

# **Evaluating the Limping Child:** A Rheumatology Perspective

by Reema Syed, MD

Taking a detailed history and completing a thorough evaluation will help hone in on the underlying cause. This article will review important causes of limp from the rheumatologist's viewpoint.



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#### Abstract

Children often present to health care providers for evaluation of limp. Having the knowledge of the different causes of leg pains both in the acute and chronic settings will help in diagnosis, treatment, and referrals to subspecialists in a timely manner. Taking a detailed history and completing a thorough evaluation will help hone in on the underlying cause. This article will review important causes of limp from the rheumatologist's viewpoint.

#### Introduction

A child may present to a primary care provider with a limp due to acute more urgent issues like infections or for more chronic issues like juvenile idiopathic arthritis or other rheumatologic issues. Picking up key information on history and a detailed examination will often lead a provider closer to the correct diagnosis.

# Clinical Evaluation History

Obtaining a detailed history regarding and surrounding the child's presenting symptoms will help narrow the differential diagnosis. Acuity of onset, duration of symptoms, preceding injury or infections, associated symptoms such as fevers, rashes, weight loss, fatigue are important clues to the underlying diagnosis. Inquiring about recent travel will help with infectious causes. Associated pain versus painless limp raises different possibilities.

#### Examination

Assessing a child's gait is crucial in completing an evaluation for lower extremity complaints. Is the child unable to bear weight or is the child holding the affected joint in a fixed position are important questions to address and are signs of more serious issues. If the child appears toxic or has high fevers infectious etiologies need to be excluded.

An antalgic gait signifies pathology in the hip, knee or ankle. A child with antalgic gait will take a shorter stance on the affected limb to avoid bearing weight on the diseased painful joint. A circumduction motion of the hip is also a clue of a painful joint aggravated by motion. Non-antalgic gait include toe walking for instance in leg length discrepancy or due to tight heal cords. Trendelenburg gait is another form of non-antalgic gait seen with weak abductor muscles.

The exam should not be limited to the affected joint but rather include the entire musculoskeletal system looking for tenderness, swelling, warmth, erythema, ligament laxity, and restriction in range of motion. The back and spine should also be examined as well as the skin, which may provide clues.

# SAINT LOUIS

# Causes of Acute Limp Septic arthritis

Septic arthritis is a medical emergency, which if not addressed promptly may lead to damage within one to two days.<sup>1</sup> The most commonly affected joint is the knee but other joints including the hips and other large more than small joints may be involved. The child is usually febrile with inability or refusal to ambulate with a painful limp if child does ambulate. The involved joint is warm, swollen, and erythematous, with limited range of motion. In case of the hip no swelling is found on exam and the child holds the joint in a flexed, abducted, and externally rotated position. The most common causative organism is Staphylococcus Aureus but may be different strains of streptococcus in neonates and infant, salmonella in sickle cell patients, and Neisseria gonorrhea in



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sexually active adolescents.<sup>2</sup> The affected joint fluid should be aspirated which will reveal elevated WBC > 50,000and cultures will be positive in 70% of individuals. Blood cultures will be positive in 50% of patients. Infected joints will need to be irrigated to allow for better healing. The hip joint always needs to be opened and surgically drained to prevent joint damage. Intravenous antibiotics should be started promptly followed by oral antibiotics for at least three weeks is needed for complete recovery.

# Osteomyelitis

Child will present acutely with fevers, localized pain, limping or refusal to ambulate.<sup>3</sup> The most commonly involved bone is the femur and the most common causative organism is *Staphylococcus aureus* either via blood or direct spread from adjacent infected tissue. There may be soft tissue swelling, local tenderness on palpation, and pain with resistance on range of motion of the affected joint. Blood cultures are positive in 60% of cases and bone cultures are positive in 80%. There usually is a delay in changes on plain xrays by a week or more such as soft tissue swelling, subperiosteal changes, and bone destruction.<sup>4</sup> Thus if the suspicion is high, bone scan or MRI, which is as sensitive but more specific than a bone scan, should be performed. Treatment should be started promptly with intravenous antibiotics followed by four to six weeks of oral antibiotics.

### Toxic Synovitis

Toxic synovitis is self-limited benign inflammation of the hip following a recent upper respiratory infection within the last few weeks up to four months. Child will present acutely with a painful limp, which usually resolves over seven to ten days.<sup>5</sup> Boys between the ages of three and ten years are affected more than girls. The child does not appear toxic or as ill as in septic arthritis or osteomyelitis. Low-grade fevers may be noted and child will hold the affected hip in a flexed and externally rotated position. To help differentiate septic arthritis from toxic synovitis Kocher et al. determined four clinical predictors, if present, are more in favor of septic arthritis: fevers

more than 101.3°F, blood white cell count greater than 12,000/ $\mu$ L, erythrocyte sedimentation rate (ESR) more than 40mm/hour, and inability to bear weight. Caird et al. added c-reactive protein (CRP) more than 2.5mg/dL to the criteria. Treatment is symptomatic with NSAIDs and most children recover in seven days with or without treatment though NSAIDs may shorten treatment. Seventeen percent of children may have recurrence out of which 10% may go on to develop inflammatory arthritis especially seronegative spondyloarthropathy.<sup>6</sup> Synovial fluid will reveal WBC counts between 5000 to 15000/ $\mu$ L and negative cultures. The serum WBC, ESR, and CRP are mildly elevated.

# Trauma

Limping is a common manifestation of ligament strains, muscle strains, contusions, and fractures and should be kept in mind whenever a child presents with a limp.<sup>7,8</sup> Apophysitis usually presents subacutely or is chronic but avulsion fractures may present acutely. Runners are especially prone to avulsion fracture of the anterior superior iliac spine at the insertion of the Sartorius muscle.

Compartment syndrome is one complication of fractures and may lead to severe swelling and diminished distal blood flow and should be recognized.

Traumas are addressed more often by orthopods thus beyond the scope of this paper.

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# **Chronic Causes of Limp**

# Juvenile Idiopathic Arthritis

Juvenile Idiopathic Arthritis (JIA) is noninfectious joint swelling, which persists in a child 16 years or younger for more than six weeks. The prevalence of JIA in the U.S. ranges from 1.6 to 86.1 per 100,000 persons.9 It is the most common rheumatic illness in childhood. Underlying etiology is multifactorial occurring in genetically susceptible host in the opportune environment. There are 17 loci found to be associated with JIA risk. The concordance rate in monozygotic twins is between 25-40%. Many cytokines are elevated in JIA including Tumor Necrosis Factor-alpha, Interleukin-1, Interleukin-6, which cause activation and proliferation of many inflammatory cells and adhesion molecules.

JIA is subclassifed based on the International League of Associations for Rheumatology (ILAR) into systemic onset (SoJIA), oligoarthritis with involvement of four or less joints, polyarthritis rheumatoid factor negative, polyarthritis rheumatoid factor positive, psoriatic arthritis, enthesitis related arthritis, and other arthritis.<sup>10</sup> The oligoarthritis is subclassifed further into persistent if the number of joint counts after six months of disease onset remain less than five or extended if five or more joints become involved.

SoJIA presents with quotidian fevers with daily spikes up to  $40^{\circ}$  and evanescent salmon colored rash which becomes prominent with fevers. Patient has polyarthralgia or arthritis. Most of these children appear very ill with systemic manifestation such as lymphadenopathy, hepatosplenomegaly, pericarditis, or serositis. Infections and malignancy must be ruled out. Though limp may occur it is not as prominent a feature of SoJIA as are the systemic symptoms and thus this subtype of JIA will not be discussed further.

#### Oligoarticular JIA

Oligoarticular JIA is the most common form of chronic inflammatory arthritis in childhood making up 40% of all JIA. Four or less joints are involved at onset, which may persist after six months of disease or extend to involve five or more joints. Oligo-JIA is mainly seen in younger kids two to four years of age and is three times more prevalent in girls than boys. The most commonly involved joint is the knee and the most common presentation is joint swelling and limp.<sup>11, 12</sup> The child may have morning stiffness manifested as refusal to walk in the morning or slow awkward gait. Examination of the affected joint(s) reveals swelling and may be warm with pain on range of motion. Unlike in SoJIA labs overall are normal with usually normal blood cell counts, inflammatory markers (ESR, CRP), and ferritin. ANA is positive in 70-80% of oligo-JIA patients

and increases the risk of silent uveitis. The rheumatoid factor (RF) and anti-cyclic citrullinated peptides (anti-CCP antibodies) are mostly negative in oligoarticular JIA.

# Complications Seen in Oligoarticular JIA Uveitis

Uveitis specifically involving the anterior chamber is seen in 45% of oligo-JIA patients. Risk factors to developing uveitis besides the positive ANA include earlier onset of disease < four years of age and female gender.<sup>13</sup> The course of joint disease does not correlate with eye disease. Five percent of patients present with eye disease prior to developing joint disease, which is linked with poorer prognosis. If a child is diagnosed with JIA especially oligoarticular JIA they must be referred to ophthalmology for evaluation for uveitis.<sup>14</sup> In patients with positive ANA follow-ups with ophthalmology should be every three months for three to four years then spaced out to every six months for another three years then annually. Those with a negative ANA need to be followed by ophthalmology every six months. Untreated uveitis may lead to visual loss, band keratopathy, posterior synechiae, and formation of cataracts, glaucoma, and abnormal intraocular pressures.

#### Leg-Length Discrepancy

Oligoarticular JIA when left untreated or if there is a delay in treatment may lead to leg-length discrepancy since the large joints of the lower extremities are mostly involved. Due to inflammation, the ossification centers mature prematurely causing premature fusion of the growth plate and growth arrest.15

# **Treatment of Oligo-JIA**

American College of Rheumatology treatment guidelines were originally made in 2011 which were further updated in 2013.<sup>16,17</sup> Patients with oligo-JIA are started on nonsteroidal anti-inflammatory drugs (NSAIDs) on a regimented schedule. Most children respond very well to NSAIDs. Some patients respond partially and other develop extended oligoarthritis with five or more involved joints and require disease modifying anti-rheumatic drugs (DMARDs). First line DMARDs is methotrexate, which is overall very well tolerated. A few patients continue to have active disease despite use of NSAIDs and DMARDs and require biologic DMARDs specifically anti-TNF agents: etanercept, adalimumab, infliximab, anti-IL 6 agent: tocilizumab, or CTLA-4 fusion protein: abatacept. In case of monoarticular arthritis not responding to NSAIDs, a corticosteroid injection to the affected joint may be very beneficial.

#### Polyarthritis JIA

Polyarthritis RF positive patients present with symmetric small and large joint involvement. This subclass SCIENCE OF MEDICINE |



appears in older adolescent females and appears similar to adult rheumatoid arthritis.<sup>18</sup>

Polyarthritis RF negative patients have similar joint involvement as oligoarthritis patients except for the number of joints involved at disease onset is more than five. This is the second most prevalent form of JIA and similar to oligoarthritis ANA positive patients are at risk for silent uveitis especially if the ANA is positive.<sup>18</sup> These patients present with joint pains, swelling, stiffness lasting for more than one hour, and with limp if lower extremity joints are involved.

For treatment corticosteroids are very often needed initially in polyarthritis patients since these patients are in a lot of pain with restriction or difficulty in activities of daily living. Most patients will need DMARDs and many require biologic agents since disease persists despite use of NSAIDs and DMARDs.

### Lyme Arthritis

Arthritis in Lyme disease is the second most common manifestation after erythema migrans and occurs in two third of patients. It is caused by borrelia burgdorferi and transmitted by tick bites. It is most prevalent in northeastern states from Northeastern Virginia to Maine; Midwestern states mainly Wisconsin and Minnesota, and Northern California. School age children are the most affected age group. Most patient do not recall a history a tick bite. Arthralgia develops in the early phase of disease; the onset of arthritis may be delayed by months to years.<sup>19</sup> Initially the arthritis is episodic but over time become more prolonged and recurrent. Diagnosis is made by obtaining lyme cultures or PCR in the blood, synovial fluid, or synovial tissue. Western blot assays may show 2 IgM or 5 IgG bands which is consistent with lyme disease. It should be kept in mind that IgG titers may remain positive for years after adequate treatment and cannot be used to assess treatment response or failure.

The recommended treatment is either intravenous ceftriaxone for 14 days or oral amoxicillin or doxycycline for four weeks.<sup>3</sup> NSAIDs may be used for symptomatic relief of pain and swelling.

#### Legg-Calve-Perthes

Legg-Calve\_Perthes (LCP) disease in another cause of chronic limp due to idiopathic avascular necrosis of the femoral head epiphysis. It occurs in five to seven-year olds and affects boys four times more than girls. Bilateral hip involvement is seen only in 10% of cases occurring in an asynchronous fashion. LCP tends to be self-limited as the initial interruption of blood supply in the lateral epiphyseal vessels, which supplies the femoral head, is eventually restored. The child presents with painful limp insidious in onset and on examination has limited hip range of motion. There are no preceding infections as seen in toxic synovitis, or fevers and/or rashes as may be seen in SoJIA or malignancy. Risk factors for worse outcome include older age of onset, more advanced changes at initial presentation, and hip deformities after remodeling.

Initial stages of LCP as seen on imaging reveal a dense and sclerosed femoral head epiphysis. A subchondral fracture line "crescent line" may be seen which demarcates the extent of the infarction.<sup>4</sup> Later in this initial stage the femoral epiphysis begins to lose height. The second stage is fragmentation phase, when fissures perpendicular to the articular surface appear within the dense physis. During this phase the articular cartilage hypertrophies and the femoral head may be displaced outside the acetabulum. The next phase is the regenerative phase during which new bone forms and healing begins, but if the femoral head has been displaced then there will be chronic deformation. During this phase there is neovascularization or recannulation of the existing vessels, which may result in early epiphyseal closure. The final stage is the healed stage. Most hips heal without sequelae or residual deformities. Factors for poor prognosis and early osteoarthritis include enlarged femoral head, short femoral neck, and significantly deformed femoral head. The factors contributing to poor prognosis include obesity, progressive loss of hip motion, adduction contracture, and longer duration between disease onset and to healing phase.

Treatment is initially symptomatic with NSAIDs to control pain, joint protection by limiting activities, and physical therapy. These patients need to be referred to orthopedic surgeons for further assessment and long-term management.

#### Slipped Capital Femoral Epiphysis (SCFE)

SCFE is another cause of chronic limp and is due to the displacement of capital femoral epiphysis from the metaphysis of the femur due to abnormality in the physis. This is an orthopedic emergency. SCFE occurs typically in an obese older child or adolescent 10-14 years of age.<sup>20</sup> Boys are affected more than girls in SCFE. It is divided into stable and unstable. In stable SCFE, which is the more common presentation occurring in 90% of cases, a child is able to bear weight and has better prognosis in the long run. Unstable SCFE, when the child is unable to bear weight, has much worse outcome because of potential severe avascular necrosis. On examination the child has an antalgic gait with out-toeing and limited internal rotation of the hip. They may complain of hip, thigh, or knee pain. A third of patients will have bilateral SCFE. This disease is associated

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with endocrinopathies such as hypothyroidism or growth hormone deficiency. Radiographic imaging is diagnostic and if found, patient should be immediately admitted to the hospital for orthopedic evaluation and surgical management. They should be made non-weight bearing immediately until after surgical intervention.

# Osgood Schlatter Disease (OSD)

OSD is apophysitis of the tibial tuberosity and may cause limping but more pain at the tibial tuberosities that is exacerbated by activities. OSD occurs more often in older boys 9-14 years of age but may occur in girls at a younger age.<sup>4</sup> On examination child will have point tenderness at the tibial tubercles with possibly some soft tissue swelling or bony prominence of the tubercle. Treatment is symptomatic with NSAIDs, restriction of activities, and icing at area. Symptoms resolve when the tibial tubercle closes around age 15 years.

# Benign Joint Hypermobility Syndrome (BJHS)

BJHS presents mainly with joint pains and some limp. The affected child may have lower extremity pain especially later afternoon or at the end of the day. Many patients will have nocturnal pain, which awakens them from sleep, which is relieved with rubbing, heat, and NSAIDs. One in five girls and one in ten boys are hypermobile. Three-quarters of these hypermobile children have musculoskeletal pain. Increased range of motion of the knees or ankles due to ligament laxity may lead to repeated microtrauma to tendons and early osteoarthritis.<sup>21</sup> These patients tend to be clumsy due to impaired proprioception. Recurrent joint subluxation, papyraceous scars, loose skin, lens abnormality, and increased capillary fragility should raise the suspicion for Ehlers Danlos or Marfan's syndrome. Patients with trisomy 21 also tend to be hypermobile in most joints leading to premature osteoarthritis and ligamentous injuries. Treatment is based on muscle strengthening for joint protection. Isometric and strengthening exercises, proprioception training, and avoiding over stretching are key to protecting the joints and minimizing pain. Water therapy is very beneficial for strengthening.

# Summary

Children may present acutely or chronically with a limp. Acute infectious causes such as septic arthritis and osteomyelitis must be treated immediately to preserve joint anatomy and function. Oligoarticular JIA may present with limp; delay in diagnosis may lead to significant morbidity with leg length discrepancy or silent uveitis.

SCFE is an orthopedic emergency, which required

immediate surgical intervention. On the other hand LCP disease is usually a self-limited disease requiring symptomatic treatment. A good history and examination will help narrow down the differential diagnosis.

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# Disclosure

None reported.