key points

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Juvenile spondyloarthropathies (JSpA) are a heterogeneous group of inflammatory disorders that present in children under the age of 16 years. Unlike other arthritides, they are not defined by the presence of rheumatoid factor (RF) or antinuclear antibody (ANA), but are associated with the presence of HLA-B27 in up to 50% of JSpA patients, signalling a role in the pathogenesis of JSpA. As the symptoms of JSpA differ from those of adult spondyloarthropathy, diagnosis can be challenging. Up to 18% of cases of ankylosing spondylitis can begin in adolescence, but there is a mean interval from symptom onset to formal diagnosis of eight years.

JSpA is an umbrella term encompassing enthesitis- related arthritis (ERA), juvenile onset ankylosing spondylitis (JoAS), juvenile psoriatic arthritis (JPsA), IBD-related arthritis and reactive arthritis. Initial assessment of any childhood arthropathy should include a full blood count with differentials and inflammatory markers (CRP and ESR), which will be normal in most cases of JSpA or may reflect a mild inflammatory response and anaemia relating to chronic disease. RF and ANA are characteristically negative. Children with JSpA should be tested for HLA-B27. A faecal calprotectin test may be considered if there are symptoms of gut inflammation.

Children with suspected JSpA should be referred via their GP to a tertiary paediatric rheumatology centre for specialist multidisciplinary input from a team including paediatric rheumatologists, paediatric ophthalmologists, specialist nurses, physiotherapists, occupational therapists and psychologists. The goals of therapy include preventing joint damage and achieving remission. Uveitis should be urgently referred for specialist ophthalmology review.

NSAIDs are a first-line therapy that can provide both anti-inflammatory and analgesic effect and may be initiated by GPs in children with clinical arthritis or enthesitis, while awaiting formal specialist review. NSAID monotherapy should be ceased after two months if it does not induce disease remission. Naproxen is generally the preferred NSAID. Toxicity monitoring includes full blood count, creatinine, liver function and urinalysis. TNF inhibitors, such as adalimumab, infliximab and etanercept, have demonstrated efficacy in arthritis, enthesitis, and the symptomatic treatment of axial disease in JSpA.

All children with JSpA should maintain active follow-up with both their GP and a paediatric rheumatology service. The GP plays a key role in facilitating medication toxicity monitoring, as well as communicating with the specialist team if the patient experiences a flare. Monitoring of disease activity in JSpA may be undertaken using the JSpA disease activity score. Regular blood tests are required to monitor medication toxicity, but generally will not indicate disease flare or quiescence. Families of children with JSpA should be educated regarding the warning signs for uveitis to ensure they seek prompt review.