Limb Pain in Childhood

David D. Sherry, MD, FAAP*

The questions below should help focus the reading of this article.

1. What are typical physical examination findings in children with joint hypermobility?
2. What clinical features are helpful in differentiating between toxic synovitis and septic arthritis?
3. In patients with normal radiographs, what causes of limb pains are likely to be detected by bone scans?
4. In an adolescent with psychologic problems and dysfunction of an extremity, what are helpful clinical features in differentiating between reflex neurovascular dystrophy, psychogenic pain, and conversion reactions?

Limb pain during childhood is common, and its causes range from the benign, such as joint hypermobility, to the potentially fatal, such as infection or tumor. Clues to the diagnosis are usually evident in the history, which can then give direction to the physical examination and, if necessary, laboratory investigations. Children with limb pain may be either ill or well (Table 1).

TABLE 1. Causes of Limb Pain in Childhood

- Ill children with limb pain
  - With severe joint pain
    - Infectious
    - Neoplastic
    - Rheumatic/immune-mediated
  - Without specific joint pain
    - Infectious
    - Neoplastic
    - Rheumatic
    - Metabolic
- Well children children with limb pain
  - Traumatic
  - Mechanical
  - Neoplastic
  - Hematologic
  - Infectious
  - Rheumatic
  - Psychophysiologic
  - Miscellaneous

Assistant Professor of Pediatrics, University of Washington, Director, Clinical Pediatric Rheumatology, Children's Hospital & Medical Center, 4800 Sand Point Way, NE, Seattle, WA 98105.

ILL CHILDREN WITH LIMB PAIN

Severe Joint Pain

The majority of ill children with severe limb pain experience an acute onset and may have associated symptoms, such as rash, fever, previous infection (especially skin infection, sore throat, or diarrhea), conjunctivitis, dysuria, or weakness (frequently interpreted by the family as tiredness). The severe nature of the pain usually renders the limb immobile or prevents even light weight-bearing. Erythema over the joint is an important sign because it is rare in chronic arthritic conditions (Table 2). The most important possible diagnoses include: infection, neoplasia, gastrointestinal disorders, rheumatic diseases, and cardiopulmonary disease.

Infection. Septic arthritis (with or without osteomyelitis) is the most common cause of severe monarticular pain. The absence of fever does not exclude a bacterial process. This is particularly true of septic sacroilitis and osteomyelitis of the pelvic and vertebral bones. Sudden onset of severe joint pain with marked swelling (and frequently erythema) is characteristic and typically involves the knee, hip, and ankle. The most common agent is *Staphylococcus* in older children and *Haemophilus influenzae* in younger children. Multiple joint infection can occur with *H influenzae* and the *Neisseria* species.

Septic sacroilitis can be occult and have an indolent course. Although the pain may radiate to the hip or leg, the most intense pain is elicited with movement of the sacroiliac joint. This is effected by either compressing the iliac wings medially or by flexing and externally rotating the hip while pushing downward on the knee when the patient is supine. If septic sacroilitis is suspected, the sacroiliac joint should be palpated by digital rectal examination.

EDUCATIONAL OBJECTIVE

26. The pediatrician should have knowledge to make an appropriate evaluation of a 9-year-old boy with a limp and pain in his left knee and thigh, differentiating among transient synovitis, septic arthritis (including Lyme disease), osteomyelitis, Legg-Calvé-Perthes disease, spondyloarthropathy, juvenile arthritis, rheumatic fever, tumor, slipped capital femoral epiphysis, and a conversion reaction (Topics, 90/91).

Signs and symptoms of septic arthritis or osteomyelitis cf the foot 2 to 3 weeks after a puncture wound deserve special mention. Frequently, the puncturing instrument comes through the shoe, and the foreign material must be arduously sought. In this case, *Pseudomonas aeruginosa* is a likely infecting organism.

Bone scans are frequently helpful when plain radiographs are normal in infections, occult fractures, diskitis, reflex neurovascular dystrophy, Legg-Calvé-Perthes disease, and osteoid osteoma.

Certain laboratory findings may also be helpful in determining whether or not severe joint pain is caused by infection. A bone scan may be diagnostic long before radiographic changes have occurred. In the case of septic arthritis, the synovial fluid usually has more than 50 000 white blood cells per cubic millimeter. The synovial fluid glucose level normally can be up to 40 mg/dL less than the serum level; lower levels suggest infection as opposed to inflammation from rheumatic causes. Measurement of synovial fluid protein concentration is unhelpful. Unfortunately, organisms are cultured from only 50% to 70% of septic joints. Therefore,

TABLE 2. Conditions With Which Joint Erythema May Occur

- Septic arthritis
- Serum sickness
- Kawasaki syndrome
- Acute rheumatic fever
- Leukemia
- Systemic lupus erythematosus
- Reiter syndrome (reactive arthritis)
- Juvenile rheumatoid arthritis (extremely rare)

with the clinical findings of septic arthritis, full antibiotic therapy should be administered. The erythrocyte sedimentation rate is a useful guide to monitor therapeutic response.

Neoplasia. Either leukemia or neuroblastoma can appear as a single inflamed joint, usually the hip. Bone pain, especially over the tibia, is common. These conditions occur in younger children, and the age of onset peaks between 2 and 7 years of age. A normal complete blood count does not eliminate these diagnosis considerations; a bone marrow examination or bone scan will be indicated in an obligate number of sick children with limb pain.

Gastrointestinal Disorders. The occurrence of gastrointestinal symptoms with joint disease (usually the lower extremity) is suggestive of inflammatory bowel disease. Most patients will have bloody diarrhea, fever, and weight loss by the time arthritic manifestations are present. Previous infectious gastroenteritis, especially those caused by *Salmonella, Shigella, Yersinia,* and *Campylobacter* can be followed within 2 to 4 weeks by a reactive arthritis, including Reiter syndrome. Reiter syndrome consists of arthritis, urethritis (which may not cause dysuria but only a sterile pyuria), and conjunctivitis (transient and frequently appearing early). Fever is common, and there may be an associated rash, most commonly on the glans penis (small, painless ulcers) or soles of the feet (small vesicles progressing to hyperkeratotic papules). The signs and symptoms of Reiter syndrome may not necessarily be synchronous; it may take a week or more to arrive at the diagnosis.

Rheumatic Diseases. Acute rheumatic fever is seen between the ages of 5 and 15 years. It follows a Group A β-hemolytic streptococcal infection by 2 to 4 weeks and is associated with a host of systemic symptoms, the most frequent being arthritis, carditis, and fever. The Jones criteria must be met in order to make a definite diagnosis (Table 3). The arthritis is quite severe causing exquisite pain, swelling, and frequently erythema. The arthritis is migratory, lasting up to a week in any single joint.

Systemic lupus erythematosus is more common in female adolescents than in male adolescents, and it is associated early with arthritis or arthralgia. When arthritis is present, it usually causes severe joint pain and can cause joint erythema.

Systemic onset juvenile rheumatoid arthritis typically is associated with high spiking fevers and marked systemic symptoms, including debilitating myalgia and arthralgia. The arthritis may appear days to weeks after the onset of systemic symptoms and is not as severe as the arthritis of the conditions just mentioned. It can be monarticular but more frequently is polyarticular.

Serum sickness, which follows 2 to 4 weeks after exposure to an antigenic (such as the antibiotic cefaclor), may be associated with rash, fever, and peripheral swelling from edema or arthritis.

In a younger child, Kawasaki syndrome can cause limb and joint pain, but typically it occurs days to weeks after fever, rash, peripheral edema and redness, mucocutaneous involvement, lymphoadenopathy, and conjunctivitis. An early clue to Kawasaki syndrome is the presence of iritis on slit lamp examination. This symptom is found in up to 80% of cases during the first week of illness.

Nonsteroidal anti-inflammatory medications will not obscure serious illness such as malignancy, infection, or arthritis.

Henoch-Schoenlein purpura is associated with arthritis. However, children generally are not very ill, except for low-grade fever. Signs and symptoms may develop over a week or two and may last as long as 8 weeks. These include: arthritis; nonthrombocytopenic purpura, which may be preceded by urticaria, in dependent regions of the body (typically, below the waist and elbows); sharp, intermittent abdominal pain, occasionally associated with hematochezia; and microscopic hematuria. Children 4 to 7 years old are usually affected. Treatment is supportive; nonsteroidal anti-inflammatory medication relieves the arthritis. Rare severe sequelae include intussusception and renal failure.

TABLE 3. Revised Jones Criteria for the Diagnosis of Acute Rheumatic Fever*

Major Manifestations	Minor Manifestations
Carditis	Fever
Polyarthritis	Arthralgia
Chorea	Previous rheumatic fever
Erythema marginatum	Previous rheumatic heart disease
Subcutaneous nodules	

* Evidence of a previous streptococcal infection (increased anti-streptococcal antibodies, positive culture, or recent scarlet fever) is required, except when the current problem is a late complication, such as Sydenham chorea or low-grade carditis. Two major criteria or one major criterion with two minor criteria are required to fulfill Jones criteria.

Cardiopulmonary Disease. Most children with new onset hypertrophic pulmonary osteoarthropathy are quite ill. They have painful widespread arthritis and may not have pulmonary symptoms. The etiology is usually pulmonary neoplasia, such as lymphoma. New onset clubbing is an early finding and may be present months before they become ill. A chest radiograph should be obtained.

Absence of Specific Joint Pain

Not all children with limb pain will have their pain limited to the joints. Careful questioning may lead the physician to suspect muscle, tendon, or bone disease. It is important to con-

sider that referred pain may be described as limb pain when the pathology is proximal to the pain. The most common are lateral thigh pain (with or without medial knee pain) arising from the hip and spinal pathology in association with leg complaints.

Infection. Limb pain that can be hard for both patient and physician to localize can occur with osteomyelitis, especially in the pelvis. Plain radiographic studies may be normal; if osteomyelitis is suspected, a bone scan is indicated. Pyogenic myositis is most common in the southern United States and tropics and is most commonly due to *Staphylococcus aureus*. It can involve any muscle; and frequently the biceps, quadriceps, gluteal muscles, and pectoralis are affected. If an abscess is present, surgical drainage and antimicrobial therapy are required.

Diskitis can occur at any age; the average age of onset is 6 to 7 years. Most children have severe back or leg pain and refuse to walk. High fever is uncommon, but most have elevated erythrocyte sedimentation rates. Diskitis is most likely infectious, especially due to *S aureus*, although positive cultures are obtained in only 60% of the patients. Treatment is bedrest using a spica cast or traction and the administration of antistaphylococcal parenteral antibiotics for 4 weeks.

Neoplasia. Limb pain due to leukemia or neuroblastoma may not necessarily cause arthritis, as previously described.

Rheumatic Diseases. Several of the conditions discussed above may result in limb pain or arthralgia before frank arthritis is present. Systemic onset juvenile rheumatoid arthritis frequently is associated with severe myalgia and arthralgia, as is acute rheumatic fever. In systemic juvenile rheumatoid arthritis, the neck frequently is involved and may mimic meningitis. Bilateral calf pain is an early sign of polyarteritis nodosa, although other signs and symptoms may develop, such as abdominal pain, fever, hematuria, hypertension, and rash over a period of days to months.

Muscle pain can be a feature of these rheumatic diseases and is a prominent feature of dermatomyositis and polymyositis. Multiple viruses may cause a transient myositis. Severe muscle pain or cramping after exercise may indicate a metabolic dysfunction, such as McArdle syndrome (first appearing as inability to relax the muscle after exercise), carnitine palmitoyl transferase deficiency, phosphofructokinase deficiency, and Fabry disease. Elevated levels of serum creatinine kinase or other muscle enzymes are found with most of the conditions mentioned.

WELL CHILDREN WITH LIMB PAIN

The majority of children who have limb pain are otherwise well on initial examination. They may have intermittent or continuous symptoms, and the pain may be well localized or quite vague. At times, gait abnormalities are the primary concern rather than pain.

Trauma. Traumatic injuries are common to long bones and joints. Traumatic joint effusions are not associated with any of the signs of inflammation. The synovium does not feel thick and the fluid is bland, accumulating during 12 to 24 hours after either a direct blow or forced abnormal motion. Chronic effusions can be associated with overuse, such as chondromalacia patella. Bleeding into the joint is much more irritating, and effusions develop within 2 hours. The work-up should include investigations for fractures or other major structural damage. Hemophilia can cause frequent, repeated hemarthrosis. With or without trauma, hemarthrosis is a feature of pigmented villonodular synovitis. This is a benign tumor of the synovium; however, it can be invasive locally, is the source of repeated bleeding into the joint, and causes marked inflammation. Adolescents and young adults are most commonly affected, and the knee is the most frequent site. Biopsy is diagnostic; synovectomy is therapeutic, although it may need to be performed repeatedly. Rarely, a synovial hemangioma will be the source of repeated hemarthrosis. This will cause a periarticular mass that may decrease when the limb is elevated. Because plain radiographs are not helpful, computed tomography or magnetic resonance images would be indicated.

Foreign body synovitis (of the kind seen when a thorn is imbedded in the joint, for instance) may lead to chronic, monarticular synovitis. There is an asymptomatic period after the implantation of a few days or longer, followed by acute synovitis that may alternately remit and relapse. Synovial fluid is inflammatory and contains 10 000 to 50 000 white blood cells/mm^3; occasionally, birefringent organic material may be seen under polarized light microscopy. Definitive diagnosis and treatment is excision biopsy and thorough synovectomy.

Joint hypermobility is a frequent cause of nocturnal limb pain in young children and is amiable to physical therapy.

Joint hypermobility is a common cause of evening leg pains, frequently located in the popliteal fossa. It typically occurs in younger children and is defined by having four of the following five criteria: thumb abducts to touch forearm, fingers hyperextend so they are parallel to the forearm, elbows or knees hyperextend greater than 10°, and the child can touch palms to the floor with knees extended. Pain may be more intense after certain activities. Treatment consists of reassurance and discontinuation of secondary gain; some children may benefit from a physical therapy program to strengthen the knee musculature (being careful not to hyperextend the knee). Many of these children sit in a W configuration. Many of these children also have flat feet; flexible flat feet should not hurt, although increases in the valgus of the ankle can place increased stress on the posterior tibial muscles and cause severe nocturnal pain. A medial wedge in the shoe frequently is therapeutic. Inflexible flat feet is a condition indicative of tarsal coalition, which will become symptomatic in half of the patients beginning in early adolescence. These children experience repeated peroneal muscle

spasm and lateral calf pain. Severe cases require surgical correction.

Not uncommonly, 3- to 5-year-old children experience benign nocturnal musculoskeletal pains, or so-called growing pains. These occur more frequently in girls than in boys and are intermittent and localized to the calf, thigh, popliteal fossa, or groin; rarely do they involve the arms. Stretching exercises, reassurance, time, and the removal of secondary gain are the only measures that are needed.

Lyme Disease. One to 2 months after infection with the Lyme spirochete, arthritis or arthralgia may develop. It is generally short-lived and episodic, but may cause symptoms through a period of years. Exposure to endemic areas and a history of erythema chronicum migrans are helpful, although many children report no rash. Lyme serology, especially for IgM antibodies, is diagnostic; tetracycline, erythromycin, and penicillin have been shown to be effective.

Septic Arthritis. The hip poses an especially difficult diagnostic problem because of its unique anatomy and because it is too deep for adequate inspection and palpation. Even in the child who appears well, early septic arthritis warrants careful consideration and will be indicated by extreme pain and hip muscle spasm with any attempt to move the hip.

Toxic synovitis is most likely a viral synovitis or postviral reactive arthritis, and it frequently follows an upper respiratory tract infection. This is the most common cause of hip pain in childhood, and it occurs commonly in children 5 to 10 years of age; boys are affected more often. Toxic synovitis is invariably unilateral. The onset of pain is acute and may radiate to the groin, thigh, or knee, which is typical for hip pathology. Some children have low-grade fever and elevated acute phase reactants, in which case joint aspiration for study and culture are indicated. Most children are much less symptomatic than those with septic arthritis, and they experience a gradually improving course in 1 to 2 weeks (although in rare cases improvement takes months). Treatment is rest and, occasionally, traction. Nonsteroidal anti-inflammatory medication will be of symptomatic help. Because Legg-Calvé-Perthes disease is a rare late sequela of toxic synovitis, follow-up radiographs should be obtained 3 to 6 months after resolution.

Idiopathic *Legg-Calvé-Perthes disease*, caused by repeated infarction of the femoral epiphysis, is characteristically insidious, leading to an antalgic limp with joint stiffness. Pain may increase with activity and be referred to the groin, thigh, or knee. The hip will be tender to palpation and limited in flexion, abduction, and internal rotation. At rest, therefore, the leg will be flexed and externally rotated. Boys are more frequently affected than girls; this condition is rare in blacks.

Radiographic changes are usually evident with radiolucency in the midportion of the femoral head; from the lateral view, irregularity of the anterior epiphysis is usually found. In later stages of Legg-Calvé-Perthes disease, one may see radiographic progression to total collapse of the femoral head. Early in this process, a bone scan will show decreased uptake, whereas inflammatory conditions such as arthritis or toxic synovitis cause increased uptake. If the bone scan is equivocal, magnetic resonance images should be obtained. Treatment is dependent on radiographic stage and age and should be directed by an experienced orthopedic surgeon. If bilateral Legg-Calvé-Perthes disease is found, other radiographs should be obtained to assess for multiple epiphyseal dysplasia. Aseptic necrosis of the hip may occur with sickle cell disease, Gaucher disease, systemic lupus erythematosus, or as a complication of corticosteroid therapy.

There is both an acute and chronic presentation of slipped capital femoral epiphysis. This condition typically affects adolescent boys and may be bilateral in up to 30% of cases. The acute form appears upon initial examination as a fracture with severe pain, which prevents weight-bearing and limits motion after minor trauma, such as running or jumping. The chronic form is more common, causing thigh and knee pain with progressive limitation of hip motion. The specific loss of hip motion depends on the direction of the slip. If the slip is medial, the leg adducts to keep the head properly aligned in the acetabulum. On examination, the hip generally externally rotates as one tries to flex it. Radiographs should include a frog-leg lateral positioning to visualize a posterior slippage. Prompt surgical management is required in all cases.

Protrusio acetabuli may result in hip pain, decreased hip range of motion, or both. Adolescent girls are more likely to develop this condition, and it is bilateral in half of cases. As the acetabulum sinks into the pelvis, a flexion contracture with increased lordosis develops, along with a loss in external rotation. Radiographs are diagnostic and show the femoral head at first crossing the teardrop line of Kohler and then bulging into the pelvis.

Young adolescent girls also seem to be more likely to develop idiopathic chrondrolysis of the hip. The symptoms of this condition include chronic anterior hip pain, which is typically unilateral and may radiate to the knee. Diagnosis is by radiograph, which shows regional osteoporosis, lateral overgrowth of the femoral head, and marked narrowing of the joint space (less than 3 mm). Treatment is symptomatic with a nonsteroidal agent, range of motion exercises, and decreased weight-bearing. In many cases, the femoral head will fuse to the acetabulum; surgical reconstruction or replacement may be required.

An unfortunate cause of hip pain is undiagnosed or inadequately treated congenital dislocation of the hip. This condition is six times more common in girls and involves the left hip 1.5 times more frequently than the right (bilateral disease is seen in 30%). Patients with congenital dislocation of the hip report hip pain (or referred thigh or knee pain), progressive limp with shortening of the involved leg, a Trendelenburg gait, and gradually increasing limitation of hip range of motion. Progressive osteoarthritis ensues. Radiographs reveal an oblique, shallow acetabulum with a dislocated, small, and deformed femoral head. At this point, surgery is the only treatment option. Prevention, by early detection and splinting, is critical. After the first few weeks of life, the characteristic hip click felt with ab-

duction in a flexed position disappears; thus, the presence of limited abduction, abnormal thigh creases, shortening of the femur, or instability of the hip should be investigated radiographically or, if necessary, by computed tomography or magnetic resonance imaging.

Hip pain (or other limb pain) in otherwise well-appearing children may also be caused by rheumatic disease, such as juvenile rheumatoid arthritis, or seronegative spondyloarthropathy. Isolated hip pain is rare with juvenile rheumatoid arthritis. Other joints—especially the knees, ankles, and elbows—are typically involved when children have pauciarticular juvenile rheumatoid arthritis. The same is true of children with polyarticular juvenile rheumatoid arthritis, who are additionally at risk for synovitis of the smaller joints of the hand, foot, neck, and jaw. These two entities most often affect girls (especially between the ages of 1 and 3 years) and adolescents. The spondyloarthropathies also often cause hip pain, and here boys predominate. However, additional sites of inflammation are usually present, especially in the lower extremities; most common are inflamed entheses (sites of tendon and ligament attachments to bone). HLA-B27 is present in 70% of the cases.

Osteoid osteomas can be located anywhere; most frequently they are in the shafts of the long bones (especially the femur) and the posterior elements of the spine. Vague, severe pain is the predominant symptom, and it may respond dramatically to aspirin. Radiographs show characteristic dense, sclerotic lesions. Small lesions are easily seen on a bone scan. Surgical removal, if complete, is curative.

Localized bone pain in a young child (less than 10 years of age) may be indicative of eosinophilic granuloma, which would be visualized as a unifocal lytic lesion within the diaphyseal medullary cavity. Depending on the lesion, surgery, radiotherapy, or chemotherapy may be necessary.

Malignant tumors, such as osteosarcoma and Ewing sarcoma, cause bone pain over the lesion, with or without swelling. Both osteosarcoma and Ewing sarcoma occur more commonly in boys in their second decade. Metastatic lesions may be heralded by constitutional symptoms, including weight loss, fever, and weakness. Both are aggressive tumors requiring equally aggressive combined therapy.

Hemoglobinopathies, such as sickle cell anemia and thalassemia, may cause bone and joint pain. Anemia is an early clue; hemoglobin electrophoresis is diagnostic.

Localized or widespread pain may be seen with reflex neurovascular dystrophy. Most children who have this condition are 9- to 16-year-old girls; the lower extremity is affected predominantly. Pain is the prominent feature reported and may last months. In half of those who have reflex neurovascular dystrophy, minor trauma precedes the pain; the pain, however, becomes increasingly severe with time. In addition to pain, this condition is characterized by hyperesthesia (and occasionally numbness), an incongruent affect when experiencing severe pain, marked compliancy about the degree of pain and dysfunction, and signs of autonomic dysfunction that may include cyanosis, coolness, diffuse edema, increased perspiration, and decreased pulse. Almost all children with this condition have some degree of psychologic overlay. Laboratory evaluation reveals normal acute phase reactants, normal or disuse osteoporosis on radiographs, and abnormal uptake on bone scan (typically decreased but can be increased). Treatment is aggressive physical therapy, which focuses on function and desensitizing hyperesthetic areas. Psychologic assessment is helpful, because almost all children with this condition require individual or family psychotherapy.

An adolescent with severe disability, hyperesthesia, complaining of excruciating pain with a cheerful affect most likely has reflex neurovascular dystrophy.

There is a larger population of children without overt signs of autonomic dysfunction who are identical to those with reflex neurovascular dystrophy, except that half have intermittent symptoms. Diagnostically, they fulfill criteria for psychogenic pain. Conversion reactions do not cause pain but dysfunction, such as paralysis.

Mechanical problems frequently lead to pain. Overuse syndromes, such as tendonitis and bursitis, are site-specific and are detected by specifically testing for each by inspection and palpation and by stressing the involved muscle or tendon by testing muscle strength and range. Chondromalacia patella, or patellofemoral pain syndrome, is the most common and most difficult to diagnose. Adolescent girls are prone to develop this; patients with chondromalacia patella report knee pain, crepitus, locking, or stiffness. Characteristically, pain results from pressure to the patella, either by quadriceps contraction (such as going down stairs) or by the physician (by exerting downward pressure on the patella while the knee is slightly flexed or by pushing downward and caudally on the patella while the patient first relaxes and then contracts the quadriceps). Gradual but intensive strengthening of the quadriceps, especially the vastus medialis (which contracts only during the last 10° of knee extension), is vital to a satisfactory outcome. Nonsteroidal agents may help. Attention to other knee mechanics is important, especially to patella tracking and the Q angle (the angle of a line drawn from the anterior-superior iliac crest and the middle of the patella and a line from the middle of the patella to the midtibial tubercle), which should be no more than 15°. Surgery may be required.

Tibial pain in athletes is a special concern and can arise from a variety of sources. Shin splints are caused by periosteal inflammation along the posterior-medial border of the tibia; pain is elicited by active plantar flexion and inversion of the foot. Rest, with or without nonsteroidal medication, followed by nonimpact exercises usually brings relief.

Anterior and lateral pain brought on by exercise may be caused by a chronic compartment syndrome. During exertion, the pressure within the compartment increases, and pro-

longed postexercise decompression leads to pain and even neurologic symptoms, such as numbness, weakness, or temporary foot drop. The diagnosis must be confirmed by dynamic pressure testing of the compartment in question.

Stress fractures of the tibia are not uncommon in athletic adolescents, particularly in female adolescents and especially in runners. Initial radiographs will be normal, but the lesion will be evident on bone scan. Discrete, tender swelling just over the fracture site will be found upon examination. Rest with close follow-up to ensure compliance and the development of a graduated exercise program is usually all that is required.

Osgood-Schlatter syndrome is caused by microavulsion fractures of the tibial tuberosity at the site of insertion of the infrapatella tendon. This syndrome usually develops in children and adolescents 11 to 15 years of age during a spurt of growth. Unilateral disease occurs in three quarters of those affected, and boys develop this syndrome more often than girls. Rest suffices for most, although the very exuberant child may need casting. A pseudoarthrosis may form and require surgical intervention.

In addition to Osgood-Schlatter syndrome, traumatic osteochondroses include osteochondrosis dissecans and Sever disease. Osteochondrosis dissecans refers to the fracturing off of a bit of subchondral bone, which gradually works through the cartilage and causes mild aching pain and swelling. This typically occurs in the knee of 10- to 20-year-old males, but may occur in the elbow, hip, or ankle. Sever disease involves the Achilles tendon insertion into the os calcis in 6- to 10-year-old children and is marked by heel pain.

Aseptic necrosis, in addition to Legg-Calvé-Perthes disease, can involve the tarsal navicular (Köhler disease), the carpal lunate (Kienböck disease), or the second or third metatarsal head (Frieberg disease). Local tenderness results; diagnosis is radiographic. Treatment is either surgical or supportive, until symptoms subside.

THE PRACTICE CASE

Problem

A 9-year-old boy complains of unilateral knee and thigh pain.

History

Present Illness. The current overall state of health is a useful and important piece of information. If the boy is ill, this would heighten suspicion of infection, which could be of the hip, pelvis, femur, or knee. Other significant points include: duration of pain (acute or chronic); previous similar episodes (involving same or different joint); recent or past trauma and the nature of injury; presence of joint limitation, morning stiffness, or swelling; changes in color or temperature of the extremity; relationship of pain to activities, including whether or not he stops any activity because of pain; intensity of the pain, including whether or not the leg can bear weight, and whether or not the pain causes him to cry or waken at night; history of previous infections (sore throat), upper respiratory tract symptoms, diarrhea) or use of medication; and other symptoms (abdominal pain, dysuria, conjunctivitis, rash, fever, mucous membrane lesions) or other musculoskeletal pain (heel, low back, metatarsal, tibial tuberosity).

Family History. This is frequently positive when dealing with the seronegative spondyloarthropathies. Parents rarely reveal the presence of low back pain if they feel it is traumatic in origin. The physician should ask if any family member has been treated by a doctor or chiropractor for backache, even if that person believes he or she knows the reason for it; falling off of a roof 20 years ago may not be the reason the pain persists. A history of enthesopathy is hard to obtain; it may be helpful to ask about the presence of Osgood-Schlatter syndrome or heel pain. Significant familial diseases include acute iritis, inflammatory bowel disease, collagen vascular diseases, psoriasis, and ankylosing spondylitis.

Social History. Inquiry into family dynamics, recent life events, school problems, and other psychosocial factors may be important in diagnosing reflex neurovascular dystrophy, psychogenic musculoskeletal pain, or in the future management of patients with chronic disease.

Physical Examination

General Examination. A careful, thorough examination will help narrow the differential diagnosis, especially in the case of an ill child. In our 9-year-old, the presence of a rash suggests Henoch-Schoenlein purpura, Reiter syndrome, systemic onset juvenile rheumatoid arthritis, psoriasis, or systemic lupus erythematosus.

Hip pain frequently radiates to the thigh and knee, therefore, always examine the joints above and below the point of pain.

Joint Examination. Our 9-year-old boy's history of thigh pain would increase suspicion of hip disease. Thigh pain may arise from the muscle, femur, or suprapatellar bursa. Inspection of the leg position when flexed, abducted, and externally rotated may indicate hip pathology. Swelling, range of motion, temperature, color, tenderness including hyperesthesia should be documented. Always check the joint above and below and, for those patients with lower extremity symptoms, also check the entheses, especially metatarsal heads, plantar fascial insertion, and the patella at 2, 6, and 10 o'clock. Leg length should be measured from the anterior-superior iliac spine to the middle of the medial malleoli. Increased length is evidence of chronic inflammation, which causes overgrowth; decreased length may signal hip dislocation. Thigh muscle mass should be compared; chronic problems cause wasting. The vastus medialis should be inspected specifically in those with knee pain, and the Q

TABLE 4. Laboratory Investigations for Limb Pain

Laboratory Study	Conditions Evaluated
Antinuclear antibodies	Juvenile rheumatoid arthritis
	Systemic lupus erythematosus
Biopsy	Tumor, including pigmented villonodular synovitis
	Chronic synovitis
	Sarcoid
Bone marrow examination	Leukemia
Bone scan	Diskitis
	Infection
	Inflammation
	Legg-Calvé-Perthes disease
	Occult fracture or tumor
	Reflex neurovascular dystrophy
C-Reactive protein	Infection
	Juvenile rheumatoid arthritis
Complete blood cell count	Hemoglobinopathies
	Infection
	Juvenile rheumatoid arthritis
	Leukemia
	Systemic lupus erythematosus
Erythrocyte sedimentation rate	Infection
	Inflammation
HLA-B27	Reiter syndrome
	Spondyloarthropathies
Immunoglobulin levels	Hypogammaglobulinemia
	IgA deficiency
Lyme titer	Lyme disease
Radiographs	Acetabuli protrusio
	Aseptic necrosis
	Chrondrolysis
	Dislocated hip
	Fractures
	Osteochondritis dissecans
	Slipped capital epiphysis
	Tumor
Rheumatoid factor	Juvenile rheumatoid arthritis
Streptococcal antibody titers	Acute rheumatic fever
Synovial fluid	
Fat globules	Fracture
Blood	Hemangioma
	Pigmented villonodular
	Synovitis
	Trauma
White blood cell count	
Less than 2 000	Trauma
2 000–50 000	Juvenile rheumatoid arthritis
	Reactive arthritis
	Reiter syndrome
	Spondyloarthropathies
	Systemic lupus erythematosus
More than 50 000	Infection
	Juvenile rheumatoid arthritis
	Reactive arthritis
	Reiter syndrome
	Spondyloarthropathies
	Systemic lupus erythematosus
Birefringent particles	Thorn synovitis
Urinalysis	Henoch-Schoenlein purpura
	Reiter syndrome
	Systemic lupus erythematosus

angle measured in those with patella pain.

Toxic synovitis would be typical for this age, sex, and location. An acute slippage of the femoral capital epiphysis would render this boy's hip immobile, and a chronic slippage would reduce range of motion markedly. For a child this age, Legg-Calvé-Perthes disease is a possibility. Possible, but less likely, would be spondyloarthropathy; juvenile rheumatoid arthritis would be even more rare. Other possibilities include osteoid osteoma and idiopathic chrondrolysis.

Initial Evaluation. Hip radiographs are essential, including both anterior-posterior and frog-leg lateral views. If the child is ill, the most important test would be joint aspiration for culture, cell count, and glucose. Additional studies in a child with hip disease that may be indicated, depending on the severity of illness, include: complete blood count, erythrocyte sedimentation rate, urinalysis, antinuclear antibodies, rheumatoid factor, HLA-B27, blood culture, and bone marrow aspirate (Table 4). A bone scan should be obtained promptly if bone disease is suspected (eg, osteomyelitis or Legg-Calvé-Perthes disease).

Any child who stops walking needs prompt attention until an answer is found.

Diagnosis. This 9-year-old boy had a one-day history of left knee and thigh pain, causing an antalgic gait. He was otherwise well without fever, rash, recent infection, or preceding trauma. Review of systems, family history, and psychosocial history were unrevealing. Physical findings were limited to decreased flexion, adduction, and internal rotation of the hip. He was not tender over the spinous processes, sacroiliac joints, or pelvic bones. Investigations included normal blood counts and urinalysis, slightly elevated erythrocyte sedimentation rate (25 mm/h), and normal hip radiographs and bone scan. In light of this information, toxic synovitis of the hip is the most likely diagnosis.

Initial Treatment. Initial therapy would be supportive and directed toward decreasing pressure within the hip joint and relief of pain. Bedrest, close follow-up, and anti-inflammatory medication is usually sufficient and will not obscure chronic, septic, or neoplastic processes.

A final note: Any child who refuses ambulation needs prompt attention and, if the diagnosis is not apparent, close follow-up or hospitalization is indicated.

Self-Evaluation Quiz

1. A 4-year-old girl complains of pain in her popliteal fossae in the evenings. Which of the following findings would *not* be consistent with joint hypermobility as a cause of her symptoms?

A. Thumb abducts to touch forearm.
B. Feet are flat and inflexible.
C. Fingers hyperextend so that they are parallel to forearm.
D. Elbows and/or knees hyperextend more than 10°.
E. Child has ability to touch palms to floor with knees extended.

2. A 7-year-old boy has an acute onset of pain in his right hip. There is no history of trauma or fever. Other than some limited range of motion in the hip joint, he appears well and the physical examination is unremarkable. The *most likely* diagnosis is:

A. Legg-Calvé-Perthes disease.
B. Slipped capital femoral epiphysis.
C. Protrusio acetabuli.
D. Septic arthritis.
E. Toxic synovitis.

3. In a patient with normal radiographs, bone scans are *least likely* to be helpful in the presence of:

A. Benign nocturnal musculoskeletal pains of childhood.
B. Occult fractures.
C. Diskitis.
D. Osteoid osteoma.
E. Legg-Calvé-Perthes disease.

4. A 14-year-old girl has had problems with her right arm for 1 month. Each of the following would be consistent with a diagnosis of reflex neurovascular dystrophy, *except*:

A. Increasingly severe pain through time.
B. Hyperesthesia.
C. Paralysis without pain.
D. Signs of autonomic dysfunction.
E. Abnormal uptake on bone scan.

SUGGESTED READING

Cassiday JT, Petty RE. *Textbook of Pediatric Rheumatology.* 2nd ed. New York, NY: Churchill Livingstone; 1990

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Gershwin ME, Robbins DL, eds. *Musculoskeletal Diseases of Children.* New York: Grune & Stratton; 1983

Miller ML, ed. Pediatric rheumatology. *Pediatr Clin North Am.* 1986;33:1015-1264

Morrissy RT, ed. *Lovell and Winter's Pediatric Orthopaedics.* 3rd ed. Philadelphia, PA: J.B. Lippincott Company; 1990

Schumacher HR Jr, ed. *Primer on the Rheumatic Diseases.* 9th ed. Atlanta, GA: Arthritis Foundation; 1988

EDUCATIONAL OBJECTIVE

91. The pediatrician should have an appropriate awareness of the cardiac defects associated with Marfan syndrome. (Recent Advances, 90/91)

Cardiovascular Involvement in Marfan Syndrome

The Clinical Course and Echocardiographic Features of Marfan's Syndrome in Childhood. Geva T, Hegest J, Frand M. *Am J Dis Child.* 1987;141:1179–1182.
Mitral Valve Dysfunction in the Marfan Syndrome: Clinical and Echocardiographic Study of Prevalence and Natural History. Pyeritz RE, Wappel MA. *Am J Med.* 1983;74:797–807.
Echocardiographic Study of Cardiac Abnormalities in Families of Patients With Marfan's Syndrome. Pan CW, Chen CC, Wang SP, et al. *J Am Coll Cardiol.* 1985;6:1016–1020.

Mitral valve problems and aortic root dilatation are nearly universal in patients with Marfan syndrome. Using echocardiography, Pan et al demonstrated mitral prolapse in 12 of 12 and aortic root dilatation in 10 of 12 patients with Marfan syndrome. These lesions are also present in over one fourth of the patients' first degree relatives. Geva et al found echocardiographic features of mitral prolapse in 25 of 25 patients with Marfan syndrome, and aortic root dilatation in 20 (80%).

The studies by Geva et al and by Pyertiz and Wappel demonstrate that mitral valve dysfunction and aortic root dilatation are progressive. Morbidity and mortality are more likely to be due to mitral insufficiency in young subjects and to aortic dissection or aortic insufficiency in older patients.

Comment: Although these studies may be criticized because of selection bias (undoubtedly many of the patients were referred because of a murmur and were subsequently diagnosed as having Marfan syndrome), there is no denying the high incidence of mitral prolapse and aortic root dilatation in patients with Marfan syndrome. There is consensus that symptoms are often present in infancy and childhood, that they are progressive, and that athletic participation is contraindicated even in the absence of symptoms. There is disagreement about the beneficial effects of long-term β-adrenergic blockade and optimal timing of surgical intervention. (Howard P. Gutgesell, MD, University of Virginia Medical School)