

QUESTIONS AND ANSWERS ABOUT JUVENILE RHEUMATOID ARTHRITIS

What Is Arthritis?

Arthritis means joint inflammation, and refers to a group of diseases that cause pain, swelling, stiffness and loss of motion in the joints. "Arthritis" is often used as a more general term to refer to 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. Children can develop almost all types of arthritis that affect adults, but the most common type of arthritis that affects children is juvenile rheumatoid arthritis.

What Is Juvenile Rheumatoid Arthritis?

Juvenile rheumatoid arthritis (JRA) is arthritis that causes joint inflammation and stiffness for more than 6 weeks in a child of 16 years of age or less. Inflammation causes redness, swelling, warmth, and soreness in the joints, although many children with JRA do not complain of joint pain. Any joint can be affected and inflammation may limit the mobility of affected joints.

Doctors classify three kinds of JRA by the number of joints involved, the symptoms, and the presence or absence of certain antibodies in the blood. (Antibodies are special proteins made by the immune system.) These classifications help the doctor determine how the disease will progress.

- **Pauciarticular** (*paw-see-are-tick-you-lar*): Pauciarticular means that four or fewer joints are affected. Pauciarticular is the most common form of JRA; about half of all children with JRA have this type. Pauciarticular disease typically affects large joints, such as the knees. Girls under age 8 are most likely to develop this type of JRA.

Some children have special proteins in the blood called antinuclear antibodies (ANAs). Eye disease affects about 20 to 30 percent of children with pauciarticular JRA. Up to 80 percent of those with eye disease also test positive for ANA and the disease tends to develop at a particularly early age in these children. Regular examinations by an ophthalmologist (a doctor who specializes in eye diseases) are necessary to prevent serious eye problems such as iritis (inflammation of the iris) or uveitis (inflammation of the inner eye, or uvea). Many children with pauciarticular disease outgrow arthritis by adulthood,

although eye problems can continue and joint symptoms may recur in some people.

- **Polyarticular:** About 30 percent of all children with JRA have polyarticular disease. In polyarticular disease, five or more joints are affected. The small joints, such as those in the hands and feet, are most commonly involved, but the disease may also affect large joints. Polyarticular JRA often is symmetrical, that is, it affects the same joint on both sides of the body. Some children with polyarticular disease have a special kind of antibody in their blood called IgM rheumatoid factor (RF). These children often have a more severe form of the disease, which doctors consider to be the same as adult rheumatoid arthritis.
- **Systemic:** Besides joint swelling, the systemic form of JRA is characterized by fever and a light pink rash, and may also affect internal organs such as the heart, liver, spleen, and lymph nodes. Doctors sometimes call it Still's disease. Almost all children with this type of JRA test negative for both RF and ANA. The systemic form affects 20 percent of all children with JRA. A small percentage of these children develop arthritis in many joints and can have severe arthritis that continues into adulthood.

How Is Juvenile Rheumatoid Arthritis Different From Adult Rheumatoid Arthritis?

The main difference between juvenile and adult rheumatoid arthritis is that many people with JRA outgrow the illness, while adults usually have lifelong symptoms. Studies estimate that by adulthood, JRA symptoms disappear in more than half of all affected children. Additionally, unlike rheumatoid arthritis in an adult, JRA may affect bone development as well as the child's growth.

Another difference between JRA and adult rheumatoid arthritis is the percentage of people who are positive for RF. About 70 to 80 percent of all adults with rheumatoid arthritis are positive for RF, but fewer than half of all children with rheumatoid arthritis are RF positive. Presence of RF indicates an increased chance that JRA will continue into adulthood.

What Causes Juvenile Rheumatoid Arthritis?

JRA is an autoimmune disorder, which means that the body mistakenly identifies some of its own cells and tissues as foreign. The immune system, which normally helps to fight off harmful, foreign substances such as bacteria or viruses, begins to attack healthy cells and tissues. The result is inflammation—marked by redness, heat, pain, and swelling. Doctors do not know why the immune system goes awry in children who develop JRA. Scientists suspect that it is a two-step process. First something in a child's

genetic makeup gives them a tendency to develop JRA; and then an environmental factor, such as a virus, triggers the development of JRA.

What Are the Symptoms and Signs of Juvenile Rheumatoid Arthritis?

The most common symptom of all types of JRA is persistent joint swelling, pain, and stiffness that typically is 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. JRA commonly affects the knees and joints in the hands and feet. One of the earliest signs of JRA may be limping in the morning because of an affected knee. Besides joint symptoms, children with systemic JRA have a high fever and a light pink rash. The rash and fever may appear and disappear very quickly. Systemic JRA also may cause the lymph nodes located in the neck and other parts of the body to swell. In some cases (less than half), internal organs including the heart, and very rarely, the lungs may be involved.

Eye inflammation is a potentially severe complication that sometimes occurs in children with pauciarticular JRA. Eye diseases such as iritis and uveitis often are not present until some time after a child first develops JRA.

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

Does Juvenile Rheumatoid Arthritis Affect Physical Appearance?

Some children with JRA may look different because they have growth problems. Depending on the severity of the disease and the joints involved, growth in affected joints may be too fast or too slow, causing one leg or arm to be longer than the other. Overall growth may also be slowed. Doctors are exploring the use of growth hormones to treat this problem. JRA also may cause joints to grow unevenly or to one side.

Children with JRA also may look different because of medication. Corticosteroids, a type of medication sometimes used to treat JRA, can result in weight gain and a round face. When the doctor stops giving the medication, these side effects may disappear.

How Is Juvenile Rheumatoid Arthritis Diagnosed?

Doctors usually suspect JRA, along with several other possible conditions, when they see children with persistent joint pain or swelling, unexplained skin rashes and fever, or swelling of lymph nodes or inflammation of internal organs. A diagnosis of JRA also is considered in children with an unexplained limp or excessive clumsiness.

No one test can be used to diagnose JRA. A doctor diagnoses JRA by carefully examining the patient and considering the patient's medical history and the results of laboratory tests that help rule out other conditions.

- **Symptoms:** One important consideration in diagnosing JRA is the length of time that symptoms have been present. Joint swelling or pain must last for at least 6 weeks for the doctor to consider a diagnosis of JRA. Because this factor is so important, it may be useful to keep a record of the symptoms, when they first appeared, and when they are worse or better.
- **Laboratory Tests:** Laboratory tests, usually blood tests, cannot by themselves provide the doctor with a clear diagnosis. But these tests can be used to help rule out other conditions and to help classify the type of JRA that a patient has. Blood may be taken to test for RF or ANA, and to determine the erythrocyte sedimentation rate (ESR).
 - ANA is found in the blood more often than RF, and both are found in only a small portion of JRA patients. The RF test helps the doctor tell the difference among the three types of JRA.
 - ESR is a test that measures how quickly red blood cells fall to the bottom of a test tube. Some people with rheumatic disease have an elevated ESR or "sed rate" (cells fall quickly to the bottom of the test tube), showing that there is inflammation in the body. Not all children with active joint inflammation have an elevated ESR.
- **X Rays:** X rays are needed if the doctor suspects injury to the bone or unusual bone development. Early in the disease, some x rays can show cartilage damage. In general, x rays are more useful later in the disease, when bones may be affected.
- **Other diseases:** Because there are many causes of joint pain and swelling, the doctor must rule out other conditions before diagnosing JRA. These include physical injury, bacterial infection, Lyme disease, inflammatory bowel disease, lupus, dermatomyositis, and some forms of cancer. The doctor may use additional laboratory tests to help rule out these and other possible conditions.

Who Treats Juvenile Rheumatoid Arthritis? What Are the Treatments?

A pediatrician, family physician, or other primary care doctor frequently manages the treatment of a child with JRA, often with the help of other doctors. Depending on the patient's and parents' wishes and the severity of the disease, the team of doctors may include pediatric rheumatologists (doctors specializing in childhood arthritis), ophthalmologists (eye doctors),

orthopaedic surgeons (bone specialists), and physiatrists (rehabilitation specialists), as well as physical and occupational therapists.

The main goals of treatment are to preserve a high level of physical and social functioning and maintain a good quality of life. To achieve these goals, doctors recommend treatments to reduce swelling; maintain full movement in the affected joints; relieve pain; and identify, treat, and prevent complications. Most children with JRA need medication and physical therapy to reach these goals.

Several types of medication are available to treat JRA:*

*Brand names included in this fact sheet are provided as examples only, and their inclusion does not mean that these products are endorsed by the National Institutes of Health or any other Government agency. Also, if a particular brand name is not mentioned, this does not mean or imply that the product is unsatisfactory.

- **Nonsteroidal anti-inflammatory drugs (NSAIDs):** Aspirin, ibuprofen (Motrin, Advil, Nuprin) and naproxen or naproxen sodium (Naprosyn, Aleve) are examples of NSAIDs. They often are the first type of medication used. Most doctors do not treat children with aspirin because of the possibility that it will cause bleeding problems, stomach upset, liver problems, or Reye's syndrome. But for some children, aspirin in the correct dose (measured by blood test) can control JRA symptoms effectively with few serious side effects.

If the doctor prefers not to use aspirin, other NSAIDs are available. For example, in addition to those mentioned above, diclofenac and tolmetin are available with a doctor's prescription. Studies show that these medications are as effective as aspirin with fewer side effects. An upset stomach is the most common complaint. Any side effects should be reported to the doctor, who may change the type or amount of medication.

- **Disease-modifying anti-rheumatic drugs (DMARDs):** If NSAIDs do not relieve symptoms of JRA, the doctor is likely to prescribe this type of medication. DMARDs slow the progression of JRA, but because they take weeks or months to relieve symptoms, they often are taken with an NSAID. Various types of DMARDs are available. In the past, doctors prescribed hydroxychloroquine, oral and injectable gold, sulfasalazine, and d-penicillamine; however, doctors are now much more likely to use methotrexate for children with JRA.
- **Methotrexate:** Researchers have learned that this type of DMARD is safe and effective for some children with rheumatoid arthritis whose symptoms are not relieved by other medications. Because only small doses of methotrexate are needed to relieve arthritis symptoms,

potentially dangerous side effects rarely occur. The most serious complication is liver damage, but it can be avoided with regular blood screening tests and doctor followup. Careful monitoring for side effects is important for people taking methotrexate. When side effects are noticed early, the doctor can reduce the dose and eliminate side effects.

- **Corticosteroids:** In children with very severe JRA, stronger medicines may be needed to stop serious symptoms such as inflammation of the sac around the heart (pericarditis). Corticosteroids like prednisone may be added to the treatment plan to control severe symptoms. This medication can be given either intravenously (directly into the vein) or by mouth. Corticosteroids can interfere with a child's normal growth and can cause other side effects, such as a round face, weakened bones, and increased susceptibility to infections. Once the medication controls severe symptoms, the doctor may reduce the dose gradually and eventually stop it completely. Because it can be dangerous to stop taking corticosteroids suddenly, it is important that the patient carefully follow the doctor's instructions about how to take or reduce the dose.

In addition to medications, physical therapy is an important part of a child's treatment plan. Exercise can help to maintain muscle tone and preserve and recover the range of motion of the joints. A physical therapist can design an appropriate exercise program for a person with JRA. The physical therapist also may recommend using splints and other devices to keep joints growing evenly.

How Can the Family Help a Child Live Well With JRA?

JRA affects the entire family who must cope with the special challenges of this disease. JRA can strain a child's participation in social and after-school activities and make school work more difficult. There are several things that family members can do to help the child do well physically and emotionally.

- Treat the child as normally as possible.
- Ensure that the child receives appropriate medical care and follows the doctor's instructions. Many treatment options are available, and because JRA is different in each child, what works for one may not work for another. If the medications that the doctor prescribes do not relieve symptoms or if they cause unpleasant side effects, patients and parents should discuss other choices with their doctor. A person with JRA can be more active when symptoms are controlled.
- Encourage exercise and physical therapy for the child. For many young people, exercise and physical therapy play important roles in treating JRA. Parents can arrange for children to participate in activities that

the doctor recommends. During symptom-free periods, many doctors suggest playing team sports or doing other activities to help keep the joints strong and flexible and to provide play time with other children and encourage appropriate social development.

- Work closely with the school to develop a suitable lesson plan for the child and to educate the teacher and the child's classmates about JRA. (See the end of this fact sheet for information about [Kids on the Block, Inc.](#), a program that uses puppets to illustrate how juvenile arthritis can affect school, sports, friends, and family.) Some children with JRA may be absent from school for prolonged periods and need to have the teacher send assignments home. Some minor changes such as an extra set of books, or leaving class a few minutes early to get to the next class on time can be a great help. With proper attention, most children progress normally through school.
- Explain to the child that getting JRA is nobody's fault. Some children believe that JRA is a punishment for something they did.
- Consider joining a support group. The American Juvenile Arthritis Organization runs support groups for people with JRA and their families. Support group meetings provide the chance to talk to other young people and parents of children with JRA and may help a child and the family cope with the condition.

Do Children With Juvenile Rheumatoid Arthritis Have To Limit Activities?

Although pain sometimes limits physical activity, exercise is important to reduce the symptoms of JRA and maintain function and range of motion of the joints. Most children with JRA can take part fully in physical activities and sports when their symptoms are under control. During a disease flare, however, the doctor may advise limiting certain activities depending on the joints involved. Once the flare is over, a child can start regular activities again.

Swimming is particularly useful because it uses many joints and muscles without putting weight on the joints. A doctor or physical therapist can recommend exercises and activities.

What Are Researchers Trying To Learn About Juvenile Rheumatoid Arthritis?

Scientists are investigating the possible causes of JRA. Researchers suspect that both genetic and environmental factors are involved in development of the disease and they are studying these factors in detail. To help explore the role of genetics, the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS) has established a research registry for families in

which two or more siblings have JRA. NIAMS also funds a Multipurpose Arthritis and Musculoskeletal Diseases Center (MAMDC) that specializes in research on pediatric rheumatic diseases including JRA.

Research doctors are continuing to try to improve existing treatments and find new medicines that will work better with fewer side effects. For example, researchers are studying the long-term effects of the use of methotrexate in children.

Where Can People Get More Information About the Research Registry and MAMDC?

For more information about the Research Registry, contact:

Edward Giannini, MD
Children's Hospital Medical Center - PAV 2-129
University of Cincinnati, College of Medicine
Cincinnati, OH 45229
513/636-7634 or 513/636-4495
E-mail address: btague@one.net
World Wide Web address: <http://www.jraregistry.org>

For information about the MAMDC, contact:

David Glass, MD
Children's Hospital Medical Center - PAV 2-129
University of Cincinnati, College of Medicine
Cincinnati, OH 45229-2899
513/636-8854
E-mail address: glasd0@chmcc.org
World Wide Web address: <http://www.cinciMAMDC.org>

Where Can People Get More Information About Juvenile Rheumatoid Arthritis?

- **American Juvenile Arthritis Organization (AJAO)**
1330 West Peachtree Street
Atlanta, GA 30309
404/872-7100
800/283-7800
World Wide Web address: <http://www.arthritis.org>
- **National Institute of Arthritis and Musculoskeletal and Skin Diseases Information Clearinghouse**
National Institutes of Health
1 AMS Circle
Bethesda, MD 20892-3675
301/495-4484 Fax: 301/587-4352 TTY: 301/565-2966

NIAMS Fast Facts: health information available 24 hours a day by fax,

call 301/881-2731 from a fax machine telephone.
World Wide Web address: <http://www.nih.gov/niams>

National Institute of Arthritis and Musculoskeletal and Skin Diseases Information Clearinghouse is a public service sponsored by the NIAMS that provides health information devoted to childhood rheumatic diseases. The organization has information about JRA, support groups, and pediatric rheumatology centers around the country.

- **Kids on the Block, Inc.**
9385-C Gerwig Lane
Columbia, MD 21046
410/290-9095
800/368-KIDS (5437)

Kids on the Block, Inc., is an educational program that uses puppets to show how JRA can affect school, sports, friends, and family. A package is available (for a fee) that includes a set of large puppets that represent a diverse group of children, as well as audiocassettes, a training guide, four different program scripts, props, followup activities, and other resources. The program is designed so that anyone can be puppeteer, and workshops to train puppeteers are available.

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The National Arthritis and Musculoskeletal and Skin Diseases Information Clearinghouse (NAMSIC) is a public service sponsored by the NIAMS that provides health information and information sources. The NIAMS, a part of the National Institutes of Health (NIH), leads the Federal medical research effort in arthritis and musculoskeletal and skin diseases. The NIAMS sponsors research and research training throughout the United States as well as on the NIH campus in Bethesda, MD, and disseminates health and research information.

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