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Approach to Acute Limb Pain in Childhood

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Author Disclosure

Drs Tse and Laxer did not disclose any financial relationships relevant to this article.

Objectives After completing this article, readers should be able to:

1. Recognize the clinical presentation of a septic joint.
2. Understand the pathogenesis and management of septic arthritis.
3. Discuss other causes of acute limb pain in children.
4. Develop an approach for the initial assessment and management of a child presenting with limb pain.

Introduction

In a pediatric setting, physicians frequently are faced with a child presenting with acute limb pain. The differential diagnosis of acute limb pain may include a variety of causes (Table 1), each requiring differing treatments. In certain circumstances, the condition must be diagnosed immediately and appropriate therapy instituted quickly to prevent serious sequelae. Working through the cases discussed in this review offers an approach for the diagnosis and management of acute limb pain in childhood.

Case 1

A 13-month-old boy presents with a 3-day history of irritability and refusal to move his left leg. He had a cold 2 weeks ago and intermittent fever. He has no history of trauma, otherwise is in good health, and has received all of his immunizations. On physical examination, the boy has a fever (temperature, 100.4°F [38°C]); is nontoxic, although apprehensive and lying with his left leg held flexed and externally rotated; has no rash; has pain and decreased range of movement (ROM) of his left hip; has a normal left knee; and refuses to bear weight on his left leg.

The diagnoses of septic arthritis and osteomyelitis should be considered initially for any child presenting with fever and limb pain because they are medical emergencies that can result in rapid joint or bone destruction if not recognized and treated immediately. However, the differential diagnosis also includes other infection-related etiologies (eg, toxic synovitis, reactive arthritis, Lyme disease), malignancies, and inflammatory arthropathies (eg, systemic juvenile idiopathic arthritis [JIA]). Lyme disease often can imitate JIA (see Case 2). A history and physical examination that reveal signs of severe pain, pain that awakens a child at night, or refusal to bear weight suggest more serious problems, such as acute infection or tumor.

Septic Arthritis

Septic arthritis is a medical emergency that results from direct entry of bacteria into the joint following a puncture injury, hematogenous seeding of bacteria into the joint space, or contiguous spread from adjacent infections (osteomyelitis, cellulitis). Hematogenous osteomyelitic spread is seen most commonly in neonates and infants because a network of blood vessels traverses from the metaphysis to the epiphysis, allowing bacteria and pus to cross into the joint space. Following the first postnatal year, the vessels become obliterated by the physis formation, which reduces the chance of infection spreading into the joints in older children. In contrast, more than 50% of neonates who have osteomyelitis have associated septic arthritis. (1)

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Table 1. Differential Diagnosis of Childhood Limb Pain

Infection/Infection-related

- Septic arthritis
- Osteomyelitis
- Reactive arthritis
- Rheumatic fever
- Lyme disease
- Toxic synovitis

Trauma/Overuse

- Fracture
- Soft-tissue injury
- Osgood-Schlatter disease
- Hypermobility

Malignancy

- Leukemia
- Neuroblastoma
- Bone tumors

Hematologic

- Hemophilia
- Sickle cell anemia

Inflammatory

- Juvenile idiopathic arthritis
- Systemic lupus erythematosus
- Henoch Schönlein purpura

Orthopedic/Mechanical

- Slipped capital femoral epiphysis
- Legg-Calvé-Perthes disease

Noninflammatory

- Growing pains
- Fibromyalgia
- Reflex sympathetic dystrophy
- Conversion reaction

Potential infectious agents responsible for septic arthritis vary according to the age of presentation (Table 2). The most common causative organism is *Staphylococcus aureus*, followed by nongroup A beta-hemolytic streptococci. (1)(2) *Haemophilus influenzae* type B (Hib) is less prevalent since the introduction of the Hib vaccine. Additionally, organisms such as group B streptococci and gram-negative pathogens should be considered in neonates, *Salmonella* in patients who have sickle cell disease, and *Neisseria gonorrhoeae* in sexually active adolescents.

Entry of pathogenic organisms into the joint space evokes an acute inflammatory response, resulting in an

intense synovitis. Leukocytes accumulate rapidly in the joint and release cytokines (tumor necrosis factor-alpha [TNF-alpha], interleukin-1 [IL-1]) and proteases that can destroy the articular cartilage. The inflammatory response is very aggressive and often causes rapid joint space loss and destruction, even after eradication of the offending organism. Septic arthritis of the hip in children also is associated with a high risk of ensuing avascular necrosis due to increased joint pressure compressing the blood vessels that supply the cartilage and femoral head.

Septic arthritis typically presents as an acute monoarthritis, with erythema, warmth, swelling, and intense pain on passive movement. The pain can be so severe that it causes a pseudoparalysis of the involved limb. The joint involved most commonly is the knee, but other joints, including the hip, ankle, wrist, elbow, shoulder, and small joints, can be affected. It is important to know that signs of erythema and warmth do not occur in children whose hips are involved. Fever is present in up to 70% of cases (2) and may be accompanied by chills.

The definitive diagnostic test for septic arthritis is aspiration of the joint and analysis of the synovial fluid. The synovial fluid characteristically is cloudy or turbid, has a very high white blood cell (WBC) count (50 to $300 \times 10^3/\text{mCL}$ [50 to $300 \times 10^9/\text{L}$], predominantly neutrophils), and is positive on Gram stain in approximately 50% of cases. Culture of the synovial fluid is positive in up to 70% of cases, with a corresponding positive blood culture in 40% to 50%. Specific media may be needed to isolate and identify potential pathogens in the synovial fluid (eg, *N gonorrhoeae* in adolescents).

The peripheral WBC count is elevated (predominance of neutrophils), along with the markers of inflammation (eg, erythrocyte sedimentation rate [ESR], C-reactive protein [CRP]). When compared with ESR, CRP has been found to be more accurate, especially as a negative predictor of disease. (3) Imaging studies are not diagnostic for septic arthritis but are helpful in supporting a clinical suspicion of the disease. Radiologic manifestations may not be apparent until 10 days into the illness and can include osteopenia, marked joint space loss, and soft-tissue swelling. Ultrasonography is useful in detecting joint effusions. Bone scans, computed tomography (CT) scans, and magnetic resonance imaging (MRI) can be considered in ambiguous cases of septic arthritis because they are more sensitive than radiographs, especially for early septic arthritis. CT scans and MRI are good at demonstrating joint effusions, soft-tissue swelling, and abscesses.

Septic arthritis requires prompt treatment with antibiotics. Empiric antibiotic therapy should be adminis-

Table 2. Microorganisms Involved in Septic Arthritis/Osteomyelitis and Choice of Empiric Antibiotics

Age	Potential Organisms	Recommended Empiric Antibiotic Therapy
Neonate	Group B <i>Streptococcus</i> , <i>Staphylococcus aureus</i> , gram-negative bacilli	Cloxacillin + gentamicin*
Infant (1 to 3 mo)	<i>Streptococcus</i> sp, <i>Staphylococcus</i> sp, <i>Haemophilus influenza</i> , pathogens as per neonates	Cefuroxime; cefotaxime*
Child	<i>S aureus</i> , <i>S pneumoniae</i> , group A <i>Streptococcus</i>	Cefazolin*
Adolescent	As above; also <i>Neisseria gonorrhoeae</i>	Ceftriaxone or cefixime + azithromycin*
Sickle cell disease	As above; also <i>Salmonella</i>	Cefotaxime*
Puncture wound of foot	As above; also <i>Pseudomonas</i>	Piperacillin + gentamicin*

*In regions that have a high prevalence of community-acquired methicillin-resistant *Staphylococcus aureus* (CA-MRSA), consultation with an infectious disease expert is warranted, and empiric antibiotic treatment with vancomycin should be considered.

tered intravenously (Table 2) and changed to more specific therapy once the pathogen has been identified and antibiotic sensitivities have been determined. With clinical improvement, antibiotics can be changed to the oral route for 3 additional weeks of therapy. When the hip, shoulder, or knee is involved, assessment and additional management by orthopedic surgeons may be necessary. Joint drainage can be beneficial to reduce intra-articular pressure and to remove bacterial debris, inflammatory cytokines, enzymes, adhesions, and necrotic tissue that can contribute to damage of the affected joint. Drainage is performed by using closed needle aspiration, arthroscopy, or open surgical drainage, which always is used to manage septic arthritis of the hip.

Recently, children who had septic arthritis and were treated according to a clinical practice guideline were found to have improved efficiency in their management (higher compliance with appropriate laboratory tests and choice of antibiotics, reduced rate of initial radiologic imaging with bone scans, faster change to oral antibiotics [after 72 h in uncomplicated disease]) and shorter hospital stays without a significant difference in adverse outcomes. (4) A randomized, double-blind study suggested that the addition of low-dose dexamethasone for 4 days as adjuvant therapy could reduce residual joint dysfunction and shorten the duration of symptoms in children whose hematogenous septic arthritis was documented. (5) The effects presumably resulted from attenuation of the intense inflammatory response and production of damaging cytokines and proteases.

The outcome of septic arthritis depends on the timing of diagnosis, initiation of antibiotics, adequacy of drainage, virulence of the pathogen, and host factors. The

single most important prognostic factor for a good outcome is early treatment. Favorable outcomes are associated with treatment that was initiated within 4 days of symptom onset; poor functional outcomes are associated with a delay of 5 or more days before treatment is begun. Prompt recognition and initiation of therapy along with assurance of patient compliance with the antibiotic course are essential in preventing the severe joint destruction seen in septic arthritis (Fig. 1) as well as associated deformities, decreased ROM, leg length discrepancies, and disabilities.

Osteomyelitis

Infectious osteomyelitis results from hematogenous spread or direct invasion of pathogens into the bone. In some cases, the osteomyelitis may be precipitated by trauma. The infectious agents responsible are similar to those described for septic arthritis.

Children who have osteomyelitis typically present with fever and localized pain. Careful examination may reveal erythema, swelling, pinpoint tenderness of the affected bone, or decreased ROM due to muscle splinting. When the lower extremities are involved, children often limp. The bones involved most commonly are the femur, followed by the tibia, humerus, fibula, radius, calcaneus, and ilium.

The diagnosis of osteomyelitis is based on the history and physical examination findings and is supported by positive technetium-99 methylene diphosphonate bone scan findings and bone culture. Bone scans have been reported to be very sensitive (sensitivity of 85% to 100%) in diagnosing osteomyelitis as well as detecting other affected sites. MRI has been shown to be as sensitive but

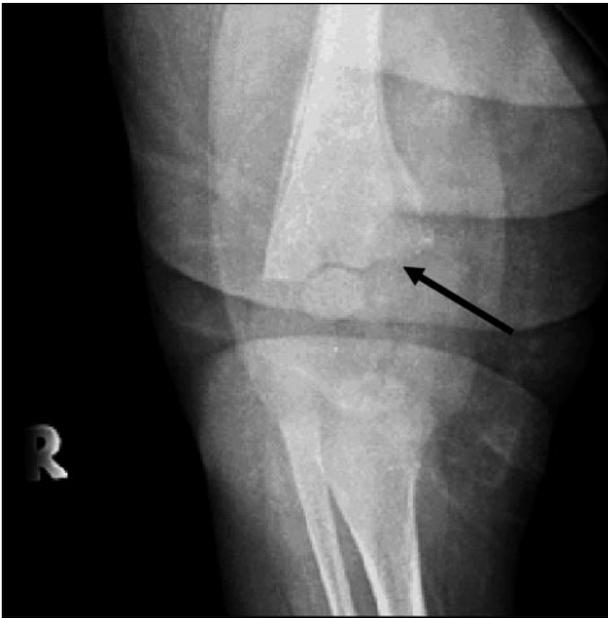


Figure 1. Radiograph of a septic knee joint. Linear periosteal reaction is extensive around the distal femoral metaphysis. Bone destruction is noted around the distal femoral metaphysis posteromedially (arrow).

more specific than bone scans in detecting osteomyelitis. (6) Bacterial pathogens have been recovered from the blood in up to 60% of patients and from affected bone in up to 80%. (7) The peripheral WBC count is elevated, as are markers of inflammation, as seen in septic arthritis. Plain films are useful in excluding other diagnoses but also may demonstrate signs of soft-tissue swelling, subperiosteal changes, and bone destruction. However, these are late findings (≥ 7 d) and, therefore, not helpful in the early diagnosis.

Osteomyelitis requires prompt treatment with appropriate antibiotics, administered intravenously initially, followed by an oral course (4 to 6 wk total duration). The response to treatment is determined by clinical improvement as well as by normalization of inflammatory markers. Most patients have a good prognosis, but complications include recurrence, chronic osteomyelitis, and growth abnormalities (eg, leg length discrepancy). To achieve a favorable outcome, the clinician must verify patient compliance in completing the full course of antibiotics.

Transient or Toxic Synovitis

Transient synovitis is a benign, self-limited disorder that has no known cause. It is postulated to be linked to

infection (acute or postinfectious process; no pathogens have been identified) and possibly to trauma. Often, there is an association with a recent upper respiratory tract infection. Synovitis is a frequent cause of painful limp in childhood, accounting for 30% of all nontraumatic limps. In general, a brief period of sterile inflammation results in a joint effusion and symptoms that resolve over 7 to 10 days.

Boys ages 3 to 8 years (typically < 4 y) are affected most commonly, presenting with a sudden onset of a painful hip or, less commonly, complaints of discomfort in the knee. However, the pain is not as pronounced as that seen in children who have septic arthritis or osteomyelitis. Most patients are afebrile or have only a low-grade fever. The child otherwise appears well and is capable of ambulating, but usually with a limp. When the hip is involved, the limb usually is held in a position of flexion and external rotation. Mildly restricted ROM of the joint is demonstrated on physical examination.

Transient synovitis is diagnosed following the exclusion of other disorders, especially septic arthritis and osteomyelitis. The WBC count tends to be normal, but the markers of inflammation can be mildly elevated. Imaging studies often show evidence of a joint effusion. A recent algorithm developed to help differentiate septic arthritis from transient synovitis of the hip showed four clinical predictors associated more highly with septic arthritis: history of fever, inability to bear weight, ESR greater than 40 mm/h, and WBC count greater than $12 \times 10^3/\text{mcL}$ ($12 \times 10^9/\text{L}$). (8)

Unlike septic arthritis, transient synovitis is self-limited and is managed with conservative measures such as bed rest and anti-inflammatory medications. A small double-blind, placebo-controlled, randomized study in patients who had transient synovitis of the hip showed that ibuprofen shortened the duration of symptoms, although 80% of patients in both groups recovered by 7 days. (9) Joint aspiration generally is unnecessary and should be reserved for patients in whom the suspicion of a septic joint is high.

The prognosis for transient synovitis is good, although some long-term studies have noted a small number of patients who subsequently developed Perthes disease, raising the suspicion of a link between the two diseases. However, this association simply may be coincidental because most patients who have transient synovitis of the hip do not develop this complication, and radionuclide scanning of the hips in these patients does not demonstrate any evidence of vascular insufficiency.

Case 2

A 2-year-old girl presents with a swollen, painful right knee. She continues to walk, but her parents notice that she is slightly less active, especially in the morning. She has no fever, preceding history of trauma, or infection. Her general health is excellent. Her physical examination results include right knee effusion with limited ROM, a warm knee without erythema, and no fever. The girl can walk and run, but she has a right-sided limp.

This case is most consistent with a diagnosis of JIA. Most pediatricians would label this disorder as juvenile rheumatoid arthritis (JRA), but the nomenclature of chronic arthritis is changing to reflect the heterogeneity of the disease (Table 3). Because the child is well and has no evidence of fever or preceding bacterial or viral symptoms, infectious or postinfectious causes of arthritis are less likely. One exception is Lyme disease, which can impersonate JIA, especially if the patient resides in or has traveled to an endemic area. Transient synovitis still can be considered in the differential diagnosis, but the age of presentation, lack of a prodromal illness, and presence of morning stiffness point to a diagnosis of JIA. This case demonstrates the classic characteristic of chronic inflammatory pain that typically is insidious, accompanied by stiffness on awakening in the morning or following prolonged inactivity, and is improved with activity or a warm

bath. The pattern clearly differs from that of a child whose pain is mechanical and who often has symptoms that worsen with increased activity, are relieved with rest, and are associated with other complaints such as locking or giving way of the involved joint.

Juvenile Idiopathic Arthritis

JIA is comprised of arthritis in at least one joint that persists for at least 6 weeks, with an age of onset of younger than 16 years. A joint effusion or at least two of the following findings define arthritis: stress pain, limited ROM, or increased warmth. The prevalence of JIA has been reported to be 16 to 150 per 100,000 persons.

The exact cause for JIA is not known and may be related to a combination of host and environmental susceptibility factors. Immune dysregulation leads to the recruitment and activation of inflammatory cells to the joint lining or synovium. The production of inflammatory mediators (prostaglandins, thromboxanes, leukotrienes) and cytokines (TNF-alpha, IL-1) leads to upregulation of adhesion molecules, further recruitment of inflammatory cells, activation of osteoclasts, proliferation of fibroblasts and synoviocytes, and production of matrix metalloproteinases, all contributing to tissue inflammation, remodeling, cartilage degradation, and in some cases, bony erosions.

The previous subtypes of JRA according to the American College of Rheumatology classification system were comprised of arthritis involving up to four joints (pauciarticular JRA), five or more joints (polyarticular JRA), or association with fever and rash (systemic onset JRA). The International League of Associations for Rheumatology (ILAR) classification of JIA into seven subtypes was proposed to facilitate grouping of patients who have juvenile arthritis into more homogenous populations for study purposes and enhanced communication among clinicians and researchers. (10) The seven subtypes are listed in Table 3. Pauciarticular JRA has been replaced by oligoarticular JIA, which may be persistent (involving no more than four joints) or extended (ie, involving five or more joints after a period of 6 mo). Polyarticular JIA has been divided into two forms, based on the presence or absence of rheumatoid factor (RF). The definition of systemic onset JIA remains the same. The two additional subtypes are arthritis associated with psoriasis (psoriatic arthritis) and enthesitis (enthesitis-related arthritis). Enthesitis is defined as inflammation at the site of the tendon, ligament, or joint capsule insertion into the bone. The subtype "other arthritis" is used for conditions that either do not meet criteria for any other

Table 3. International League of Associations for Rheumatology (ILAR) Classification Criteria for Juvenile Idiopathic Arthritis

General Findings

- Onset <16 y
- Arthritis in at least one joint
- Duration of arthritis \geq 6 wk
- Exclusion of other causes of arthritis

Subtype Categorization

Subtype of arthritis determined according to first 6 mo of disease

- Systemic
- Oligoarthritis—persistent
 - extended
- Polyarthritis—rheumatoid factor—negative
- Polyarthritis—rheumatoid factor—positive
- Psoriatic arthritis
- Enthesitis-related arthritis
- Other arthritis

subtype or fulfill criteria for more than one subtype at the same time.

Oligoarticular JIA is the most common subtype, usually presenting in girls between the ages of 1 and 3 years and involving large joints such as the knees, ankles, wrists, or elbows. Hip involvement is very unusual and should direct the clinician to look for other causes of limb pain. Morning stiffness often is reported. Many children, particularly those who have antinuclear antibodies (ANAs), are at risk of developing anterior uveitis, which almost always is asymptomatic. It is important, therefore, that all children who have JIA undergo ophthalmologic screening regularly.

Children who have polyarticular involvement tend to have symmetric involvement of the small and large joints in the upper and lower extremities. Although most children are RF-negative, those who are RF-positive typically are females who present in adolescence and follow a course similar to that of an adult who has rheumatoid arthritis, with an early onset of erosive synovitis, a chronic course, and frequent development of rheumatoid nodules.

Patients who have systemic onset JIA have arthritis associated with daily spiking fevers and an evanescent rash (salmon pink macules) but also may have features of hepatosplenomegaly, pericarditis, serositis, and lymphadenopathy.

Psoriatic arthritis commonly involves the knees but also the small joints of the hands and feet. Careful attention should be paid to identifying the typical rash, which manifests as erythematous, scaly lesions over the extensor surfaces of the elbows or knees, scalp, postauricular area, and umbilicus. Some patients also may have nail changes (pits, subungual hyperkeratosis, onycholysis) or swollen, sausage-shaped digits or toes (dactylitis).

Older boys make up the majority of the enthesitis-related arthritis subtype and characteristically present with asymmetric involvement of the joints of the lower limbs. Enthesitis usually affects the tendon insertions in the heel, plantar fascia, or insertions around the patella. The condition of some of these patients may progress to ankylosing spondylitis or other human leukocyte antigen B27-associated diseases.

JIA is diagnosed from the history and clinical examination. Initial screening tests include a complete blood count, inflammatory markers (ESR, CRP), ANA, and RF. Radiographs can be used to exclude other causes of joint disease and may demonstrate JIA features of joint effusions, joint space loss, osteopenia, advanced maturation, or erosions. Symptomatic relief can be obtained with anti-inflammatory drugs. Referral to a pediatric

rheumatologist should be made for further management. The treatment for arthritis depends on the involvement and subtype of JIA present and may include anti-inflammatory drugs, intra-articular corticosteroid injections, and disease-modifying agents (methotrexate, sulfasalazine) and biologics (anti-TNF agents) in conjunction with an appropriate physiotherapy program. With prompt recognition and initiation of therapy, the prognosis of JIA has improved and is associated with better functional outcomes than reported previously.

Lyme Disease

Lyme disease is caused by infection with the spirochete *Borrelia burgdorferi*, which is transmitted by tick bites. In North America, Lyme disease is most prevalent in the northeastern, midwestern, and southern and western coastal areas of the United States and in Ontario, Canada. School-age children are affected most commonly, with equal involvement of boys and girls. Most patients are asymptomatic and may not remember having been bitten by a tick. Therefore, it is important to obtain a history of residence or travel to a Lyme-endemic area.

Arthritis is the second most frequent presentation of Lyme disease, following the cutaneous signs of erythema migrans (painless enlarging erythematous rash with central clearing). Other manifestations of Lyme disease include meningitis, cranial nerve palsies, carditis, and ocular involvement (eg, conjunctivitis, keratitis, uveitis, choroiditis, optic neuritis). Arthralgias usually develop in the early phase, but the onset of arthritis may occur months to years after the original infection. Initially, the arthritis is episodic, but it may evolve to a recurrent and prolonged condition. Two thirds of children present with monoarthritis of the knee, (11) but oligoarticular involvement of the large joints and, rarely, a polyarthritis of the small joints also can occur.

The diagnosis of Lyme arthritis is based on a history, physical examination, and laboratory tests to document infection with *B burgdorferi*. Direct tests include culture, stains, or polymerase chain reaction to detect the pathogen in blood, synovial fluid, or synovial tissue. Indirect serologic testing can be carried out via enzyme-linked immunoassay, immunofluorescence, hemagglutination, or Western blotting. The immunoglobulin G titers to *B burgdorferi* can remain positive for years and, therefore, cannot be used to monitor treatment response or failure.

The recommended management of Lyme arthritis is antibiotics administered either intravenously (ceftriaxone 50 mg/kg per day for 14 d) or orally (amoxicillin or

doxycycline for 4 wk). Anti-inflammatory drugs also can be given to alleviate the pain and inflammation of the involved joints. Preventive measures in endemic areas to avoid tick bites include wearing appropriate clothing (long-sleeve shirts and long pants) and using tick repellents.

In contrast to adults, the prognosis of Lyme arthritis in children generally is good, and symptoms resolve over time without permanent damage to the bone or cartilage of the joints.

Growing Pains

Although the child in Case 2 clearly exhibits chronic inflammatory joint pain, growing pains also can occur in this age group and should be considered in the differential diagnosis. Growing pains are intermittent nonarticular pains occurring in childhood and are diagnosed by exclusion based on a typical history and normal physical examination findings. The cause of the pain is unknown, but the condition generally is regarded as benign. Growing pains may occur in any growing child but usually present between the ages of 3 to 10 years. The pain typically occurs at night and frequently is limited to the calf, thigh, or shin. Unlike inflammatory joint pain, the discomfort is short-lived and relieved with heat, massage, or mild analgesics. The child otherwise is healthy and is asymptomatic during the day, having no functional limitations. There may be a history of growing pains in the family. Importantly, the physical examination never is associated with physical findings such as swelling, redness, warmth, or fever. Management of growing pains

consists of reassurance and supportive measures and typically does not require any further investigations.

Case 3

A 10-year-boy presents with a 6-day history of left-sided limp. He complains of pain involving the left thigh and knee. He denies fever, a preceding illness, and trauma and otherwise is in good health. On physical examination, he is afebrile, obese, and walks with a painful limp. He has pain on stress and decreased internal rotation and abduction in the left hip. The findings on examination of his left knee are normal.

Hip pain presenting in an older child can have a variety of causes. Following the exclusion of traumatic injury, the most common cause in this age group is slipped capital femoral epiphysis. It is important to recognize this diagnosis because it is an emergency that requires orthopedic consultation and, in most cases, fixation to prevent further slippage. Transient synovitis is less likely in the absence of a preceding history of infection and also is an uncommon disease at this age. Avascular necrosis of the hip or Legg-Calvé-Perthes disease also should be in the differential diagnosis. Hip involvement as the presenting complaint for JIA is unusual in young patients but can occur in older adolescent boys who have enthesitis-related arthritis.

Slipped Capital Femoral Epiphysis (SCFE)

SCFE is a noninflammatory condition in which the femoral head is displaced from the femoral neck (Fig. 2).



Figure 2. Radiographs of slipped capital femoral epiphysis. The left panel demonstrates displacement of the femoral head from the femoral neck in the left hip. Orthopedic correction includes realignment and surgical fixation with a central screw and is depicted in the right panel.

This condition commonly affects overweight boys between the ages of 10 and 14 years. SCFE also can be associated with endocrine disorders such as hypothyroidism or pituitary deficiencies (eg, growth hormone deficiency); additional endocrine evaluation is recommended in the presence of disease onset in those younger than 10 years of age, in a child who is short for his or her age, or if hypogonadism is present.

A child who has SCFE may report a preceding history of trauma and often presents with pain and an inability to walk (acute “slip”). However, in the setting of a subacute or chronic slip, the symptoms frequently are insidious, with complaints of pain in the affected hip or a limp. The physical examination may demonstrate a limb held slightly flexed and externally rotated. Passive internal rotation of the hip often is limited and painful.

SCFE is diagnosed radiographically and should include imaging of both hips because SCFE can be bilateral

at diagnosis in up to 30% of cases. (12) The slip is classified as mild if the downward slip of the femoral head is less than one third the diameter of the femoral head and severe if the slip is greater than this amount. On diagnosis, urgent referral to an orthopedic surgeon is required. The child should not bear weight and should be prescribed crutches until assessed by orthopedics. Treatment is surgical fixation with a central screw or with bone graft epiphysiodesis. Most patients have a good prognosis, but they can be at risk for acute chondrolysis or avascular necrosis of the hip. Close follow-up is necessary because the contralateral hip can be involved in up to one third of cases.

Legg–Calvé–Perthes Disease (Perthes)

Perthes disease is self-limited, resulting from avascular necrosis of the capital femoral epiphysis. Different causes have been suggested, including trauma and developmen-

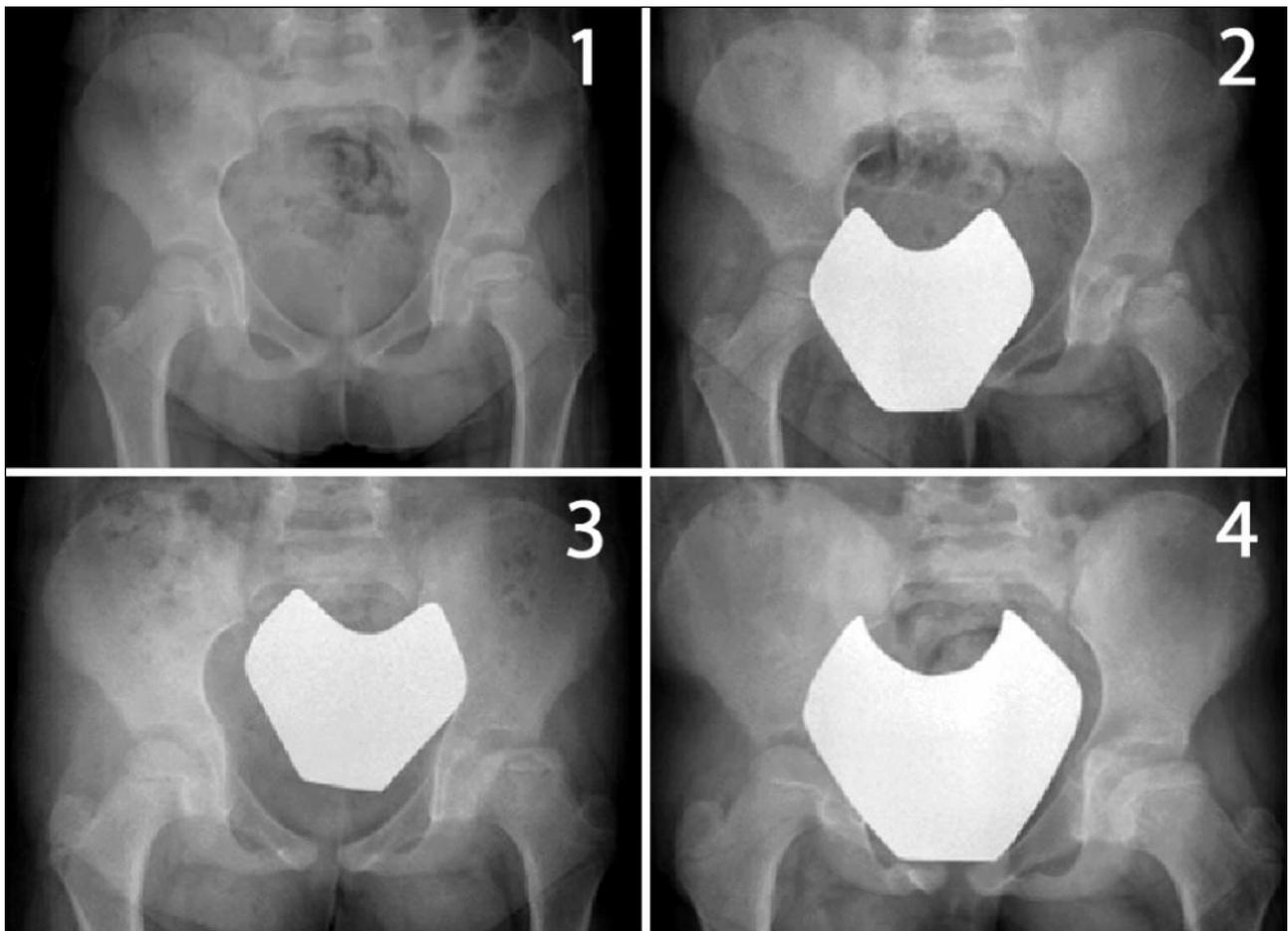


Figure 3. Radiographs of various stages of Legg–Calvé–Perthes disease. Progressive changes of the left proximal femur include Stage 1: initial joint space widening and irregularity of the physis, Stage 2: fragmentation, Stage 3: reossification, and Stage 4: healing.

tal, inflammatory, and coagulation abnormalities. The changes to the proximal femur are hypothesized to arise from repeated interruptions of the vascular supply to the femoral heads. Corresponding pathologic findings can be reproduced from experimental infarctions to the femoral heads in laboratory animals. Recent studies supporting this hypothesis have demonstrated the association of prothrombotic conditions in such patients who have an increased frequency of protein C and protein S deficiencies as well as factor V gene defects. Although underlying prothrombotic conditions can increase the risk of developing Perthes diseases, further studies are required to characterize this association. It is during the revascularization phase that deformation of the epiphysis can arise, which subsequently causes morbidity and increased risk of degenerative arthritis. These changes may be heightened by any weight-bearing forces or muscular stresses transmitted across the acetabular rim.

Perthes disease frequently occurs in boys between 4 and 10 years of age and commonly affects the hips. Children present with a limp, pain, and reduced hip ROM. Radiographs may appear normal at presentation but eventually show progressive changes through four stages: 1) initial joint space widening and irregularity of the physis, 2) fragmentation, 3) reossification, and 4) healing (Fig. 3). MRI often is more helpful and sensitive than radiographs in detecting early disease.

Treatment is aimed at maintaining containment of the femoral head within the acetabulum, which can be achieved conservatively with abduction splints or casts or surgically with an osteotomy of the proximal femur.

In general, children who present before the age of 5 or 6 years do better, which may be related to the extended period of time allowed for remodeling of the femoral head and acetabulum. Another prognostic factor is the extent of epiphyseal necrosis present, with patients demonstrating less than 50% necrosis having a good outcome. (13) Bilateral involvement has been reported in 24% of affected children, with the contralateral hip being involved within 2 years of disease onset, although one study demonstrated that one third of patients presenting with bilateral hip involvement had concordant hip disease with identical staging in both hips throughout the disease course. (14) Girls tended to have a higher prevalence of bilateral hip disease. However, bilateral disease was not necessarily associated with more severe disease or a worse prognosis.

Summary

It is evident from the three cases presented that many disorders present as acute limb pain in childhood. A de-

tailed history and physical examination are critical in the initial assessment, with attention paid to the nature of the pain, presence of limp, weight-bearing status, morning stiffness, systemic symptoms (fever, rash, weight loss, fatigue), history of past medical illnesses, travel, or positive family history (arthritis, bleeding disorders, sickle cell anemia, inflammatory bowel disease, psoriasis). The important aspects of the physical examination include assessing the joints (swelling, erythema, warmth, tenderness, deformity, ROM), adjacent structures (bones, tendons, muscles, skin), gait, and leg length discrepancy, in addition to performing a full neurologic examination. The extent of investigations is determined from the information gained through the history and physical examination; in some cases, no additional testing (ie, growing pains) is required. In the context of atypical features, basic screening laboratory tests and radiographs of the affected site (Table 4) should be obtained. Additional investigations can be performed to arrive at a clinical diagnosis. Suspected cases of septic arthritis require aspiration of the affected joint, with the synovial fluid being sent for Gram stain, culture, and analysis. Fever, redness, moderate-to-severe pain, pinpoint pain or tenderness, and weight loss are clues to more serious causes of limb pain that require additional investigations

Table 4. Preliminary Investigations Suggested for Evaluation of Acute Limb Pain

Basic Screening

- Complete blood count
- Differential count
- Blood smear
- Erythrocyte sedimentation rate
- C-reactive protein
- Radiographs

Further Investigations

- Blood (antinuclear antibody, rheumatoid factor, culture, viral/bacterial serology, creatine kinase, partial thromboplastin time, sickle cell screen, immunoglobulins, complement)
- Urinalysis
- Synovial fluid (cell count, Gram stain, culture)
- Tuberculin skin test
- Imaging (bone scan, ultrasonography, computed tomography scan, magnetic resonance imaging)
- Bone marrow aspiration
- Slitlamp examination of eyes

and referrals to specialists (rheumatologists, orthopedic surgeons, neurologists, hematologist/oncologists).

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Suggested Reading

- Goldmuntz E, White PH. Juvenile idiopathic arthritis: a review for the pediatrician. *Pediatr Rev.* 2006;27:e24–e32. Available at: <http://pedsinreview.aappublications.org/cgi/content/full/27/4/e24>

PIR Quiz

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5. An infant who has osteomyelitis involving the metaphyseal area of the bone is at increased risk of developing septic arthritis of the ipsilateral joint. The reason for this added risk is the traversing blood vessels that extend from the metaphysis to epiphysis, allowing the organisms that cause the osteomyelitis to spread to the joint space. At what age in a child's development do these vessels disappear, thereby reducing this risk of both osteomyelitis and septic arthritis in the same area?
 - A. 6 months.
 - B. 1 year.
 - C. 2 years.
 - D. 3 years.
 - E. 4 years.
6. All joints are at risk for developing septic arthritis with rapid destruction of the joint cartilage if the infection is not recognized and treated promptly. The joint that is at the *highest* risk for avascular necrosis if prompt drainage and antibiotic therapy is not provided is the:
 - A. Ankle.
 - B. Elbow.
 - C. Hip.
 - D. Knee.
 - E. Wrist.
7. A 2-year-old boy has had a temperature to 102.2°F (39°C) for the past 2 days and is unwilling to bear weight on his right leg. Physical examination reveals tenderness of his distal right femur. The combination of tests or procedures that would *best* help to confirm a clinical diagnosis of osteomyelitis is:
 - A. Blood culture and complete blood count.
 - B. Magnetic resonance imaging and C-reactive protein.
 - C. Plain radiograph and erythrocytic sedimentation rate.
 - D. Technetium-99 methylene diphosphonate bone scan and bone culture.
 - E. Ultrasonography and blood culture.
8. Seven different classifications for juvenile idiopathic arthritis now are recognized. Of the following, the type that is *most* commonly associated with anterior uveitis and requires close ophthalmologic follow-up is:
 - A. Enthesitis-related arthritis.
 - B. Oligoarthritis-ANA positive.
 - C. Polyarthritis-RF-negative.
 - D. Psoriatic arthritis.
 - E. Systemic onset arthritis.
9. A 6-year-old boy is experiencing a right-sided limp that has resulted in his increasing reluctance to walk to school over the past month. Physical examination reveals some limitation of motion of his right hip on external rotation. You suspect Legg-Calvé-Perthes disease. Of the following, the test that is *most* sensitive in the early identification of this condition is:
 - A. Computed tomography scan.
 - B. Magnetic resonance imaging.
 - C. Plain radiography.
 - D. Technetium-99 methylene diphosphate bone scan.
 - E. Ultrasonography.

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