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Juvenile Arthritis

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Questions and Answers about Juvenile Arthritis

This publication contains general information about juvenile arthritis (JA). It describes what juvenile arthritis is and how it may develop. It also explains how juvenile arthritis is diagnosed and treated. At the end is a list of key words to help you understand the terms used in this publication. If you have further questions after reading this publication, you may wish to discuss them with your doctor.

What Is Juvenile Arthritis?

“Arthritis” means joint inflammation. This term refers to a group of diseases that cause pain, swelling, stiffness, and loss of motion in the joints. Arthritis is also used more generally to describe the more than 100 rheumatic diseases that may affect the joints but can also cause pain, swelling, and stiffness in other supporting structures of the body such as muscles, tendons, ligaments, and bones. Some rheumatic diseases can affect other parts of the body, including various internal organs. **Juvenile arthritis (JA)** is a term often used to describe arthritis in children. Children can develop almost all types of arthritis that affect adults, but the most common type that affects children is juvenile idiopathic arthritis.

What Is Juvenile Idiopathic Arthritis?

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Key Words

Both **juvenile idiopathic arthritis (JIA)** and **juvenile rheumatoid arthritis (JRA)** are classification systems for chronic arthritis in children. The juvenile rheumatoid arthritis classification system was developed about 30 years ago and had three different subtypes: polyarticular, pauciarticular, and systemic-onset. This classification system is rarely used today. More recently, pediatric rheumatologists throughout the world developed the juvenile idiopathic arthritis classification system, which includes more types of chronic arthritis that affect children. This classification system also provides a more accurate separation of the three juvenile rheumatoid arthritis subtypes.

Prevalence statistics for juvenile arthritis vary, but according to a 2008 report from the National Arthritis Data Workgroup, about 294,000 children age 0 to 17 are affected with arthritis or other rheumatic conditions.¹

¹According to the National Arthritis Data Workgroup, the actual number of new cases of JA is higher than previously reported because the statistic includes conditions not previously captured, as cited in Helmick CG, Felson DT, Lawrence RC, Gabriel S, Hirsch R, Kwoh CK, et al.; National Arthritis Data Workgroup. Estimates of the prevalence of arthritis and other rheumatic conditions in the United States. Part I. *Arthritis Rheum.* 2008 Jan;58(1):15-25.

What Is Juvenile Idiopathic Arthritis?

Juvenile idiopathic arthritis is currently the most widely accepted term to describe various types of chronic

arthritis in children.

In general, the symptoms of juvenile idiopathic arthritis include joint pain, swelling, tenderness, warmth, and stiffness that last for more than 6 continuous weeks. It is divided into seven separate subtypes, each with characteristic symptoms:

1. **Systemic arthritis (formerly known as systemic juvenile rheumatoid arthritis).** A patient has arthritis with, or that was preceded by, a fever that has lasted for at least 2 weeks. It must be documented as an intermittent fever, spiking for at least 3 days, and it must be accompanied by at least one or more of the following:
 - Generalized enlargement of the lymph nodes.
 - Enlargement of the liver or spleen.
 - Inflammation of the lining of the heart or the lungs (pericarditis or pleuritis).
 - The characteristic rheumatoid rash, which is flat, pale, pink, and generally not itchy. The individual spots of the rash are usually the size of a quarter or smaller. They are present for a few minutes to a few hours, and then disappear without any changes in the skin. The rash may move from one part of the body to another.
2. **Oligoarthritis (formerly known as pauciarticular juvenile rheumatoid arthritis).** A patient has arthritis affecting one to four joints during the first 6 months of disease. Two subcategories are recognized:
 - *Persistent oligoarthritis*, which means the child never has more than four joints involved throughout the disease course.
 - *Extended oligoarthritis*, which means that more than four joints are involved after the first 6 months of the disease.
3. **Polyarthritis—rheumatoid factor negative (formerly known as polyarticular juvenile rheumatoid arthritis—rheumatoid factor negative).** A patient has arthritis in five or more joints during the first 6 months of disease, and all tests for rheumatoid factor (proteins produced by the immune system that can attack healthy tissue, which are commonly found in rheumatoid arthritis and juvenile arthritis) are negative.
4. **Polyarthritis—rheumatoid factor positive (formerly known as polyarticular rheumatoid arthritis—rheumatoid factor positive).** A patient has arthritis in five or more joints during the first 6 months of the disease. Also, at least two tests for rheumatoid factor, at least 3 months apart, are positive.
5. **Psoriatic arthritis.** Patients have both arthritis and psoriasis (a skin disease), or they have arthritis

and at least two of the following:

- inflammation and swelling of an entire finger or toe (this is called dactylitis)
- nail pitting or splitting
- a first-degree relative with psoriasis.

6. **Enthesitis-related arthritis.** The enthesis is the point at which a ligament, tendon, or joint capsule attaches to the bone. If this point becomes inflamed, it can be tender, swollen, and painful with use. The most common locations are around the knee and at the Achilles tendon on the back of the ankle. Patients are diagnosed with this juvenile idiopathic arthritis subtype if they have both arthritis and inflammation of an enthesitis site, or if they have either arthritis or enthesitis with at least two of the following:

- inflammation of the sacroiliac joints (at the bottom of the back) or pain and stiffness in the lumbosacral area (in the lower back)
- a positive blood test for the human leukocyte antigen (HLA) *B27* gene
- onset of arthritis in males after age 6 years
- a first-degree relative diagnosed with ankylosing spondylitis, enthesitis-related arthritis, or inflammation of the sacroiliac joint in association with inflammatory bowel disease or acute inflammation of the eye.

7. **Undifferentiated arthritis.** A child is said to have this subtype of juvenile idiopathic arthritis if the arthritis manifestations do not fulfill the criteria for one of the other six categories or if they fulfill the criteria for more than one category.

What Causes Juvenile Arthritis?

Most forms of juvenile arthritis are autoimmune disorders, which means that the body's immune system—which normally helps to fight off bacteria or viruses—mistakenly attacks some of its own healthy cells and tissues. The result is inflammation, marked by redness, heat, pain, and swelling. Inflammation can cause joint damage. Doctors do not know why the immune system attacks healthy tissues in children who develop juvenile arthritis. Scientists suspect that it is a two-step process. First, something in a child's genetic makeup gives him or her a tendency to develop juvenile arthritis; then an environmental factor, such as a virus, triggers the development of the disease.

Not all cases of juvenile arthritis are autoimmune, however. Recent research has demonstrated that some people, such as many with systemic arthritis, have what is more accurately called an autoinflammatory condition. Although the two terms sound somewhat similar, the disease processes behind autoimmune and autoinflammatory disorders are different.

When the immune system is working properly, foreign invaders such as bacteria and viruses provoke the body to produce proteins called antibodies. Antibodies attach to these invaders so that they can be recognized and destroyed. In an autoimmune reaction, the antibodies attach to the body's own healthy tissues by mistake, signaling the body to attack them. Because they target the self, these proteins are called autoantibodies.

Like autoimmune disorders, autoinflammatory conditions also cause inflammation. And like autoimmune disorders, they also involve an overactive immune system. However, autoinflammation is not caused by autoantibodies. Instead, autoinflammation involves a more primitive part of the immune system that in healthy people causes white blood cells to destroy harmful substances. When this system goes awry, it causes inflammation for unknown reasons. In addition to inflammation, autoinflammatory diseases often cause fever and rashes.

What Are Its Symptoms and Signs?

The most common symptom of all types of juvenile arthritis is persistent joint swelling, pain, and stiffness that is typically worse in the morning or after a nap. The pain may limit movement of the affected joint, although many children, especially younger ones, will not complain of pain. Juvenile arthritis commonly affects the knees and the joints in the hands and feet. One of the earliest signs of juvenile arthritis may be limping in the morning because of an affected knee. Besides joint symptoms, children with systemic juvenile arthritis have a high fever and a skin rash. The rash and fever may appear and disappear very quickly. Systemic arthritis also may cause the lymph nodes located in the neck and other parts of the body to swell. In some cases (fewer than half), internal organs including the heart and (very rarely) the lungs, may be involved.

Eye inflammation is a potentially severe complication that commonly occurs in children with oligoarthritis but can also be seen in other types of juvenile arthritis. All children with juvenile arthritis need to have regular eye exams, including a special exam called a slit lamp exam. Eye diseases such as iritis or uveitis can be present at the beginning of arthritis but often develop some time after a child first develops juvenile arthritis. Very commonly, juvenile arthritis-associated eye inflammation does not cause any symptoms and is found only by performing eye exams.

Typically, there are periods when the symptoms of juvenile arthritis are better or disappear (remissions) and times when symptoms "flare," or get worse. Juvenile arthritis is different in each child; some may have just one or two flares and never have symptoms again, while others may experience many flares or even have symptoms that never go away.

Some children with juvenile arthritis have growth problems. Depending on the severity of the disease and the joints involved, bone growth at the affected joints may be too fast or too slow, causing one leg or arm to be longer than the other, for example, or resulting in a small or misshapen chin. Overall growth also may be slowed. Doctors are exploring the use of growth hormone to treat this problem. Juvenile arthritis may also cause joints to grow unevenly.

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