve Transfer

When all of the contributing variables are considered and recovery potential from repair or graft is predicted to be minimal, nerve transfer may offer a better outcome. A nerve transfer refers to dividing an expendable donor nerve at a distal location and connecting it to the distal receiving end of a nerve that has been injured at a more proximal location [3]. The regenerating axons transferred from the donor nerve will arrive at the terminal receptors of the recipient nerve long before any regenerating axons from the proximal injury site. For example, outcome studies indicate no recovery of intrinsic muscle function in the hand following an ulnar nerve avulsion defect proximal to the elbow. Partial reinnervation of the hand intrinsic muscles can be accomplished by transferring the distal anterior interosseous nerve (AIN) to the ulnar motor branch. The junction of the nerve transfer is much closer to the progressively deteriorating motor end plates. Axons from the AIN can arrive within 12 months, whereas regeneration from proximal to the elbow would require more than 2 years. Nerve transfer is primarily used to restore motor function but can also be performed to restore critical sensory functions such as the ulnar digital nerve of the thumb and radial index finger. Principles of nerve transfer include tension-free coaptation, matching axon counts, and pure motor or pure sensory pairing. Nerve transfers continue to be studied with certain combinations demonstrating better outcomes than others. Some nerve transfers do not conflict with subsequent procedures, but for others, the donor nerve supplies the same muscle group that would otherwise be used in a tendon transfer to solve the same original deficit. If the nerve transfer fails, the corresponding tendon transfer has also been eliminated as an option.

77.6.6 Tendon Transfer

Before the advent of nerve transfers, motor deficits following nerve injury were managed by tendon transfers. Tendon transfer involves distal division of an expendable muscle-tendon unit with rerouting and connection to the recipient tendon associated with the lost motor function. Just like nerve transfers, some tendon transfers inherently work better than others. Principles of tendon transfer include matching excursion and power, a straight line of pull, one transfer for one function, and using muscles that naturally contract in the same phase of movement. One of the most reliable transfers, for a non-reconstructible radial nerve injury, restores digital extension using a wrist flexor muscle. Debate continues as to which is best in cases where either a nerve transfer or a tendon transfer could be used. Although a successful nerve transfer reinnervates the original muscle-tendon unit, nerve transfer has greater potential for complete failure. Also, nerve transfers must still be accomplished within the time window for motor recovery, but tendon transfers can be performed long after the original injury.

Take-Home Message

- Nerve lacerations should be taken emergently to the operating room for direct repair.
- Most closed nerve injuries should be initially followed nonsurgically and methodically evaluated with a timed plan for intervention in the absence of recovery.
- If a tension-free direct repair of undamaged nerve tissue cannot be achieved, nerve graft or nerve transfer is indicated.
- Tendon transfer remains the ultimate salvage for late-presenting cases or other circumstances precluding useful reinnervation.

Summary

Traumatic nerve injury immediately deprives the patient of the motor or sensory functions served by that nerve. Lesser degrees of closed mechanism injury typically recover with patience and time. Open lacerations are repaired immediately, but ultimate recovery is influenced by multiple variables. Delayed nerve reconstructions using more complex techniques of nerve graft and transfer demonstrate even further diminished outcomes.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. What is the difference between a Sunderland I and II injury?
   (a) The nerve cell survives in a Sunderland I injury and clinical recovery occurs with re-myelination, but Sunderland II injuries require axon regeneration
   (b) The nerve cell survives in a Sunderland II injury and clinical recovery occurs with re-myelination, but Sunderland I injuries require axon regeneration
   (c) Sunderland I injuries result in complete nerve cell death, leading to no clinical recovery
   (d) Sunderland II injuries cause immediate clinical recovery without the need for re-myelination
2. How long after closed nerve injury should the initial NCS/EMG be performed?
   (a) After 3 weeks, the EMG changes appear that distinguish a Sunderland I from higher grade injury
   (b) After 1 week, the EMG changes appear that distinguish a Sunderland I from higher grade injury
   (c) The EMG must be performed immediately after injury to accurately diagnose nerve damage
   (d) NCS/EMG is not a reliable method for diagnosing closed nerve injuries
3. When repairing a nerve laceration, what is the main indication to use a graft?
   (a) Excess tension or gap at the repair site
   (b) Signs of infection or association with exposed fractures or gap at the repair site
   (c) Grafts are mainly used for cosmetic purposes and are not essential for nerve repair
   (d) Grafts are never recommended for nerve repair due to the increased risk of complications
4. In nerve repair or reconstruction, which variable has the greatest impact on outcome?
   (a) Young patient age has the greatest impact on clinical recovery
   (b) Sex has the greatest impact on clinical recovery
   (c) The severity of the initial nerve injury has the most significant influence on outcome
   (d) Genetic factors are the primary determinant of clinical recovery following nerve repair
5. What is the main advantage of tendon transfer over nerve transfer?
   (a) There is no time limit following injury to perform a tendon transfer
   (b) It requires less costs and days of hospitalization
   (c) Tendon transfers have a higher risk of complications compared to nerve transfers
   (d) Tendon transfers can only be performed on upper limbs, limiting their applicability

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Canalicular Syndromes of the Upper Limbs

78

Ismail Turkmen and Muhlik Akyürek

Overview
This chapter includes canalicular syndromes that cause entrapment neuropathies in the upper limbs. Cervical rib syndrome, scalenus anterior syndrome, thoracic outlet syndrome, suprascapular nerve syndrome, carpal tunnel syndrome, anterior interosseous syndrome, pronator teres syndrome, supracondylar process syndrome, cubital tunnel syndrome, Guyon canal syndrome, posterior interosseous syndrome and other entrapment neuropathies of the upper limbs are described in this chapter.

78.1 Introduction

Several canicular syndromes occur in the upper limb as a result of the anatomical position and the path of nerves supplying the muscles of the arm and forearm. Many of these occur in regions which involve tight areas that the nerve traverses and are common sites of entrapment resulting in pathology. Understanding the anatomy of these nerves with a focused neurological examination can allow diagnosis and appropriate management. In this chapter, we discuss the main canicular pathologies that present in the general orthopaedic clinic with a focus on the three most common canicular syndromes: carpal tunnel syndrome (CTS) (Fig. 78.1), cubital tunnel syndrome and posterior interosseous nerve (PIN) syndrome.

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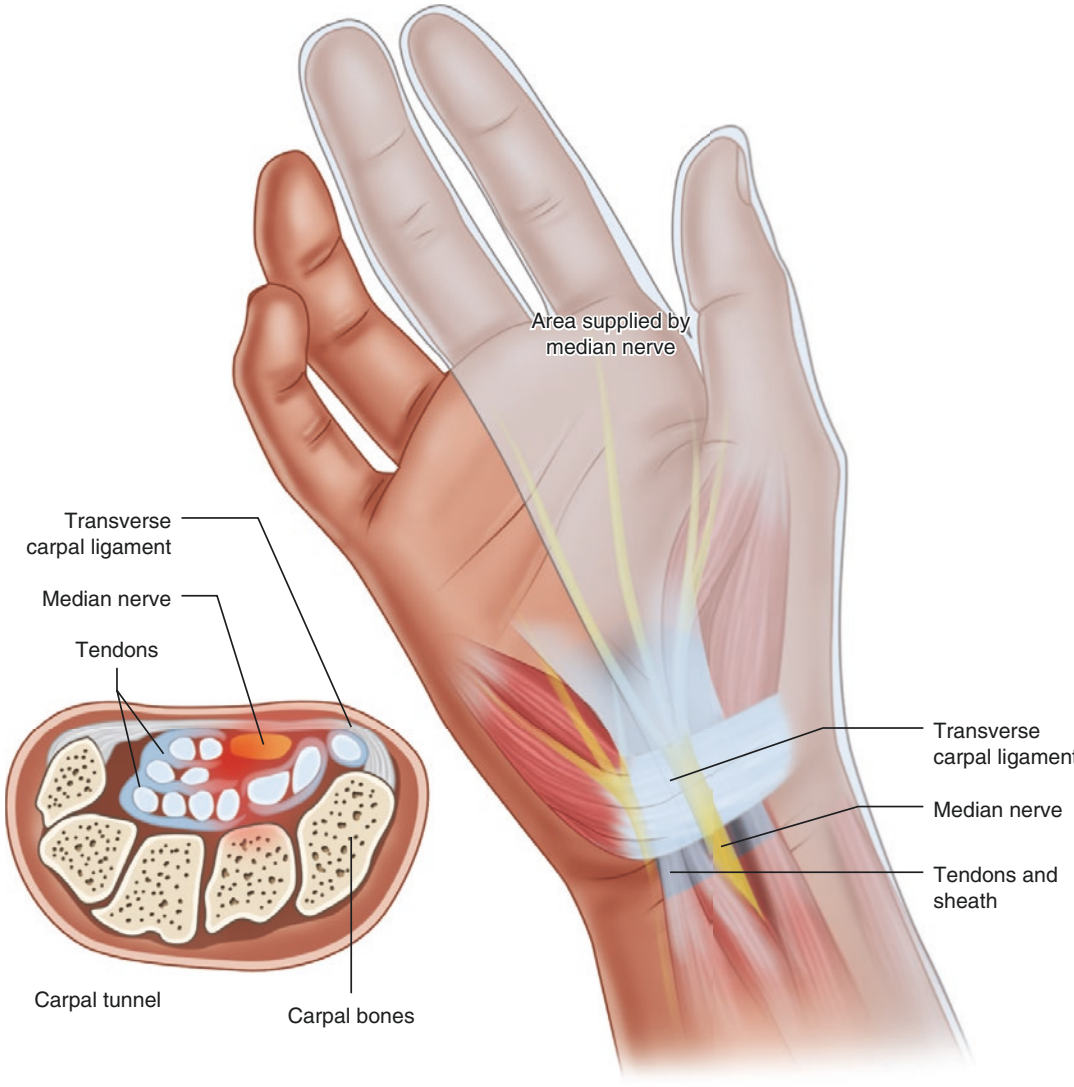

Fig. 78.1 Carpal tunnel syndrome

78.2 General Approach to Canalicular Syndromes

A thorough neurological and orthopaedic examination, coupled with a detailed history, can usually allow the nerve in question to be determined. This requires an interrogation of symptom progression and the location of affected regions. Particular attention is paid to the duration of symptoms, i.e. whether insidious or acute in nature. Associated symptoms include weakness of certain muscles pertaining to the nerve and paraesthesia in a specific dermatome. In the majority of cases, this can be associated with certain movements, which may exacerbate some of the symptoms.

Further assessment of the nerve is carried out by two approaches: nerve conduction studies and magnetic resonance imaging (MRI) to assess if there is localised pressure on the nerve as a result of inflammation or anatomical constraints around the nerve. Evidence of neuropraxia can be ascertained through nerve conduction studies, which will show abnormal signalling when the nerve is stimulated. These are typically reserved for patients that have borderline clinical history suggestive of a canicular disorder

and may help with the underlying diagnosis of the condition with a view towards surgical management. Use of MRI can allow identification of the structures surrounding the nerve in question for surgical planning in the presence of stenosis. This has largely superseded the use of ultrasonography due to better visualisation of the affected anatomy.

78.3 Anatomy of the Upper Limb Nerves

Five nerves are responsible for the innervation of the muscles of the arm, forearm and arm: the axillary, musculocutaneous, radial, median and ulnar nerves. Each of these derives from the brachial plexus, a combination of intertwining anterior rami nerve roots from C5 to T1, which divide sequentially into trunks, divisions, cords and finally nerve proper. It should be noted that several nerves also exit from the brachial plexus prior to reaching the cords and may contribute to symptomology affecting the upper shoulder and the ability to move the pectoral girdle but are beyond the scope of this chapter. A summary of the five nerves is shown in Table 78.1.

Upper extremity nerve entrapments are summarised in Figs. 78.2 and 78.3

Table 78.1 The terminal and sensorimotor innervation of the upper limb nerves

Nerve	Roots involved	Muscle supply	Movement	Cutaneous
Musculocutaneous nerve	C5–C7	Coracobrachialis, biceps brachii, brachialis	Flexion and supination at elbow	Anterolateral aspect of forearm
Axillary nerve	C5–C6	Deltoid, long head of triceps and teres minor	Abduction, flexion and extension of shoulder	Lateral aspect of upper arm "regimental patch"
Median nerve	C6–T1	Flexors of the forearm **except** FCU and lateral half of FDP, lateral two lumbricals, thenar eminence	Wrist flexion, forearm pronation, distal phalanx flexion, opposition and flexion of thumb	Thumb + radial two and a half fingers of the palm in the anatomical position
Ulnar nerve	C8–T1	Medial half of FDP, FCU, intrinsic muscles of the hand	Ulnar deviation of the wrist, fine finger movement	Ulnar one and a half digits of the palm
Radial nerve	C5–T1	Triceps brachii, all extensors of posterior compartment of forearm	Arm and wrist extension	Posterior forearm and dorsal radial three and a half digits

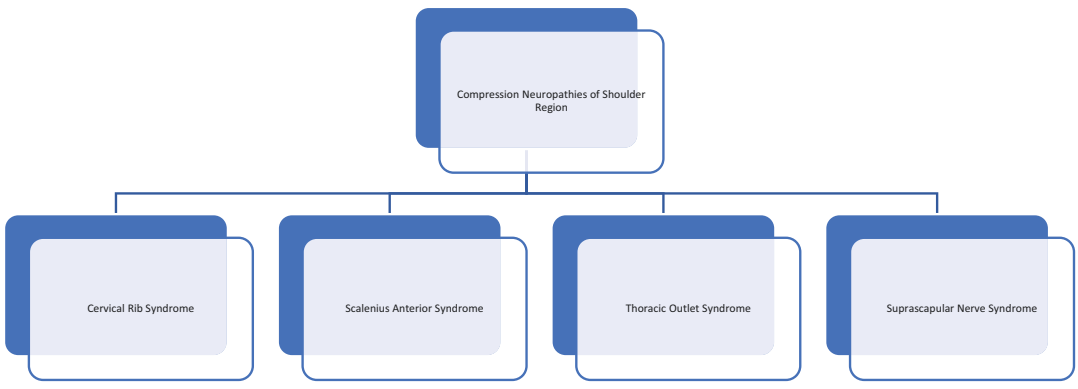

Fig. 78.2 Canalicular syndromes of the shoulder region

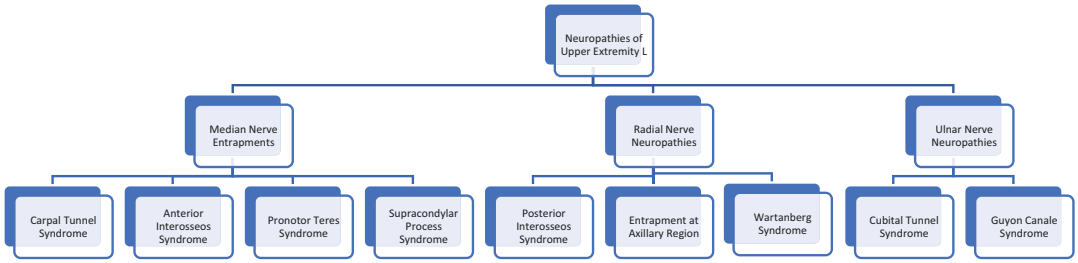

Fig. 78.3 Canalicular syndromes of upper extremity

78.4 Musculocutaneous Nerve

78.4.1 Overview

Isolated compressive musculocutaneous nerve neuropathy has been sparsely described in the literature and is uncommonly seen in the orthopaedic or neurological clinic. It has been noted that isolated injuries can occur as a result of strenuous exercises involving repetitive elbow flexion and extension (such as weight lifting and rowing) or repeated pronation and supination. The typical site of compression is at the aponeurosis of the biceps muscle between the tendon and the fascia of the brachialis muscle.

78.4.2 Anatomy

The musculocutaneous nerve arises from the lateral cord of the brachial plexus at the level of the inferior border of the axilla with the majority of its contribution from the anterior rami of roots C5 and C6. At this level, it supplies the coracobrachialis muscle and subsequently pierces it to supply both the brachialis and brachioradialis muscles. It continues to traverse down the mid-arm to reappear laterally superficial to the biceps tendon where it terminates as the lateral cutaneous nerve of the forearm, providing sensation to the lateral aspect of the forearm. The musculocutaneous nerve is usually found 2 cm lateral to the biceps tendon.

78.4.3 Presenting Features

Typical presentations include paraesthesia over the lateral forearm in keeping with neuropathy affecting the lateral cutaneous nerve of the forearm along with weakness in flexion and supination of the elbow. Diminished bicipital tendon reflexes may also be noted. Provocation of symptoms occurs with supination and pronation movements.

78.4.4 Treatment Options

The majority of these injuries occur as a result of repetitive strenuous activity, and cessation of this leads to complete symptom control in the majority of cases. Steroid injection is indicated in some cases to reduce inflammation around the nerve. Surgical decompression has been championed in the few cases seen in the literature but is dependent on the anatomical features seen. Partial resection of the lateral margin of the bicipital aponeurosis has shown to provide some benefit in patients who have neuropathy of the lateral cutaneous nerve of the forearm.

78.5 Axillary Nerve

78.5.1 Overview

Axillary nerve neuropathy typically occurs in the context of fractures of the proximal humerus and shoulder dislocation resulting in traction injuries

of the nerve. The main canalicular syndrome in this region involves compression of the nerve as a result of obliteration of the quadrilateral space, resulting in the phenomenon known as quadrilateral space syndrome (QSS). Due to the lack of literature and misdiagnosis of this particular condition, there is a lack of data to show how common this condition is in the general population, but it is well established to occur in the young patient in their dominant arm following repetitive overarm action. It is thought to be a result of a culmination of hypertrophy of the muscles making up the quadrilateral space as well as the development of fibrous bands following microtears associated with repetitive overarm movement, particularly shoulder abduction and external rotation. Patients may also have concomitant digital ischaemia related to posterior circumflex humeral artery (PCHA) occlusion as this also transmits through the space with the axillary nerve.

78.5.2 Anatomy

The axillary nerve derives from the posterior cord of the brachial plexus and has the majority of its contribution from the anterior rami of the C5 nerve root with a minor contribution from C6. Lying posteriorly to the axillary artery, the nerve exits the axilla below the inferior border of the subscapularis muscle and through the quadrilateral space, a site of compression of the nerve.

The nerve passes medial to the surgical neck of the humerus and divides into an anterior and posterior bundle along with a small articular branch, which supplies the glenohumeral joint. The anterior bundle loops around the surgical neck, innervating the anterolateral aspect of the shoulder. The posterior bundle is involved in innervation to the posterior aspect of the deltoid muscle and teres minor. This terminates as the upper lateral cutaneous nerve of the arm and provides sensation to this region. Damage to this branch results in the "regimental patch" anaesthesia seen in axillary nerve palsy.

78.5.3 Presenting Features

Presentation typically consists of the young patient with the dominant arm affected. Poorly localised, vague shoulder pain aggravated by flexion and external rotation of the arm is the main feature. Atrophy of the deltoid muscle may be seen in severe cases and may be associated with weakness in abduction and external rotation of the arm. Paraesthesia involving the lateral aspect of the arm has also been described. Digital ischaemia is commonly reported in association with PCHA occlusion within the quadrilateral space due to the anatomical proximity of the artery and nerve.

78.5.4 Investigations and Diagnosis

Given the vague nature of the symptomology of this condition, a thorough neurological and vascular examination must be performed in the outpatient setting. Features of both warrant investigation by use of MRI and/or magnetic resonance angiography (MRA). This helps delineate several differential diagnoses including rotator cuff injury, C5/C6 cervical root radiculopathy, thoracic outlet syndrome, ganglion, paralabral cyst and glenoid labral cyst. Evidence of cystic structures around the quadrilateral space may be amenable to arthroscopic decompression in some instances. Electromyography studies are usually normal but can be useful in ruling out differential pathologies including Parsonage-Turner syndrome and thoracic outlet syndrome. MRA allows assessment of vascular occlusion of the PCHA, which may be amenable to thrombolysis or vascular transposition.

78.5.5 Treatment Options

Variability in treatment options has been documented in the literature due to the scarcity and probable underdiagnosis of quadrilateral space syndrome. A propensity towards conservative management through use of non-steroidal anti-

inflammatory medication, physiotherapy and rest has been shown to improve symptoms in several cases. Symptomatic relief by use of local anaesthetic and steroidal injection has been suggested as both diagnostic and therapeutic management.

Surgical management options are limited but may be indicated if extrinsic causes exist including arthroscopic decompression of paralabral and glenoid labral cysts. Identification of fibrous bands on MRI may warrant surgical exploration if symptoms do not improve after 6 months of conservative management.

78.6 Median Nerve

78.6.1 Overview

There are three canalicular syndromes that commonly affect the median nerve: carpal tunnel syndrome (CTS) distally, and both anterior interosseous nerve syndrome (AINS) and pronator teres syndrome (PTS) proximally. All three provide a different subset of presenting symptoms, with CTS being most common by far. CTS occurs in approximately 3.8% of the general population with an incidence rate reported at 276 per 100,000 per year. However, the condition is probably underreported, and subacute cases are likely to be present in the community.

The most common aetiology is idiopathic with peak incidence between the ages of 40 and 60 and having a predilection towards the female sex. The latter is thought to be a result of the smaller bony anatomy compared to their male counterparts. Although the majority is idiopathic, work-related factors include repetitive movements of the wrist that may contribute to the development of the condition. Those with an underlying inflammatory condition, such as rheumatoid arthritis, obesity, diabetes and body states such as pregnancy, are known to increase the risk of carpal tunnel syndrome. These may be transient as is the case in pregnancy or patients that rapidly lose weight but can also be chronic. Diabetic patients are at a greater than twofold risk. New genome-wide association studies are now pointing towards genetic susceptibility as a cause for developing the condition.

78.6.2 Anatomy

The median nerve is an amalgamation of the medial and lateral cords and receives contributions from the anterior rami of roots C6–T1. It is crucial in allowing manipulation of the forearm and use of hand (in particular thumb opposition). It traverses antero-medially to the humerus in the arm, lateral to the brachial artery at first until mid-humerus where it crosses over to lie medially. While descending down the arm, the nerve does not give any branches.

At the elbow, the median nerve dives in between the two heads of pronator teres, a key area in which compression of the nerve can occur. The anterior interosseous nerve (AIN) divides in this region and travels deep onto the interosseous membrane to supply part of flexor digitorum profundus, flexor pollicis longus and pronator quadratus. The median nerve proper continues between flexor digitorum superficialis (FDS) and flexor digitorum profundus (FDP) to supply all the flexors of the forearm except for flexor carpi ulnaris before entering the carpal tunnel, a site of compression of the nerve. Prior to entering this enclosed region and transmitting into the hand, the median nerve gives off a clinically important branch, the palmar cutaneous branch of the median nerve at approximately 4–8 cm proximal to the volar wrist crease. This branch continues radially to the median nerve proper to supply the sensation to the palm and base of the thenar eminence.

The carpal tunnel is a space formed by the transverse carpal ligament superiorly and the concavity of the proximal carpal row inferiorly. In this tight space, the four tendons of FDP and FDS, one tendon of FPL and median nerve all transmit into the hand. This is the most common region of compression of the median nerve and is the culprit in the development of carpal tunnel syndrome.

The median nerve exits the tunnel to terminate as the palmar digital branches responsible for the sensory innervation of the radial volar three and a half digits and motor input to the lateral two lumbricals along with the recurrent motor branch of the median nerve which innervates the muscles of the thenar eminence.

Median nerve neuropathies consist of carpal tunnel syndrome, anterior interosseous nerve syndrome, pronator teres syndrome and supracondylar process syndrome.

Median nerve innervates from C6–T1 spinal nerve roots. It consists of the upper-middle-lower trunk of the brachial plexus. The most common compressive neuropathy in the upper limb is **carpal tunnel syndrome** (CTS). CTS is a condition that takes place with compression of the median nerve under the carpal transverse ligament at the wrist level. The main complaints are pain, tingling and numbness in the first three fingers of the hand. Both hands are generally affected. Patients present with the complaint of numbness that appears at night or in the early morning. Although CTS is usually idiopathic, some systemic diseases such as diabetes, thyroid disorders, pregnancy and occupational conditions can also be considered among the causes of CTS. Carpal tunnel syndrome diagnosis can be made clinically. EMG is often useful in the diagnosis. In the early stage of the disease, patients with moderate symptoms have normal electrophysiological test. To diagnose CTS, some provocative manoeuvres, such as Phalen and Tinel tests, are available. Phalen test is the occurrence of paraesthesia by keeping the wrist in flexion for 1 min. Tinel test is the occurrence of paraesthesia with the pressure applied on the carpal tunnel while the wrist is in extension. In CTS, as the motor fibres begin to be affected over time, atrophy begins to occur in the lumbrical muscles of the second and third fingers. Complaints of hand weakness and incompetence (dropping something from the hand, inability to do fine hand skills) also begin to come into view in the patient. Prolonged use of the wrist in flexion or extension position further aggravates the symptoms. Conservative approaches or surgical treatment can be preferred in CTS treatment. The aim of treatment is to reduce the compression force on the median nerve. Conservative approaches aim to reduce inflammation in the flexor tendon synovium. The first step of conservative treatment is splint treatment to rest inflamed tissues. Patients are recommended to use these splints that keep the wrist and the metacarpophalangeal (MCP) joint in neutral position for 1 month. Other conservative treatment methods, non-steroidal anti-inflammatory drugs, steroid injection, physical therapy modalities (most commonly ultrasound, paraffin, iontophoresis, transcutaneous electrical nerve stimulation, laser, massage, tendon and exercises) and hand therapy applications are the most commonly used methods. When symptoms begin to subside, stretching exercises involving the wrist and fingers are added to the programme. If no significant improvement is seen after 8–12 weeks despite conservative treatments, surgical treatment is indicated. Carpal tunnel release is performed in surgical treatment. Carpal tunnel release surgery is applied in two different ways, open or endoscopic, in order to create more space in the carpal tunnel for the median nerve and reduce the pressure by releasing the transverse carpal ligament (Fig. 78.4). In terms of hand rehabilitation, endoscopic surgeries appear to be more advantageous because of less scar formation and no incision on

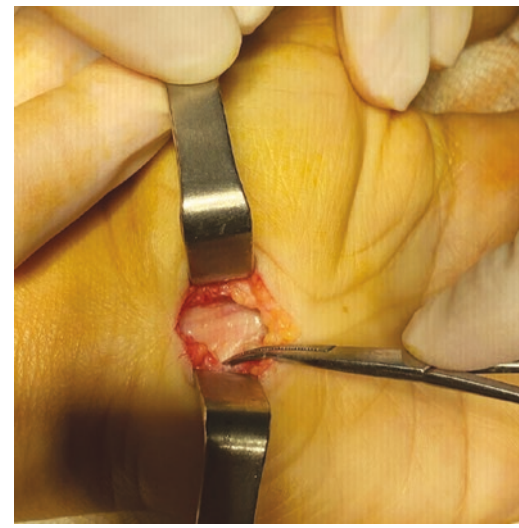

Fig. 78.4 Transverse carpal ligament release

the carpal tunnel. The time to return to work after endoscopic surgery is much shorter in this way. Postoperative hand therapy applications should be started as soon as possible. After both open and endoscopic carpal tunnel release surgeries, finger movements are begun within the first day, while wrist movements are usually begun within 1 week.

78.7 Ulnar Nerve

78.7.1 Overview

Both cubital tunnel syndrome and Guyon canal syndrome are common canalicular ulnar neuropathies. Many people have experienced features of the disease when accidentally hitting the medial elbow, resulting in the "funny-bone" sensation. Cubital tunnel syndrome is the second most common canicular pathology of the upper limb, with a prevalence of 1.8–5.9%. Typically, it is due to compression of the nerve at the elbow within the cubital tunnel. Other areas where the ulnar nerve can compress at the elbow include the arcade of Struthers, medial epicondyle, medial intermuscular septum and deep flexor aponeurosis.

Guyon canal syndrome is rare and is associated with chronic compression of the canal against bicycle handlebars at the wrist (hence given the eponymous name bicycler's neuropathy).

78.7.2 Anatomy

The ulnar nerve arises from the medial cord, which itself derives from the anterior rami of the C8 and T1 roots. This travels in the medial arm towards the elbow, medial to the medial epicondyle of the humerus. The nerve traverses into the anterior compartment of the forearm and initially gives off two muscular branches, with one supplying flexor carpi ulnar and the second supplying the deeper medial half of flexor digitorum profundus. It continues to traverse down the forearm until about 5 cm proximal to the wrist where two further sensory branches appear: the dorsal and palmar cutaneous branches of the ulnar nerve. The dorsal cutaneous branch supplies the sensory innervation to the ulnar one and a half digits of the dorsal surface of the hand, while the palmar cutaneous branch provides sensation to the ulnar one and a half digits of the palm.

The ulnar nerve lies superficial to the flexor retinaculum of the wrist to enter the second area of compression, which results in ulnar nerve canalicular symptoms, Guyon canal. This region, similar to the carpal tunnel, transmits both the ulnar nerve and ulnar artery towards the and compression here results in Guyon canal syndrome. This terminates as the deep and superficial branches of the ulnar nerves, both of which are involved in the motor innervation of the majority of the intrinsic muscles of the hand.

Ulnar nerve neuropathies are cubital tunnel syndrome and Guyon canal syndrome. **Cubital tunnel syndrome** is the second most common impingement syndrome after carpal tunnel syndrome. It develops as a result of the ulnar nerve being trapped at the elbow level. Causes include systemic diseases, prolonged tourniquet application, medial epicondyle fracture, cubitus valgus deformity, repetitive elbow flexion-extension or prolonged elbow casting. When the elbow joint is fully flexed, the cubital tunnel is in the narrowest position; this causes ulnar nerve to be compressed. The complaint is usually in the form of pain radiating to the medial of the forearm, sensory abnormalities in the dorsal and palmar aspects of the hand and motor weakness in the intrinsic muscles of the hand. In further stages, claw hand deformity (hyperextension of the MP joints of the fourth and fifth fingers, flexion of the PIP and DIP joints) may occur. In patients with mild-to-moderate compression, conservative treatment is recommended. Static long-arm splint

is used in conservative treatment to limit the flexion of the elbow joint. After 6 months of conservative treatment if there is no relief in symptoms, surgery can be considered (Fig. 78.5). After cubital tunnel decompression surgery, rehabilitation programme splinting, stretching and strengthening exercises should be begun.

Guyon canal syndrome is the entrapment of the ulnar nerve within the Guyon canal. Ganglion cyst and inflammatory diseases are the common causes of impingement. The first-line treatment is conservative.

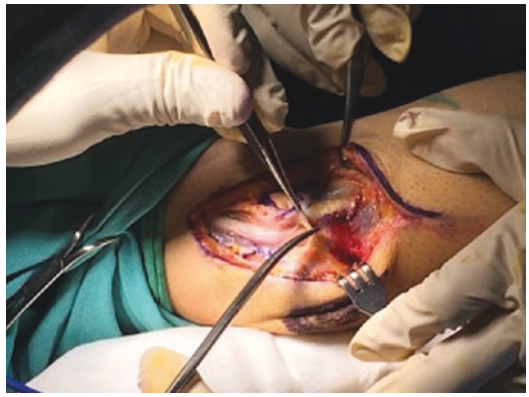

Fig. 78.5 Ulnar nerve decompression of a patient with cubital tunnel syndrome

78.8 Radial Nerve

78.8.1 Overview

Radial tunnel syndrome (RTS) (Fig. 78.6) and posterior interosseous nerve syndrome (PINS) are the two main compressive neuropathies affecting the radial nerve. PINS is more common and has an average incidence of 3 per 100,000. It is reported to be twice as common in males and is associated with overuse of forearm muscles in occupations that involve heavy lifting.

RTS is much less common and, until recently, was unestablished due to underdiagnosis. The clinical features are subtle, resulting in a high threshold for patients seeking medical attention. The pathology is believed to be caused by compression at the five sites mentioned later as well as the posterior interosseous nerve at the radial tunnel.

78.8.2 Anatomy

The radial nerve is the most important nerve for both the arm and posterior compartment of the forearm, providing motor innervation to the mus-

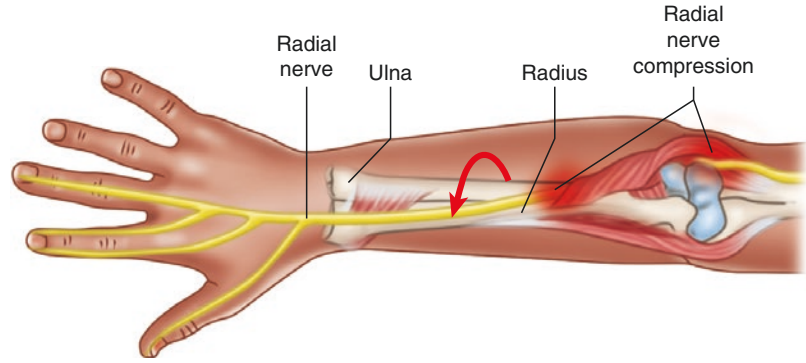

Fig. 78.6 Radial nerve compression syndrome

cles of the posterior compartment. The radial nerve is the major branch of the posterior cord of the brachial plexus and receives contributions from roots C5–T1. The nerve descends posteriorly through the triangular interval inferior to the axilla to follow the radial groove of the humerus and supplies the triceps muscle in the arm.

At the elbow, the nerve moves anterior to the lateral epicondyle of the humerus to enter the antecubital fossa. Here, it divides into a deep and superficial branch. The deep branch winds laterally around the radius between the two heads of the supinator to become the posterior interosseous nerve, the motor nerve responsible in the innervation of all the muscles of the posterior compartment of the forearm. Compression between the two heads of the supinator can cause neuropathy, resulting in PINS.

Five typical areas affect the radial nerve: the proximal edge of extensor carpi radialis brevis, the distal supinator margin, the arcade of Frohse at the proximal aspect of supinator, the leash of Henry around the radial head and the fibrous tissue making up the anterior capsular of the radiocapitellar joint.

Radial nerve neuropathies consist of entrapment at the axillary region, posterior interosseous nerve syndrome and Wartenberg syndrome.

Entrapment at the axillary region may be seen as a result of pressure on the radial nerve in the axillary area as a result of external compression. Weakness can be seen in the triceps and extensor muscles. Tumoral causes and trauma are also effective in compression. The radial nerve can also be compressed on upper arm. It may occur when the radial nerve is exposed to pressure while rotating at the proximal humerus. It may be seen as a result of the arm being in a bad position during sleep. Fractures of the humerus, tumour and overuse of the triceps muscle are the other causes of the compression. Conservative treatment is preferred.

The radial nerve is divided into two: the superficial sensory branch and the posterior interosseous motor branch at the radial head level. The posterior interosseous nerve can be compressed in the proximal forearm or anterior to the elbow capsule or within the supinator muscle. The reasons for the radial nerve entrapment at these points are radial head fractures, tumours and repetitive pronation-supination movements. In **posterior interosseous syndrome** (PIS) syndrome, innervation of the wrist extensors is preserved because the innervation of these muscles is at the level of the elbow joint. Finger extensors, thumb extensors and abductors may have paralysis. There is no sensory deficit. Lateral epicondylitis may pose a diagnostic dilemma to the physicians. In lateral epicondylitis syndrome, pain concentrates on the lateral epicondyle and increases with the extension of the wrist. In PIS syndrome, pain is exacerbated by the resistant extension of the third finger and radiates to the lateral side of the arm. Also, resistant supination movement causes pain. An arm splint may be preferred to reduce pain and inflammation in conservative treatment. Classical massage to be applied around the elbow is an effective method to reduce oedema and pain. In addition, other classical physical therapy agents such as the TENS device can be used to reduce pain. Radial nerve shifting exercises should be started as early as possible. Strengthening exercises should be added to the program as symptoms subside. If conservative treatment is not successful, decompression surgery is considered. After surgery, splinting, physical therapy modalities and exercises should be started. **Wartenberg (superficial radial cutaneous nerve compression) syndrome** is a condition in which the superficial sensory branch of the radial nerve is under pressure. During the course of the nerve in the forearm, the nerve can be subjected to pressure in any area. Tumours, effusions, watch straps, bracelets and callus caused by radial fractures may cause the radial nerve to be entrapped. Pain and paraesthesia on the back of the hand and in the radial part of the hand are the symptoms of the disease. The treatment is conservative and rarely requires surgery.

Cervical rib syndrome is caused by the compression by the cervical rib or C7 transverse process to the C8 and T1 roots of the brachial plexus, resulting in sensory and motor sensation losses in the hand. Generally, sensory defects are seen in the inner surface of the forearm and the fourth

and fifth fingers. Pain, atrophy of the intrinsic muscles of the hand and vasomotor changes are common symptoms. If conservative treatment fails, surgical treatment is considered. **Scalenus anterior syndrome** occurs when the brachial plexus compresses as it passes through the m.scalenus. Vascular insufficiency and claudication may be seen in the ipsilateral extremity. **Thoracic outlet syndrome (TOS)** is a complex symptom group that includes neural, arterial and venous disorders of the upper extremity. The lower trunk of brachial plexus, subclavian artery and vein compresses between the clavicle and the first rib. The main complaint may be pain, and pain may spread from the neck and shoulder to the hand. It is accompanied by paraesthesia of the inner surface of the forearm and hand. Complaints increase in the position that require working above the shoulder level. Besides, there may be cold intolerances, colour variations or Raynaud's phenomenon. When conservative treatment fails, surgical treatment should be considered. The main goal in the surgical treatment of the above-mentioned syndromes is to reduce the pressure on the nerve. For this purpose, tenotomies or rib excisions may be applied by open or percutaneous/endoscopic methods.

The suprascapular nerve originates from the C5–C6 nerve roots. It is a peripheral nerve that leaves the upper trunk of the plexus and has motor and sensory fibres. It passes from the suprascapular notch, under the transverse scapular ligament to the back of the scapula, through the supraspinous fossa of the scapula and innervates the supraspinatus and infraspinatus muscles. The suprascapular groove—due to its anatomical structure—is the point where the nerve is vulnerable to external factors, and nerve damage often reveals in this region. If the suprascapular nerve is compressed around the scapular notch, both the supraspinatus and infraspinatus muscles are affected. If compression occurs around the spinoglenoid notch, only the infraspinatus muscle is affected (Fig. 78.1). **Suprascapular nerve compression** appears as a result of compression by the suprascapular ligament. Compression is mostly caused by the ganglion and is more common in the spinoglenoid notch. The ganglion cyst ensues after a capsulolabral injury. The ganglion cyst causes infraspinatus muscle denervation. If both the supraspinatus and infraspinatus muscles are affected, Parsonage-Turner syndrome—in differential diagnosis—should be considered on condition that there is no compression in the scapular notch. Electrophysiological tests and radiological imaging are as important as clinical examination in the diagnosis of suprascapular nerve syndrome. Shoulder external rotation is affected in most of the cases. Ganglion cyst due to intra-articular superior labrum tear—diagnosed by MRI—is typical presentation. Conservative treatment includes USG-guided aspiration of the cyst, local anaesthetic and steroid injections. In surgical treatment, open or arthroscopic suprascapular nerve release and repair if there is a concomitant labral tear are recommended.

Anterior interosseous syndrome is the compression of the anterior interosseous nerve, which is the motor branch of the median nerve. Patients suffer from pain in the proximal part of the forearm that increases with activity. Since the flexor pollicis longus and second finger flexor digitorum profundus muscles do not work, the distal interphalangeal joint flexion cannot be realised. Due to Martin-Gruber anastomosis, paralysis may also occur in the intrinsic muscles of the hand. The treatment is conservative; physical therapy applications, avoiding repetitive forearm movements, splinting and injections are recommended.

Pronator teres syndrome occurs as a result of the compression of the median nerve between the two heads of the pronator teres muscle or the pressure of the fibrous bands. Symptoms increase with wrist flexion and forearm pronation. Pain is localised medial to the forearm. Paraesthesia and sensory problems are observed in the first three fingers of the hand, which is the dermatome area of the median nerve. Conservative treatment is generally preferred in its treatment. Conservative treatments are similar to CTS. Nerve decompression is applied in surgical treatment. **Supracondylar process syndrome** is formed by the compression of the median nerve between the Struthers ligament and the bony prominence in

the distal humerus. In diagnosis, it is important to identify bony prominence in the distal humerus on conventional radiographs and to have pain radiating from the distal humerus to the hand. Surgery may be recommended for those who do not have relief with conservative treatment.

Take-Home Message

- Canalicular syndromes may cause symptoms depending on compression of the peripheral nerve at certain points along its anatomical course for several reasons.
- The most common compressive neuropathy in the upper limb is carpal tunnel syndrome.
- If no significant improvement is seen after 6 months despite conservative treatments, surgical treatment is indicated.
- Clinical examination, nerve conduction tests and radiologic tests (MRI, USG) are the tools for diagnosis.
- After severe compression to the nerve in canalicular syndromes, long-lasting epineural oedema is followed by endoneurial oedema and fibrosis, which may lead to irreversible loss of function.

Summary

Canalicular syndromes (entrapment/compression neuropathies) are characterised by motor, sensory and autonomic deficits that occur as a result of compression of the peripheral nerve at certain points along its anatomical course for different reasons. In general, while the reason of the compression is occupational factors in active-young individuals, metabolic/hormonal factors are the reason in the middle aged. Compression or similar terms are used to indicate the reason of disease. The onset of the problem does not originate from the nerve itself; however, external mechanical factors affect the condition. In entrapment neuropathies, the diagnosis is made on the basis of symptoms and diagnostic tests. Nerve conduction tests and radiologic images are used, as well. Conservative treatment is generally preferred in the treatment of canalicular syndromes. Surgical interventions should be considered in cases where nonsurgical treatment does not respond, principally if muscle atrophy or motor deficit has occurred.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Which is the most common ulnar nerve entrapment?
   (a) Cubital tunnel syndrome
   (b) Carpal tunnel syndrome
   (c) Median entrapment
   (d) Radial entrapment
2. Which entrapment neuropathy results in shoulder external rotation deficit?
   (a) Suprascapular nerve syndrome
   (b) Supraspinatus nerve syndrome
   (c) Minor teres nerve syndrome
   (d) Brachial nerve syndrome
3. To diagnose carpal tunnel syndrome, which clinical examination manoeuvres are characteristic?
   (a) Phalen and Tinel tests
   (b) Napoleon test
   (c) Spurling test
   (d) Neer test
4. Lateral epicondylitis may pose a diagnostic dilemma to the physicians for …:
   (a) Posterior interosseous nerve syndrome
   (b) Anterior interosseous nerve syndrome
   (c) Ulnar entrapment
   (d) Radial entrapment
5. Wartenberg syndrome is an entrapment of …:
   (a) Superficial radial cutaneous nerve compression
   (b) Median nerve
   (c) Ulnar nerve
   (d) Posterior interosseus nerve

Further Reading

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Canalicular Syndromes of the Lower Limbs

79

M. Esat Uygur

Overview
This chapter includes canalicular syndromes that cause entrapment neuropathies in the lower limbs. Piriformis syndrome, obturator neuropathy, meralgia paresthetica, iliacus syndrome, tarsal tunnel syndrome, anterior tarsal tunnel syndrome, jogger's foot, Baxter's neuropathy, and other entrapment neuropathies of the lower limbs are described in this chapter.

79.1 Introduction

Canalicular syndromes are defined as nerve compression pathologies that cause entrapment neuropathies in nerves. Numbness, neuropathic pain characterized by burning and tingling, paresthesia and atrophy, weakness, or loss of motor or sensory function usually occur because of chronic compression of the nerve.

Patients affected by entrapment neuropathy syndromes have different types of clinical manifestations (Table 79.1). The complaints vary according to the affected nerve. Piriformis syndrome is the most common cause of sciatic nerve entrapment around the hip. Seventy percent of nondiscal sciatica cases are caused by piriformis syndromes. The incidence of lateral femoral cutaneous nerve (LFCN) entrapment is nearly 4.3/10,000 persons/year, and more men than women are affected. Peroneal nerve entrapments occur more frequently in men, and 21.7% of cases occur due to iatrogenic injuries. On the other hand, entrapment neuropathies in the foot are more common in women than in men, probably because of their footwear habits.

The pathogeneses of canalicular syndromes mainly involve the compression of a nerve because of space-occupying lesions or extrinsic factors. Individuals with rheumatologic comorbidities are more susceptible to neuropathies because they often develop tenosynovitis, rheumatoid nodules, and limb deformities. Alignment impairments may also cause neuropathy due to tractional or impingement mechanisms.

Knowledge on the anatomy of nerves is important for determining the possible pathologies of canalicular syndromes. The sciatic nerve originates from L4, L5, S1, S2, and S3 nerve roots. It exits the pelvis from the greater sciatic foramen. The sciatic nerve moves across to the anterior side of the piriformis muscle and courses downward. Piriformis syndrome is caused by the entrapment of the sciatic nerve under the piriformis muscle (Fig. 79.1). The femoral nerve, which lies under the psoas muscles coursing in the pel-

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Table 79.1 Major properties of canalicular syndromes of the lower limbs

Canalicular syndrome (affected nerve)	Site of compression	Possible aetiology	Sensory deficit	Motor deficit
Piriformis syndrome (Sciatic n.)	Great sciatic notch	• Hypertrophy of the piriformis muscle • Inflammation of the piriformis muscle (infection, inflammatory process) • Fibrous adhesive band of the piriformis muscle • Spasticity due to cerebral palsy • Posttraumatic hematoma • Local ischemia	Unilateral buttock, trochanteric or posterior thigh pain	Infrequent
Iliacussyndrome-(Femoral n.)	Pelvis and groin	• Iatrogenic injury • Traumatic injury • Enlargement of the iliopsoas muscle • Distended iliopsoas bursa	Paresthesia at anterior thigh, medial calf and medial foot	Weakness and atrophy at quadriceps muscle
Saphenous neuropathy (Saphenous n.)	Adductor canal	• Traumatic injury • Iatrogenic injury • Stretching injury after knee instability	Paresthesia at the medial leg, medial foot, great toe	None
Obturator neuropathy (Obturator n.)	Obturator foramen	• Iatrogenic injury • Pelvic fractures • Hematomas • Tumors • Fibrous band secondary to chronic adductor tendinitis and osteitis pubis	Groin and medial thigh pain	Weakness of adductor muscles
Meralgia paresthetica (Lateral femoral cutaneous n.)	Inguinal ligament or tensor fascia lata	• Avulsion fracture of ASIS • Pelvic or retroperitoneal tumors • Stretching of nerve (due to prolong leg hyperextension) • Iatrogenic (during harvesting an autograft from iliac bone) • External compression (tight clothes, belts…)	Burning pain, numbness, tingling at lateral thigh	None
Peroneal neuropathy (Common peroneal n.)	Around fibular head	• Prolonged immobilization (surgery, coma, overdose); • Space-occupying lesions (osteochondromas, tumors, ganglion/synovial cysts, varicosities) • Traumatic injury following fibular head fracture, knee dislocation or knee surgery • Post-traumatic compartment syndrome • Hypertrophy of the short head of the biceps femoris muscle	Dysesthesias in the proximal third lateral leg	Lateral and Anterior compartment muscles of the lower leg

Table 79.1 (continued)

Canalicular syndrome (affected nerve)	Site of compression	Possible aetiology	Sensory deficit	Motor deficit
Anterior tarsal tunnel syndrome (DPN)	1. Superior and inferior extensor retinaculum, under EHB 2. First and second tarsometatarsal joint	• Tight shoelaces • Bony prominences, dorsal osteophytes, fracture sequels at midfoot, os intermetatarseum • Space-occupying lesions • Hypertrophy of EHB	Predominantly sensorial at the foot Dorsum of first webspace at distal metatarsal	EDB (if the entrapment occurs at proximal)
Tarsal tunnel syndrome (Tibial n.)	Flexor retinaculum	• Space-occupying lesions (ganglia, the most common) • Osseous spurs, sequel of calcaneus fractures • Varicosities	Plantar aspect of the foot	Plantar muscles
Jogger's foot (MPN)	Between navicula and AbH or between AbH and knot of Henry	• High medial arch • Dynamic hindfoot valgus, excessive pronation while running	Medial plantar heel and medial plantar 2/3 of the foot	FDB FHB AbH First Lumbrical muscles
Baxter's neuropathy (ICN, 1st branch of LPN.)	Between AbH and quadratus plantae	• Calcaneal fracture, direct medial trauma • Hypertrophy of the AbH and Quadratus plantae muscles (especially in runners) • Presence of accessory muscles or bursae • Phlebitis of calcaneal venous plexus • Hypermobile flat foot	1/3 Lateral sole of the foot	FDB QP Abductor digiti minimi
Superficial peroneal nerve entrapment (SPN)	Anterolateral mid-cruris where the nerve exits from deep fascia of the lateral compartment	• Iatrogenic, after ankle arthroscopy • Direct trauma, fibular fractures, ankle sprains • Syndesmotic injury • Chronic ankle instability	Along the lateral aspect of the lower leg Dorsum of the foot (sparing DPN area-dorsum of the 1st webspace)	None

AbH Abductor hallucis, *ASIS* anterosuperior iliac spine, *DPN* deep peroneal nerve, *EDB* extensor digitorum brevis, *EHB* extensor hallucis brevis, *FDB* flexor digitorum brevis, *FHB* flexor hallucis brevis, *ICN* inferior calcaneal nerve, *LPN* lateral plantar nerve, *MPN* medial plantar nerve, *n* nerve, *SPN* superficial peroneal nerve, *QP* quadratus plantae

vis, originates from the L2, L3, and L4 nerve roots. It exits from the pelvis under the inguinal ligament. Iliacus syndrome is a femoral neuropathy that is caused by an entrapment under the inguinal ligament (lacuna musculorum). The obturator nerve, which originates from the L2, L3, and L4 nerve roots, descends through the medial border of the psoas muscle at the level of the pelvis. Obturator neuropathy can be caused by obturator nerve entrapment at the level of the pelvic exits. The LFCN originates from the L2 and L3 nerve roots, travels to the lateral side, and passes beneath the psoas muscle. Entrapment can occur, as the nerve passes under the inguinal ligament when it exits the pelvis. The LFCN exits the pelvis immediately medial to the anterior-superior iliac spine (ASIS) and is susceptible to compression by belts. In addition, iatrogenic nerve injuries may occur while an autograft is being obtained from the iliac bone.

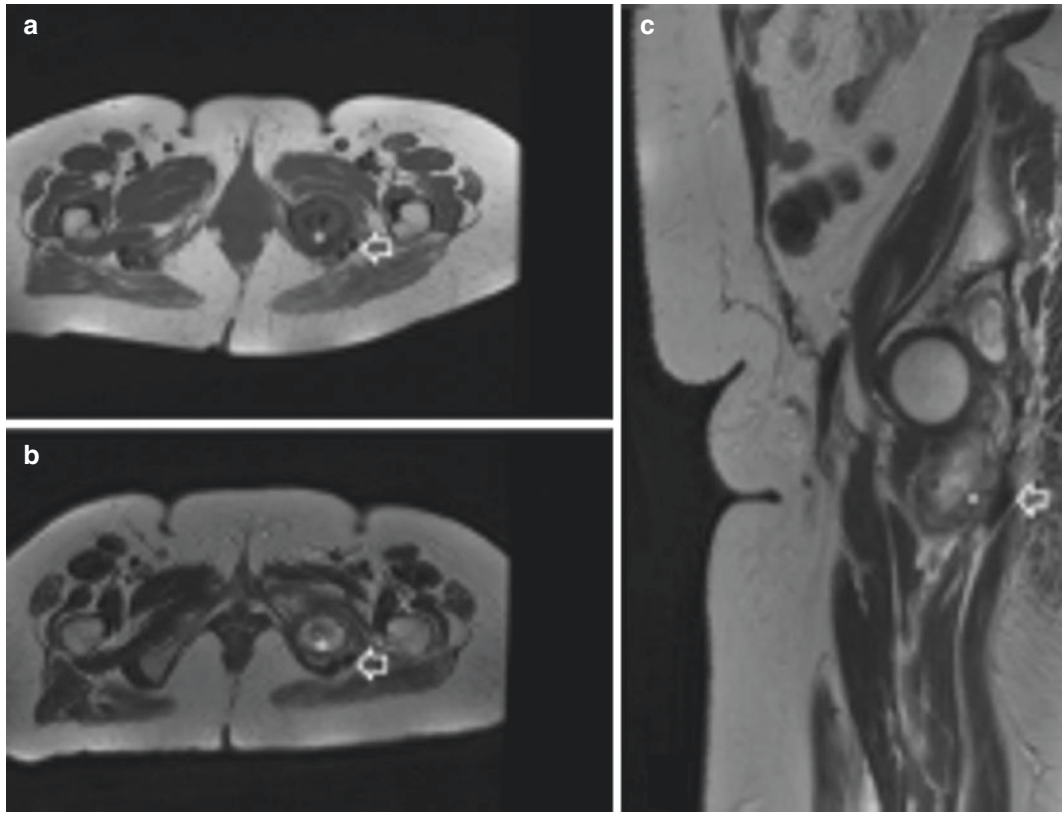

Fig. 79.1 A T_1-weighted axial magnetic resonance image (MRI) (**a**), T_2-weighted axial MRI (**b**), and T_2-weighted sagittal MRI view (**c**) of plasmacytoma (asterisk) originated from ischial tuberosity causing piriformis syndrome in a 60-year-old female patient. The sciatic nerve is demonstrated by arrows

The sciatic nerve consists of both the common peroneal nerve and tibial nerves. From the proximal to distal direction, the nerve passes from the posterior end of the femoral head capsule to the posterior side of the thigh. The tibial and peroneal nerves separate from each other at the level of distal thigh. Tibial neuropathies can occur in the popliteal fossa. Immediately distal to the fibular head, the common peroneal nerve separates into the superficial and deep branches. The superficial peroneal nerve (SPN) innervates the ankle evertors and provides sensory nerve branches from the lateral aspect of the lower leg and dorsum of the foot; the deep peroneal nerve (DPN) enters the anterior leg compartment and innervates the tibialis anterior and extensor hallucis longus (EHL), which are ankle and first toe extensors. The terminal branch of the DPN provides sensory innervation from the first webspace in the forefoot. The most common compressive neuropathy in the lower limb is peroneal neuropathy. In terms of chronic nerve entrapment, the peroneal nerve is most commonly affected around the fibular head. Since the DPN includes motor fibers, it affects the ankle and toe extensors. Mostly, DPN compressions result in drop foot, but occasionally, it also causes drop toe (Fig. 79.2).

The tarsal tunnel is a fibro-osseous tunnel consisting of deep and superficial aponeuroses (flexor retinaculum), which are located posterior and inferior to the medial malleolus. The flexor digitorum longus and flexor hallucis longus tendons and the posterior tibial artery and veins also pass through this tunnel. The tibial nerve can be compressed in this tunnel by a soft-tissue mass (most commonly a ganglion cyst) (Fig. 79.3), muscle hypertrophy, or vascular abnormalities.

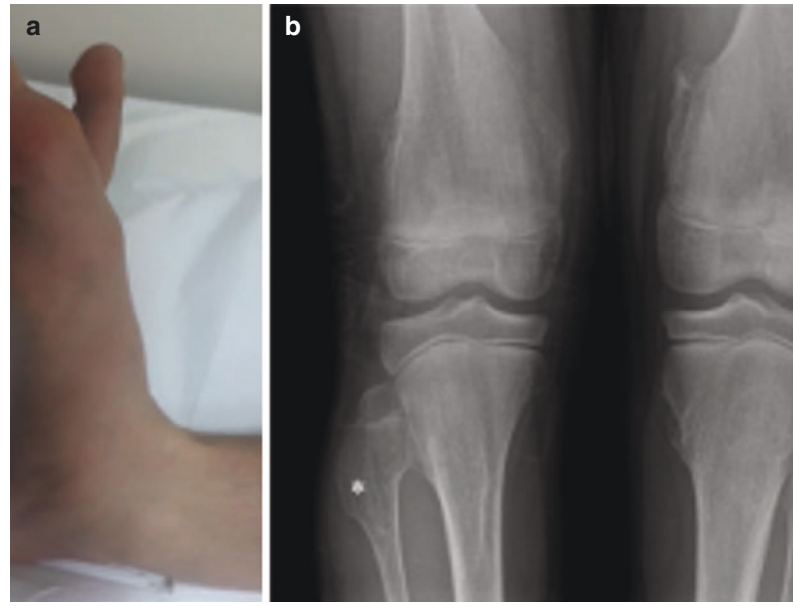

Fig. 79.2 A direct radiograph (**a**) of an osteochondroma (asterisk) around the fibular head that led to drop toe and (**b**) lack of extension in first toe because of deep peroneal nerve entrapment in a 14-year-old boy

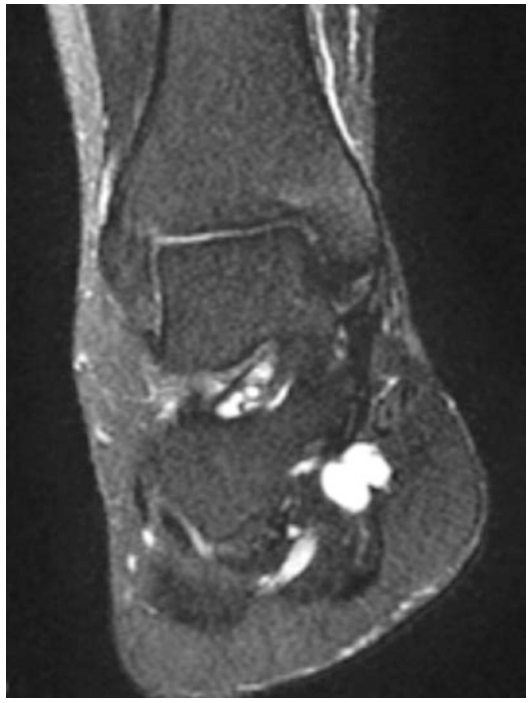

Fig. 79.3 A T_2-weighted coronal magnetic resonance imaging section of a ganglion mass causing tarsal tunnel syndrome in a 56-year-old female patient. Ganglia constitute the most common type of mass causing compression neuropathy of the tibial nerve

The tibial nerve separates into three branches within the tarsal tunnel: the medial calcaneal nerve (provides sensation from the medial region of the heel), medial plantar nerve (MPN), and lateral plantar nerve (LPN). Medial plantar nerve syndrome, which is also known as jogger's foot, refers to the compression of the nerve between the abductor hallucis muscle (AbH) and the knot of Henry or between the AbH and navicular bone. The first branch of the LPN is the inferior calcaneal nerve (Baxter's nerve). Entrapment usually occurs because of compression of the hypertrophied AbH muscle or thickened plantar fascia.

In general, patients report paresthesia, tingling, and burning pain at the innervation area of the associated peripheric nerve. Depending on the branch of the nerve and site of compression, atrophy, weakness, and sensorial and/or motor deficits may also occur.

The symptoms often worsen with prolonged traction and compression of the nerve, which means that the symptoms progress with activity and weight-bearing. However, in piriformis syndrome, prolonged sitting in the cross-legged position, and common peroneal neuropathy, prolonged compression (the use of a tourniquet or

positional compression during surgery) may cause neuropathy symptoms. Prolonged walking in tarsal tunnel syndrome, Baxter's neuropathy, and jogger's foot and tight belts in meralgia paresthetica can aggravate symptoms. Prolonged plantar flexion often exacerbates the symptoms of anterior tarsal tunnel syndrome. As the foot remains in plantar flexion all night, patients experience more severe pain and a more severe tingling sensation in the middle of the night, which cause them to wake up. On the other hand, any type of mass, such as osteophytes, myositis ossificans, tumoral lesions, muscle hypertrophy, and varicosities, can cause compression of the associated nerve. The most common type of soft-tissue mass that causes tarsal tunnel syndrome is a ganglion cyst.

In meralgia paresthetica, the burning pain radiates to the anterolateral side of the thigh; in piriformis syndrome, the pain is felt in the lateral-posterior region of the buttocks. Iliacus syndrome causes numbness and paresthesia anterior to the thigh, in the medial calf, and medial to the toe. In obturator neuropathy, groin pain or pain in the medial end of the thigh is expected. Saphenous neuropathy causes paresthesia on the medial side of the lower leg, foot, and great toe. In tarsal tunnel syndrome, there is pain on the medial side of the ankle and heel, and neuropathy radiates to the medial and plantar sole of foot; in Baxter's neuropathy, the neuropathic area is the medio-plantar side of the heel. In anterior tarsal tunnel syndrome, patients often complain of a burning sensation at the dorsum of the foot and paresthesia in the first webspace in the forefoot. Occasionally, in tarsal tunnel syndrome and Baxter's neuropathy, the neuropathy extends not only distally but also proximally through the lower leg, and this phenomenon is known as the Valleix phenomenon.

The physical examination for piriformis syndrome should start with inspection. First, both limbs are compared with each other. Although sciatic nerve deficits affect the posterior thigh muscles and all of the muscles under the knee, neurologic deficits are rarely observed. Tenderness may be detected over the piriformis muscle. While unilateral symptoms (buttock, trochanteric, or posterior thigh pain) are aggravated by sitting, walking, or squatting, they are relieved by lying down. The inspection may reveal iliacus syndrome; as the femoral nerve innervates the quadriceps muscle, in cases of entrapment, thigh atrophy may be present. As the saphenous nerve is a sensory nerve, it obtains sensory innervation from the medial thigh, leg, and foot; typical neuropathic symptoms are seen in affected areas in cases of entrapment. If the femoral nerve is entrapped at the groin, quadriceps atrophy, extension weakness of the knee, and paresthesia in the medial thigh, knee, and foot are expected. On the other hand, if paresthesia is predominant without atrophy, saphenous nerve entrapment may be present, which is the entrapment of the femoral nerve within the adductor canal. Therefore, the site of compression may be predicted by a physical examination of the femoral nerve. If there is adductor weakness in addition to groin pain or medial thigh pain, obturator neuropathy may be present. When patients report burning pain, numbness, and tingling on the lateral side of the thigh and symptoms are relieved with sitting, one should consider meralgia paresthetica.

In foot and ankle conditions, the physical examination should start with inspection for any protrusion, mass, or varicosities. Patients should be examined while walking, standing, and lying or sitting. Peroneal neuropathy may present with steppage gait, which reveals a motor deficit in the ankle extensors. As the DPN is mainly a motor nerve, compression of the nerve around the fibular head mostly causes drop foot or drop toe. First webspace neuropathy should also be checked for sensory deficits. For the differential diagnosis, the tibialis posterior muscle strength should also be examined. If both the ankle extensors and inverters do not function properly, the region of the neuropathy may be located more proximally. On the other hand, if the ankle extensors do not function properly but the tibialis posterior functions properly, the site of compression is most likely near the fibular head. Checking hindfoot alignment during walking and standing may also reveal the reason for the neuropathy. Excessive hindfoot valgus causes tractional neuropathy on the tibial nerve and Baxter's nerve. The specific

examination used for assessing the canalicular syndrome is the Tinel test, which is a percussion test of the nerve. For the diagnosis of tarsal tunnel syndrome, the Tinel sign test is performed behind the medial malleolus. However, percussion should be performed while the patient is standing (neutral ankle valgus) and while the patient is sitting cross-legged (valgus push of the ankle). Performing the Trepman test (applying compression with the examiner's thumb while the ankle is positioned in inversion and plantar flexion) in addition to the Tinel test increases the sensitivity and specificity for the diagnosis of tarsal tunnel syndrome. Pes cavus patients are more susceptible to MPN compression. In jogger's foot, the Tinel sign is positive beneath the navicular bone, through which the MPN passes. Hyperpronation and the reverse Phalen's maneuver (forced dorsiflexion and eversion) often exacerbate pain in the LPN. The Tinel sign test should be performed at the dorsum of the foot to screen for anterior tarsal tunnel syndrome, and plantar flexion of the foot may elicit neuropathic symptoms due to increased compression on the DPN.

For the diagnosis of entrapment syndrome, the first diagnostic imaging method that should be used is plain radiography, with proper planning and execution. In cases of peroneal neuropathy, bone tumors (mostly osteochondromas following ganglion cysts) may be detected around the fibular head (Fig. 79.2). In tarsal tunnel syndrome, any bony spurs or osteophytes that are present are investigated on anteroposterior, lateral, and oblique foot radiographs. Dorsal osteophytes in the midfoot may be detected on the lateral and oblique radiographs as a possible cause of anterior tarsal tunnel syndrome. If bony prominences are suspicious, a computed tomography scan should be performed. However, some soft-tissue lesions, such as tumoral lesions and muscle hypertrophies, can easily be detected with magnetic resonance imaging (MRI). If the prediagnosis is a ganglion cyst, there is no need for a contrast agent; otherwise, MRI with gadolinium is useful for identifying the lesion characteristics. MRI is also useful to indicate fatty degeneration in particular muscles related to motor deficits. Ultrasound imaging is also helpful for the diagnosis of soft-tissue lesions. Electrodiagnostic studies may be useful in differentiating entrapment neuropathy from radiculopathy or peripheric neuropathy. Nerve conduction and needle electromyography studies may be useful for localizing the site of denervation. Local anesthetic injections may also play a role in the differential diagnosis and symptomatic transient pain relief.

While the first-line treatment of a canalicular syndrome is usually nonoperative, to prevent neurofibrosis, decompression should be performed before the case progresses to the late stage and becomes irreversible. Local anesthetic injections can be helpful for both diagnosis and symptomatic pain relief. Physical therapy for hip lesions, footwear modifications, shoelace lacing technique adjustments, and use of an orthosis can be helpful for foot and ankle lesions before surgical intervention. The operative treatment includes the release of the nerve and removal of the bone or soft-tissue mass, if present. The mass that was identified to be compressing the nerve should be excised. After excision, a Hemovac drain, cast, or bulky elastic bandage should be applied to avoid hemorrhage around the nerve. Endoscopic tarsal tunnel release has also been reported in the literature; however, one should be aware that it can lead to inadequate release and iatrogenic nerve injuries.

Take-Home Message

- Knowledge on the anatomy of the nerve is important for identifying possible pathologies of an entrapped nerve.
- Canalicular syndromes may cause atrophy and motor and/or sensory deficits, depending on the nerve affected and compression site.
- As entrapment of the saphenous nerve, lateral femoral cutaneous nerve, and superficial peroneal nerve is purely sensorial, these neuropathies are isolated sensory neuropathies.
- The typical symptoms of entrapment neuropathy include paresthesia, tingling, and burning pain.

- The Tinel test can be used on most nerves.
- Direct radiography, MRI and electromyography studies may be helpful for diagnosis.
- The first-line treatment of a canalicular syndrome is usually nonoperative. However, to avoid neurofibrosis, decompression should be performed before the case progresses to the late stage and becomes irreversible.

Summary

A canalicular syndrome is defined as an entrapment neuropathy of a nerve. Numbness, neuropathic pain characterized by burning and tingling, paresthesia and atrophy, weakness, or loss of motor and/or sensory function usually occur because of chronic compression of the nerve. As the location of the symptoms is helpful in identifying the particular nerve affected, knowledge on the anatomy and distribution of nerves is important for diagnosing canalicular syndromes. To exacerbate the symptoms of a neuropathy, the Tinel test may be performed in different positions on a particular nerve, as the test is mainly a percussion test performed on a nerve. Imaging techniques are useful for diagnosis. Local anesthetic injections are also helpful for both diagnosis and symptomatic pain relief. While the first-line treatment of an entrapment neuropathy is usually nonoperative, care should be taken to prevent neurofibrosis; decompression needs to be performed before the case progresses to the late stage and becomes irreversible.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. A long duration in which position causes the symptoms of piriformis syndrome?
   (a) Sitting cross-legged
   (b) Leg extended
   (c) W-sitting
   (d) It is indifferent, because it is due to the stiffness of the chair
2. After inguinal surgery, a patient complained of paresthesia and burning pain on the anterolateral side of the thigh. What is the possible diagnosis, and which nerve may be affected?
   (a) Meralgia paresthetica; the lateral femoral cutaneous nerve
   (b) Meralgia paresthetica; the medial femoral cutaneous nerve
   (c) Piriformis syndrome; sciatic nerve
   (d) Gluteal nerve syndrome; gluteal nerve
3. The entrapment of which nerve is particularly common in patients with pes cavus deformity?
   (a) The medial plantar nerve
   (b) The femoral nerve
   (c) The posterior tibialis nerve
   (d) The anterior tibialis nerve
4. Prolonged flexion of the ankle during sleep may elicit the symptoms of anterior tarsal tunnel syndrome. Which nerve is associated with this pathology?
   (a) The deep peroneal nerve
   (b) The posterior tibialis
   (c) The anterior tibialis
   (d) The medial plantar nerve
5. A 47-year-old female patient underwent ankle arthroscopy 2 months ago. Aberrant scar tissue was observed in her ankle, which is related to an anterolateral arthroscopic portal. She also complained of numbness in the dorsum of her foot. Which type of nerve entrapment do you think caused this scenario?
   (a) Superficial peroneal nerve injury
   (b) Posterior peroneal nerve injury
   (c) Tibialis posterior nerve injury
   (d) Tibialis anterior nerve injury

Further Reading

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Answers to Questions

Chapter 1

1	a
2	a
3	a
4	a
5	a

Chapter 2

1	b
2	a
3	a
4	a
5	a

Chapter 3

1	b
2	d
3	b
4	d
5	d

Chapter 4

1	b, e
2	a, d
3	a, e
4	e
5	e
6	a, b, c

Chapter 5

1	b, c, d
2	b, c, d
3	b, c
4	b
5	a, b, d, e

Chapter 6

1	a, b, e
2	b, d
3	b, d, e
4	b, d, e
5	c, d

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Chapter 7

1	b, e
2	c, d, e
3	c, e
4	b, e
5	e
6	c, d

Chapter 8

1	a
2	a
3	a
4	a
5	a

Chapter 9

1	a
2	a
3	a
4	a
5	a

Chapter 10

1	a
2	a
3	a
4	a
5	a

Chapter 11

1	a
2	b
3	a
4	a
5	c

Chapter 12

1	a
2	b
3	a
4	c
5	b

Chapter 13

1	a
2	a
3	a
4	a
5	a

Chapter 14

1	a
2	a
3	a
4	a
5	a

Chapter 15

1	a
2	d
3	d
4	a
5	b

Chapter 16

1	a
2	a
3	a
4	a
5	a

Chapter 17

1	a
2	d
3	a
4	b
5	c

Chapter 18

1	a
2	a
3	a
4	a
5	a

Chapter 19

1	a
2	a
3	a
4	a
5	a

Chapter 20

1	a
2	a
3	a
4	a
5	a

Chapter 21

1	a
2	a
3	a
4	a
5	a

Chapter 22

1	b
2	a
3	d
4	b
5	b

Chapter 23

1	a
2	a
3	a
4	c
5	d

Chapter 24

1	b
2	a
3	d
4	b
5	b

Chapter 25

1	c
2	b
3	d
4	a
5	d

Chapter 26

1	a
2	a
3	a
4	a
5	a

Chapter 27

1	a
2	a
3	a
4	a
5	a

Chapter 28

1	a
2	a
3	a
4	a
5	a

Chapter 29

1	a
2	a
3	a
4	a
5	a

Chapter 30

1	a
2	a
3	a
4	a
5	a

Chapter 31

1	a
2	a
3	a
4	a
5	a

Chapter 32

1	a
2	a
3	a
4	a
5	a

Chapter 33

1	a
2	a
3	a
4	a
5	a, b

Chapter 34

1	a
2	a
3	a
4	a
5	a

Chapter 35

1	a
2	a
3	a
4	a
5	a

Chapter 36

1	a, b, c
2	a, b, c
3	a
4	a
5	a

Chapter 37

1	b
2	e
3	c
4	c
5	e

Chapter 38

1	a
2	a
3	a
4	c
5	d

Chapter 39

1	a
2	b
3	a
4	d
5	b

Chapter 40

1	a
2	a
3	a
4	a

Chapter 41

1	a
2	a
3	a
4	a
5	a

Chapter 42

1	a
2	a
3	a
4	a
5	a

Chapter 43

1	a
2	b
3	a
4	a
5	a

Chapter 44

1	a
2	a
3	a
4	a
5	a

Chapter 45

1	c
2	b
3	c
4	c
5	c

Chapter 46

1	a
2	a
3	a
4	a
5	a

Chapter 47

1	c
2	b
3	c
4	b
5	a

Chapter 48

1	a
2	a
3	a
4	a
5	a

Chapter 49

1	a
2	d
3	c
4	b
5	a

Chapter 50

1	d
2	c
3	b
4	b
5	a

Chapter 51

1	c
2	d
3	a
4	e
5	d

Chapter 52

1	b
2	c
3	b
4	d
5	c

Chapter 53

1	d
2	b
3	b
4	c
5	d
6	c
7	a

Chapter 54

1	c
2	c
3	c
4	b
5	d

Chapter 55

1	a
2	a
3	a
4	a
5	a

Chapter 56

1	a
2	a
3	a
4	a
5	a

Chapter 57

1	a
2	a
3	a
4	a
5	a

Chapter 58

1	a
2	a
3	a
4	a
5	a

Chapter 59

1	b
2	d
3	a
4	d
5	b

Chapter 60

1	e
2	a
3	a
4	a

Chapter 61

1	a
2	a
3	a
4	a
5	a

Chapter 62

1	a
2	a
3	a
4	a
5	a

Chapter 63

1	a
2	a
3	a
4	a
5	a

Chapter 64

1	a
2	a
3	d
4	e
5	f

Chapter 65

1	d
2	b
3	a
4	d
5	c

Chapter 66

1	d
2	b
3	d
4	d
5	a

Chapter 67

1	a
2	a
3	a
4	a
5	a

Chapter 68

1	d
2	d
3	b
4	c
5	b

Chapter 69

1	c
2	c
3	d
4	a
5	d

Chapter 70

1	b
2	c
3	c
4	c
5	a

Chapter 71

1	b
2	c
3	c
4	c
	e

Chapter 72

1	a
2	a
3	a
4	a
5	a

Chapter 73

1	a
2	a
3	a
4	a
5	a

Chapter 74

1	a
2	a
3	a
4	a
5	a

Chapter 75

1	c
2	d
3	a
4	a
5	a

Chapter 76

1	c
2	a
3	a
4	d
5	b

Chapter 77

1	a
2	a
3	a
4	a
5	a

Chapter 78

1	a
2	a
3	a
4	a
5	a

Chapter 79

1	a
2	a
3	a
4	a
5	a

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