Juvenile Spondyloarthritis
A Distinct Form of Juvenile Arthritis

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KEYWORDS

• Juvenile • SpA • Juvenile idiopathic arthritis • Ankylosing spondylitis
• Enthesitis-related arthritis • Psoriatic arthritis

KEY POINTS

- Juvenile spondyloarthritis (SpA) is distinct from other forms of childhood arthritis because of the male predominance, later age of onset, and involvement of the entheses and axial skeleton.
- Some forms of juvenile SpA are associated with psoriasis or bowel inflammation.
- Juvenile SpA is an immune-mediated inflammatory disease strongly linked to HLA-B27.
- The microbial environment is believed to play a role in SpA pathogenesis.
- Most children do not achieve remission off medication within 5 years of diagnosis.

INTRODUCTION

Spondyloarthritis (SpA) is an umbrella term for a group of heterogeneous conditions occurring in adults and children, which differ from other types of inflammatory arthritis in genetic predisposition, pathogenesis, and outcome. SpA is characterized by enthesitis and other features listed in **Box 1**. These conditions are not associated with rheumatoid factor (RF), the marker associated with adult rheumatoid arthritis and the RF-positive polyarticular juvenile idiopathic arthritis (JIA) subset. Instead, they are strongly associated with the presence of HLA-B27. SpA can involve the axial skeleton, which can lead to abnormal bone formation with eventual ankylosis of the spine, resulting in substantial disability.

Nomenclature and classification for juvenile SpA is problematic for several reasons. The current International League of Associations for Rheumatology (ILAR) classification criteria of JIA does not recognize SpA as a distinct entity. Most childhood SpA in the ILAR

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Box 1
Features of juvenile spondyloarthritis

- Male predominance
- Lower extremity arthritis
- Tenderness at the insertion sites of tendons and ligaments into bone (enthesitis)
- Bowel inflammation
- Spine and sacroiliac joint inflammation
- Symptomatic anterior uveitis
- Psoriasis
- Dactylitis (sausage digits)
- HLA-B27 allele

criteria is classified as enthesitis-related arthritis (ERA), with other SpA patients falling under the categories of psoriatic arthritis (PsA) and undifferentiated arthritis (**Box 2**). A major limitation of the ILAR criteria is that they do not specifically recognize the presence of axial disease. In addition, ERA and PsA are mutually exclusive; if patients have psoriasis and fulfill criteria for ERA, they are considered to have undifferentiated arthritis.

Other conditions not specifically addressed by the ILAR classification that are considered SpA include

- Inflammatory bowel disease (IBD)-related arthritis
- Reactive arthritis
- Juvenile ankylosing spondylitis (AS)

Box 2
International League of Associations for Rheumatology classification criteria

Enthesitis-related arthritis

Inclusion criteria
Arthritis and enthesitis, *OR* arthritis *or* enthesitis plus ≥2 of the following:
- Inflammatory lumbosacral pain or sacroiliac joint tenderness
- HLA-B27 positivity
- Onset of arthritis in a male patient older than 6 years
- Acute anterior uveitis
- First-degree relative with HLA-B27–associated disease

Exclusion criteria
- RF positivity ≥2 occasions at least 3 months apart
- Systemic JIA
- Personal history of or first-degree relative with psoriasis

Psoriatic arthritis

Inclusion criteria
Arthritis and psoriasis, *OR* arthritis and ≥2 of the following:
- Dactylitis (sausage digit [**Fig. 1**])
- Nail pitting or onycholysis (**Fig. 2**)
- First-degree relative with psoriasis

Exclusion criteria
- Arthritis in an HLA-B27–positive male patient hat started after age 6
- Personal or family history of HLA-B27–associated disease
- RF positivity ≥2 occasions at least 3 months apart