Juvenile Spondyloarthritis: An Updated Overview

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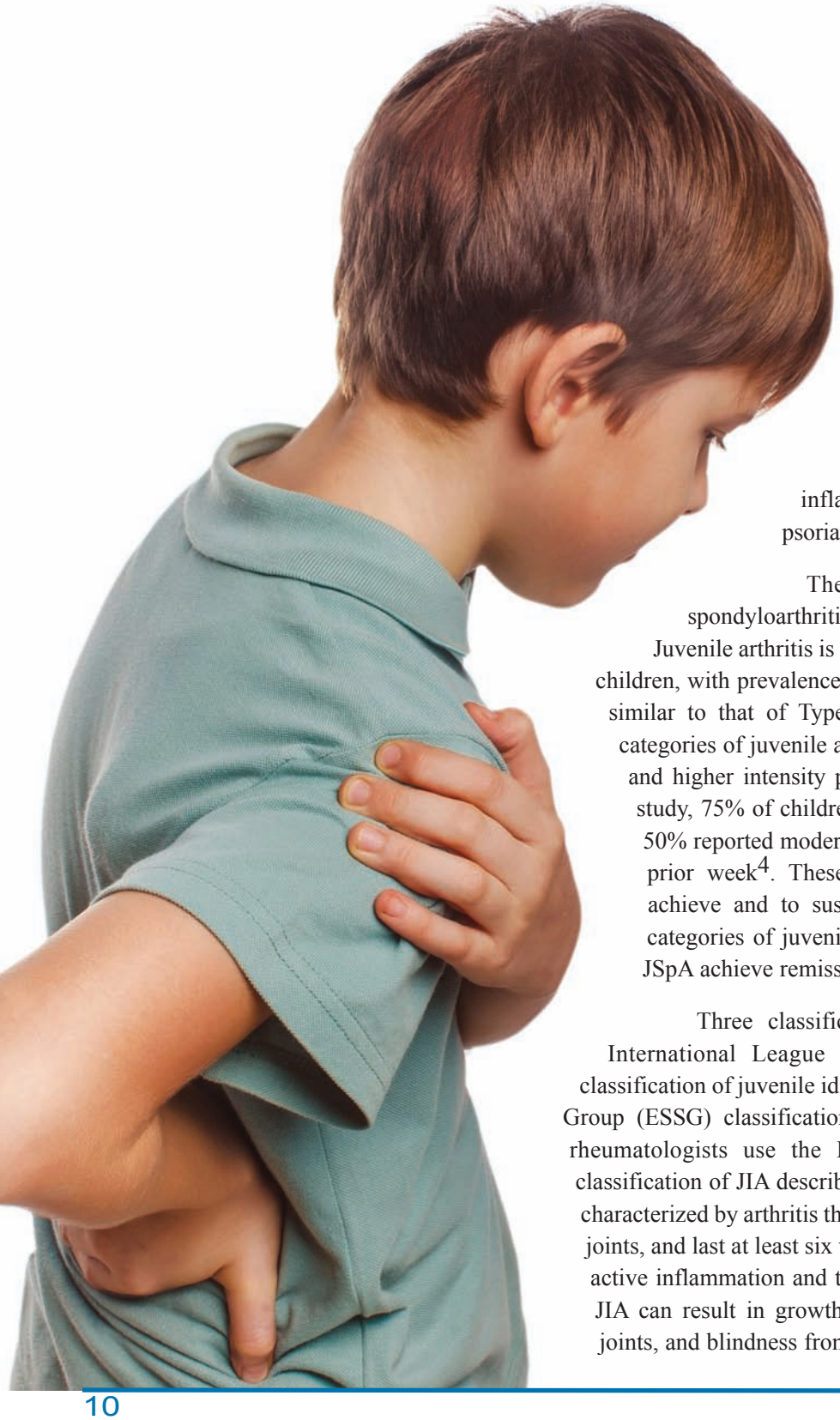
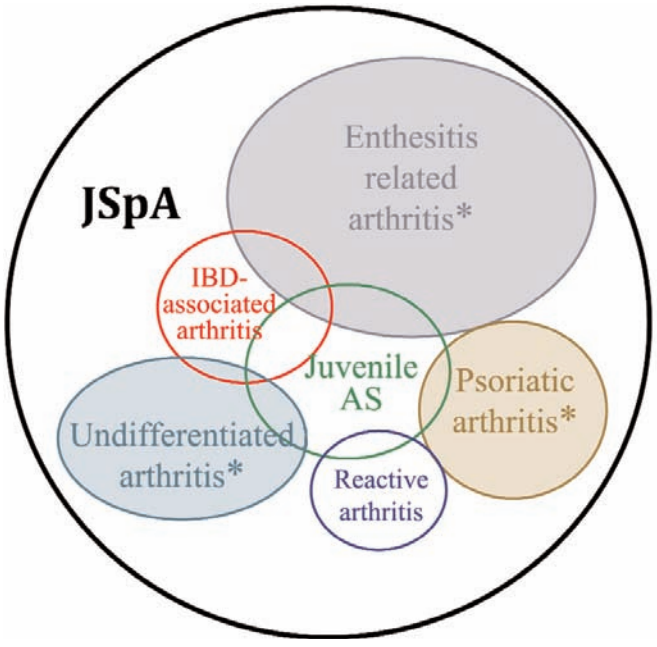


Spondyloarthritis (SpA) is a group of chronic inflammatory conditions characterized by arthritis, enthesitis, dactylitis (sausage-like swelling of the fingers or toes), acute and painful eye inflammation, HLA-B27 positivity, inflammatory back pain, and sacroiliitis. The term SpA encompasses ankylosing spondylitis (AS), undifferentiated SpA, inflammatory bowel disease associated arthritis, psoriatic arthritis, and reactive arthritis.

The term juvenile SpA (JSpA) refers to spondyloarthritis that starts during childhood (before age 16). Juvenile arthritis is the most common rheumatologic disease among children, with prevalence estimates ranging from 1-4 per 1,000 children, similar to that of Type I diabetes mellitus. In comparison to other categories of juvenile arthritis, children with JSpA have more frequent and higher intensity pain as well as poorer health status[1-3]. In one study, 75% of children with JSpA had moderate or severe pain, and 50% reported moderate or severe impairment of well-being over the prior week[4]. These children and adolescents are less likely to achieve and to sustain disease remission than those with other categories of juvenile arthritis[5,6]. Less than 20% of children with JSpA achieve remission within five years of diagnosis[7].

Three classification systems used for JSpA include: The International League of Associations for Rheumatology (ILAR) classification of juvenile idiopathic arthritis (JIA); the European SpA Study Group (ESSG) classification; and the Amor criteria. Of these, pediatric rheumatologists use the ILAR classification most often. The ILAR classification of JIA describes a clinically heterogeneous group of diseases characterized by arthritis that begin before age sixteen, involve one or more joints, and last at least six weeks. The goals of JIA treatment are to control active inflammation and to prevent long-term damage. Poorly controlled JIA can result in growth disturbances, loss of range of motion of the joints, and blindness from chronic eye inflammation.



*** = Also a JIA Category**

Most children with JSpA fall into the categories of enthesitis related arthritis (ERA), psoriatic arthritis, and undifferentiated arthritis. *Editor's note: These three conditions fall under both the JIA and JSpA classifications. (See above diagram)*

• Enthesitis-related arthritis is diagnosed in children who have arthritis and enthesitis *or* either arthritis or enthesitis plus at least one of the following: lower back pain or sacroiliac tenderness, HLA-B27 positivity, a parent with a history of SpA, onset of arthritis in a male older than six years, or acute and painful eye inflammation.

• Children with psoriatic arthritis have arthritis and psoriasis *or* arthritis plus at least two of the following three characteristics: nail pitting or onycholysis (separation of the nail from the nailbed), sausage-like swelling of the fingers or toes, or a parent with psoriasis.

• Children who are categorized as having undifferentiated arthritis don't fulfill any of the JSpA *or* JIA categories, or fulfill criteria of more than one category.

Other conditions not explicitly accounted for in the ILAR JIA classification but that are traditionally thought of as JSpA include: juvenile ankylosing spondylitis (JAS), reactive arthritis, and inflammatory bowel disease associated arthritis.

• JAS is AS that starts prior to age 16; 10-20% of adults with AS have symptom onset during childhood.

• Reactive arthritis is arthritis that occurs following a gastrointestinal infection; reactive arthritis can be a singular event or may progress to chronic JSpA. The classic triad of painful urination, painful eyes, and arthritis seen in adults is much less common in children.

• Inflammatory bowel disease associated arthritis is also considered part of the SpA group of diseases; as many as one-quarter of children and adolescents with inflammatory bowel disease develop arthritis

Other categories of JIA *not* considered under the umbrella of JSpA include: Oligoarthritis (arthritis in four or fewer joints); rheumatoid factor positive and rheumatoid factor negative polyarticular arthritis; (arthritis in five or more joints in the presence or absence of rheumatoid factor); and systemic arthritis (arthritis with a characteristic fever pattern and salmon colored rash, often associated with full body inflammation).

JIA categories (non overlapping)

1. Oligoarticular
2. Polyarticular RF+
3. Polyarticular RF-
4. Psoriatic arthritis *
5. Enthesitis related arthritis *
6. Systemic arthritis
7. Undifferentiated arthritis *

* Included under umbrella term JSpA

Children and adolescents with JSpA tend to have more peripheral arthritis than adults with SpA. The arthritis typically involves joints in the lower extremities in an asymmetric fashion. The presence of hip arthritis and arthritis of the small joints of the mid-foot are highly suggestive of the diagnosis. As with adult SpA, children can also develop arthritis of the lower back (sacroiliitis) or spine. Prior studies report that as many as two-thirds of children with JSpA develop arthritis of the lower back or spine within 10 years of diagnosis[8]. HLA-B27 positivity in these children increases the likelihood of developing lower back arthritis, though many children who are HLA-B27 positive never develop JSpA. The presence of lower back pain is not as helpful in JSpA as in adults with SpA in signaling the onset of sacroiliitis.

Current treatment recommendations for children with JSpA are based on those developed for adults with SpA and for all

STAY INFORMED: JSpA

categories of JIA. The 2011 American College of Rheumatology Treatment recommendations for juvenile arthritis group children with JSpA with the other JIA categories[9]. Treatment suggestions are based primarily upon the number of active joints. Methotrexate and sulfasalazine are two commonly used drugs that have established efficacy for peripheral arthritis in children. Anti-tumor necrosis factor (anti-TNF) medications also have demonstrated efficacy in children with JSpA for peripheral arthritis and enthesitis[10-17]. Additionally, anti-TNF medications are effective in the symptomatic treatment of lower back arthritis; the efficacy for halting progression of structural damage is more controversial[11]. It remains unclear which patients need anti-TNF medications as not all patients have progressive disease. New and promising drugs that are being evaluated in adults with SpA, including ustekinumab, secukinumab, and apremilast, have not yet been evaluated in children but remain on the horizon.

We still have a long way to go to understand the causes and optimal treatment of JSpA. Additional studies on the evolution of disease (particularly lower back arthritis) and optimal therapy for peripheral and spinal disease in children and adolescents are greatly needed in order to improve both short and long-term outcomes in this condition.

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