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Chapter 432

Bone, Joint, and Muscle Problems

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An estimated 20% to 30% of children's primary-care office visits involve skeletal problems. This chapter is designed to help in the initial understanding of the broad range of orthopedic conditions that are encountered in children. Conditions isolated to one body region are presented first, followed by generalized musculoskeletal conditions. Table 432-1 lists the differential diagnosis of certain signs and symptoms and is meant to aid in the organization of a diagnostic plan.

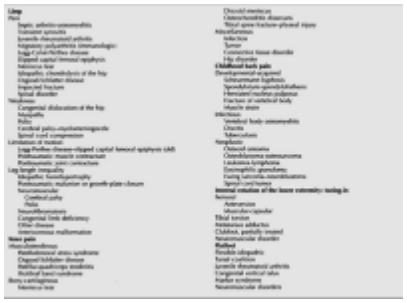

TABLE 432-1. Differential diagnosis of common pediatric symptoms

Because some orthopedic terminology may be unfamiliar, a few definitions are in order. The term *congenital* refers to a condition that is present at birth, regardless of whether it is inherited. *Developmental* refers to problems that occur or increase with time and growth, irrespective of their causes. *Physis* refers to the growth plate (Fig. 432-1); therefore, the *epiphysis* is that portion of a bone “on top of” the physis (i.e., nearer to the joint), the *metaphysis* is the widened portion of the shaft adjacent to and arising from the growth plate, and the *diaphysis* is the narrow portion of a tubular bone midway between two physes. The Greek root words *genu*, *coxa*, and *pes* refer to the knee, hip, and foot, respectively. They are used in such terms as *genu recurvatum* (hyperextensible knee), *coxa plana* [flat femoral head (i.e., Perthes disease)], and *pes planus* (flat feet). When two bones or two fracture fragments form an angle, they are in varus when the apex of the angle points away from the midline of the body, and they are in valgus when it points toward the midline (Fig. 432-2). Alternatively, angulation may be stated by the direction of the apex (i.e., genu valgus is medial angulation of the lower extremity at the knee). *Dislocation* refers to complete loss of contact of two joint surfaces, and it is specified by the direction

of displacement of the most distal part. For example, most shoulder dislocations are anterior because the humeral head comes out in front of the scapula. *Subluxation* is an incomplete dislocation. *Abduction* refers to movement away from the midline; *adduction* denotes movement toward the midline.

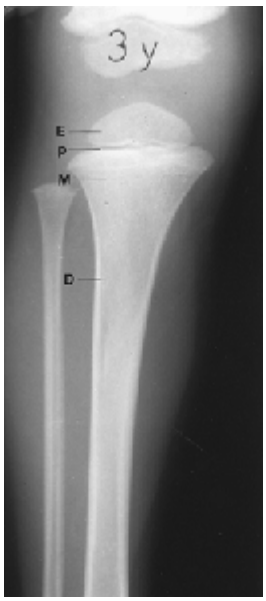

Figure 432-1. Regions of a long bone. D, diaphysis; E, epiphysis; M, metaphysis; P, physis.

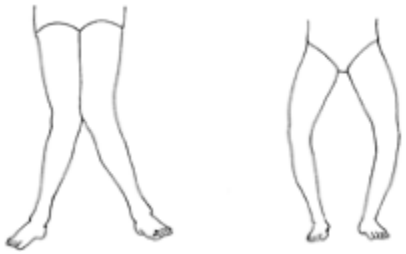

Figure 432-2. Examples of varus and valgus. **A:** Genu valgus, as apex of angulation is toward midline. **B:** Genu varus.

Regional Abnormalities

Hip and Femur

Developmental Dysplasia of the Hip

The normal hip develops from a single region of cartilage. Its characteristic spherical shape results from reciprocal contact between the femur and acetabulum during growth. Loss of

this contact may occur as a result of abnormal *in utero* positioning; such neuromuscular abnormalities as cerebral palsy, arthrogryposis, and Larsen syndrome; or intrinsic underdevelopment of the otherwise normal hip. The earlier this loss of relationship occurs, the more marked are the femoral and acetabular abnormalities that result; the later it is corrected, the less the remodeling potential.

The cause of congenital dislocation of the hip in an otherwise normal child is multifactorial. Mechanical factors play a role; thus, the frequency is increased greatly in fetuses with breech presentation, a factor in 30% of all cases of developmental dysplasia of the hip (DDH) in firstborn children and in infants with oligohydramnios. The left hip is involved more commonly than is the right. These factors are associated with increased forces across the hip or positioning in adduction and hyperflexion, causing the femur to be directed out over the edge of the acetabulum. Hormonal factors may play a role, as generalized ligamentous laxity occurs around the time of birth, caused by increased circulating estrogens and relaxin. Probably for this reason, the incidence of DDH is sixfold greater in girls than in boys. Evidence for hereditary control of these and other factors lies in the fact that more than 20% of patients have a positive family history.

In order of their increasing severity, the three degrees of hip dysplasia are subluxatable, dislocatable, and dislocated. In the *subluxatable* hip, the femoral head rests in the acetabulum and can be dislocated partially during the examination. The *dislocatable* hip can be dislocated fully with manipulation but is located normally when a baby is at rest. In the *dislocated* hip, the femoral head rests in the dislocated position. The combined incidence of these three conditions is 1 in 60 births; the incidence of true dislocation is only 1 to 2 per 1,000 births. A change in terminology from *congenital dislocation of the hip* to *DDH* has become accepted widely. The term *dysplasia* better describes the spectrum of severity of this disorder, which ranges from slight malformation to full dislocation of the hip. The term *developmental* acknowledges that some cases cannot be detected at birth and may occur later. The pathologic anatomy includes capsular laxity. The acetabulum becomes shallow as a result of lack of concentric contact with the femoral head. A false acetabulum may form where the femoral head contacts the lateral wall of the pelvis above the normal location. The outer rim of the acetabulum becomes rounded during the period when the femoral head is able to slide in and out of the acetabulum. The movement over this ridge is felt as the “clunk” of Ortolani and Barlow tests. The femur remains rotated anteriorly (anteverted) as the head rests against the lateral iliac wall.

Physical examination remains the key to the diagnosis of DDH. The signs in the newborn period usually include instability without significant fixed deformity; in later months, untreated dislocation becomes more fixed, and less instability and more limitation of certain motions occur. Specifically, Barlow and Ortolani signs should be sought in newborns. These

signs are defined as positive when the hip can be dislocated and relocated, respectively. Affected children should be relaxed when the tests are performed, and only one hip should be examined at a time. The pelvis should be stabilized with one hand while the femur is controlled with the other hand, with fingers placed on the greater and lesser trochanters (Fig. 432-3). With adduction and pressure directed posteriorly, the femur can be felt to

slide out in a posterosuperior direction over the rounded limbus in the abnormal hip (Barlow sign; see Fig. 432-3A) and then back in with abduction, causing a dull clunk to be heard (Ortolani sign; see Fig. 432-3B). Thus, the Barlow and Ortolani signs, which are the sensations of dislocation and relocation, are alternate phases of the same process of hip instability.

Figure 432-3. Barlow and Ortolani tests, performed with fingers on the lesser and greater trochanters, examining only one hip at a time. **A:** Barlow test: adduction and posterior pressure may produce a “clunk” of subluxation or dislocation. **B:** Ortolani test: abducting and “lifting” hip back into place. **C:** In children older than 3 to 6 months, Barlow and Ortolani tests often will be negative despite dislocation because of diminished laxity; the most important finding in this age group may be limitation of abduction.

A common error made in DDH diagnosis is examining both hips at once, which impairs sensitivity. Another error is mistaking insignificant soft tissue “clicks” for the more important and palpable “clunk.” These innocent clicks may result from movement of fascia over the greater trochanter or may originate from the meniscus or the patella. Routine screening of neonates in the last three decades has resulted in a dramatic increase in the early diagnosis of DDH and, thus, more successful treatment of the disorder.

Approximately 60% of all unstable hips seen in newborns normalize spontaneously within the first 2 to 4 weeks after birth as perinatal laxity resolves. A severely dysplastic hip may have negative results on examination because of the lack of an acetabular shelf. Not all hips can be reduced at birth, however, presumably because of fixed-joint contracture. Some cases of dysplasia are believed to develop after birth. For this reason, most large series have shown that not all abnormal hips can be detected by screening, even when it is performed by skilled examiners. Ultrasonography is a useful tool in examining a newborn's hip, but it may result in overdiagnosis of dysplasia if performed on all newborns. The precise indications for ultrasonography vary from region to region, but they usually involve some combination of abnormal physical examination or presence of risk factors.

The signs of a dysplastic hip change with time. If the hip remains dislocated, usually it cannot be relocated by the time an affected baby is 6 months old. Findings of asymmetry, such as limitation of abduction (see Fig. 432-3C) and apparent shortening of the thigh, are more sensitive at this time. This last sign, known as *Allis sign* or the *Galeazzi sign*, is best noted by comparing the lengths of the two flexed thighs when they are held together. Asymmetry of skin folds by itself is an unreliable finding. When children begin to walk, a positive Trendelenburg sign is noted: When weight is borne on the unstable side, the pelvis inclines to the other side.

Radiographs are not the preferred imaging method before age 6 months because of a lack of apparent bony changes during this time, except in infants with teratologic conditions. Many centers use ultrasonography, but accurate interpretation of these studies

requires extensive experience and should be done by a pediatric radiologist or orthopedist. Ultrasonography is indicated if the neonatal examination is abnormal or questionable and may be used later to guide treatment. After age 6 months, the increasing ossification of the femur renders ultrasonography less reliable, and plain films are preferred. Anteroposterior (AP) radiography of the pelvis may show cephalad and lateral migration of the femur with a break in Shenton's line (Fig. 432-4), delayed appearance of the femoral ossific nucleus, a shallow and more vertical acetabulum, and later formation of a false acetabulum.

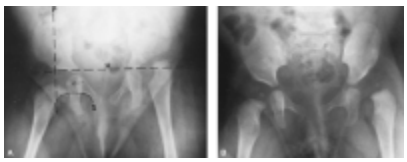

Figure 432-4. **A:** Radiographic examination of congenitally dislocated hip. The femoral head ossific nucleus should be within the lower inner quadrant formed by Perkin's vertical line (*P*) at the outer edge of the acetabulum and Hilgenreiner's horizontal line (*H*). The nucleus appears at age 5 months, on the average. Shenton's line (*S*) is the arc of the femoral neck, which should continue smoothly into the superior pubic ramus. This is a teratologic hip dislocation; note extreme height and rounded false acetabulum. **B:** A subtler example of congenital dislocation of the left hip.

Treatment involves different measures at different ages. The aim of all therapy is to restore contact between the femoral head and the acetabulum. Because of the high percentage of patients who experience spontaneous improvement of lax hip capsules in the early perinatal period, most orthopedists recommend observing a hip that is subject to subluxation and reexamining it at age 3 to 4 weeks. Dislocated hips should be treated at the time of diagnosis. Usually, the initial treatment is a brace that holds the hip in flexion and abduction, such as the Pavlik harness, which allows some motion while it holds the hip reduced. The alignment should be checked by radiography in 1 to 2 weeks. The brace is worn until the results of clinical and radiologic examinations are normal, an interval equal to approximately one to two times an affected child's age at diagnosis. If treatment is begun after affected children have reached age 6 months, usually they are too large and strong to tolerate the brace. At that point, reduction is performed under general anesthesia. Traditionally, this procedure has been preceded by a period of traction to stretch out the soft tissues, but the necessity of this stretching has been questioned. Closed reduction is attempted first and usually is checked with an arthrogram. If it is successful, the reduction is held in a spica cast for a number of months. If closed reduction is unsuccessful, open reduction should be performed. Such reduction involves tightening the lax superior capsule and releasing the tight psoas tendon and inferior capsule, allowing the femoral head to be brought down to its appropriate location.

If extensive distortion of the bones (i.e., a shallow acetabulum or a rotated femur) has occurred, a femoral or pelvic osteotomy and open reduction might be indicated. This condition is more common in patients who are older than 2 years.

Possible complications include persistent dysplasia from failure of normal development, recurrent dislocation, and avascular necrosis of the femoral head. The latter condition, which is the most serious complication of DDH, is caused by obstruction of the epiphyseal vessels by excess pressure or capsular stretch. Avascular necrosis is more likely to occur when a hip is reduced in a patient who is older than 6 months and requires excessive traction or abduction to reduce the hip.

The earlier treatment is carried out, the better is the resultant hip development and the safer is each of the steps in treatment. Thus, careful, methodical early screening can decrease the need for complex orthopedic procedures later.

Transient (Toxic) Synovitis of the Hip

Transient (toxic) synovitis of the hip is a diagnosis of exclusion; it is a self-limited condition that represents the most common cause of an irritable hip in children. The usual clinical presentation is a painful limp or hip pain of acute or insidious onset, usually occurring unilaterally. The most common age range for the condition is 2 to 6 years, but it has been described in patients ranging from ages 1 to 15. Spasm occurs on testing of hip range of motion, particularly with internal rotation. The temperature, white blood cell (WBC) count, and erythrocyte sedimentation rate may be normal or slightly elevated. The cause of the condition is unknown; an immune mechanism or viral infection is postulated. The differential diagnosis should include septic arthritis, osteomyelitis, and Legg-Calvé-Perthes disease, which usually is associated with a subchondral crescent of lucency or further changes in the femoral head on radiography. Juvenile monoarthritic rheumatoid arthritis and slipped capital femoral epiphysis (SCFE) also should be considered. Admission to the hospital, observation, and possible early aspiration should be undertaken if septic arthritis cannot be ruled out. Treatment consists of bed rest, with analgesic agents provided as needed for 2 to 7 days. Sometimes, if the diagnosis is clear, therapy can be accomplished on an outpatient basis with frequent follow-up. Persistence of the symptoms beyond 1 week should prompt reevaluation, although bed rest for as long as 1 month occasionally has been required.

Legg-Calvé-Perthes Disease (Coxa Plana)

Legg-Calvé-Perthes disease first was differentiated from tuberculosis within a decade after the popularization of radiography, but its cause still is unknown. More recent evidence indicates that some cases may be caused by a subclinical hypercoagulable state, such as a deficiency of antithrombotic factors S or C or a decrease in fibrinolysis. The condition is characterized by ischemic necrosis of the proximal femoral epiphysis with later resorption. The amount of the femur that is rendered ischemic varies and affects the outcome. Ischemia is followed by reossification with or without collapse of the femoral head. Legg-Calvé-Perthes disease usually, but not exclusively, affects children between ages 4 and 8. Boys are affected four times as often as are girls. As a group, affected patients have

slightly shorter stature and delayed bone age as compared to their peers. Fifteen percent of all cases are bilateral.

Usually, the clinical presentation of this disorder is a limp (i.e., an abductor lurch) with minimal pain of either short or long duration. The pain is not as acute or severe as that of transient synovitis or septic arthritis. Motions that are especially limited include internal rotation and abduction. Internal rotation is performed with the patient supine and the hip flexed, and the angle to which the leg may be rotated laterally is measured. These movements may be resisted by mild spasm or guarding. In the earliest stage, radiographic results may be normal or reveal that the affected femoral epiphysis is slightly smaller as compared to the contralateral side, as a result of its failure to grow after becoming avascular. Later, a narrow crescentic lucency, seen best on the lateral view, may be observed; it is the result of a tiny fracture of the subchondral bone. This view reveals the extent of bone involved (Fig. 432-5A). In some cases, revascularization may occur without collapse; in others, revascularization of the femoral head is accompanied by progressive resorption and deformation, often with lateral and superior migration (Fig. 432-5B). Reossification follows, and the femoral head continues to grow (Fig. 432-5C). Whether this further growth occurs spherically depends on the patient's age, the amount of collapse, and the method of treatment.

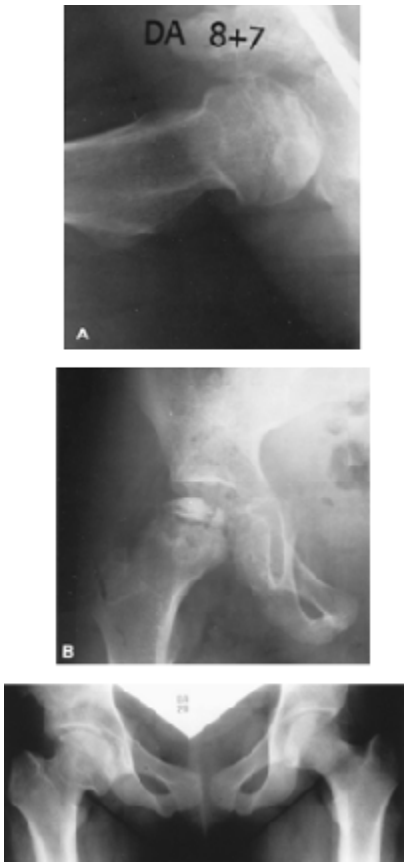

Figure 432-5. **A:** Early Legg-Calvé-Perthes disease, showing subchondral “crescent.” **B:** Later, resorption and apparent collapse of the femoral head are evident. **C:** Finally, restoration of the spherically ossified femoral head is seen here at age 20, although the femoral neck is slightly short.

The differential diagnosis should include transient synovitis, septic arthritis, hematogenous

hemoglobinopathy, Gaucher disease, hypothyroidism, and the epiphyseal dysplasias. Often, the latter two conditions are temporally symmetric bilaterally, whereas Legg-Calvé-Perthes disease is not.

Treatment follows two principles: containment of the femoral head within the acetabulum and maintenance of range of motion. During the vulnerable phase, the avascular portion of the femoral head is less likely to become deformed severely and is more likely to reconstitute spherically if it is *contained* within the “mold” of the acetabulum by abduction. Children younger than 6 years or those who have involvement of less than one-half the femoral head may be observed without active treatment if a full range of motion is preserved because this range signals containment, and patients in this age group have a good prognosis. Aggressive treatment is indicated for patients who have involvement of more than one-half the femoral head and are older than 6 years.

Containment may be achieved by the use of an orthosis or by surgery. Orthoses produce abduction with or without internal rotation. The orthosis used most commonly is the Scottish Rite brace, which does not extend below the knees (Fig. 432-6). Affected children are allowed to perform any activity that is possible in the brace. The orthosis should be worn until early reossification is seen. Generally, surgical treatment is more effective in achieving containment than is the brace. Either a femoral osteotomy to redirect the involved portion within the acetabulum or an innominate osteotomy or shelf procedure may be performed. The femoral osteotomy may cause slight shortening and an increased likelihood of a limp, but it can be controlled more precisely. The two procedures produce approximately equal results. Surgery does not speed the healing of the femoral head but causes it to reossify in a more spherical fashion. Generally, children with Legg-Calve-Perthes disease have intermittent mild aching in the hip for 1 to 2 years until reossification is complete, but then they are virtually asymptomatic throughout childhood. Symptoms may develop later in adulthood because of the lack of sphericity of the femoral head, depending on the degree of this deformation.

Figure 432-6. Scottish Rite brace for Legg-Calvé-Perthes disease produces containment by abduction and allows free knee motion.

Slipped Capital Femoral Epiphysis

SCFE is a growth-plate disorder that occurs near the age of skeletal maturity; it involves a three-dimensional displacement of the

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epiphysis (posteriorly, medially, and inferiorly). In other words, the femur is rotated externally from under the epiphysis. The cause appears to involve both mechanical and biological factors. Usually, SCFE occurs without severe sudden force or trauma. Mechanically, in most affected children, increased stress occurs as a result of obesity and abnormal retroversion (posterior rotation) of the femoral head and neck. The periosteum at this age is thin and less able to resist the shearing forces. Possible biological causes include hormonal factors and delayed growth-plate maturation, which may account for the associated obesity. Increased growth hormone levels have been associated with decreased physeal shear strength, and hypothyroidism has been found in some cases. Usually, SCFE occurs during the growth spurt and before menarche in girls. The condition is rare, with a frequency of 1 in 100,000 to 8 in 100,000. It is more common in men and in blacks. Approximately one-fourth to one-third of all affected children experience bilateral involvement but usually not simultaneously.

The clinical presentation varies with the acuity of the process. Most affected children exhibit a limp and endure varying degrees of aching or pain. The discomfort may be in the groin, but often it is referred to the thigh or knee. This variance may cause confusion: Many patients are dismissed for an apparent knee complaint with no obvious cause, only to have the true hip pathology discovered later with worsening of the slip. This paradoxic distribution of pain is attributed to referral within the femoral nerve distribution, which involves both the hip and knee joints. Some patients have acute, severe pain and inability

to walk or move the hip. Abduction, internal rotation, and flexion are the motions that are most limited. A characteristic finding is external rotation of the hip with flexion, which is caused by the preexisting retroversion and the slip itself (Fig. 432-7). Apparent limb shortening as a result of the proximal displacement of the metaphysis may be present.

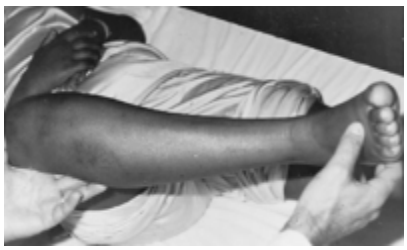
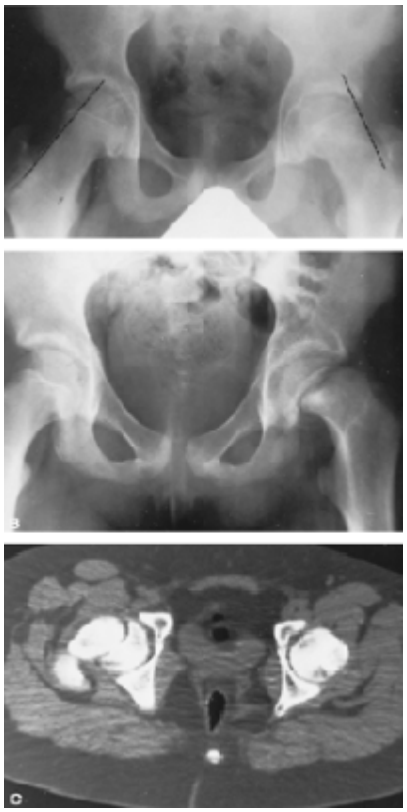


Figure 432-7. In slipped capital femoral epiphysis, the hip rotates externally as it is flexed by the examiner.

The earliest radiographic findings are widening and irregularity of the growth plate and osteopenia of the femur. Later, displacement of the epiphysis occurs and is seen best on the frog-leg lateral view of the pelvis. On the AP view, a line drawn through the upper margin of the narrowest portion of the neck should intersect at least 20% of the epiphysis (Fig. 432-8). This point is important because, with remodeling during chronic slipping, a step-off at the junction of the epiphysis and metaphysis may be absent. The severity of the slip is graded as mild (<33%), moderate (33% to 50%), or severe (>50%). Later changes may include avascular necrosis of the epiphysis or chondrolysis (i.e., joint-space narrowing).

Figure 432-8. Radiographic findings in slipped capital femoral epiphysis. **A:** A line drawn along the superior-lateral femoral neck intersects less than the normal 20% of the epiphysis on the left (affected) side. **B:** A more severe slip, showing that the femoral neck subluxates laterally and superiorly with respect to the epiphysis. **C:** Computed tomographic scan most clearly shows the direction of the slip. This figure shows *in situ* fixation with a single screw, the preferred method for slips of mild to moderate degree and even many cases of severe degree.



Treatment centers on preventing further slippage, usually by placing affected patients immediately at bed rest and obtaining a prompt orthopedic consultation. Surgery is intended to stabilize the upper femur and to cause the growth plate to close. Realignment of the slip is not safe in chronic cases because the forces necessary to accomplish realignment may produce avascular necrosis by disrupting the blood supply to the epiphysis. The gold standard of treatment is screw fixation *in situ*. The pins should not penetrate the joint. Osteotomy of the proximal or distal neck to correct the deformity has been performed occasionally, but it carries a high risk of avascular necrosis. The contralateral

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side should be monitored by affected patients' parents for symptoms of SCFE and should be pinned early if such symptoms occur. Long-term follow-up reveals no *early* degenerative change unless chondrolysis or avascular necrosis occurs; each has an incidence of 1% to 5% and produces disability during adolescence. However, even in the absence of these complications, degenerative joint disease may occur in middle age.

Increased Femoral Anteversion

Increased femoral anteversion is one of a spectrum of torsional deformities that affect the alignment of the knee and foot with the body. The differential diagnosis of toeing-in includes this disorder and internal tibial torsion and foot deformities, such as metatarsus adductus (see Table 432-1). Increased anteversion of the femur is defined as an increase

in the angle between the plane of the femoral neck and the plane of the knee (posterior femoral condyles; Fig. 432-9). Normally, this angle is approximately 30 degrees at birth and declines to 15 degrees by age 10. Femoral anteversion persists in some neuromuscular conditions, presumably as a result of lack of remodeling forces. The type discussed here is isolated idiopathic femoral anteversion.

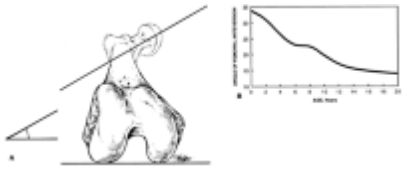


Figure 432-9. **A:** Femoral anteversion is defined as the rotation of the femoral neck forward (in comparison with the distal condyles), as seen in this view down the axis of the femur. **B:** The curve shows the normal decrease in femoral anteversion with age.

On physical examination, affected patients appear to toe in unless compensatory external tibial torsion is present. The patellae also face medially (“squint”). Internal rotation of the hip is much greater than is external rotation in both flexion (supine) and extension (prone). Usually, anteversion is not clinically significant unless external rotation at the hip is less than 15 degrees.

Radiographically, the femoral head and neck appear to be relatively straight on an AP film, with the patella forward. This is a one-plane projection of a two-plane deformity. Computed tomography (CT) is the best device for measuring femoral anteversion directly.

The natural history of femoral anteversion is benign. In a few patients, it may contribute to patellar malalignment. Anteversion later in life has been found to be unrelated to arthritis of the hip or knee. Anteversion does not impair function. Treatment of increased anteversion consists of observation, at least until affected patients are age 8, and restriction from prolonged W-sitting (with the knees touching and the legs folded under), which may impair remodeling. Instead, affected children should sit in the tailor position (with the feet tucked under and the knees out to the side). Such braces as cables and bars are not effective in derotating the femur, and no orthotic method of treatment affects anteversion. In fact, most cases need no treatment.

Femoral osteotomy, proximally or distally, is the only truly effective therapy. It should be performed rarely, however, and only in children who are older than 8 and have functional disability as a result of patellar malalignment or, rarely, a persistent concern regarding their appearance.

Knee

Extensor Mechanism Disorders (Patellofemoral Problems)

The patellofemoral joint is subject to repeated high loads of laterally and posteriorly directed forces. Numerous conditions involving this joint have been described in children and adolescents, and they are treated by attempting to improve the basic forces.

Chondromalacia refers specifically to the appearance of softening and degeneration of the patellar cartilage. Patellar subluxation refers to partial lateral displacement of the patella. The terms *patellofemoral stress syndrome*, *patellar malalignment*, and *excessive lateral pressure syndrome* refer to the abnormal mechanics causing stress concentration and pain.

The patellofemoral force may be as great as 2.5 times body weight and is greatest in flexion. The average tibiofemoral angle is angled approximately 6 degrees outward, which the patella must follow. The quadriceps-patella mechanism itself is angled away from the midline of the body, as measured by the Q (quadriceps) angle from the anterosuperior spine to the patella to the tibial tubercle (Fig. 432-10A). These high forces and asymmetric loads cause minor variations to become significant, especially when repeated over high numbers of cycles as a part of daily living. Possible factors contributing to patellar pathology (Fig. 432-10B) include increased outward angulation of the knee, abnormal rotation in the form of increased anteversion of the femur or external torsion of the tibia, a high patella (“alta”), abnormal shape or development of the quadriceps, or flattening of the femoral groove. Laxity of the medial side of the patellar restraints contributes to subluxation or dislocation. Normally, women have slightly greater genu valgum than do men. Usually, the aforementioned factors cause greater stress on the lateral side of the patella and, sometimes, decreased medial patellofemoral contact. Cartilage degeneration occurs as a result of the decreased contact, beginning in the deep layers centrally and medially and becoming visible later.

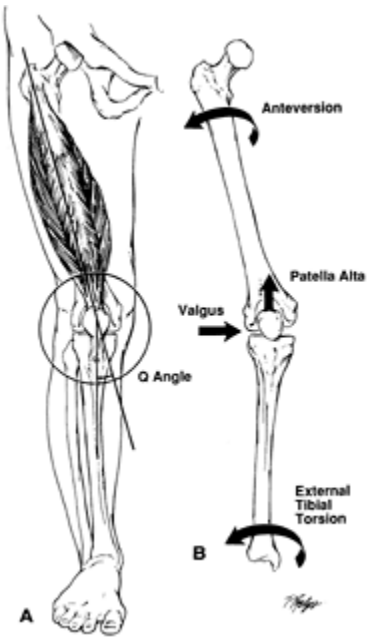


Figure 432-10. **A:** The Q angle, measured from anterior superior iliac spine to center of patella to tibial tubercle. **B:** Factors that may contribute to excess patellofemoral stress.

Clinically, problems with the patella cause aching that is greatest in the anteromedial knee region, on the medial side or

center of the patella. Usually, this pain is worse with stair climbing or prolonged sitting, as flexion increases patellofemoral force. Crepitus may be felt, but it may be painless in some patients and is not pathologic in itself. “Catching” or “locking” may be noted and might represent pain-induced inhibition or mechanical phenomena. A feeling of “giving way” may be described by the patient, especially with subluxation of the patella.

On physical examination, the most reliable way to test patellar tenderness is by direct compression of each facet against the femur. Palpation under the patella is not diagnostic. Contraction of the quadriceps and patella against resistance is nonspecific because it may be painful even in normal persons. Effusion is present only if patellar degenerative changes or extreme overuse has occurred. Reproducing patellar subluxation with laterally directed pressure may cause apprehension. The Q angle, femoral anteversion, and tibial torsion should be checked. Usually, radiographic results are nonspecific, but lateral displacement or tilt of the patella may be seen occasionally on the sunrise view.

The natural history of patellofemoral stress disorders is that they are common in patients between the ages of 10 and 20, but often they become less symptomatic later. Usually, they do not progress to osteoarthritis.

The differential diagnosis includes a synovial fold or “plica” that may snap over the medial femoral condyle, a medial meniscus tear, tendinitis of the quadriceps or patellar tendon, or osteochondritis dissecans of the patella or distal femur.

Treatment consists of altering the abnormal stresses that are occurring. Modification may include decreasing activities performed with the knees flexed, especially those that cause pain (i.e., stair climbing, prolonged sitting, and bicycling). Temporary rest from sports and the use of nonsteroidal antiinflammatory agents may be necessary. Exercises to strengthen the medial (stabilizing) part of the quadriceps include resisted extension from 0 degrees to 30 degrees, most practically by lifting weights within this range or extending the knee on a pillow, flattening it. Hamstrings and rectus femoris muscles, if they are tight, should be stretched to decrease preload on the extensors. Arch supports may help if severe flexible flatfoot is contributing to tibial torsion. Surgical measures include release of a tight lateral patellar retinaculum, medial soft tissue tightening, tibial tubercle transfer, or correction of genu valgum, knee anteversion, or patella alta if it is severe; all produce satisfactory pain relief in 75% to 90% of patients.

Patellar dislocations may be acute, recurrent or, rarely, habitual. Almost always, they occur in the lateral direction. Acute dislocations are associated with significant swelling and medial knee pain and with a history of significant outward or rotating force. They should be treated for extension with a lateral knee immobilizer until symptom-free, except in skeletally mature patients with bony avulsion. Recurrent subluxation is common, causes less pain and swelling, and often occurs with minimal force. Usually, an associated extensor mechanism abnormality is present. A realignment operation (as described) is the only effective way to stop frequent and bothersome episodes.

Variously, Osgood-Schlatter disease, patellar tendinitis (jumper's knee), and quadriceps

tendinitis are manifestations of excessive, repetitive stresses on the extensor mechanism. They are listed here in order of decreasing frequency in children.

Osgood-Schlatter Disease

Osgood-Schlatter disease is a traction-induced inflammation of the tibial tubercle. It is a reaction of the bone and cartilage of this region to high stress. The tibial tubercle is a downward extension of the proximal tibial epiphysis. It develops an ossification center in patients between ages 9 and 13, but it does not ossify completely until they are 15 to 17 years old. Within this age range, repetitive stresses gradually can deform the outer surface of the tubercle plastically, causing it to enlarge and become locally inflamed (Fig. 432-11). Tenderness and swelling are localized to this region. Symptoms are worse with running, jumping, or kneeling. Treatment involves decreasing activity to a tolerable level and occasionally using a knee immobilizer, crutches, and ice after activity in severe cases. The patient may be vulnerable to recurrence of symptoms for up to 2 years until the tubercle matures. If affected children and their families are informed of this likelihood, individual regulation of activities can be effective. Usually, activities of daily living and even some sports are tolerated, using daily stretching of tight quadriceps and hamstrings and occasional antiinflammatory agents. Complete avulsion of the tubercle is extremely rare and seems to be related more to sudden stress than to apophysitis.

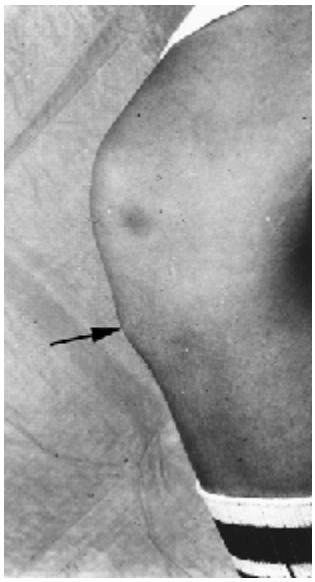


Figure 432-11. Osgood-Schlatter “disease” often produces an enlargement of the tibial tubercle (*arrow*).

Patellar Tendinitis

Inflammation at the origin of the patellar tendon (which is at the inferior pole of the patella) is known as *patellar tendinitis* and is related to the same type of overuse as that seen in Osgood-Schlatter apophysitis. Most often, it is seen in basketball players and also is called *jumper's knee*. The duration of pain serves as a guide to the severity of involvement. Pain that is present during both rest and activity is more worrisome than is pain that occurs only after activity. Treatment is the same as that for Osgood-Schlatter disease. Warm packs

before and cold packs after activity also may be beneficial. Rarely, pain may occur at the proximal pole of the patella; in this case, the condition is termed *quadriceps tendinitis*. Treatment is the same as that described for Osgood-Schlatter apophysitis.

P.2110

Tibiofemoral Disorders

Popliteal Cysts

Popliteal cysts in children are localized behind the knee on the medial side (Fig. 432-12). They are firm and nontender, vary in size with activity, and transilluminate in a darkened room. They occur most commonly in boys younger than age 9. Unlike those in adults, usually these cysts in children are not associated with any intraarticular pathology, and they tend to resolve spontaneously with time. Actually, the recurrence rate is higher after surgical excision. The origin of these cysts is a slitlike communication between the knee joint and the gastrocnemio-semimembranous bursa.

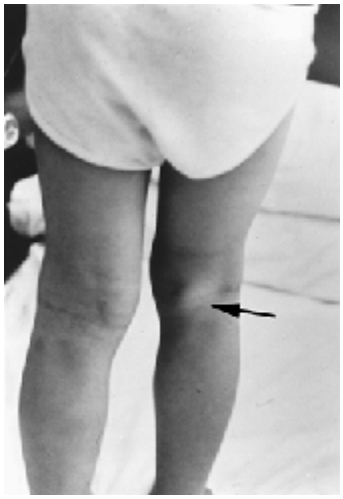


Figure 432-12. Popliteal cyst (*arrow*) in typical (medial) location behind right knee of this child.

Discoid Meniscus

An acquired flattening of the lateral meniscus is known as *discoid meniscus*. In some cases, this flattening occurs as a result of the absence of normal peripheral attachments. Often, such symptoms as pain, clicking, and locking develop in the absence of trauma in children from the age of 2 to adulthood. The meniscus should be trimmed or excised if symptoms become severe. If the attachments are intact, no removal is necessary unless a tear is seen.

Genu Varum

Genu varum, or “bowed leg” of up to 20 degrees, is normal in children until age 18 months (Fig. 432-13). Normally, it does not increase significantly after walking begins. After the child reaches the age of 24 months, genu valgum normally develops. Radiographs are

indicated if genu varum is present after this age or is progressive after the age of 1 year, if it appears to be severe, or if it occurs in a high-risk group, such as obese black children who walk early. Radiographic findings of benign genu varus include symmetric bowing of the tibia and femur, a normal-appearing growth plate without narrowing or step-off, and a generalized, rather than focal, outward bowing (Fig. 432-14).

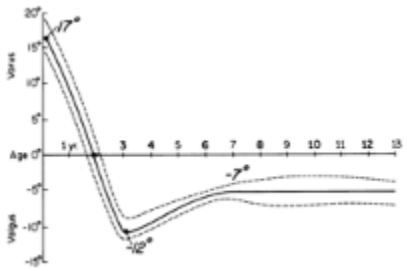


Figure 432-13. Normal change in the tibiofemoral angle during growth. (Reproduced with permission from Salenius P. Development of the tibiofemoral angle in children. *J Bone Joint Surg* 1975;57A:260.)

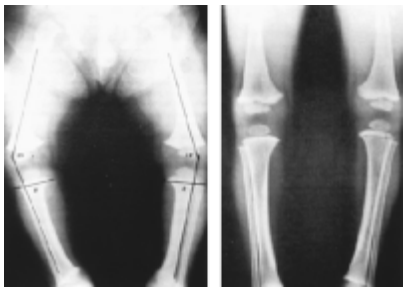


Figure 432-14. Note improvement in physiologic bowing between age 18 months **(A)** and 24 months **(B)** without treatment.

Treatment involves observation to verify resolution. Measurement of the angle on physical examination should be performed with the child standing and may be accomplished also by measuring the distance between the femoral condyles or of the AP tibiofemoral angle. These methods are not as accurate as are radiographs, but they are a practical way of observing change in patients when the presumptive diagnosis is physiologic genu varum.

The differential diagnosis of physiologic genu varum includes Blount disease, rickets, posttraumatic growth-plate disturbance, enchondromatosis, achondroplasia, and other skeletal dysplasias. The ratio of physiologic bowing to Blount disease is more than 1,000:1.

Tibia Vara (Blount Disease)

Tibia vara, also known as *Blount disease*, is an idiopathic, probably mechanical deficiency in the medial tibial growth plate that may be unilateral or bilateral. Initially, it may present in two different age groups: infants and adolescents.

Almost always, untreated infantile tibia vara is progressive and, in addition to exhibiting outward angulation, includes

flexion, internal rotation and, often, abnormal lateral knee laxity. Radiography demonstrates progressive depression of the medial metaphysis, the growth plate, and the epiphysis and, eventually, fusion of the medial metaphysis to the epiphysis in severe cases. A helpful early distinction in tibia vara is the focal nature of the change, with sharp angulation of the proximal tibial metaphysis resulting in a metaphyseal-diaphyseal angle of 11 to 16 degrees or more, measured as shown in Figure 432-15. It is a specific sign because such localized angulation occurs in fewer than 5% of children with physiologic varus but is seen in essentially all those with Blount disease.

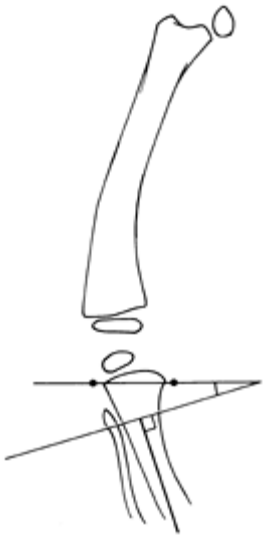


Figure 432-15. Metaphyseal-diaphyseal angle. The angle is formed by the line of the tibial shaft and the line between the medical and lateral beaks of the proximal tibial metaphysis. (Redrawn with permission from Levine AM. Physiologic bowing and tibia varum. *J Bone Joint Surg* 1982;64A:1159.)

Night-brace treatment, though not formally proven to be effective, usually is used for mild but definite cases of Blount disease. Valgus rotational osteotomy of the tibia is indicated if the angulation persists beyond 3 years. Recurrence is common if treatment begins after age 4, if the epiphysis is fragmented, or if an affected child is obese. Persistent tibia vara leads to early degenerative change.

Adolescent tibia vara has its onset in children older than age 9. It is most common in obese boys. Probably, it is caused by decreased growth of the medial tibial physis resulting from excessive medial stresses. Radiography shows medial femoral and tibial bowing. Bracing is not practical in these obese adolescents. Treatment involves osteotomy to realign the limb or lateral growth-plate closure to allow growth to “catch up” medially.

Genu Valgum

Genu valgum of the knee is normal in children older than age 2, reaches a mean of 12

degrees at age 3, and remains approximately constant at a mean of approximately 7 degrees in boys and 9 degrees in girls after age 8. Night bracing may be helpful in children with angulation exceeding 20 degrees. If the angle remains greater than 15 degrees when the child reaches age 10, early growth-plate stapling or later osteotomy of the affected region may be indicated. Valgus of the proximal tibia often follows medial metaphyseal fractures, but frequently it corrects spontaneously.

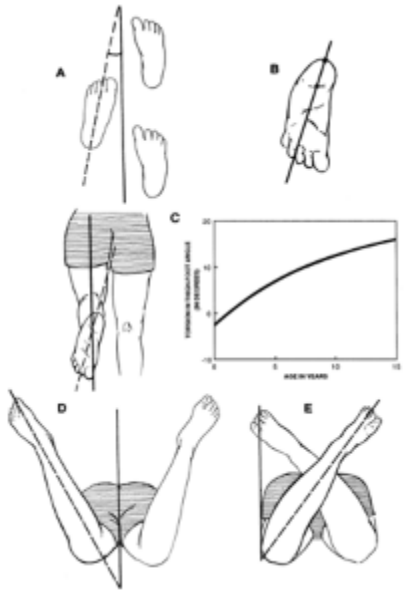
Tibia

Internal Tibial Torsion

Internal tibial torsion is the most common cause of toeing-in in children between ages 1 and 3. Tibial torsion is determined by measuring the angle between the foot and the thigh with the

ankle and knee positioned at 90 degrees. Normally, the foot rotates externally with age (Fig. 432-16). The differential diagnosis includes metatarsus adductus, femoral anteversion, and neuromuscular disorders. For making these distinctions, the foot as well as the hip should be examined (see Fig. 432-16A–C). Tibial torsion improves naturally with growth, but this improvement often takes years. Because of our improved knowledge of the benign natural history of this condition, bracing with such devices as the Denis-Browne bar is used only rarely. Studies have shown that braces cannot apply significant rotational force to the tibia because the corrective force is taken up in the foot, knee, and hip joints. The improvement that previously was attributed to the brace is primarily the result of normal growth patterns. Correction is a slow process and often frustrates parents. Knowledge that braces were used heavily in the past, reinforced by grandparents and friends, often drives anxious parents to visit the doctor to ensure that they are not missing a golden opportunity to avoid problems. Attendant physicians should be confident in allaying such anxiety and should advise them in some such fashion as, “Your child is toeing-in because of inward twisting of the tibia, or leg bone. This is a normal stage in resolution of the position in the womb. Some children have more toeing-in than others. Although such children occasionally may catch a foot when they run, this will improve. As the bone grows longer, it will grow straighter. It may take a few years to correct. Doctors used to use braces for this, but we found that children were getting better by themselves. You can reassure friends and relatives that the child will outgrow it.” Use of the graph shown in Figure 432-16C may prove convincing. Although very little evidence substantiates the efficacy of a brace or of any orthotic method, it has been a very widely used treatment that now is declining. Minor persistent internal torsion has not been shown to be detrimental.

Figure 432-16. Assessment of torsional deformities. **A:** Angle of progression (the angle between the foot and the line of gait)—summation of femoral, knee, tibial, and foot relationships. **B:** Assessment of metatarsus adductus. Normally, the heel bisector falls between the second and third toe space. **C:** Thigh-foot angle and its variation with age. This is a reflection of tibial torsion. Measurement of



internal **(D)** and external **(E)** rotation of the hip. If external rotation is less than 20 degrees, “in-toeing” may come from femoral anteversion or hip capsular contractures. (Redrawn with permission from Staheli LT, et al. Lower-extremity rotational problems in children. *J Bone Joint Surg* 1985;67A:41.)

External Tibial Torsion

External tibial torsion is less common. Few data address the course of the condition, no treatment is indicated, and some spontaneous improvement can be expected.

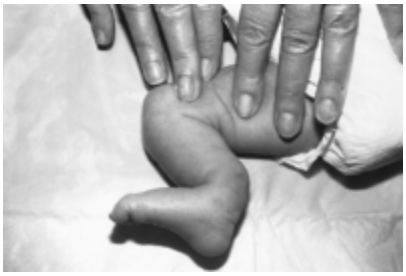
Anterolateral Bowing

Mild anterolateral bowing of the tibia is common in infancy and should be observed to ensure that spontaneous straightening occurs. Focal sclerotic defects in the tibia may be seen with severe anterolateral bowing. Fractures are present or develop in affected tibias (congenital pseudarthrosis), and some affected patients may be found to have neurofibromatosis. If severe anterolateral bowing is present but the tibia is not fractured, it should be braced for protection. If the bone is fractured, attempts to gain union by electrical stimulation, vascularized fibula grafting, and bone grafting with rod stabilization have success rates that range from 50% to 75%. Anterolateral bowing may be seen also with congenital absence of the fibula.

Posteromedial Bowing

Posteromedial bowing (Fig. 432-17) of the tibia is more benign, usually straightens by the time the child reaches age 4, and is not associated with fracture. Commonly, between 2 and 6 cm of shortening is seen by maturity, however. Treatment involves stretching the tight dorsiflexor muscles and the use of length equalization as indicated.

Figure 432-17. Posteromedial bow of the tibia in a 2-month-old child. This condition almost always resolves without treatment. Note the bow in the bone itself, which distinguishes it from a calcaneovalgus foot.



Limb-length Inequality

Limb-length differences are screened best by palpating the heights of the iliac crests with the hips and knees straight. Any

discrepancy may be confirmed with a tape measure held between the anterior superior iliac spine and the inferior edge of the medial malleolus. Most commonly, apparent differences in the lengths of the lower limbs in children are due to measurement error, as affected patients have difficulty in lying still and in holding hips and knees straight. In the normal population, differences of 1 cm between the two sides are not uncommon findings.

Discrepancies of up to 2 cm in adults (or a proportionately smaller amount in children) have been shown to have no ill effects on gait or joints and do not need treatment. Larger discrepancies should be confirmed with a radiographic film (scanogram) or with a CT scan. Causes of true, significant discrepancy greater than 2 cm may include hemihypertrophy, hemiatrophy, coxa vara, hip dysplasia, or growth-plate damage, to name a few. Because hemihypertrophy has been associated with Wilms tumor in some cases, these patients should be examined with abdominal ultrasonography two to three times per year. Treatment of significant limb-length inequalities may include a lift, an epiphyseodesis of the long side if growth remains, or a surgical shortening or lengthening procedure.

Foot

Metatarsus Adductus

Metatarsus adductus, or isolated idiopathic adduction of the metatarsals, is known also as *metatarsus varus* or *C-foot*. In contrast to conditions seen in clubfoot, in metatarsus adductus the hindfoot is normal or angled outward slightly. The ankle joint itself has normal dorsiflexion and plantar flexion (Fig. 432-18). The probable cause of this condition is medially directed intrauterine pressure. Children with metatarsus adductus also may have an increased incidence of other molding deformities, such as congenital dislocation of the hip or torticollis. Metatarsus adductus deformity should be differentiated from skewfoot, which involves severe outward deviation of the hindfoot, treatment of which is much more difficult. A rough measure of the degree of adduction can be obtained by determining the position of an imaginary line that would bisect the sole of the hindfoot. Normally, the line falls between the second and third toes; in patients with severe adduction, it is lateral to

the fourth toe (see Fig. 432-18B).

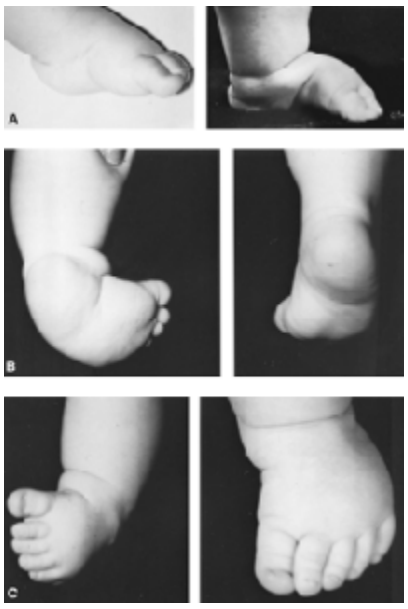


Figure 432-18. Comparison of clubfoot (*left*) and metatarsus adductus (*right*). **A:** Lateral view, showing the equinus present only in clubfoot. **B:** Posterior view, showing the hindfoot varus in clubfoot but not in metatarsus adductus. **C:** Anterior view, showing adduction in both feet, with the varus also present in clubfoot.

The natural history of untreated metatarsus adductus is spontaneous correction in 85% of children, with the persistence of mild adduction in 10% and more pronounced adduction in 5%. In one longitudinal study of 2,000 feet in newborns followed until maturity, no patients had symptomatic adduction in adulthood. Those cases that will resolve spontaneously cannot be predicted, even on the basis of severity or rigidity.

Usually, manipulative correction is successful anytime during the first 8 months of life. Thus, the author's preferred treatment is observation with stretching for the first 6 to 8 months, followed by corrective casts or splints if the condition persists beyond this time. The casts are changed every 1 to 2 weeks until the defect is corrected clinically; then a “holding cast” is applied for 2 weeks. Osteotomy for very late-presenting adduction in children older than age 3 rarely is necessary.

Clubfoot

Talipes equinovarus congenita, or clubfoot, is a more complex disorder involving not only metatarsal adduction but abnormalities of the hind part of the foot, including malrotation of the calcaneus under the talus and equinus (plantar flexion) of the ankle. The incidence is 1 in 1,000, and it is more common in men than in women. Clubfoot may be unilateral or bilateral. Its cause is unknown but appears to be related to a primary defect of local connective tissue or a very early insult to the leg muscles or tarsal bones. Muscle biopsy results are abnormal, which is consistent with the observation that the leg muscles are underdeveloped, even in treated cases.

Physical examination reveals a small foot, and the combination of deformities often results in a 90-degree rotation of the forefoot in all planes so that the leg and foot truly resemble the shape of a club. A deep crease is present on the medial border of the foot. The

deformity may be correctable to the neutral position initially only in the neonatal period, and the range of motion in all planes is limited. Radiography shows an abnormal parallelism of the talus and calcaneus, but they are not necessary in the typical case.

Neuromuscular disorders (especially lipomeningocele, myelomeningocele, or arthrogryposis) may produce similar deformities. Also, the conditions of diastrophic dwarfism and Freeman-Sheldon ("whistling face") syndrome includes a deformity similar to clubfoot.

Clubfoot ranges from a mild, "postural," and easily correctable condition to one that is severe and resistant to treatment. A trial of cast correction is indicated in all cases, however. This treatment is most successful when started in the perinatal period when ligamentous laxity is greatest, with the casts being changed every few days. Overall, casting is effective in approximately

one-third to one-half of all patients. Surgery is indicated in the others and involves complete release of all bony malalignments and tendon contractures; it is performed most commonly between the ages of 6 and 12 months.

Calcaneovalgus Foot

A calcaneovalgus foot is seen commonly in newborns; it appears dramatic but is in fact benign and self-resolving. In this condition, the foot is dorsiflexed so extremely that the dorsum of the foot is pressed against the tibia (Fig. 432-19). The cause of the disorder is intrauterine molding. Actually, the alignment of the bones of the foot internally is fine and can be verified by observing the presence of an arch and by bringing the foot to a neutral position, a movement that should not require much pressure. The toes should have normal movement. This condition should be differentiated from posteromedial bowing of the tibia, which causes the foot to assume the same direction but with a more proximal apex to the deformity and a bowing in the distal tibia. The natural history is benign. The ankle and foot stretch out naturally with time, and no cast is needed. No known sequelae accompany this condition.

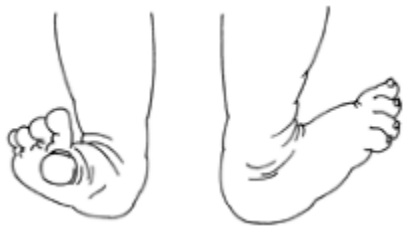


Figure 432-19. Calcaneovalgus foot shortly after birth, displaying significant valgus and dorsiflexion.

Flatfoot (Pes Planovalgus)

The condition called *flatfoot* must be divided into flexible and rigid types. The flexible type is very common in children and usually causes no symptoms. Development of the arch of the foot occurs spontaneously during the first 8 years of life in most children. The arch of the foot is restored when weight bearing is relieved. Inward-outward motion is normal. In contrast, rigid flatfoot may be caused by tarsal coalition, a vertical talus, neuromuscular imbalance (which occasionally also may be flexible), or arthritis of the foot. These conditions should be considered in the differential diagnosis.

The cause of the usual type of flexible flatfoot is ligamentous laxity, with mild secondary bony changes. No primary muscle abnormality exists. Occasionally, a tight heel cord may contribute by pulling the foot into greater outward angulation. Treatment is not indicated in asymptomatic cases of flexible flatfoot; prospective studies have shown that no orthotic or special shoe can produce a lasting change in pediatric flatfoot. Such devices may be indicated for rigid or neuromuscular flatfoot but not in asymptomatic children who have flexible flatfoot. The heel cord should be stretched if it is tight. Rarely is soft tissue reconstruction or osteotomy indicated.

General principles that a physician should stress to parents when asked about shoes are summarized in an article by Staheli. In short, shoes are primarily for protection; “corrective shoes” have no effect on flatfoot; and shoes should be flat, flexible, porous, and high-topped to prevent them from slipping off the foot. These characteristics are available in most reasonably priced footwear found in regular shoe stores.

Tarsal Coalition

Tarsal coalition is the failure of complete separation of hindfoot bones, with possible persistence of a variously fibrous, cartilaginous, or bony bridge or coalition between two of them. This anomaly is transmitted in an autosomal dominant fashion and is present in approximately 5% of the population. Many individuals with tarsal coalition are asymptomatic. The presence of symptoms seems to be related to the degree of outward angulation that is present, which places more shear strain on the abnormal junction.

Usually, the diagnosis is made during the second decade after an ankle “sprain,” with persistent pain or the spontaneous onset of pain in the ankle. The probable reason for this presentation is that the ossification that occurs at this time, near the point of skeletal maturity, renders the coalition stiffer. The hindfoot shows limitation of the inward-outward motion, but it is tender to palpation. Sometimes pain is manifested over the peroneal muscles, which contract excessively to stabilize the foot.

Oblique radiography of the foot can illustrate reliably the most common type of coalitionthe calcaneonavicular barif it has ossified. If it is fibrous or cartilaginous, a bony connection may be absent, but an irregularity of the cortices might be seen. Negative radiographic results combined with clinical suspicion indicate the need for a coronal CT of the foot to search for a talocalcaneal coalition, which is the next most common type of deformity.

With rest or casting, many cases of tarsal coalition will stabilize and become painless. If pain persists, the coalition can be excised if it is not large and if no degenerative change

has occurred. If these conditions are not met, fusion of the hindfoot is indicated.

Spine

Generally, back problems in children fall into two categories: those associated with spinal deformity and those associated with pain in various parts of the back. When these conditions exist in the same child, determining the cause of the pain is more urgent than is treating the deformity.

Childhood Back Pain

Although popular wisdom formerly held that back pain in children almost always had a serious underlying cause, more recent studies have shown that nonspecific back pain is common in children, with only one-fifth eventually being given a diagnosis of a specific cause. Nevertheless, potential problems should be ruled out. Careful neurologic examination is necessary in evaluating children for back pain, as is an assessment of spinal flexibility and deformity. The differential diagnoses listed in Table 432-1 are the conditions encountered most commonly.

Musculoligamentous Pain. All the components of a child's spine (i.e., discs, ligaments, muscles, and joint capsules) are flexible and conform easily to the extreme spinal positions that are encountered daily in the school yard or on the playing field. After the child reaches approximately 12 years, generally the spine loses some of its flexibility and, during the teenage years, further “stiffening” may take place. Muscular or ligamentous back pain almost never is observed in children who are younger than 10 years. This diagnosis should be reserved for an older child (often involved in a new physical activity) who has lumbar area pain for which no other specific cause can be elucidated. To warrant this diagnosis, affected children should have pain localized to the lumbar area, a normal neurologic examination, normal results on radiography of the lumbar spine and, in some instances, normal bone scan results.

Once the diagnosis has been made, the treatment of musculoligamentous pain involves rest from any activity that causes the pain. The use of ice in the first 24 hours after onset is helpful;

thereafter, usually heat is more efficacious. Once the pain has resolved, exercises should be used to strengthen the abdominal musculature and the lumbar muscles before sports activity is resumed. Often, a lumbosacral corset is helpful in the acute stage and for a few months thereafter, when affected children are participating in sports, to protect the low back and its muscles from the extremes of spinal movement. The long-term use of a corset or other brace rarely is needed if the diagnosis of musculoligamentous back pain is correct. Once low-back pain has resolved, attendant physicians should stress to affected children that warming up before participating in sports activities is more important for them than for their teammates. The persistence of pain requires further investigation for unusual causes of back pain.

Spondylolysis and Spondylolisthesis. Spondylolysis is the most common cause of back pain to yield an anatomic diagnosis in children. Usually, it is a stress fracture of the pars

interarticularis segment of the vertebra. This thin segment of bone between the facet joints is subjected to high forces, especially with marked lordosis of the lumbar spine or with heavy lifting. The overall incidence in the general population is approximately 6%. Most of these stress fractures likely occur in the early school years, though symptoms occur most frequently when children are in their early teenage years. A much higher frequency of spondylolysis occurs in children who participate in gymnastics, wrestling, and weight-lifting activities, at times approaching 20% for participants in these sports.

Most commonly, symptoms include pain in the lumbar area after or during a sports activity and a concomitant limitation of lumbar spine motion. If affected children have a chronic spondylolysis, often the pain is intermittent; if the spondylolysis is acute, the pain is more severe the first time it is noted. Occasionally, radiation of pain along the sciatic nerve distribution into the lateral calf or dorsum of the foot may be present.

The physical examination may not be remarkable. Usually, limitation of lateral spine flexion toward the side of the spondylolysis occurs; often, it is associated with limited forward flexion from back pain, not only hamstring tightness as in some other conditions. Back pain may be produced by straight-leg raising, but radiation of pain into the legs with this maneuver is rare. The results of the neurologic examination are normal.

Often, the diagnosis can be made by lumbar spine radiographs (Fig. 432-20). The most common location of spondylolysis is at L5, with L4 being the next most frequent site. Often, spondylolysis can be visualized by AP and lateral radiographic views, but usually oblique views are more definitive. In addition, oblique views reveal the unilateral or bilateral nature of the defect. If a lytic defect is observed, the age of the lesion should be determined. Generally, the spondylolysis is old if the defect has sclerotic edges. A technetium Tc 99m bone scan with single-photon emission CT or pinhole collimator views is helpful to determine the age of the stress fracture. If the scan is cold and sclerotic edges are present on radiography, obtaining bone union nonoperatively will not be possible. If the scan shows increased uptake at the lytic area and radiography reveals no sclerotic edges, however, the stress fractures might heal if the child is placed in a body jacket brace or cast.

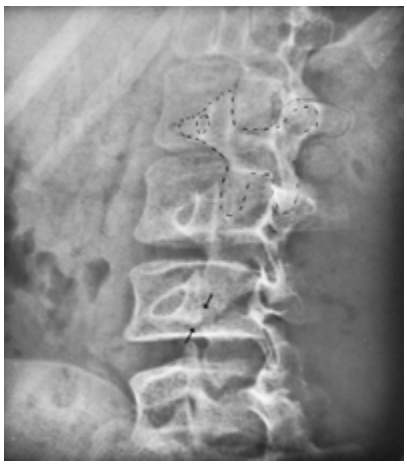


Figure 432-20. Spondylolysis. Oblique film of lumbar spine shows defect in pars interarticularis of L4 (*arrows*). Note that the posterior elements of a vertebra resemble a Scottie dog in this view, as outlined. The nose, eyes, ears, neck, and body are the transverse process, pedicle, pedicle superior articular process, pars interarticularis, and lamina, respectively. Spondylolysis appears as a break in the “neck” region.

If the scan shows no increased uptake, the treatment of spondylolysis is similar to that of a

musculoligamentous problem. Initially, rest from activity is instituted. Often, a lumbosacral corset is helpful for a few weeks until pain resolves. Some teenagers with spondylolysis prefer to wear the corset during sports activities as added protection even after acute back pain resolves. Children should be followed with serial radiography during the growing years to rule out development of a progressive slip. Generally, fusion for spondylolysis without spondylolisthesis is not needed. With the exception of occasional episodes of low-back pain, teenagers are able to participate in any sport that does not lead repetitively to back pain after playing.

Spondylolisthesis occurs in some children who have spondylolysis. This condition results from forward slipping of a superior vertebra on the inferior vertebra, most commonly slipping of L5 on the sacrum (Fig. 432-21). Worsening of this slip coincides with growth of the spine and generally subsides once growth is completed. Because the slip of the vertebra progresses forward, the posterior elements (i.e., the spinous process and inferior facets) remain behind, attached to the adjacent vertebrae by ligaments. The combination of excessive motion of the posterior elements and forward vertebral slipping may lead to irritation of L5 or S1 nerve roots. Because this slipping usually is slow, the nerve root irritation may present only as progressive tightening of the hamstrings. Affected children may note difficulty in touching their toes or reaching objects on the floor. If one side of the spine is affected more than the other, scoliosis also may be present.

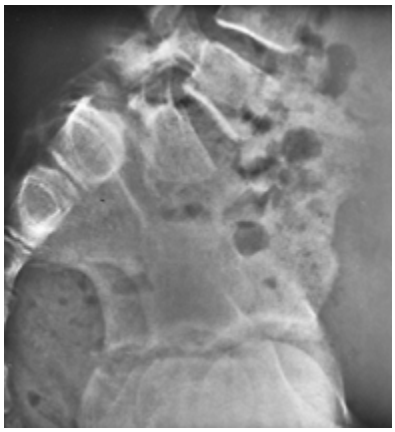


Figure 432-21. Severe vertebral slips, as shown here, lead to hamstring spasm and leg pain as well as to back pain. Surgery is needed.

On physical examination, the most striking finding is limitation in straight-leg raising because of the hamstring spasm. Radiation of pain into the calf or foot with straight-leg raising may indicate more advanced nerve root irritation. At times, the ankle jerk reflex is diminished.

A definite diagnosis can be made on the basis of plain radiography, most easily in the lateral view. The extent of slip should be estimated between grade 1 and grade 4 (i.e., grade 1, up to 25%; grade 2, up to 50%; etc.). If the slip is greater than 50%, posterior lumbosacral fusion is indicated. If the slip is less than 50%, initial management is directed toward relief of back pain and hamstring spasm, using rest and corset therapy as with spondylolysis. If the pain does not respond to conservative treatment, fusion may be needed. If the pain improves with conservative treatment, a corset may be used for sports

activities, and follow-up lateral lumbosacral radiography at 6- to 9-month intervals is recommended until growth is complete or a worsening slip can be identified. If progression of the slip occurs, fusion is indicated.

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The use of a brace may prevent further slipping. If affected children do not have hamstring tightness and are pain-free, usually activities do not need to be restricted, provided such children and their parents are aware that periodic back pain is likely to occur. No evidence substantiates that increased physical activity causes an increase in vertebral slip.

Intervertebral Disc Herniation. Herniation of the intervertebral disc is a common cause of back and leg pain in middle-aged adults. In this age group, disc protrusion occurs posteriorly, with the protruded disc compressing the nerve roots or the cauda equina. If similar forces are applied to the spine of skeletally immature children, the disc will not always rupture posteriorly, but a fracture of the ring apophysis of the vertebral body may occur, causing the extrusion of disc material anteriorly (Fig. 432-22) or into the vertebral body itself (Schmorl nodes). Most affected children have back pain without radiation into the leg or calf. If nerve-root pain also occurs, a small avulsion fracture of the ring apophysis posterolaterally might be present in a position that causes nerve-root compression. The diagnosis of an old disc injury in children can be confirmed by radiographic findings of a narrowed lumbar disc adjacent to a vertebral end plate that has some irregular pattern of ossification.

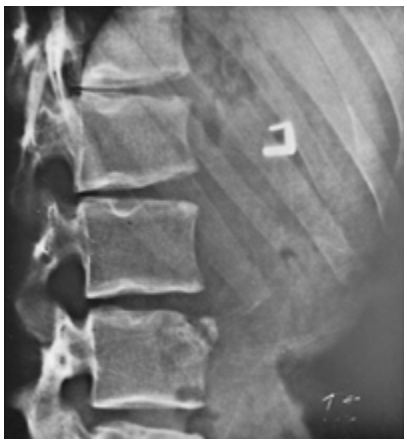

Figure 432-22. A disc rupture in skeletally immature children will lead to disruption of the apophysis or vertebral growth area, as shown here.

If no neurologic defect is present, treatment consists of symptomatic care, usually rest until the pain resolves. If the condition is the result of a vehicular accident, evaluation for the development of an ileus should be performed. If both leg pain and back pain are present, magnetic resonance imaging (MRI) or CT-myelography should be performed to localize any neural compression, which could be relieved by surgical treatment.

Discitis. Discitis may present in a wide variety of ways. Severe back pain with limitation of back movements is common in older children, whereas younger children simply may refuse to walk or may limp. Usually, the cause of this disc inflammation is bacterial infection. The vascular anatomy of the growing disc varies from that of the adult, and the common

bacteremias of childhood can infect the disc more readily than the vertebral body itself. Approximately 50% of affected children have positive blood culture results at the time of their acute pain, with *Staphylococcus aureus* being the organism identified most commonly. Despite this finding, milder forms of discitis often appear to resolve without the need for antibiotics.

The most striking finding on physical examination in discitis patients is marked stiffness of the spine that is notable with attempts at flexion. Often, fever is present. The results of the neurologic examination are normal. In the early stages, radiographs of the spine appear normal. Usually, a few weeks after the onset of pain, narrowing of a single disc may be seen on radiography. Often, the sedimentation rate and WBC count are elevated. Virtually always, a technetium Tc 99m bone scan reveals increased uptake at the involved level and should be performed whenever discitis is suspected. If the bone scan results are positive and the age and clinical presentation are typical, generally needle aspiration or open biopsy of the involved disc is not necessary.

Treatment decisions in cases of discitis revolve around choosing between antibiotics or a body jacket brace or cast. If a positive blood culture result has been obtained, antibiotics should be used for 3 to 6 weeks. We prefer to use antibiotics in children who have a significant pain or a positive bone scan result, even if no bacteremia has been demonstrated. Bed rest should be instituted at the time of presentation if significant spasm is present. If the spasm persists for more than a few days, a body jacket brace or cast will allow for immobilization and ambulation on a limited basis. Rarely does discitis develop into vertebral osteomyelitis with local bone destruction. Usually, the involved vertebral bodies fuse together eventually after the infection resolves.

Spinal Cord Tumors. Back pain and limitation of spinal movement may be seen also as the presenting problem in patients with spinal cord tumors, even without a demonstrable neurologic deficit. In one large series of spinal cord tumors, the presenting complaint was back pain or scoliosis in almost one-third of the children. The hallmark of the physical examination is severe limitation of forward flexion of the spine. Pain may be worsened by neck flexion. Neurologic changes may be very subtle, and detection may be difficult.

In patients with back pain and marked limitation of spinal motion, especially if scoliosis also is present, MRI or CT-myelography is needed if the bone scan and plain radiography do not elucidate the cause. The most common tumor is an ependymoma. Treatment is neurosurgical. If the tumor is benign and can be removed, the pain, scoliosis, and back stiffness generally resolve.

Spinal Deformity

Approximately 5% of children will experience some degree of spinal deformity as they grow. Scoliosis, a lateral curvature of the spine, is the most common condition, although increased thoracic kyphosis or round back is not rare. School screening programs for spinal deformity, which are mandated in many states, have served to increase the public's awareness of these conditions. Whereas formal school programs generally are targeted toward children in the fifth or sixth grades, routine evaluation of the back should be a

feature of each child's annual examination.

Scoliosis. Scoliosis is a lateral curvature of the spine. The two forms of scoliosis are postural and structural. Postural scoliosis results from spinal factors outside the spine, such as leg-length discrepancy or hip disorders. In these cases, if the leg lengths are equalized or if the child sits, the spine becomes straight, indicating that no structural change has occurred. Structural scoliosis is of greater concern, because it involves not only a lateral spinal curvature but a rotation of the vertebrae involved in the lateral curve.

Although numerous conditions are associated with scoliosis, the most common groups include idiopathic (80%), congenital (5%), neuromuscular (10%), and miscellaneous (5%) disorders. The miscellaneous disorders encompass connective tissue disorders, genetic diseases, and other, less common, conditions.

Congenital scoliosis is present at birth, though often the diagnosis is not made at that time (Fig. 432-23). It may be associated with other birth defects or may present as an isolated condition. Because the genitourinary system arises embryologically from the same region as that of the spine, approximately 30% of children with a congenital spinal deformity have an associated genitourinary abnormality. Because the most common anomaly is unilateral renal agenesis, sonography or intravenous pyelography should be performed on all patients who have congenital scoliosis or kyphosis. Although active treatment of unilateral kidney absence may not be necessary, an important adjunct is appropriate cautioning against the child's participation in contact sports that may lead to kidney injury. The treatment of congenital scoliosis consists of serial radiographic follow-up to determine whether the deformity is worsening. If no curve progression occurs, generally further treatment is not needed. If worsening of 5 to 10 degrees or more is documented, surgical fusion is necessary, regardless of an affected child's age. Brace treatment may be useful to prevent worsening of curves above or below the congenital scoliosis, but it is not indicated for the congenital scoliosis itself.

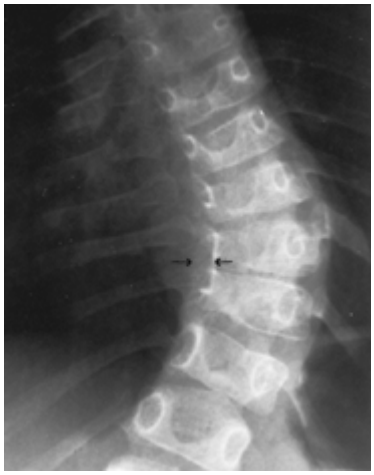

Figure 432-23. Congenital scoliosis results from incomplete vertebral segmentation *in utero* (*arrows*). Frequently, genitourinary abnormalities are associated with these bony deformities.

Neuromuscular scoliosis is spinal deformity associated with a wide variety of neurologic or muscular diseases, such as cerebral palsy, muscular dystrophy, myelomeningocele, and poliomyelitis. Spinal curvature secondary to muscular imbalance classically is C-shaped

and extends to include the pelvis (Fig. 432-24), which usually is not the case in idiopathic scoliosis. Scoliosis is present more often and tends to worsen most quickly in patients who do not walk because of their neuromuscular disease. With continued progression, sitting balance becomes impaired further, and the child may need to use one arm or hand to assist in sitting. Treatment centers on preservation of sitting ability and pulmonary function. Although wearing a brace often is useful, frequently surgical fusion is indicated to preserve function.

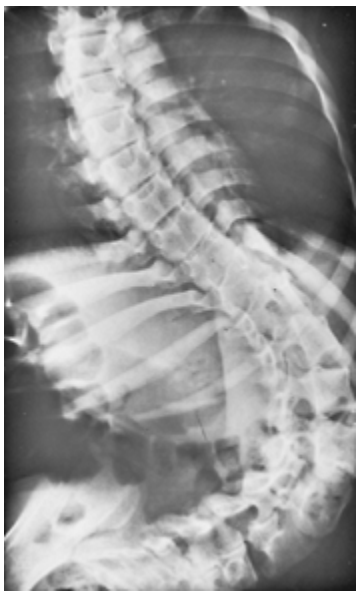

Figure 432-24. Sitting anteroposterior radiograph of a child with cerebral palsy and severe scoliosis. Note the C-shaped curve and the pelvic tilt characteristic of neuromuscular scoliosis.

Generally, idiopathic scoliosis is found in otherwise healthy children. Although idiopathic scoliosis requiring treatment is approximately eight times more frequent in girls than in boys, the incidence of mild curves is approximately equal between the genders.

A family history of curvature of the spine is found in as many as 70% of all children with scoliosis, though the exact mode of inheritance has not been determined definitely. Although the cause of idiopathic scoliosis remains elusive, a combination of growth asymmetry and postural imbalance is

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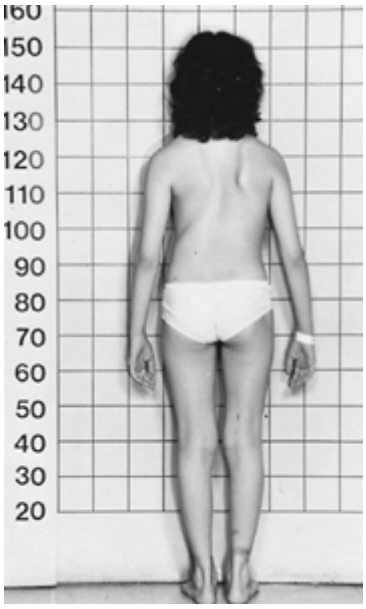
believed to be important. Minor abnormalities in the postural control center in the brainstem have been demonstrated in children with mild scoliosis. Once the curve begins to develop in response to this impaired postural feedback, growth asymmetry likely occurs. Growth is slower when increased pressure is exerted on the growth areas. Because more pressure is exerted on the concave growing areas than on the convex side, the convexity grows more quickly, leading to increasing curve size. This theory accounts for the observation that curves worsen most during the rapid adolescent growth spurt, which is the time when most of these curves are diagnosed. Muscles, discs, and bone appear to be normal in the young patient with idiopathic scoliosis.

The key to early detection of scoliosis is careful assessment of the entire trunk for asymmetry. Affected children should be examined with their back clearly exposed. The

examination should include evaluation of shoulder height, scapula position and prominence, waistline symmetry, and levelness of the pelvis (Table 432-2). Asymmetry in any of these areas may indicate a scoliosis (Fig. 432-25). In approximately 50% of children with uneven shoulder height, no spinal deformity is present on radiography. To define further whether a structural scoliosis is present, affected children should be examined bending forward (Fig. 432-26). The view from the caudal aspect allows ready detection of prominence of the thoracic ribs, whereas further bending or viewing from the head down is better for suspected lumbar curves. Both thoracic and lumbar regions should be checked. This “forward-bending” test is very sensitive in demonstrating the vertebral rotation that takes place in a structural scoliotic curve. However, it is not highly specific, and small degrees of rotation occur in patients without any significant scoliosis. A means of quantitating the trunk rotation is needed. The amount of rib hump can be measured by means of an inclinometer placed at the apex of the curve with an affected child bending forward. If the inclinometer measurement is 5 degrees or less, the scoliosis rarely is significant and radiographs usually are not needed. If the inclinometer reading exceeds 6 to 7 degrees, standing posteroanterior and lateral radiography is indicated for better assessment.

Examine in swimming suit or similar clothing to expose back. Observe asymmetry on trunk examination: shoulder height, scapular height, waistline equality, levelness of pelvis, leg-length difference, forward bending (both side and front-back). Measure rib prominence with inclinometer (optional). Assess skeletal maturity (e.g., age of menses onset). Obtain standing posteroanterior radiograph of the spine if asymmetry is seen. Measure using Cobb method. Recommend follow-up or treatment (none if the curve is less than 25 degrees and growth is complete). If growth is not complete and the curve is less than 25 degrees, obtain repeat radiographs in 4 to 15 months (see text). If scoliosis of more than 25 degrees is seen and growth is not complete, consider a brace. If scoliosis of more than 40 degrees is seen, consider surgery.	TABLE 432-2. Spinal deformity evaluation

	Figure 432-25. In examining for scoliosis, asymmetry of the trunk (shoulders, scapular height, waist area, pelvic height) should be noted carefully.



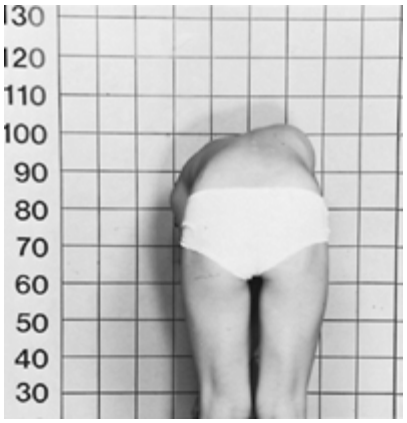


Figure 432-26. The forward-bending examination will detect even very small curvatures. The prominence is produced by chest-wall asymmetry, the result of vertebral-body rotation in the curved segment of the spine.

The magnitude of the scoliosis is measured radiographically by the Cobb method (Fig. 432-27). This measurement always should be performed on an erect posteroanterior spine radiograph. The error of measurement for this method is approximately 5 degrees. Because no active treatment is needed until the curve reaches 25 degrees, the time estimate for a follow-up radiograph, once the diagnosis has been made, is 25 minus the present curve magnitude. This result provides an estimate of the number of months that may pass until another radiograph is indicated. For example, if an affected child has a scoliosis of 15 degrees, waiting approximately 10 months before repeating the posteroanterior radiograph to check for progression is appropriate. This time estimate is based on the premise that during the adolescent growth spurt, annual curve progression is 5 to 10 degrees or approximately 1 degree per month.

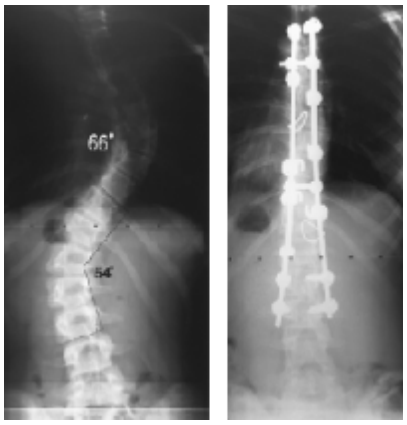


Figure 432-27. A standing posteroanterior radiograph of the spine is the correct film to use in quantitating the magnitude of scoliosis. **A:** The Cobb method of measurement is used routinely and is obtained as shown on this radiograph. **B:** The postoperative result after spinal correction and fusion in the same patient.

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Completion of growth or skeletal maturity can be assessed most accurately with bone age radiography of the hand and wrist. From the clinical standpoint, girls who have been menstruating for 2 years essentially have completed their spinal growth.

The treatment of scoliosis is based on three fundamental principles:

- Curves of more than 25 degrees are likely to increase if an affected child still is growing.
- Curves of 40 to 50 degrees are likely to increase even after growth is complete.
- Some degree of clinical pulmonary restriction may begin to be noticeable in thoracic curves of more than approximately 75 degrees.

If affected children are skeletally mature and have a curvature of less than 25 degrees, no further evaluation or treatment of scoliosis is needed. If the scoliosis is 25 degrees or more and such children still are growing, generally brace treatment is recommended and is successful in approximately 80% of the patients who actually wear the brace as prescribed (Fig. 432-28). Spinal exercises alone will not be successful in stopping curve progression. Once the brace treatment begins, it is continued until growth is complete. Usually, the brace is worn 18 to 23 hours daily. Physical activity is not limited by scoliosis, and affected children often can participate in sports activities while wearing their brace. Wearing a brace is considered successful if it prevents further progression rather than providing correction of the curve, as long-term follow-up studies have shown that the final size of the curve is virtually the same as before brace treatment begins. Despite some earlier controversy about the efficacy of bracing, prospective randomized studies published within the last few years have shown bracing to be an effective treatment method.

Figure 432-28. This child is wearing a low-profile brace currently recommended for most types of curves. It is

	worn under clothing and is not noticeable.

Although children and parents often are dismayed by our inability to straighten the spine nonoperatively, if curves can be kept at less than 35 to 40 degrees by the time growth is completed, most cases of scoliosis will not worsen in adult life. If the thoracic curve is greater than 50 degrees or the lumbar curve is greater than 40 degrees at the time growth is completed, usually progression will continue at a rate of approximately 1 degree annually, and often surgery will be recommended.

Surgical treatment is recommended for curves that are greater than 40 degrees, particularly in children who are not fully grown. Usually, the surgical treatment used consists of instrumentation of the curved area of the spine, combined with posterior spinal fusion of the instrumented area (see Fig. 432-27). Generally, correction of the scoliosis is at least 50%. Failure of fusion occurs in only approximately 1% of affected teenagers. Fusion is complete by 6 months after surgery, at which time such teenagers can return to almost all physical activities, except tackle football, wrestling, and gymnastics. Teenagers should be encouraged to return to activity, including physical education class in school, to deemphasize the psychological potential for disability after this surgery.

If the thoracic scoliosis exceeds 50 degrees, patients commonly have diminished vital capacity and residual lung volumes on pulmonary function testing. Arterial blood gas levels and forced expiratory volume in 1 second are normal except in children with severe curves. Vital capacity is decreased further if a thoracic lordosis is associated with the scoliosis. Even with surgical correction of the scoliosis, pulmonary function postoperatively will change little because of the persistence of chest wall or rib deformities that have occurred as a result of the scoliosis. Therefore, scoliosis should be prevented from progressing to this point if possible.

Pain is rare in adolescents who have idiopathic scoliosis. Although it may result from degenerative changes that are present by the time such patients are middle-aged, pain that occurs during

adolescence is an indication for further evaluation. Such patients should be questioned in

detail about the nature of the pain. If it is severe, limits activities, or requires frequent analgesics, a workup is indicated. If the neurologic examination is normal, a technetium Tc 99m bone scan should be performed to screen for discitis, stress fracture, osteoid osteoma, or other bone tumors. If spinal flexion is limited and a neurologic deficit is discovered, MRI or CT-myelography is necessary to rule out intraspinal pathology. Although all these conditions may cause scoliosis, the curvature will straighten as soon as its underlying cause is treated. Therefore, physicians should evaluate patients thoroughly for treatable causes of scoliosis before making a diagnosis of idiopathic scoliosis and instituting brace treatment or recommending spinal fusion.

Kyphosis. Normal spinal sagittal contours consist of lordosis in the cervical and lumbar spinal segments to balance the kyphosis that is present in the thoracic area. Sometimes, the term *kyphosis* is used to describe those abnormal conditions in which increased rounding of the back is present in the thoracic or thoracolumbar area. Usually, parents complain about a child's posture. Assessment of apparent excessive kyphosis should include a forward-bending examination, viewed from the side, to determine whether the back is flexible or rigid in the rounded segment (Fig. 432-29). The kyphosis may be discovered to be a rib prominence associated with a scoliosis. Similarly, mild to moderate scoliosis is seen commonly with moderate and marked kyphosis, so careful examination for both conditions is necessary.

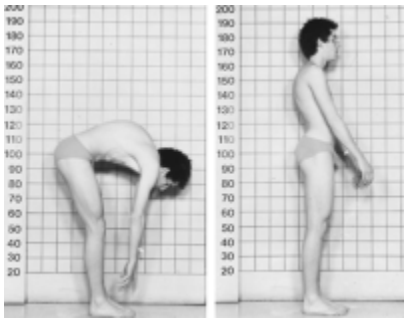


Figure 432-29. The examination for kyphosis also should include a forward-bending test to help to determine the rigidity and severity of the kyphosis, which may be hidden more easily in the upright position.

The least serious of these conditions is postural round back. This is seen most commonly in the preadolescent years. It occurs more often in children who are taller than their peers and in girls whose breasts have developed earlier than have their friends'. This condition is a flexible, increased kyphosis that can be straightened voluntarily by the child and can be corrected well with hyperextension positioning. This group of spinal deformities can be treated with exercises alone. Active hyperextension of the trunk and sit-ups to decrease lumbar lordosis are useful in improving trunk control. As long as no fixed deformity is established, as an affected teenager's body image improves, so will the rounding of the upper back.

Usually, a more fixed and less flexible thoracic or thoracolumbar kyphosis is called *Scheuermann disease*. This condition occurs most commonly in teenage boys. Attempts to

correct this kyphosis passively are unsuccessful, and often a large lumbar lordosis is associated with it. Lateral radiography of the spine will demonstrate irregularity of numerous disc spaces and anterior vertebral body wedging (Fig. 432-30). To establish the diagnosis

of Scheuermann kyphosis radiographically, at least 5 degrees of wedging in three adjacent vertebrae should be demonstrated. The Cobb method also is used to measure the amount of kyphosis present. Normally, the amount of kyphosis from T3 to T12 is between 20 and 45 degrees. If the kyphosis is present in the thoracolumbar area, which normally appears straight on lateral radiography, measurements greater than 10 degrees are abnormal.

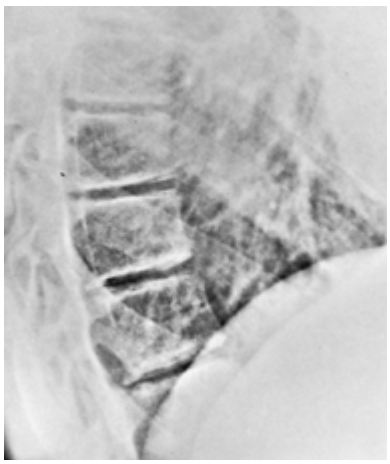


Figure 432-30. Lateral spine radiograph demonstrating the disc irregularities and anterior vertebral body wedging seen in Scheuermann disease.

If wedging is present, little correction can be achieved with thoracic spine hyperextension; if the lateral thoracic kyphosis is 55 to 60 degrees, bracing is indicated while the child is still growing. A Milwaukee brace, which uses a neck ring in addition to trunk pads, is the gold standard, but for kyphoses low in the thoracic spine, a more unobtrusive underarm brace may be used. Unlike scoliosis, in which little correction results from bracing, in kyphosis, approximately 50% improvement can be anticipated after 1 year of full-time brace wear. Once this degree of correction is obtained, generally nighttime brace wear is sufficient until growth is complete.

Increased thoracic kyphosis does not cause abnormalities in pulmonary function. The principal problem seen later in Scheuermann disease is pain in the low thoracic spine after an affected patient has been standing for some time. If the kyphosis exceeds 70 degrees by the time such a patient has stopped growing, spinal instrumentation and fusion, as with scoliosis, can provide excellent correction with a significant improvement in appearance.

Congenital kyphosis is less common than is congenital scoliosis but almost always requires early spinal fusion surgery. If the congenital kyphosis progresses unchecked, spinal cord compression at the apex of the kyphosis is common. As with congenital scoliosis, evaluation for associated genitourinary abnormalities should be performed.

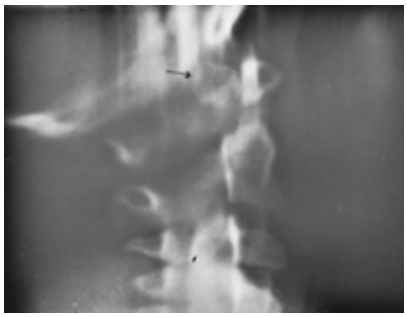
Cervical Spine

Evaluation of the cervical spine, because of its many normal variations on radiography, often is confusing. As seen on lateral cervical spine radiography, the anterior and superior corner of each vertebral body normally is the last part to ossify, sometimes giving the appearance of a small compression fracture. Full ossification and development of the odontoid process are not complete in young children and may give the appearance of being maldeveloped. The spine of children younger than age 10 is much more flexible than that of teenagers or older adults. As much as 3 mm of anterior movement of C2 on C3 with flexion is normal in this group, whereas no such movement should be present in adults. In fact, under experimental conditions, the newborn spine can stretch about 5 cm (2 in.) before it fails, whereas the adult spinal cord can stretch only 1.25 cm (0.5 in.) before it ruptures. Because of this difference in elasticity, infants who are involved in automobile accidents may sustain spinal cord injury without apparent spinal fracture. The proper use of car-seating supports for these very young children decreases the risk of these devastating injuries (see Chapter 91, Injury Prevention and Control).

Children with Down syndrome comprise a special group that commonly has instability of the atlantoaxial region. If this instability persists unrecognized, spinal cord compression with myelopathy may result, leading to leg weakness and lessened walking ability. Lateral cervical flexion-extension radiography should be performed at age 5 years in all children who have Down syndrome and are involved in activities involving forceful flexion of the neck or impact to the head. Although approximately 15% of such children will have some evidence of atlantoaxial instability, the majority do not need fusion surgery but can have periodic follow-up by neurologic examination. If the first radiograph reveals increased laxity (>5 mm distance between the odontoid and the atlas), films should be repeated every 2 years. If no laxity is seen, repeat radiography is not recommended as long as no signs of spasticity or symptoms of neck pain occur. Atlantoaxial posterior fusion is recommended if a neurologic deficit or excessive instability (>8 mm) is present.

Instability of the upper cervical spine may result also from os odontoideum or from odontoid hypoplasia. Os odontoideum occurs most commonly as the result of an early childhood fall that causes a fracture through the synchondrosis of the odontoid process. This unrecognized fracture develops into a fibrous nonunion (Fig. 432-31) that gradually becomes unstable over the ensuing months and years. Usually, the diagnosis is made when an affected patient is being evaluated for neck pain or other head or neck trauma. Neurologic symptoms rarely are present, but generally atlantoaxial fusion is indicated to stabilize this region and to protect the spinal cord from sudden, catastrophic injury that may result in death. After fusion is accomplished, such children will be able to participate in normal activities, although mild to moderate limitation of head rotation will be present. Odontoid dysplasia occurs periodically, but it is associated most often with genetic disorders, such as Morquio syndrome and spondyloepiphyseal dysplasia congenita. Generally, fusion is necessary.

Figure 432-31. An unsuspected injury to the synchondrosis of the odontoid in early childhood may lead to high cervical instability due to an os odontoideum. This child required atlantoaxial fusion and is now



asymptomatic.

Torticollis

Most commonly, torticollis is present at or near the time of birth and results from a contracture of one of the sternocleidomastoid muscles. An affected child's head will be tilted toward the side of the contracture, with the chin rotated away from the contracted side because the origin of the contracted muscle is on the mastoid process. The cause of torticollis is not well defined, but the incidence is higher in children with breech presentation and forceps delivery. Commonly, a fusiform, firm mass is palpable in the body of the contracted muscle. Often, affected children have plagiocephaly, or asymmetry of face and skull development. If the neck range of motion can be returned to normal by age 1 year, this facial asymmetry will disappear. If the torticollis is untreated until later in childhood, the eyes and ears never will become level.

Cervical spine radiography should be evaluated to ensure that the position of the head is not the result of congenital spine abnormalities, such as hemivertebrae. If the bony cervical spine is normal, stretching exercises should be instituted shortly after birth. These exercises are designed to stretch the contracted sternocleidomastoid muscle and should be taught to parents of

affected children by a knowledgeable physical therapist. Although one of the parents should be asked to do these stretching exercises at home, initial weekly checkups by the therapist or the physician can help to ensure compliance. If a significant contracture persists by the time such patients reach age 1 despite stretching exercises, surgical treatment to lengthen the sternocleidomastoid muscle is appropriate. Even after surgical release, some stretching and (at times) bracing will continue to be needed as the child grows.

Torticollis may present later in childhood after an upper respiratory infection or trauma. Torticollis that occurs after an upper respiratory infection is thought to result from retropharyngeal edema that leads to malposition at the atlantoaxial level, causing a rotatory deformity. Similarly, after muscular neck trauma, affected children may have a persistent torticollis for several days or weeks, secondary to an unsuspected rotatory subluxation at the atlantoaxial level. If torticollis from either of these causes persists, affected children should be treated with traction, followed by either bracing or atlantoaxial fusion. The likelihood that surgical fusion will be necessary increases with the duration of

symptoms, so prompt treatment is required.

Klippel-Feil Syndrome

Failure of normal vertebral segmentation in the cervical spine is known as *Klippel-Feil syndrome*, defined as congenital fusion of at least two cervical vertebrae (Fig. 432-32). In the milder forms, when only one or two levels are involved, diagnosis may be delayed until the teenage years and, even then, may be made only when the neck is examined radiographically for other reasons. In more severely involved children, however, the neck is very short, and webbing appears to be present. Often, Klippel-Feil syndrome is associated with Sprengel deformity, which is failure of normal descent of the scapulae. Associated genitourinary abnormalities may be present, and sonography or intravenous pyelography is indicated when the diagnosis of Klippel-Feil syndrome is made. Little specific treatment is available for this syndrome. Because of the congenital fusion of several segments of the cervical spine, instability may occur at the levels that move. If this instability is excessive or if neurologic deficits are present, fusing the unstable segment is necessary. Surgical fusion may be needed also in adult life for degenerative changes at the moveable segments.

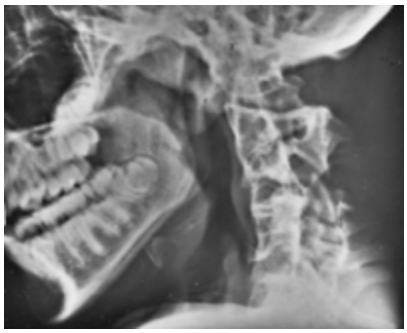


Figure 432-32. Klippel-Feil syndrome results from incomplete segmentation of the cervical vertebrae. Often, Sprengel deformity of the scapulae is associated with this syndrome.

Particularly in more involved cases, contact sports should be avoided, because any neck injury in a child with Klippel-Feil syndrome is more likely to be serious as a result of the limited flexibility of the cervical area.

Upper Extremity

Congenital and developmental abnormalities of the upper extremities of children are less common than those of the lower extremities, perhaps partly because of the greater stresses that are imposed on the lower extremity *in utero* and later during standing.

Obstetric (Brachial Plexus) Palsy

The brachial plexus is composed of contributions from nerve roots C5 to T1. Most severe injuries to the area involve lateral flexion of the neck or downward pressure on the

shoulder, such as may occur during a difficult delivery. Therefore, the upper portions of the plexus (C5-7) are stretched most commonly in a manner similar to that which causes the "burners" that occur during blocking maneuvers made in sports activities. This stretching causes denervation of the shoulder abductors and elbow flexors, which results in gradual joint contractures if it is not treated. These brachial plexus injuries are known as *Erb-Duchenne palsies*. The lower plexus (C7 to T1) can be affected by excessive abduction-traction and has a poorer prognosis; this is the rarest occurrence, and it is called *Klumpke palsy*. Such cases result in loss of function of the elbow extensors, wrist flexors, and finger muscles and possibly in a Horner syndrome. Occasionally, the entire plexus may be involved.

Factors associated with brachial plexus palsy include shoulder dystocia, breech position, high birth weight, and prolonged labor. The incidence is 1 to 3 in 1,000 births. The incidence and severity of this condition have declined gradually as obstetric care has improved. The site of injury may be at any level from the origin of the nerve roots to the plexus itself, but even root lesions may resolve spontaneously. On physical examination, the early typical Erb palsy appears as an arm that is rotated internally at the shoulder, extended at the elbow, and flexed at the fingers. Initially, passive range of motion should be full.

Such skeletal injuries as clavicle fractures and proximal humerus separations should be ruled out radiographically, although often they can be differentiated by guarding on testing of passive motion and the presence of Moro response. Because of the trauma, palsy and skeletal injury may coexist.

Treatment involves maintenance of motion and transfers of tendon in those rare, severe cases in which function does not return. With current obstetric practice, 92% of palsies resolve completely by the time affected infants reach 3 months of age, and 95% of such infants eventually recover fully. Physical therapy should be used initially to maintain range of motion. Splinting in most cases results in contractures, although later functional splinting of the hand may have a role. For patients with persistent weakness at age 3 months, electromyography and possibly myelography may help to identify those rare cases in which nerve grafting is required. Cases detected later may benefit from osteotomy or contracture release and tendon transfer, especially to restore shoulder external rotation.

Sprengel Deformity

Sprengel deformity, or congenital elevation of the scapula, actually represents embryonic failure of complete descent, rotation, and development of the scapula. Normally, this realignment occurs predominantly between the ninth and twelfth weeks of gestation. The cause is unknown.

On physical examination, the upper pole of the scapula may be visible in the base of the neck. Abduction is limited. The pectoralis major muscle may be underdeveloped. Scapular winging may occur as a result of serratus anterior muscle palsy. The

scapula may be connected to the vertebrae by an abnormal omovertebral bone, which is named for the two structures it connects. Such associated congenital anomalies as cervical

or thoracic vertebral fusions, anal abnormalities, or cardiac abnormalities may coexist. Treatment is indicated in moderate and severe cases to improve abduction and appearance. The most effective means of treatment involves detaching and lowering the midline origins of the rhomboid and trapezius muscles (Woodward procedure), combined with a clavicular osteotomy.

Congenital Pseudarthrosis of the Clavicle

Congenital pseudarthrosis of the clavicle is a defect in bone continuity presumably caused by pressure from the more cephalad position of the right subclavian artery. The ends of the bone are sclerotic and tapered, like an old fracture. Almost always, the disorder involves the right clavicle unless the patient has dextrocardia or a cervical rib. Bone grafting and pin fixation before age 6 may be indicated.

Radial Clubhand

Longitudinal failure of formation of many tissues on the radial side of the forearm and hand is known as *radial clubhand*. The severity of this condition varies. Approximately 50% of cases are bilateral. Associated abnormalities may include VATER (*v*ertebral defects, imperforate *a*nus, *t*rach *e*osophageal fistula, *r*adial and *r*enal dysplasia) syndrome, hydrocephalus, and clubfoot. The upper arm also may be short, and the shoulder girdle may be underdeveloped. The radial-sided muscles, radial carpal bones, thumb, and radial artery may be absent. The hand is deviated radially up to 90 degrees because it lacks its normal radial support, and the ulna may be bowed. Treatment involves centralization of the wrist on the ulna, transfer of tendon, and possible straightening of the ulna and creation of a thumb, as long as reasonable elbow flexion is present. Untreated cases are problematic cosmetically, although surprisingly they pose less functional difficulty. Congenital absence of the ulna is only one-third as common. In most cases, some remnant of the proximal ulna provides elbow stability.

Radioulnar Synostosis

Often inherited, radioulnar synostosis (fusion) results in a fixed position of forearm rotation, usually in pronation. Usually, diagnosis is delayed because affected children use other joints to compensate for the lack of forearm motion. At times, the synostosis may be only fibrous. Usually, shoulder and wrist motion can compensate for the lack of rotation. Motion to the forearm cannot be restored surgically. Rotational osteotomy to alter the position of the fused forearm should be performed only if clear-cut functional deficit can be demonstrated.

Congenital Constriction Bands (Streeter's Bands)

Congenital constriction bands most likely are the result of intrauterine encirclement by amniotic bands or the umbilical cord. They may be located anywhere, and they also may cause nerve palsies or the amputation of parts (Fig. 432-33). The bands can be released with Z-plasties after the patient reaches age 2 or urgently if they are associated with neurocirculatory compromise.

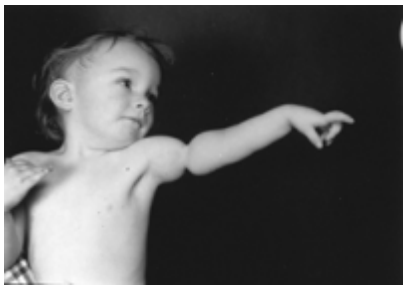


Figure 432-33. Child showing three sequelae of congenital constriction bands: deep ring around the left upper arm, paresis of the nerves to the tendons controlling the fingers, and amputation of the tips of the right fingers.

Polydactyly

Polydactyly, or the presence of an extra digit, varies in spectrum from a hypoplastic addition of soft tissue to a fully developed digit with all phalanges and metacarpals. Fifth-finger polydactyly is ten times more common in black than in white individuals. Therefore, a white child with this finding should be examined for other abnormalities, especially of the cardiovascular system. The simple, small, nonskeletal duplications can be excised or tied off. If significant skeletal stability is present, all the digits should be reexamined to determine which is the least functional, and this should be excised.

Congenital Trigger Thumb

Congenital trigger thumb presents as a clenched digit and is not always recognized at birth. Usually, it is the result of excessive tightness of the annular ligament that encircles the flexor tendon at the metacarpal head. This tightness causes swelling of the tendon, which later becomes firm. Treatment consists of 6 to 8 weeks of stretching if the condition is diagnosed early and surgical release if it persists or is diagnosed later.

Nursemaid's Elbow

Annular ligament entrapment, known as *nursemaid's elbow*, consists of elbow pain after longitudinal traction on a pronated, extended elbow in children between the ages of 2 and 7. A snap may or may not be heard. Usually, radiography shows no bony abnormality or displacement. Only one case report describes actual exploration of this pathology (Salter, 1971). This report and laboratory studies suggest that the annular ligament of the radial head slips partially over the radial head, the narrowest portion being prominent when pronated (Fig. 432-34). An elbow fracture or septic arthritis should be ruled out, especially if the mechanism of injury is a fall rather than traction. Usually, treatment is reduction by stabilizing the elbow with one hand, with a finger placed over the radial head for palpation, followed by gentle firm flexion until a click is felt. Affected children should begin using the elbow within minutes. Usually, immobilization is not carried out and is not necessary in first-time cases. It can be accomplished with 2 to 3 weeks in a cast if the episode recurs.

Parent education regarding the mechanism of this condition is most important.

Figure 432-34. “Pulled elbow” represents subluxation of the radial head through a partially torn annular ligament. Method of injury (*above*) and method of reduction (*below*).

Generalized Abnormalities

Bone Dysplasias

Osteocartilaginous Exostoses (Osteochondromas)

Osteochondromas, which can be single or multiple, are sessile or pedunculated bony masses that are located on the metaphysis, are directed away from the growth plate, and appear to move away from it over time (Fig. 432-35). These outgrowths have

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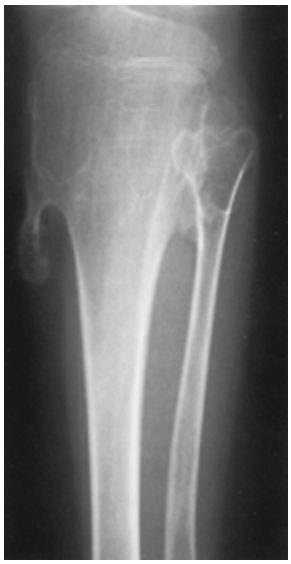
their own growth plates. Osteochondromas are thought to arise from defects in the perichondrial ring that encircles the growth plate, permitting lateral growth rather than the usual organized distal growth. Usually, the condition of multiple exostoses is distinct and is transmitted in an autosomal dominant manner, and affected persons frequently are somewhat short.

Figure 432-35. Osteocartilaginous exostoses are metaphyseal pedunculated or sessile lesions directed away from the growth plate.



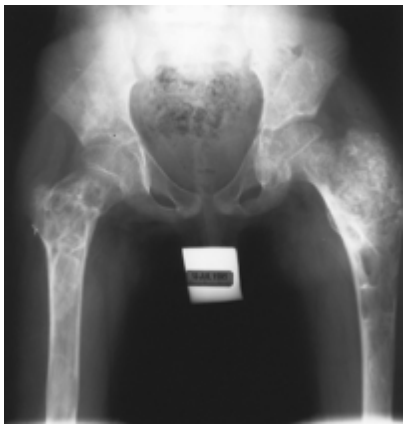
Any bone with endochondral growth may be affected, but the long bones of the extremities are involved most often. Because of asymmetric growth-plate activity, angular growth often ensues, resulting in outward angulation of the knees and ankles and in ulnar deviation of the forearm and wrist. These conditions should be corrected by partial epiphyseal stapling in young children or by osteotomy in older patients. Leg-length inequality is significant in 50% of patients.

The indication for excision of the lesions themselves is pain or compromise resulting from pressure on the tendons, nerves, or spinal cord. Malignant transformation should be suspected if continued growth occurs after skeletal maturity is reached or if new onset of pain occurs. A bone scan may be helpful because absence of uptake indicates a benign lesion; however, increased uptake does not always mean malignant change.

Fibrous Dysplasia

Fibrous dysplasia is a disorder exhibiting altered bone formation in the medulla and cortex and much fibrous tissue in the marrow. Radiographically, the bone has a uniform “ground glass” consistency and the cortex is thin and often deformed. One bone (in the monostotic form) or several bones (in the polyostotic form) can be affected. Pathologic fractures occur often, but usually heal in a normal period. Management of proximal femoral (“shepherd's crook”) bowing (Fig. 432-36) is the most difficult. Usually, deformities and fractures of the lower extremities require internal fixation, whereas those in the upper extremities require casting.

Figure 432-36. Polyostotic fibrous dysplasia produces loss of the normal trabecular pattern and thinning of the cortex and often involves the proximal femur.



Irregular café au lait spots occur in 30% of patients with the polyostotic form of fibrous dysplasia. When polyostotic lesions and café au lait spots are associated with precocious puberty, the condition is called *Albright syndrome*. Other types of endocrinopathies (thyroid, parathyroid, or adrenal problems) may occur. Malignant transformation to fibrosarcoma or osteosarcoma is rare.

Osteogenesis Imperfecta

Osteogenesis imperfecta encompasses a spectrum of diseases that are the end result of defects in collagen or proteoglycan synthesis. These diseases result in bones that have thin cortices and multiple fractures. Short stature, blue sclerae, middle-ear deafness, abnormal dentition, and thin skin may coexist. Usually, inheritance is dominant, occasionally is recessive, but frequently is the result of spontaneous mutation. Tiny fractures occur to cause bowing of long bones and scoliosis. Child abuse should be considered in the differential diagnosis, and the absence of pelvic deformities or wormian cranial bones in children who are subjected to abuse may be helpful.

Aids to mobility and preventive bracing can be very helpful in preventing fractures. Occasionally, intramedullary rods that elongate with growth are needed. Fortunately, the frequency of fractures diminishes with age.

Tumors

A complete discussion of musculoskeletal tumors is beyond the scope of this section; instead, an attempt is made to describe an appropriate differential diagnosis and evaluation.

Benign or malignant musculoskeletal tumors can be classified according to their tissue of origin (Table 432-3). The history and physical examination rarely are definitive. Many tumors become evident after trauma, when a new prominence is noted or when pathologic fracture occurs through weakened bone. For example, osteoid osteoma, a benign condition, frequently produces pain that is relieved by nonsteroidal antiinflammatory agents. Very early sarcoma may be painless. Unexpected presentations may occur, such as Ewing

sarcoma, various types of histiocytoses, and leukemia, each of which may present with fever and malaise.

Origin	Benign	Malignant
Cartilage	Chondroblastoma Enchondroma Chondromyxoid fibroma Osteochondroma	Chondrosarcoma*
Bone	Osteoid osteoma Osteoblastoma	Osteosarcoma
Marrow elements	Lipoma	Ewing sarcoma Reticulum cell sarcoma Liposarcoma* Plasma cell myeloma
Fibrous connective tissue	Desmoplastic fibroma Fibrous cortical defect	Fibrosarcoma
Skeletal muscle		Rhabdomyosarcoma
Neurogenous tissue	Neurilemma Neurofibroma	Neuroblastoma
Unclear	Giant cell tumor*	Adamantinoma

*Rarely occurs in children.

TABLE 432-3. Musculoskeletal neoplasms

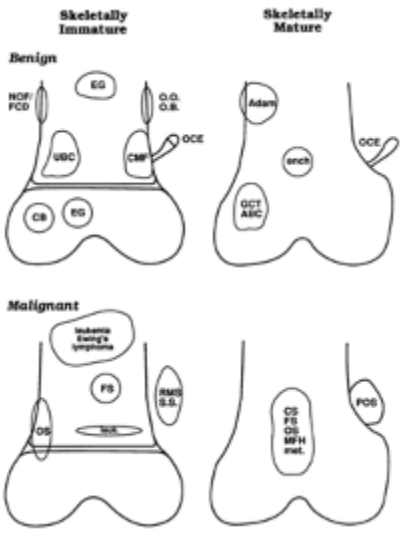
Some idea of the benign or malignant nature of a tumor can be gained from the following radiographic features. Lesions associated with rapid spread and lack of local containment should heighten the suspicion of malignancy. A vague zone of transition between the lesion and normal bone is worrisome, as is a soft tissue mass in the presence of a bone tumor. Periosteal lamellar change is a response to spread outside the cortex and may occur with benign or malignant tumors. Rapid growth is suggested when periosteal lamellation is extensive and no formation of definite new cortex occurs. Thinning of the cortex itself is not pathognomonic of malignancy; this condition occurs also with fibrous cortical defects and unicameral cysts. Internal stippling suggests calcification of a cartilage matrix; usually, fluffy opacification represents new bone formation, as in osteosarcoma. Usually, lesions crossing the epiphyseal plate are infections or malignant tumors. Leukemia presents with musculoskeletal complaints 20% of the time, and radiographic findings include osteopenia, sclerotic or lytic lesions, lucent metaphyseal bands, or periosteal new bone.

Certain general radiographic studies can be helpful. Radiographic studies must be tailored to the differential diagnosis. CT may show internal consistency, soft tissue spread, and extent of the lesion. Technetium Tc 99m bone scans reveal lesions in the remainder of the skeleton, bony involvement with soft tissue lesions, and bone turnover or activity of questionable lesions. Angiograms may be helpful to determine whether the tumor involves a vascular bundle. MRI is helpful in assessing soft tissue involvement.

The location of a lesion is meaningful, and a diagram of the location of common bone lesions is presented in Figure 432-37.

Generally, laboratory studies are not specific; the sedimentation rate and complete blood count are abnormal in several of the aforementioned tumors, and often the alkaline phosphatase level is elevated in patients with osteogenic sarcoma.

Figure 432-37. Location of tumors in immature and



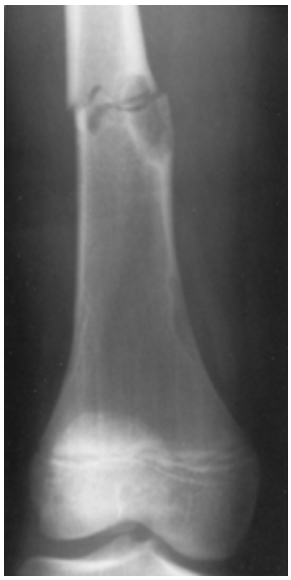
mature skeletons. Adam, adamantinoma; ABC, aneurysmal bone cyst; CB, chondroblastoma; CMF, chondromyxoid fibroma; CS, chondrosarcoma; EG, eosinophilic granuloma; ench, enchondroma; FCD, fibrous cortical defect; FS, fibrosarcoma; GCT, giant cell tumor; leuk., leukemia; met., metastasis; MFH, malignant fibrous histiocytoma; NOF, nonossifying fibroma; O.B., osteoblastoma; OCE, osteocartilaginous exostosis; O.O., osteoid osteoma; OS, osteogenic sarcoma; P, parosteal; RMS, rhabdomyosarcoma; S.S., synovial sarcoma; UBC, unicameral bone cyst.

The treatment of musculoskeletal tumors defies simplification. The most important generalization is that any patient requiring surgery should be under the care of a surgeon who has had experience in this area. Errors related to biopsy placement or specimen adequacy are three to five times more frequent in centers where the surgeons do not specialize in tumor treatment.

Unicameral Bone Cyst

Two common, benign bone tumors deserve brief mention. A unicameral bone cyst is a smooth, well-marginated lucency that is located fairly centrally in the metaphysis of children between ages 2 and 15 (Fig. 432-38). Usually, it is not recognized until a fracture occurs through the cyst. The fracture should be allowed to heal, and the lesion can be observed if it is small and is located in a bone that does not bear weight; otherwise, it can be injected with steroids or bone-inducing substance. The latter two treatments produce results equal to or better than open bone grafting. The natural history of these defects is spontaneous regression during late adolescence.

Figure 432-38. Fibrous cortical defects with pathologic fracture. Note two lesions on the medial side, within one wall of the cortex and expanding it.



Fibrous Cortical Defects

Fibrous cortical defects are well-marginated lucencies located in (and occasionally slightly expanding) the cortex. Usually, one radiographic view can show that these lesions are not central in bone (Fig. 432-39). They are present in as many as one-third of all young children at some time and disappear with age. In a weight-bearing bone, the risk of fracture is appreciable if the lesion is greater than approximately 3 cm in length and more than one-half the width of the bone. Lesions this large should be protected by limiting activities if possible or by performing bone grafting.

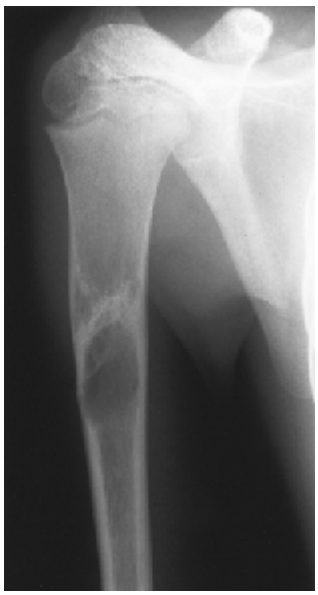


Figure 432-39. Unicameral bone cyst in the proximal humerus of a 9-year-old. Often, it is located closer to the growth plate.

Neuromuscular Disorders: Cerebral Palsy

Cerebral palsy is a collective term for a group of nonprogressive affections of the upper central nervous system. Two types of musculoskeletal problems result: disorders of control, for which little can be done, and bone and joint deformities resulting from continued muscle imbalance, which can be treated. Modifying the athetoid features that predominate in a few children is difficult, except with supportive bracing, but the more common spastic features are more amenable to modification. Discussion of such patients always should include identification of current functional problems and goals. Gait, if present, may be marked by a crouched position as a result of knee or hip flexion contractures. The ankle may tend toward plantar flexion or dorsiflexion.

Trial bracing or gait studies may help to determine which is the primary problem. Often, ankle plantar flexion can be controlled with bracing if the foot can be brought up to a right angle with the tibia when the knee is extended. If this is not possible, the tight heel cord should be lengthened; tight hamstring and hip flexors also may be lengthened when indicated. The “scissoring” of the legs seen with walking or lying down may be caused by tight adductor muscles.

Hip dislocation occurs with increasing frequency as the severity of disease involvement increases. It is the result of imbalance between the strong adductors and flexors and the weak abductors and extensors. The imbalance is acquired rather than congenital and usually occurs after affected patients reach several years of age. It should be checked every 6 to 12 months in diplegic or quadriplegic patients. Affected children are at risk for progressive hip subluxation if abduction is less than 30 degrees with the hip extended. Dislocation and subluxation cause difficulty with perineal care, balanced sitting, degenerative joint disease, pain, and increased spasm. Consequently, they should be treated aggressively. Even in patients

with severe involvement, they can be prevented by early muscle release or later osteotomy.

Scoliosis also is encountered more frequently with increasing severity of cerebral palsy. It is present in as many as 69% of severely affected children because of persistent primitive reflexes, an inclined pelvis, or asymmetric muscle tone. A brace may be prescribed if affected children have difficulty in sitting, but it is less effective in preventing worsening of the curve than it is in idiopathic scoliosis. Surgery may be necessary.

The upper extremity may be flexed at the elbow, wrist, and fingers. The decision to correct this condition is based on patients' intelligence, ability to control the hand voluntarily, and degree of sensation. The thumb may be clenched, and early bracing may be helpful, with surgery performed later if the digit has potential for use.

The benefits of physical therapy in general are debated. Positioning and hand and heel-cord stretching may produce increased range of motion. However, the severity of involvement probably is more important than is therapy in determining patients' ability to walk.

Myelodysplasia or spinal dysraphism involves malformation of the neural tube with paralysis below a certain thoracic or lumbar level of innervation. Usually, the functioning

muscles allow more control than is possible in cerebral palsy patients. The goal of orthopedic treatment is optimizing mobility and socialization, which does not always mean enabling the patient to walk. The quadriceps are the most important muscles for mobility. Severely affected children with poor intelligence and weak quadriceps muscles are more mobile in wheelchairs. In some cases, joint deformities are treated by surgical release and bracing. In contrast to cerebral palsy, usually hip dislocations are not painful and should not be reduced unless they are unilateral or affected children have good quadriceps muscles. Scoliosis also may occur, especially with higher-level spinal defects. One of the most important roles of the primary physician is to observe affected children for loss of lower extremity muscle power as they grow. This loss may be caused by tethering of the cord distally as such children grow or by a disturbance of cerebrospinal fluid pressure.

Infections

Hematogenous Osteomyelitis

The incidence and presentation of hematogenous osteomyelitis are changing after the introduction of newer imaging and treatment methods, but certain principles remain constant. The summary presented here should be coupled with that provided in Chapter 70, Osteomyelitis and Septic Arthritis, to illustrate the spectrum of treatment philosophies.

By definition, acute hematogenous osteomyelitis includes processes that have been operating for a week or less at the time of diagnosis. After infancy, this condition occurs more frequently in boys than in girls, presumably because trauma plays a role in increasing susceptibility. The peak ages of occurrence are infancy (younger than 1 year) and preadolescence (9 to 11 years). The incidence declines in adulthood because of the change in vascular supply of bone. The sites affected most commonly are the femur and tibia, each of which accounts for one-third of all cases, followed by the humerus, calcaneus, and pelvis. Any bone may be affected, however. The metaphysis is the region involved most often, and spread may occur from this point to involve any other portion. Rarely, the process may begin in the epiphysis.

Its pathophysiology explains some features of this disease. The metaphyseal vascular channels form loops near the growth plate. Blood flow is slowed, and the capillary basement membrane and reticuloendothelial system are deficient in these regions. Experimental bacteremias have been shown to produce foci of infection only in these areas. Trauma likely plays more than a circumstantial role, as experimentally traumatized areas are more susceptible to the development of osteomyelitis. Only approximately one-fourth of all cases have a demonstrable source, such as cutaneous, aural, or respiratory seeding. Direct traumatic inoculation is a different disease process.

After a focus of infection is initiated, local inflammation is followed by spread up and down the medullary canal. The growth plate in children has no bridging vessels and acts as a barrier to spread in most cases. The germinal cells are on the epiphyseal side and, therefore, are spared. In the first year of life, however, the transphyseal vessels that exist allow spread to proceed up to the epiphysis and into the joint. These facts have two implications. First, growth-plate damage is more likely during the first year of life. Second, in children of this age, septic arthritis may follow osteomyelitis in any metaphyseal location,

whereas in older children without transphyseal vessels, it occurs only in locations where the joint capsule extends over the growth plate (i.e., the shoulder, elbow, and hip). At skeletal maturity with growth-plate closure, this barrier again is eliminated, although hematogenous osteomyelitis is rare after this point. As intramedullary pressure increases, pus dissects through the haversian system, elevating the periosteum and producing a subperiosteal, then soft tissue, abscess. The elevated periosteum may be radiographically apparent within 1 to 2 weeks.

Unlike septic arthritis, the organisms involved in hematogenous osteomyelitis vary slightly with the age of affected patients (Fig. 432-40). In all age groups, the predominant organism is *S. aureus*, although *Streptococcus pneumoniae* and

Haemophilus species must be considered. *S. aureus* infection is associated with a higher recurrence rate than other organisms. *Salmonella* should be considered in patients with sickle cell anemia, although *S. aureus* infection still is more common in these patients. Blood culture results during the acute phase are positive approximately 40% to 50% of the time, and direct cultures of pus or bone are positive only 60% to 80% of the time, which may be the result of prior antibiotic use, errors in sampling or processing, or autoeradication of the organism.

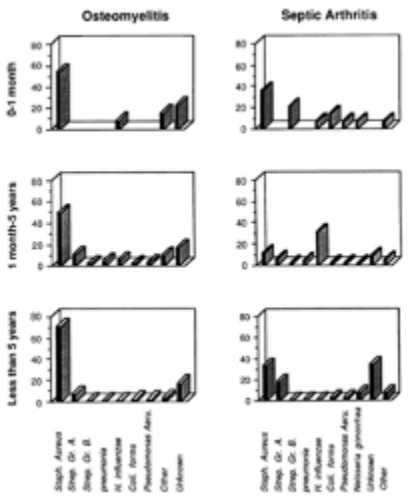


Figure 432-40. Frequency of occurrence of organisms involved in acute hematogenous osteomyelitis and septic arthritis in three age groups. (Drawn from table with permission from Jackson MA. Management of the bone and joint infections. *Pediatr Orthop* 1982;2:315.)

Clinical diagnosis remains key despite the availability of new imaging techniques. Affected children may appear well or may have systemic involvement ranging from malaise to shock. Often, refusal to bear weight is an early symptom. The very earliest sign is fever and local bone tenderness, followed later by a fluctuant mass if a subperiosteal or soft tissue abscess has developed. Spread to adjacent joints should be ruled out by palpation and range of motion evaluation. Usually, passive motion of the extremity is not resisted significantly unless a soft tissue abscess or joint involvement is present. Increased suspicion should be aroused with neonates, who more often are afebrile and first may be noted to have a swollen or motionless limb. Vertebral or pelvic osteomyelitis may present as abdominal pain and can resemble the more common septic arthritis of the hip.

The differential diagnosis primarily includes neoplasm, contusion, nondisplaced fracture, and sickle cell crisis. Elevated WBC counts and sedimentation rates are helpful but not diagnostic. Serum antibody titers may be helpful, but their sensitivity is a problem. Radiography at the earliest stage may show soft tissue swelling. Osteopenia or lysis may appear after 7 to 10 days, followed by new bone formation at the borders of the process. Bone scanning has been used widely in the last two decades, but the subtleties of its use have been recognized only recently. The tracer that is used most widely is technetium Tc 99m methylene diphosphonate because of its speed, cost, and sensitivity (Fig. 432-41). Immediate scans for flow and blood pool should be obtained, as should later skeletal images. Results of the scan may be normal in the very early stages but the procedure should be repeated after 48 hours if clinically indicated.

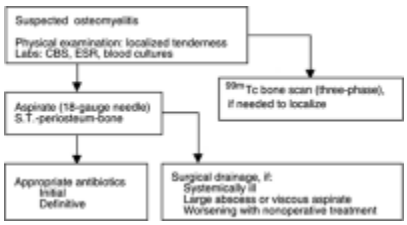


Figure 432-41. Diagnosis and treatment algorithm for hematogenous osteomyelitis. CBS, conjugated bile salts; ESR, erythrocyte sedimentation rate; S.T., soft tissue.

Cold or photopenic areas are important because they may indicate avascular sites, especially when they are accompanied by adjacent areas of increased uptake. Cellulitis may cause confusion but usually does not show bony localization on delayed images. The overall accuracy of nuclear imaging is approximately 60% to 90%. It may be much lower in neonates, however, according to some reports. Gallium citrate may be sensitive, but it requires a minimum of 24 hours; indium-labeled WBC studies require similar amounts of time, including preparation of the tracer. Because of the aforementioned limitations, radionuclide scans are not reliable in some instances, especially when the clinical diagnosis is clear. These studies have their greatest value when localization for aspiration is difficult. The role of MRI has yet to be defined.

Aspiration is indicated in all cases to identify the pathogen and, in some cases, to decompress localized purulence. It should be performed with a large-diameter needle. The anesthetic may be local, intravenous, or general, as indicated. In sequence, the extraosseous soft tissues, periosteum, and (if necessary) intramedullary canal should be assessed for purulent localization. Fluoroscopy may be useful in deep lesions if radiographic changes are evident. Experiments in animals have shown that aspiration of bone alone does not cause a bone scan result to become positive.

Treatment involves the delivery of an appropriate antibiotic to all infected tissue. Therefore, avascular abscesses may require surgical decompression if aspiration cannot accomplish

drainage. Antibiotic therapy can be divided into initial and definitive periods. In the initial phase, broad-spectrum antibiotics, including such antistaphylococcal agents as nafcillin or oxacillin (150 to 200 mg/kg/day), are indicated. Vancomycin should be used if resistance is suspected. In neonates, an aminoglycoside should be added. In children who are younger than 3 years and have osteomyelitis associated with septic arthritis, chloramphenicol or cefuroxime may be used to cover *Haemophilus influenzae*. In the definitive period, the most effective, least toxic antibiotic effective against the isolated organism should be given for 4 to 6 weeks. It may be administered by the oral route if affected patients are clinically improved and compliant and if adequate blood levels can be documented.

Surgery is reserved for cases in which affected children are systemically ill or worsening under medical treatment or in which an abscess has been demonstrated. An abscess or avascular tissue should be removed to allow antibiotic penetration, and usually the wound is closed over a drain. Complications include recurrence (10% overall; 6% at 6 months), minor growth acceleration, growth-plate damage, and fracture through weakened bone.

Subacute Osteomyelitis

Subacute osteomyelitis is a more subtle condition. No systemic signs may be evident and, in Roberts' series, fewer than one-fifth of all patients had a fever, an elevated WBC count, or a positive blood count result. Abnormal radiographic and bone scan results, however, were more common than in the acute form. Treatment follows the principles discussed.

Chronic Recurrent Multifocal Osteomyelitis

Chronic recurrent multifocal osteomyelitis is a rare syndrome involving low-grade systemic manifestations that are ongoing for several years, with reports of up to 12 areas of lytic-sclerotic juxtaepiphyseal involvement. No organism has been isolated, and treatment is supportive.

Fungal Osteomyelitis

Fungal osteomyelitis may be disseminated (sporotrichosis, candidiasis) or direct (eumycetoma). Aggressive débridement in these conditions is more important than that in bacterial infections.

Puncture Wounds

Puncture wounds to the foot are significant in that they may involve *Pseudomonas* infection, which occurs with colonization of a sock or a sneaker by this organism. The wound should be inspected, and foreign material should be removed. If the bone or joint is contaminated, débridement and antibiotic therapy for recovered organisms should be begun. Otherwise, affected

patients should be seen in 3 to 5 days or at least should be instructed to return if symptoms of infection occur.

Septic Arthritis

Slightly more common than hematogenous osteomyelitis, septic arthritis may have more disastrous long-term consequences if effective treatment is delayed. Most cases occur in infants and younger children, with nearly one-half of all affected patients being younger than age 3. A high index of suspicion for septic arthritis should be maintained in sick neonatal patients, for they show few signs. The hip is the joint involved most commonly in infants, as compared to the knee in older children. The spread may be from the bloodstream or from an adjacent osteomyelitis, especially in the hip and shoulder where the capsular insertion extends over the growth plate onto the metaphysis. Many theories, including alteration of joint fluid by toxins from both the neutrophils and the bacteria, have been advanced to explain the pathogenesis of joint destruction.

The spectrum of causative organisms in septic arthritis is somewhat broader than that of hematogenous osteomyelitis (see Fig. 432-40), which may be related to the greater frequency of this condition. Overall, *S. aureus* remains the most common causative organism. In patients between ages 1 month and 5 years, however, *H. influenzae* also is common. The use of the vaccine against this organism appears to have reduced, although not eliminated, this organism as a cause of septic arthritis in the toddler. The streptococci, *Escherichia coli*, *Proteus*, and other organisms also should be considered. The yield of organisms from aspiration is approximately 60% to 80%.

Clinical findings vary with the age of affected patients. In infants, fever, failure to feed, and tachycardia may be present. Subtle changes in position, unilateral swelling of an extremity or a joint, asymmetry of soft tissue folds, and pain with range of motion may serve as clues. In older children, the signs are more localized.

Aspiration with a large needle should be performed if any reasonable suspicion of septic arthritis exists, both for diagnosis and, in some cases, for treatment. In such deep joints as the hip, injection of radiopaque dye should be used to confirm the position of the needle, especially if the aspirated fluid is normal. This ensures that joint fluid actually was obtained, and it also helps to distinguish joint infection from septic involvement of the bursa underneath the nearby psoas muscle. Usually, the WBC count in fluid obtained from patients with septic arthritis ranges from 50,000 to 250,000, with 95% polymorphonuclear leukocytes. Elevated lactate levels may be helpful in cases in which WBC counts are borderline.

The differential diagnosis includes toxic synovitis of the hip, in which pain, fever, leukocytosis, and spasm are more moderate and do not escalate on serial observations. However, at times the two conditions are indistinguishable, and aspiration should be performed. Lyme arthritis, rheumatoid arthritis, cellulitis, traumatic synovitis, and the migratory multiple arthralgias of rheumatic fever should be considered. A sympathetic effusion also may occur from adjacent osteomyelitis.

The role of arthrotomy versus aspiration in confirmed cases of septic arthritis is controversial. The key feature is removal of deleterious enzymes and restoration of effective synovial perfusion. Because the decision not to operate requires the ability to monitor and aspirate repeatedly as needed, the use of arthrotomy probably is preferable in

deep joints that are difficult to assess, such as the hip and shoulder; in young patients in whom examination is difficult; and when the fluid obtained is viscous or has a very high WBC count.

The surgical procedure should include irrigation, drainage, and closure, which may be performed arthroscopically in the knee, shoulder, and ankle. Direct instillation of antibiotics has no benefit. Some investigators feel that the femoral metaphysis should be drilled whenever the hip is aspirated, to decompress any possible femoral osteomyelitis.

Early effective treatment is very important. The chance of achieving good results declines dramatically if treatment is initiated after the symptoms have been present for 4 days. Antibiotics should be continued for 4 to 6 weeks. Whether the joint should be immobilized or treated with continuous passive motion is controversial; however, the latter modality is practiced less commonly. Contractures should be prevented, and abduction of the hip decreases the likelihood of dislocation. Complications include permanent destruction of cartilage and, in the hip, avascular necrosis with resorption or overgrowth of the femoral head. Complications are more frequent in young infants.

Gonococcal arthritis also occurs in children. Usually, it becomes evident after the systemic and febrile phase of the illness and should be distinguished from the more common gonococcal migratory multiple arthralgia or tenosynovitis. An average of two to three joints, most commonly the wrists and knees, are affected. Treatment is aspiration and closed irrigation followed by 3 days of intravenous penicillin and 4 days of ampicillin or amoxicillin. Oral treatment alone with one of these drugs for 7 days is acceptable in compliant patients after a loading dose has been given.

Injuries

A comprehensive discussion of musculoskeletal trauma is beyond the scope of this chapter. The reader is referred to the works by Rockwood and by Rang for further information. Basic principles of injury evaluation and common injuries and emergencies are discussed here.

Children's bones differ from those of adults both biomechanically and physiologically. Mechanically, immature bone is more porous, and the pores serve to limit crack propagation. Instead of complete fractures, children often have involvement of only part of the cortex, such as in a buckle fracture from compression or a greenstick fracture from tension. The most extreme example is plastic deformation of bone without fracture. This deformity should be corrected if it is 20 degrees or greater.

Another biomechanical feature of children's skeletons is that the ligaments are stronger than is either the bone or the growth plate. Injuries that would produce dislocations or sprains in adults (e.g., elbow dislocation or medial collateral ligament tear of the knee, respectively) produce different patterns in children (i.e., supracondylar humeral fractures or femoral physeal separations, respectively; Fig. 432-42). Thus, the presence of nondisplaced fractures and separations should be sought on physical examination and radiography in children. In the knee, gentle stress radiography may show a nondisplaced separation, which should be immobilized.

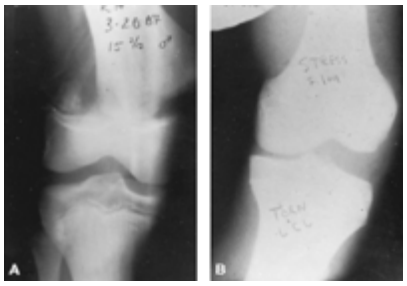


Figure 432-42. A valgus stress to the knee produces failure at the weakest spot. **A:** Growth plate in a child. **B:** The medial collateral ligament in an adult.

Physiologic differences include union rates, remodeling, overgrowth, and growth-plate injuries. Nonunion is nearly nonexistent in children, occurring only in open fractures with extensive soft tissue loss and periosteal stripping. Bone union times range from 3 weeks in infants to 3 months in adolescents. Remodeling of angulation and displacement is an impressive tendency until the early teenage years. It occurs through alterations in physeal growth and through local periosteal resorption on the convex side and deposition on the concave side. It is most effective in the metaphysis, where angulation does not create as much deformity as in the midshaft.

Compensation for any residual deformity is much better if it is in the plane of joint motion. For example, posterior angulation of a distal femur fracture can be compensated by knee flexion, whereas the knee has no ability to compensate outward (varus) angulation at this site. In the upper extremity, overlap of fracture segments is acceptable as long as the angulation is not excessive.

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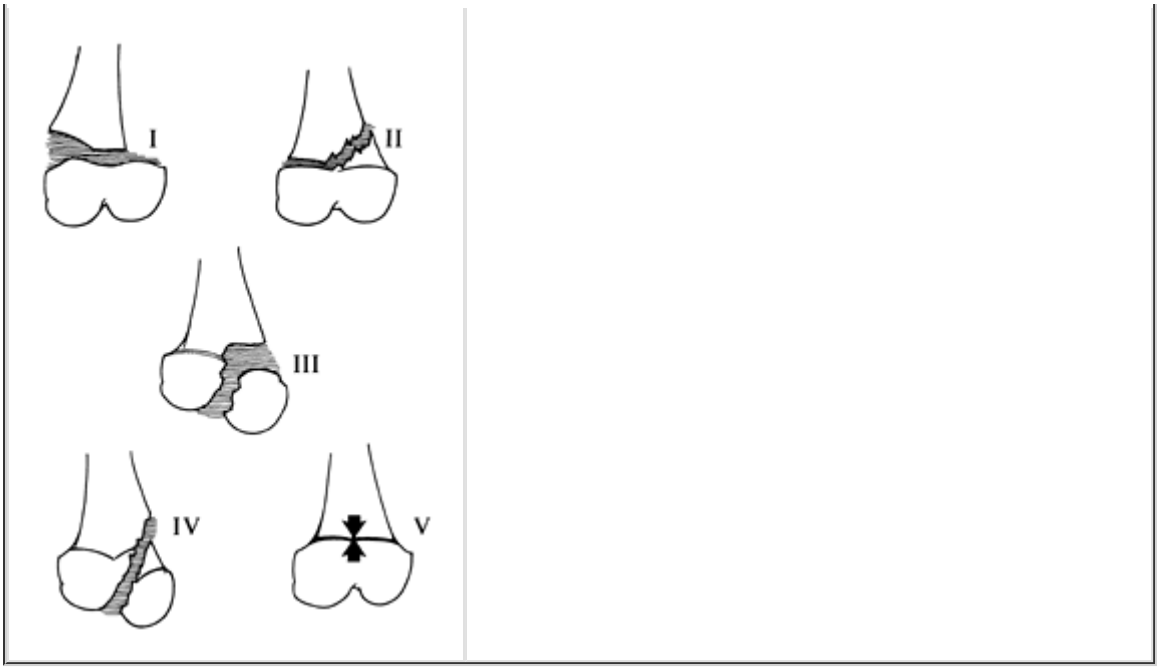
Another physiologic feature of children's fractures is growth stimulation, which occurs because of hyperemia and continues for approximately 18 months after the fracture occurs. It is most significant in the femur, where it averages 1 cm, but occasionally it becomes significant in the tibia. In contrast, growth arrest may occur if the growth plate is crushed or crossed by the fracture. The Salter-Harris classification (Fig. 432-43) was developed to predict the risk for growth arrest. It is helpful also in communicating with consulting physicians. Because types I and II do not cross the germinal layers, the risk of growth-plate damage with these injuries is minimal. In types III and IV, the fracture crosses the plate, but anatomic reduction may diminish the risk of plate closure. In type V, the crushing cannot be reversed. Combinations of types may occur, especially with irregular growth plates. Such injuries should be observed carefully because, if a limited growth-plate bar forms, it can be resected to restore normal growth.

Figure 432-43. Salter-Harris classification of fractures involving the growth plate.



Basic evaluation of specific injuries should include three features: (a) assessment of the entire limb to rule out other fractures or dislocation above or below the joint; (b) assessment of neurovascular status, including sensation and motor function and pulses, capillary refill, and temperature (the possibility of compartment syndrometissue pressure greater than capillary perfusion pressure—should be considered in the forearm or leg); and (c) assessment of the site of injury. A surprising amount can be learned regarding skeletal structures (e.g., growth plates, ligaments) by palpating carefully in an attempt to identify the injured area. For example, lateral ankle swelling may be caused by fibular growth-plate injury, ligament sprain, or peroneal tendon subluxation, and all these conditions can be differentiated by palpation. Typically, ankle sprains have tenderness around the lateral ligaments of the ankle. A mild sprain has no bony tenderness, no laxity on forward pull of the foot, and no medial tenderness. Patients with mild ankle sprain may be treated with ice, Ace wrap, and range-of-motion exercises as tolerated.

Hand Injuries

Injuries to the hand are common in children. Fractures of the phalangeal and metacarpal shafts can be splinted if they are minimally angled and stable, but rotational alignment should be checked by observing the fingernails with the fingers flexed and extended. They should be aligned similarly if no malrotation is present. “Buddy taping” helps to minimize malrotation. Growth-plate or epiphyseal fractures may be splinted if they are nondisplaced, but should be referred if they are displaced. Dorsally dislocated interphalangeal joints may be reduced and viewed radiographically to rule out fracture. If they are stable enough to allow active range of motion, they should be splinted for 3 weeks with an aluminum splint. Immobilization of the metacarpophalangeal joints should be in 50 to 90 degrees of flexion to minimize stiffness, and the interphalangeal joints should be in mild flexion of approximately 20 degrees.

One problematic injury is an avulsion of the base of the nail bed, often associated with open separation of the nearby phalangeal growth plate, which may become infected and stop growing. This condition should be distinguished from a Kirner deformity, which is a bilateral, idiopathic irregularity of the distal phalangeal growth plate.

Scaphoid Fractures

The scaphoid bone ossifies when a child is 6 years old. Scaphoid fractures may occur in children and develop nonunion if they are not recognized. They occur less often in children than in adults, however. Laceration of the palm and digits may sever a flexor tendon, and active range of motion of each joint should be checked to ensure that this has not occurred.

Forearm Fractures

Forearm fractures are very common in children. Nondisplaced buckle fractures of one or both bones should be treated with a short arm cast for 3 or 4 weeks. Greenstick fractures represent tension or rotational failures with less intrinsic stability, and they should be held in a long arm cast for 6 weeks. Completion of the greenstick fracture is not necessary as long as angulation can be controlled. With any forearm fracture, the wrist and elbow joints should be checked because dislocation may occur at one of these locations to compensate for fracture malalignment (Fig. 432-44A).

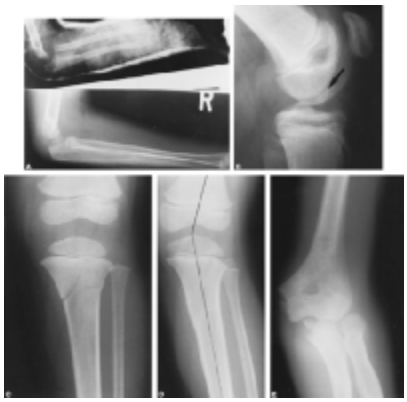


Figure 432-44. Avoiding four common pitfalls in evaluating musculoskeletal trauma. **A:** Always check the joint above and below a fracture. This Monteggia fracture of the ulna is reduced, but the radial head fracture was missed. The radial head should line up with the capitellum, as it does in the elbow on the right. **B:** In a child's swollen knee, check the lateral film for a tibial spine avulsion fracture. It may be obscured partially by the overlying femoral condyles. **C,D:** A proximal tibial fracture may result in valgus deformity even when initially reduced well. **E:** Obtain comparison views when uncertain. This round ossicle in the medial joint space appears to belong there and was missed by an orthopedist, but actually it represents the medial epicondyle trapped inside the joint.

Clavicle Fractures

Usually, the clavicle fractures at the junction of the middle and distal thirds. Neurovascular damage to the underlying brachial plexus and subclavian vessels is rare but should be considered. Usually, the periosteal sleeve is intact, and remodeling is excellent. Treatment consists of preventing movement with a sling for 4 weeks in children and 6 weeks in teenagers.

Knee Ligament Injuries

Knee ligament injuries are rare in children. Usually, femoral growth-plate separation occurs instead of collateral ligament damage. The tibial growth plate is protected because the collateral ligaments insert distal to it. Any trauma resulting in a swollen knee in a growing child (i.e., ages 7 to 13 years) should be examined to rule out avulsion of the tibial intercondylar eminence because this eminence represents the insertion of the anterior cruciate ligament and is a serious injury. It is seen best on the lateral view but may be pulled up and overlapped by the femoral condyles (Fig. 432-44B). Oblique views or positioning in extension may help.

Partial fractures of the medial proximal tibial metaphysis are notorious for later developing outward angulation as a result of medial tibial growth-plate overactivity. Care should be taken to

obtain good initial fracture alignment, and parents will accept this outward angulation better if they are forewarned that it may occur despite adequate care. Some of the angulation may decrease with time, but osteotomy may be performed near maturity if significant deformity persists (Fig. 432-44C,D). The ossification patterns of the elbow and wrist are complex. If a radiographic text is not available, comparison films of the contralateral side may clarify abnormalities (Fig. 432-44E).

Ankle Injuries

Several types of ankle injuries are common, with injuries of the distal fibula predominating. If a bone fragment is nondisplaced, diagnosis is made by palpating for tenderness at the growth plate, not by obtaining stress films. Three weeks in a short leg cast is the usual treatment; the intent is more to increase patient mobility than to decrease instability. On the tibial side, the anterolateral quadrant of the distal tibial physis is the last to close, and this area may be avulsed with rotation. It should be reduced if it is displaced.

Soft Tissue Injuries

Soft tissue trauma of the extremities may be encountered in the emergency department, and foreign penetration should be considered. Occasionally, palpation and radiography at soft tissue settings may help. Glass is seen only if it has a high lead content. Exploration for deeper foreign bodies may be very frustrating unless it is done surgically under adequate regional or general anesthesia. Contusions to areas with thick subcutaneous fat may produce permanent depression of the area secondary to fat necrosis; families of affected children should be so counseled. Injuries involving large amounts of soft tissue loss might possibly be covered with a patient's skin, which can serve as a split-thickness graft if it can be saved in cool storage.

Open Fractures, Dislocations, and Compartment Syndromes

Open fractures, dislocations, and compartment syndromes (the last in particular) should be treated as quickly as possible for best results. Open fractures allow bacteria to come in contact with damaged tissue, which is an ideal culture medium. They should be irrigated down to bone within 6 hours to decrease the rate of infection and, if extensive, should be left open for several days for drainage and further débridement unless they are small and

clean.

Dislocations of almost all major joints constitute emergencies because nerves and vessels are stretched, and further swelling can add to the problem. The ulnar, median, and radial nerves and the brachial artery are involved at the elbow, the sciatic nerve is involved at the hip, and the popliteal artery is involved at the knee. Fractures that occur near these areas have similar implications.

Compartment syndromes have been recognized increasingly in the last two decades. Such syndromes occur when injury increases the tissue pressure within a closed fascial compartment, such as the forearm, leg, and buttock, above the capillary perfusion pressure (usually 30 to 45 mm Hg), resulting in ischemia and swelling. The earliest sign is excessive pain on passive stretch of the involved muscles within the compartment, followed by sensory loss and paresthesias of the involved nerves, and weakness of the muscle. Loss of pulses is a very late sign, and it indicates that pressure has risen above large arteriolar systolic pressure. Confirmation is obtained by measurement of the pressures using a hydrostatic or electronic apparatus. Treatment involves release of the tight fascia, with later skin closure when swelling resolves (if possible) or skin grafting (if not possible).

Fractures with Malposition

Fractures with malposition may have to be stabilized so that an affected child can be transported to a consultant. Materials used for this purpose may be improvised, or plaster splints over soft wadding may be used. In general, fractures should be splinted in the position in which they present, with the exception of femur fractures, which can be placed in longitudinal traction with a splint.

Muscle Contusions

Muscle contusions occur most frequently in the quadriceps, upper arm, or shoulder muscles. They may be intensely painful. Compartment syndromes are rare in these regions. Treatment consists of limitation of hemorrhage by rest, ice packing, and elastic bandage wrapping. Active range of motion should be instituted in 1 to 3 days, but passive range of motion (i.e., stretching) should be avoided because it may cause further damage. Strength rehabilitation is instituted after motion is regained. Myositis ossificans (i.e., intramuscular calcification and ossification) may follow this injury but usually does not limit function.

Child Abuse

Approximately 1% of all children in the United States are abused. Most victims are younger than age 3. In children younger than 1 year, fracture more often than not may be nonaccidental. One-third of victims are reinjured if the initial diagnosis is missed. Fractures occur in approximately one-third to one-half of all abused children. The most common sites include the long bones, skull, and ribs. Most fractures that are specific for abuse are those of the metaphysis near the growth plate, the posterior ribs, the scapula, or the sternum. No radiographic signs are completely diagnostic of abuse. The use of radiographs to look for inconsistency and to guide the investigation into the mechanism of fracture is helpful. Although long-bone fractures may occur from a spontaneous fall out of bed or from a

counter, they are rare. Obtaining an idea of the “age” of a fracture by radiography also is useful. Periosteal new bone forms 6 to 10 days after a fracture in infants. At 10 to 14 days, blurring of the fracture lines and soft, mobile (poorly defined) callus is present. Hard callus occurs at 14 to 21 days. The differential diagnosis includes osteogenesis imperfecta, Caffey disease, syphilis, scurvy, rickets, leukemia, and congenital insensitivity to pain.

If abuse is suspected, reporting is mandatory, and the reporter is protected by law. The initial search for other fractures should be done by skeletal survey (AP and lateral views of the skull and spine, an AP view of the extremities), with bone scanning in selected cases. Usually, admission to the hospital is the best means of protecting affected children and further evaluating the family. Keeping careful records and being willing to advocate for such children may be the most important steps physicians can take to help.

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