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Joint Pain in Children, Part V: Juvenile Chronic Arthritis

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The most common cause of chronic joint disease in children is juvenile rheumatoid arthritis. Arthritis affects approximately one child in 1,000 in a given year. Fortunately, most of these cases are mild, however, approximately one child in 10,000 will have more severe arthritis that doesn't just go away. Many children have what is called an acute reactive arthritis following a viral or bacterial infection. This arthritis is often quite severe for a brief period, but usually disappears within a few weeks or months. For both children with arthritis and the doctors who care for them, public education is one of the greatest challenges. Most lay people and many doctors fail to realize that the problem exists. Many children suffer for months or years before the diagnosis of arthritis is thought of the proper treatment begun.¹

There are three major forms of juvenile rheumatoid arthritis: systemic, pauciarticular, and polyarticular. These types are distinguished from one another by their distinct modes of onset and characteristic features (see Table).

The diagnostic criteria for juvenile rheumatoid arthritis include age of onset 16 years or younger, and the involvement of one or more joints for at least three months. No extraarticular manifestations are diagnostic by themselves. No one laboratory test is diagnostic, thus tests to exclude other disease processes are useful. The test for rheumatoid factor is usually negative, except in older children. The most helpful antibody test is that for antinuclear antibodies, which are commonly positive in younger children, particularly when the mode of onset is pauciarticular. As far as initial x-ray findings, there should be only evidence of soft tissue swelling. X-rays should be taken only to rule out any other pathology, i.e., trauma or developmental anomalies. The type of signs of joint changes associated with rheumatoid are not generally present and, one hopes, will not progressive if properly managed.

Table: The three forms of juvenile rheumatoid arthritis.

Mode of Onset	Features: Articular	Systemic
Systemic	varies, only arthralgia may be present	high fevers, rash, possible pericarditis
Pauciarticular	involves no more than four joints	possible iridocyclitis
Polyarticular	bilateral & symmetric four or more joints	low-grade fever, rash, pericarditis

Pauciarticular JRA is defined by the involvement of less than four joints; it is the most common form of JRA. It often begins in a swollen knee or ankle which appears without a history of injury. There are no systemic manifestations, but there is a high incidence of chronic asymptomatic iridocyclitis associated with this type of JRA. There are two dominant subtypes of pauciarticular JRA. The more common subtype occurs in girls ages 2-3 years. The test for antinuclear antibodies is often positive and iridocyclitis is a common finding. The other subtype is most often seen in older boys. The large joints of the lower extremities are usually involved and heel pain is common. An HLA-B27 antigen test is usually positive. Other conditions should be considered when diagnosing this subtype (i.e., inflammatory bowel disease, psoriatic arthritis, ankylosing spondylitis, infection, etc.).

Aside from the pauciarticular JRA, the greatest concern with this condition is the iridocyclitis. The inflammation is not painful, but if not detected and treated it may lead to scarring of the lens and permanent visual damage. Initially this inflammation cannot be seen except by an ophthalmoscope. Iridocyclitis is more common in children with a positive ANA test; these children should be examined at regular intervals. It is recommended that children with positive ANA test be examined every three months; and all other children with JRA be examined every six months. The etiology for this condition and relationship with JRA is still unknown, but it is important to know there is a high incidence associated with JRA.

Another problem associated with pauciarticular JRA is that it may cause accelerated growth of the symptomatic extremity. This will cause a leg length discrepancy, and may lead to premature arthritis (osteoarthritis/DJD) involving the normal extremity. The accelerated growth is caused by the inflammation; if that is controlled there will be minimal accelerated growth. Most often treatment is successful with controlling the inflammation; however, if there appears to be significant leg length discrepancy, the growth of the extremities should be monitored. Shoe lifts can be used to reduce any limp that child may develop. If, however, the leg with arthritis is on radiographs 3cm or more longer than the other leg, more aggressive intervention may be necessary to prevent any damage to the normal leg.

Polyarticular JRA is the form in which four or more joints are involved. This form is more severe, because of the greater number of joints involved, and the fact that it tends to progress over time. Symptoms generally are bilateral and symmetric. Systemic manifestations include low-grade fever, rash, and pericarditis.

Polyarticular JRA can also be divided into two subtypes. The more common subtype can affect either sex but is more common in girls. Small and large joints are affected, and involvement tends to be symmetric. Tests for antinuclear antibodies and rheumatoid factor are usually negative. The other subtype mainly affects older girls. There is symmetric involvement of small and large joints, and the rheumatoid factor test is positive. This is the only subtype that is truly rheumatoid.²

Polyarticular JRA may require fairly aggressive treatment, but it's important to be extremely careful with the use of steroids. In severe cases steroid may be necessary, however the child should be given a short course of treatment with the steroid and taken off as quickly as possible. Steroid treatment for long periods of time will cause short stature and osteoporotic bones, along with other systemic side effects.

The worst form of JRA is systemic juvenile rheumatoid arthritis (Still's disease). It is characterized by high fever, rash, anemia, leukocytosis, and elevated ESR, and sometimes pericarditis. Articular symptoms are variable and arthralgia may be the only such symptoms that are present. It is very important to rule out an infection before diagnosing the patient with systemic JRA. One of the most important findings is that the fever goes away for a least part of the day. Usually the fever is high once or twice each day. At those times the child appears systemically ill; when the fever falls the child appears and feels better. This form of JRA is very unpredictable: at times, the child will have only one episode and will recover completely; other times the fevers and rash disappear, but the arthritis progresses over time and may become very severe. This form of systemic JRA can involve the internal organs, and rarely may result in life threatening disease. In addition to this systemic disease these children have an increased likelihood in iatrogenic reactions to many medications and should be monitored very carefully.

There are several other forms of arthritis which can affect children and adolescents which are often lumped together with JRA, but have different outcomes and should be considered separately. These conditions will be reviewed in the following articles in this series on joint pain in children.

One further topic I'd like to just touch on is the growing evidence that rheumatoid arthritis may be caused by an infectious agent. It is clear that autoimmunity plays a major role in the progression of RA. Most rheumatology investigators believe that an infectious agent causes RA. There is, however, little agreement

as to the responsible organism. Investigators have proposed the following infectious agents: human T-cell lymphotropic virus type I; rubella virus; cytomegalovirus; herpes virus; and mycoplasma. I will not review the theories on each of these causative agents, since this is far beyond my expertise. We should be aware that patients with RA often respond very favorably to antibiotic therapy.³ It appears that investigations are very close to discovering the true cause of at least some of the arthritides.

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