

# Juvenile Arthritis

## Fast Facts

- Arthritis in children is treatable. It is important to seek treatment from health care professionals who are knowledgeable about childhood arthritis.
- In spite of their diagnosis, most children with arthritis can expect to live normal lives.
- Some children with JIA have their disease go into remission.
- Federal and state programs may provide assistance with school accommodations or services. Ask the rheumatology team about summer camps and opportunities to meet other children with arthritis.
- Except in rare circumstances, this condition is not directly inherited from the mother or father.



About 1 child in every 1,000 develops some type of chronic arthritis. These disorders can affect children at any age, although rarely in the first six months of life. It is estimated that around 300,000 children in the United States have been diagnosed with the condition.

Growing up with arthritis can be challenging. However, with care from a team of rheumatology professionals, most children with arthritis live full and active lives and are able to do everything their peers do. There are various types of chronic childhood arthritis, which can last from several months to many years. In every instance, early diagnosis and treatment can help avoid joint damage.

There are many terms used to describe a child with chronic arthritis. These include juvenile rheumatoid arthritis, juvenile chronic arthritis, and juvenile idiopathic arthritis. While JIA is used most by specialists in pediatric rheumatology, JRA is commonly used in the United States. Juvenile arthritis may involve one or many joints and can also cause silent eye inflammation. It can also cause other symptoms such as fevers or rash.

## **+** What is juvenile idiopathic arthritis?

Several types of arthritis, all involving chronic (long-term) joint inflammation, fall under the JIA heading. This inflammation begins before patients reach the age of 16, and symptoms must last more than 6 weeks to be called chronic. JIA may involve one or many joints, and may also affect the eyes. It can cause other symptoms such as fevers or rash.

**Systemic onset JIA** affects about ten percent of children with arthritis. It begins with repeating fevers that can be 103°F or higher, often accompanied by a salmon-colored rash that comes and goes. Systemic onset JIA may cause inflammation of the internal organs as well as the joints, though joint swelling may not appear until months or even years after the fevers began. Anemia (a low red blood cell count) and elevated white blood cell counts are also typical findings in blood tests ordered to evaluate the fevers and ongoing symptoms. Arthritis may persist even after the fevers and other symptoms have disappeared.

**Oligoarticular JIA**, which involves fewer than five joints in its first stages, affects about half of all children with arthritis. Girls are more at risk than boys. Some older children with oligoarticular JIA may develop “extended” arthritis that involves many joints and lasts into adulthood. Children who develop the oligoarticular form of JIA when they are younger than seven years old have the best chance of having their joint disease subside with time. They are, however, at increased risk of developing an inflammatory eye problem (iritis or uveitis). Eye inflammation may persist independently of the arthritis. Because this eye inflammation usually does not cause symptoms, regular exams by an ophthalmologist (eye doctor) are essential to detect these conditions and identify treatment to prevent vision loss.

**Polyarticular JIA** affects five or more joints and can begin at any age. Children diagnosed with polyarticular JIA in their teens may actually have the adult form of rheumatoid arthritis at an earlier-than-usual age.

With psoriatic arthritis, children have both arthritis and a skin disease called psoriasis or a family history of psoriasis in a parent or sibling. Typical signs of psoriatic arthritis include nail changes and widespread swelling of a toe or finger called dactylitis.

**Enthesitis-Related Arthritis** is a form of JIA that often involves attachments of ligaments as well as the spine. This form is sometimes called a spondyloarthritis. These children may have joint pain without obvious swelling and may complain of back pain and stiffness. There is sometimes a family history of arthritis of the spine.

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## **+** What causes JIA?

Malfunctioning of the immune system in JIA targets the lining of the joint, known as the synovial membrane. This causes inflammation. When the inflammation is untreated, joint damage may occur.

It is not known what causes the immune system to malfunction in JIA. In rare cases (such as in psoriatic arthritis or enthesitis-related arthritis) a parent has the same form of arthritis. Dietary and emotional factors do not appear to play a role in the development of JIA.

Because the causes of JIA are unknown, no one knows how to prevent these conditions.

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## **+** How is JIA diagnosed?

JIA may be difficult to diagnose because some children may not complain of pain at first and joint swelling may not be obvious. There is no blood test that can be used to diagnose the condition. Adults with rheumatoid arthritis typically have a positive rheumatoid factor blood test, but children with JIA typically have a negative

rheumatoid factor blood test. As a result, diagnosis of JIA depends on physical findings, medical history, and the exclusion of other diagnoses.

Typical symptoms include:

- limping
- stiffness when awakening
- reluctance to use an arm or leg
- reduced activity level
- persistent fever
- joint swelling
- difficulty with fine motor activities

Other conditions that can look like JIA, including infections, childhood cancer, bone disorders, Lyme disease, and lupus also must be ruled out before a diagnosis of JIA can be confirmed.

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## **+** How is JIA treated?

The best care for children with arthritis is provided by a pediatric rheumatology team that has extensive experience and can diagnose and manage the complex needs of the child and family most effectively. The core team may consist of a pediatric rheumatologist, physical and occupational therapist, social worker, and nurse specialist. This core team can coordinate care with a child's pediatrician, adult rheumatologists, other physicians (such as an ophthalmologist or orthopedic surgeon), and other health professionals (dentist, nutritionist or psychologist) as well as reach out to schools and additional community resources to ensure that the child receives the best care possible.

The overall treatment goal is to control symptoms, prevent joint damage, and maintain function. When only a few joints are involved, a steroid can be injected into the joint before any additional medications are given. Steroids injected into the joint do not have significant side effects. Oral steroids such as prednisone (Deltasone, Orasone, Prelone, Orapred) may be used in certain situations, but only for as short a time and at the lowest dose possible. The long-term use of steroids is associated with side effects such as weight gain, poor growth, osteoporosis, cataracts, avascular necrosis, hypertension, and risk of infection.

Disease modifying drugs - commonly called DMARDs - are added as a second-line treatment when arthritis involves many joints or does not respond to steroid joint injections. DMARDs include methotrexate (Rheumatrex), leflunamide (Arava), and more recently developed medications known as biologics. The biologics include anti-tumor necrosis factor agents such as etanercept (*Enbrel*), infliximab (Remicade), adalimumab (Humira), abatacept (Orencia), anakinra (Kineret); canakinumab (Ilaris), tocilizumab (Actemra), and rituximab (Rituxan). Each of these medications may cause side effects that need to be monitored and discussed with the pediatric rheumatologist treating your child. Many of these treatments are approved for use in children as well as adults. In addition, researchers are developing new treatments.

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## **+** Living with juvenile arthritis