Every parent knows that healthy children may complain of pains in their limbs or joints, most often in the legs. Complaints that require medical attention and perhaps specialist referral are infrequent, and rheumatic diseases in childhood are comparatively rare. The two major categories are: limb pains and non-inflammatory disorders; and inflammatory disorders and arthritis. Several simple questions

Jeremy’s non-inflammatory problem

Jeremy, 11, complains of pain in the front of his left knee. He limps after playing soccer. His dad is the coach. Exam shows bony swelling and mild tenderness at the tibial insertion of the patellar tendon.

Does Jeremy have arthritis?

Lateral radiograph of Jeremy’s knee shows irregularity of the tibial tuberosity (Figure 1). He has Osgood-Schlatter (osteocondrosis). This condition will settle spontaneously over several weeks or months. Jeremy should not play soccer for a while.

Leanne’s inflammatory problem

Leanne, 13, has been unwell for 4 weeks with fatigue, fever every evening that subsides by morning, and painful swelling in her wrists, fingers, knees, and ankles. Her pharynx is clear, there are no heart murmurs, and there are no enlarged lymph nodes or abdominal viscera.

Hemoglobin: 98 g/L
Total white cell count: 24,000/L, with 80% neutrophils
Platelet count: 795,000/L
Erythrocyte sedimentation rate: 85 mm/1st hour

What is the diagnosis and the prognosis for Leanne’s illness?

The rash on Leanne’s thigh is evanescent and most easily seen in the evening when her temperature is rising (Figure 2). She has systemic-onset juvenile arthritis (Still’s disease). Leanne has been treated with anti-inflammatory and weekly methotrexate injections. Prednisone may need to be added for a brief period. Leanne’s prognosis ranges from gradual improvement over several weeks or months to progression to sustained arthritis.
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should help to clarify the nature and setting of symptoms, including whether the child is generally well or sick. Important diagnostic pointers are duration, localization to limbs or muscles or to one or more joint(s), and presence of systemic illness. Inflammatory arthritis may cause morning stiffness and will usually cause joint swelling.

Younger children may not actually complain of pain, but they or their parent(s) will observe difficulty in daily activities, such as play and walking to school. Ask older children about their sports and recreational activities: Are they or their parents highly competitive? Are they spending many hours each week in dance or gymnastics training? Do they carry heavy books in a backpack to and from school? Or are they posturally inseparable from computers and electronic games consoles?

The presence of sustained systemic illness or failure to thrive may be reason to worry about a more serious disorder, either chronic inflammatory rheumatic disease or, rarely, malignancies, such as leukemia and neuroblastoma.

What are the common non-inflammatory disorders?

Growing pains (benign limb pains)

Usually, in young children (three to seven years), pains are almost always in the legs and between the joints rather than in the joints. The child wakes an hour or two after going to sleep, receives comfort from the mother, and then sleeps throughout the remainder of the night. Sometimes children seem to learn a pattern of obtaining parental attention by complaining of leg pains.

Take a careful social history. The onset of complaints may follow family events, either
disturbing or happy, including the arrival of a new sibling. A family history of the “growing pains” is more often obtained from a parent rather than the siblings. Physical exam is usually normal.

Management requires exclusion of other orthopedic or rheumatic disorders, reassurance that growth is not painful, patience, and limited use of simple analgesics.

**Hypermobility syndrome**

Most often encountered in girls in their early teens, hypermobility syndrome is common with knee or ankle complaints. Children have increased articular flexibility, usually in many joints, but sometimes they only have hyperextensibility in the knees or increased patellar mobility. Musculoskeletal exam will show ranges of joint mobility beyond normal for age, particularly in the metacarpal-phalangeal joints, wrists, and knees. Observe the gait and the increased mobility of the patella or flat feet. Small non-inflammatory effusions may occur.
Hypermobility is frequently familial and the child’s mother may give a history of being loose-jointed in childhood, but grew out of the tendency in her early twenties. The patient’s first-degree relatives may have had scoliosis, pes planus, developmental hip dysplasia, or other musculoskeletal disorders associated with lax ligaments.

Management of hypermobility syndrome includes reassurance that the disorder is benign and that ligaments will stiffen as the child grows into the mid to late teens. Quadricep exercises are helpful to improve patellar stability and knee function. Regular swimming and occasional use of analgesics, such as acetaminophen, are helpful but anti-inflammatory drugs have limited value.

**Anterior knee pain syndromes and orthopedic disorders**

Anterior knee pain is also very common in the early teens, especially in girls. Most often the etiology of symptoms is mechanical rather than inflammatory, although small effusions may be found with patellar instability. A history of recreational and sports activities is especially important. Physical exam must include watching the child walk. A recent literature review of the evidence has concluded that exercise therapy for treatment of patello-femoral pain syndrome may help reduce pain, but its value is uncertain with respect to functional improvement.¹

Localized unilateral hip or knee region pain in the second decade may indicate an epiphyseal problem, such as slipped femoral capital epiphysis, Perthes’ disease, or Osgood-Schlatter disease. These disorders are usually, but not invariably, unilateral. Radiographs are required to confirm the diagnosis and exclude other disorders (Figure 3).
Amplified pain syndromes, back pain and fibromyalgia syndrome

There is increasing interest and concern regarding chronic musculoskeletal pain syndromes in children. Complaints tend to be diffuse, in the manner of “pain all over”. The patterns of symptoms and disorders are reminiscent of fibromyalgia syndrome in adults, and that clinical picture may be encountered in older children. Chronicity and functional limitations disproportionate to objective physical findings may be accompanied by incapacitating distress in the child and the family. Sometimes there is localized vascular abnormality in a limb with the clinical presentation of reflex autonomic dystrophy. This group of disorders has been lumped together in pediatric rheumatology within the concept of amplified musculoskeletal pain syndromes. Treatment for these children requires time and understanding, with specialized co-ordination of institutional multi-professional rehabilitation.

Table 2
Varieties of childhood arthritis, with typical features and age of onset*

<table>
<thead>
<tr>
<th>Type</th>
<th>Gender &amp; age</th>
<th>Lab test</th>
<th>Diagnosis</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Early-onset pauci-articular arthritis</td>
<td>F, 2-3</td>
<td>ANA</td>
<td>Good</td>
<td>Up to 4 joints, risk of uveitis</td>
</tr>
<tr>
<td>Late-onset</td>
<td>M, ≥ 11 years</td>
<td>HLA-B27</td>
<td>Variable</td>
<td>May progress to spondylitis</td>
</tr>
<tr>
<td>Early-onset poly-arthritis</td>
<td>F&gt;M, ≥ 5 years</td>
<td>None specific</td>
<td>Often poor</td>
<td>More than 4 joints</td>
</tr>
<tr>
<td>Late-onset poly-arthritis</td>
<td>F&gt;M, ≥ 10 years</td>
<td>Rheumatoid factor</td>
<td>Variable</td>
<td>Some progress to adult RA, but many do not</td>
</tr>
<tr>
<td>Systemic-onset (Still’s disease)</td>
<td>F&gt;M, ≥ 2 years</td>
<td>High WBC</td>
<td>Variable</td>
<td>Uncommon, and difficult to treat</td>
</tr>
<tr>
<td>Enteropathic arthritis</td>
<td>F=M, ≥ 2 years</td>
<td>None specific</td>
<td>May depend on bowel disease</td>
<td>Diagnostic clue may be in family history</td>
</tr>
<tr>
<td>Psoriatic arthritis</td>
<td>F=M, ≥ 7 years</td>
<td>None specific</td>
<td>Very variable</td>
<td>Joints often affected before skin</td>
</tr>
<tr>
<td>Lupus</td>
<td>F=M, pre-puberty, later age F&gt;M</td>
<td>ANA and specific tests for SLE</td>
<td>Variable</td>
<td>Joint complaints often not prominent</td>
</tr>
</tbody>
</table>

*In the current International League of Associations for Rheumatism juvenile idiopathic arthritis classification, pauci-articular arthritis is called oligo-arthritis, and enthesitis-related arthritis probably includes late-onset pauci-/oligo arthritis.

F: Female HLA: Human leukocyte antigen
M: Male WBC: White blood cell count
ANA: Antinuclear antibody SLE: Systemic lupus erythematosus
resources,5 and very judicious use of medications.

Low back pain is much less common in children than adults, and may have the same implications as chronic limb pain. There must be careful evaluation for significant psychosocial problems, skeletal disease or visceral pathology.

**What should I know about arthritis in children?**

Chronic juvenile arthritis in its various forms is rare and affects only one child in 1,000 up to the age of 16. The cause of most varieties of juvenile arthritis remains obscure. Perhaps infectious agents trigger sustained immunologic reactions, and there are recognized acute disease models (Table 1).

Acute rheumatic fever has not disappeared, despite the decrease in incidence in Canada during the last several decades. Streptococcal infection followed by acute arthritis, which may migrate, should raise the possibility of rheumatic fever.

Lyme disease has a limited geographic distribution in Canada. It follows a tick bite and development of the typical skin lesion, erythema chronicum migrans. Arthritis develops slowly with involvement of large joints, such as the knees, but there may be extensive polyarthritis. Neurologic and other complications can appear later. Diagnosis is based on recognition of the clinical syndrome, and serologic results must be interpreted in the context of the clinical picture.

Septic arthritis is usually seen in young or debilitated children, but not always, especially if there is a potential source of pyogenic infection. Joint aspiration must be done if there is any suspicion of joint sepsis.
Some types of childhood-onset arthritis correspond to adult-onset diseases, and some clearly do not; only 5% or less of all childhood arthritis patients progress to typical adult rheumatoid arthritis. Studies with followup into adult life have demonstrated that outcomes of some subtypes of childhood arthritis are less favourable than was thought in the past with regard to physical disability and social achievement.\textsuperscript{6,7}

Tips for evaluating arthritis

Ascertain whether the symptoms are in or between joints. Different types of arthritis differ in the typical age of onset and gender (Table 2). Children younger than five or six may not be able to localize their symptoms. Look for skin lesions or rashes, lymphadenopathy or hepatosplenomegaly, and heart murmurs or pericardial rub. Anterior uveitis is usually asymptomatic in young children and slit-lamp examination by an ophthalmologist is necessary in early-onset pauci-articular syndrome. If undetected and not treated, serious visual damage may occur. Growth delay can be due to several factors, including impaired nutrition, effects of chronic inflammatory disease, and effects of medication.

Investigation and management of childhood arthritis

Laboratory

Tests can provide non-specific evidence of inflammation and/or evidence for a specific type of juvenile arthritis. The sedimentation rate and platelet count will rise as acute phase inflammatory reactants. Rheumatoid factor testing is of minimal value in children before age eight or nine. Anti-nuclear antibody testing is also of limited value except in young children with early-onset pauci-articular syndrome, or if there are clinical features suggesting lupus. If joint aspiration is indicated, it is done with local or general anesthesia, and the synovial fluid should be examined for total and differential white cell count, as well as bacterial culture.
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Radiology

The primary indication for X-ray examination is to demonstrate orthopedic disorders, some of which can simulate chronic arthritis, e.g. slipped epiphysis, osteochondritis, Perthes’ disease, and bone or joint sepsis or neoplasia. In early stages of arthritis, routine radiographs will show only soft tissue swelling or no abnormality and, therefore, have limited diagnostic value. There may be diagnostic value in early magnetic resonance imaging investigation of childhood arthritis, but access and costs limit its routine application. The role of isotope bone scans is to investigate possible musculoskeletal sepsis.

References


Net Readings

1. Arthritis Society www.arthritis.ca
2. Arthritis Foundation (USA) www.rheumatology.org/patients

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