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**Spondylitis  
Association  
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**JUVENILE**



## **JUVENILE Spondyloarthritis**

- ❖ Ankylosing Spondylitis
- ❖ Enteropathic Arthritis
- ❖ Psoriatic Arthritis
- ❖ Reactive Arthritis
- ❖ Undifferentiated Spondyloarthritis
- ❖ Juvenile Spondyloarthritis

In this brochure, you will learn about juvenile spondyloarthritis. The following pages present the primary signs and symptoms of this form of arthritis in children and teens, and the various treatments and techniques to manage the condition. Since knowledge is key to successful management of spondyloarthritis, we would suggest that you share this booklet with other adults who play an important role in your child's life, including educators and school counselors.



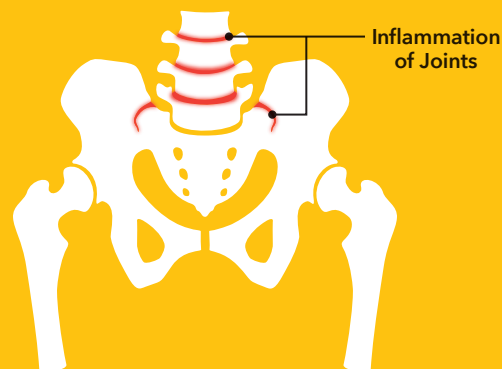
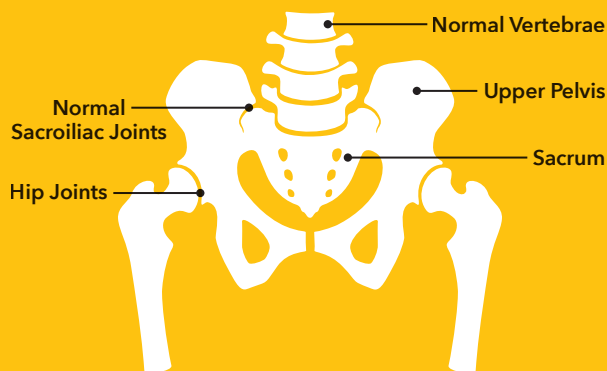
**Information in this brochure cannot replace treatment provided by health care professionals. If you have questions as you read, you may wish to consult further with your doctor.**

## What is it?

Spondyloarthritis is the name given to a group of closely related rheumatic diseases that primarily affect the sacroiliac joints between the pelvis and the spine (backbone), as well as joints along the spine. The sacroiliac joint is located where the spine attaches to the pelvic bones. It can also affect other joints such as knees, ankles, toes, hips, shoulders, and the ribcage. When it occurs before age seventeen it is called juvenile spondyloarthritis (JSpA).

**Juvenile spondyloarthritis encompasses the following closely related conditions:**

- ❖ Juvenile ankylosing spondylitis (JAS)
- ❖ Juvenile psoriatic arthritis (JPsa)
- ❖ Juvenile inflammatory bowel disease associated (enteropathic) arthritis (JEnA)
- ❖ Juvenile reactive arthritis (JReA)
- ❖ Juvenile undifferentiated spondyloarthritis (JuSpA)



JSpA accounts for about 20% of all types of childhood arthritis, and is more commonly found in boys. This is the opposite of most other forms of childhood arthritis (i.e. juvenile idiopathic arthritis, or JIA) which are more commonly found in girls.



Childhood arthritis in whole impacts up to 4 in 1,000 kids, making JSpA relatively rare. JSpA usually begins in the pre-teen or teen years, generally after the onset of puberty. While the cause is unknown, we do know that children who inherit a gene called HLA-B27 from one of their parents are more likely to develop the illness than other children. Since HLA-B27 is also found in about 8% of Caucasian children who will never develop JSpA, it cannot be used to make a diagnosis. Moreover, we know that many other genes contribute to the complicated picture of who will develop the condition. Sometimes symptoms are triggered after an infection of the intestinal or genitourinary tract, although in many cases there is no previous infection.

## What Are the Symptoms?

In children, spondyloarthritis usually starts in a leg joint such as the ankle or knee, or in the hip. It may take months or sometimes years for other joints to be affected, particularly the spine or the sacroiliac joints. In each person, the disease can behave somewhat differently. Sometimes the first symptom of JSpA is enthesitis, which is pain and tenderness due to inflammation in the ligaments and tendons where they attach to the bone. In this case the pain is not in the joint (as it is in JIA), but instead occurs near or around the joint. In children, enthesitis is most common under the heel on the bottom of the foot, behind the heel (Achilles tendon), under the toes where they attach to the foot, or around the kneecap. About 20% of juvenile ankylosing spondylitis (JAS) patients develop inflammation in the eye called uveitis (or acute anterior uveitis) or iritis. When this occurs in the setting of JAS, it usually causes pain, redness, and sensitivity to light and can occur in one or both eyes at the same time. Other complications of JAS such as damage to the heart valves is very rare.





## How is JSpA Diagnosed?

The diagnosis of JSpA is made on the basis of signs and symptoms of the disease and imaging of the sacroiliac or other joints that show characteristic abnormalities. Physicians experienced in the care of children with JSpA and other types of arthritis (pediatric rheumatologists) will ask detailed questions about local and generalized pain and stiffness, particularly whether it occurs in the back, joint symptoms, heel pain, eye problems, and other symptoms. They will do a careful examination looking for arthritis including sacroiliitis, enthesitis, loss of mobility in joints and the spine, and reduced chest expansion. Strict diagnostic criteria for JAS specifically include imaging studies (such as MRI of the sacroiliac joints that show inflammation and/or joint damage).



However, when some of the signs and symptoms of JAS are present, but not enough to make a definite diagnosis, it is possible that a diagnosis of JSpA can be made. This is sometimes referred to as 'Undifferentiated SpA' or 'Enthesitis-related arthritis,' and usually means that the patient hasn't developed the abnormal imaging findings in the sacroiliac joints. There are no specific laboratory tests for JAS or JSpA, but certain tests can be very helpful in determining the presence of risk factors such as the HLA-B27 gene, or measuring how active the disease is. Blood tests show that children with JSpA only very rarely have antinuclear antibodies (ANA) or rheumatoid factor (RF), which are typical in

some other types of juvenile idiopathic arthritis. The HLA-B27 gene is present in approximately 90% of Caucasian patients with JAS, but only about 40% of African-Americans with this disease. The HLA-B27 test is helpful, but it is not sufficient to make a diagnosis of JAS or JSpA, as it is also found in 7-8% of healthy people.

Many pediatric patients start out with a diagnosis of JSpA and eventually develop JAS (or AS as adults), but this is not always the case. It is still unclear exactly what proportion of patients with JSpA will develop AS, but it is probably close to 50%.

Sometimes JAS/JSpA can look like other diseases, and doctors must take care to make a correct diagnosis. For example, in young children JSpA sometimes looks a lot like JIA. However, since JIA is more common in girls, often associated with a positive antinuclear antibody test, and rarely involves enthesitis, these features can frequently be used to help make the correct diagnosis.

There are also certain conditions that tend to occur with JSpA such as inflammatory bowel disease (Crohn's disease and ulcerative colitis) and psoriasis (severe skin rash). These may occur in children as well as adults.





## How is JSpA Treated?

Treatment of children with JSpA is individualized according to how severe the disease is, what joints are affected, and whether there are complications. Pediatric rheumatologists are trained in the diagnosis and management of children with JSpA. However, if one is not available, an adult rheumatologist may be appropriate, particularly when the patient is older. In general, a five-part approach is used.



- 1. EDUCATION AND COUNSELING.** Children with JSpA and their families must understand that there is no cure for this disease, but that treatments continue to improve. Also, the disease symptoms may wax and wane over time, and can sometimes be mild. This makes it very difficult to evaluate the effectiveness of treatments. It is also important for patients with JSpA and their families to understand the risks and potential complications of treatment. Finally, it is important to anticipate and evaluate the psychological effects of the illness on the child and the family. Families may over protect children with JSpA and treat them as more ill than necessary.
- 2. MEDICATION.** The first type of medication used for patients with JSpA is usually a non-steroidal anti-inflammatory drug (NSAID). These drugs include naproxen (Naprosyn), ibuprofen (Advil), tolmetin (Tolectin), and in older patients, indomethacin (Indocin) or diclofenac (Voltaren); other

NSAIDS are sometimes used as well. Most pediatric rheumatologists agree that indomethacin is a particularly effective NSAID for JSpA. However, its side effects can include headaches, dizziness, stomachaches, or other nervous system effects, and therefore it may not be suitable for younger children and may not be tolerated even in teenagers. It is generally safe for children age 14 and older who recognize these adverse symptoms, and can stop taking the medication if they occur. All NSAIDS may cause gastrointestinal discomfort or pain, and occasionally ulcers when taken chronically. Children seem less susceptible to these problems than adults, but still should be closely watched and have routine blood tests. These medications should be taken with food in order to minimize stomach distress. Medications that help protect the gastrointestinal tract (such as ranitidine or proton pump inhibitors) can be prescribed if needed to control symptoms.

Newer medications known as 'biologics' block the effects of TNF- $\alpha$ , a chemical the body produces that increases inflammation. Known as TNF inhibitors, this class of medication has been shown to be highly effective in reducing or sometimes even eliminating the symptoms of SpA. We also know that they are relatively safe medications in children because they have been used for decades now in children with JIA, as well as more recently in JSpA. The main risk for people taking a biologic is that they will develop a serious infection. This makes it important for doctors to screen for certain infections before starting a biologic.

Other medications that are sometimes effective include sulfasalazine and its derivatives. Oral corticosteroids like prednisone are rarely used in JSpA, although they may be needed to control eye inflammation. Topical steroids are used to treat the eye disease of JSpA. Injection of steroids into an inflamed joint that has not responded to NSAID therapy may be highly effective.



Parents are often bombarded by stories from acquaintances, in magazines, or on the internet, of arthritis responding to various herbs. Although herbs may well contain very potent chemicals, there are no data about the effectiveness of herbs in treating arthritis, including JSpA, in children. Even more concerning is that it is not known whether these herbs are safe in children, or whether any serious interactions might occur with other medications the child is taking.

**3. PHYSICAL AND OCCUPATIONAL THERAPY.** The child with JSpA must strive to maintain range of motion in affected joints and strengthen weakened muscles in the back, abdomen, and limbs. The well-designed therapy program will involve daily activities designed to promote normal function. The three areas of emphasis are stretching, posture, and regular daily exercise. Basic stretching exercises for range of motion in the chest, back and other joints may be prescribed depending upon the specific joints and ligaments affected. Children with JAS may experience rounding of their shoulders over time, with the head being gradually thrust forward. Attention to posture will keep the head, shoulders, and back well aligned, reduce pain and fatigue, and promote effective breathing patterns.

Children with JSpA should be encouraged to participate in physical activities appropriate for their age. Regular daily activities such as walking, bicycle riding, and swimming should be encouraged, but it is important to know that these activities are not a substitute for prescribed physical therapy, particularly when the disease has already affected mobility. Decisions to pursue more vigorous physical activity such as contact sports should be made after consultation with a pediatric rheumatologist and physical therapist.

Children with JSpA can also benefit from special equipment. For example, shoe inserts may help relieve heel and foot pain, and splints for affected joints help prevent and/or treat joint contractures.





- 4. SURGERY.** Surgery for joint problems is not usually necessary for the child with JSpA. In some cases, however, a needle may need to be put into the joint space in order to remove excess fluid and test for infection, or to put medication directly in the joint. In very rare cases, there may be enough destruction in a knee or hip joint to require a joint replacement to improve function or reduce severe pain.
- 5. COMFORT MEASURES.** If NSAIDs and other anti-inflammatory medications are not adequate for pain relief, often heat and/or acetaminophen can be beneficial. Stronger pain medications are only rarely needed. In addition, because children with back arthritis may experience muscle spasm, massage may be useful, and occasionally muscle relaxants. Acupuncture is not usually recommended for children.

## Who Can Help?

Ideally, a team of health professionals is best suited to take care of children with JSpA. They will assist the pediatrician or family doctor in the evaluation and management of the disease.

### Members of the team include:

#### **PEDIATRIC RHEUMATOLOGIST –**

The pediatric rheumatologist is a physician with highly specialized training and experience in the care of children with rheumatic diseases like JSpA. In the United States, most pediatric rheumatologists are found at university medical centers and medical schools.

**NURSE –** In most medical centers, the health care team includes a nurse with special experience in the care of children with arthritis. The nurse generally has a central role in educating the child, the family, and often the child's school about JSpA.



**PHYSICAL THERAPIST –** The physical therapist will evaluate joint motion, strength, and posture. The therapist also has the responsibility to develop exercise programs for the child to be carried out at home or with a local therapist. Most physical therapists also maintain contact with physical education teachers in the school system, so that the most appropriate program will be undertaken in school for the child with JSpA.

**OCCUPATIONAL THERAPIST –** A major focus of the occupational therapists is to ensure that the child can perform well physically in school and with daily activities. In addition, the occupational therapist will teach the child and the family how to adapt to activities of daily living so that the child with severe joint disease can still function independently.

**SOCIAL WORKER OR PSYCHOLOGIST –** The potential stress of JSpA on the child and the family should never be taken lightly. Children may perceive themselves as different from their peers even if their disease is not severe. They may be even more upset if JSpA limits their activities significantly. Therefore, children and families should have ready access to psychological experts, such as child psychologists and social workers, who have experience in the evaluation and care of chronically ill children.





**NUTRITIONIST** – Good nutrition is critically important for any child with a chronic illness. Children with JSpA are at risk for becoming overweight because of decreased physical activity. Unfortunately, extra weight can also worsen the symptoms of the illness by producing more stress on affected joints.



**OTHER PHYSICIANS** – During the course of care of JSpA, children may require the services of a number of other physicians. This group might include the ophthalmologist, who would generally see the child for acute eye pain and redness. The orthopedic surgeon might become involved to evaluate and treat a very severely affected joint, particularly the hip. Doctors who specialize in rehabilitative care can assist with reaching functional goals through prescribing more intensive fitness programs, and/or specialized equipment.



## What About School?

Children with JSpA and their families should be aware that the disease may affect school life. Parents and teachers can help plan appropriate educational goals and activities to maintain as normal a routine as possible.

Teachers may be helpful in watching for the side effects of medication. The physician must watch for such problems and work to come up with medications that will not impair the child's school performance.

Children with JSpA may experience some difficulty in dealing with other children. They may need help in learning how to handle being different and the stress of any special activities such as getting up to stretch during class, or taking medication during the school day. Special preparation for physical education may also be required. For example, a child with JSpA who has been sitting all day may need to do stretches before participating in physical education.

Finally, fatigue may be a factor, decreasing the child's ability to do homework. Careful attention to a balanced schedule of school, exercise, and rest can help manage fatigue.





## What's in the Future?

JSpA is a chronic disease. It can last months or years with periods of remission when the patient may have little or no symptoms. It often persists into adulthood, and even children with limited symptoms initially such as enthesitis, may develop back problems as adults. Treatment is aimed at reducing symptoms, preventing loss of function, and preserving healthy joints. With comprehensive therapy, most persons with JSpA will continue to lead highly productive lives.

### **The Spondylitis Association of America™**

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The Spondylitis Association of America was the first, and remains the largest, resource in the U.S. for people affected by spondyloarthritis. For more than 35 years, SAA has dedicated all of its resources to funding medical research, education, advocacy, and supportive programs and services that directly benefit the spondylitis community.

By joining SAA you gain access to tools that will improve your own quality of life while also making a difference for the 2.7 million affected people throughout the nation. Join today and receive:

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- ❖ The satisfaction of knowing that you are part of an extraordinary community of patients, friends, family, and healthcare professionals dedicated to finding the cure!

There are over 100 types of arthritis. At SAA, we focus on one – yours. So that no one has to face spondylitis alone.

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