American College of Rheumatology
Empowering Rheumatology Professionals

Five Things

A Rheumatologist Needs to Know About the Adult With Juvenile Idiopathic Arthritis

1. **Juvenile idiopathic arthritis (JIA) encompasses a range of distinct phenotypes.** By definition, JIA includes all forms of arthritis of unknown cause that start before the 16th birthday. The most common form is oligoarticular JIA, typically starting in early childhood (before age 6) and affecting only a few large joints. Polyarticular JIA, affecting 5 joints or more, can occur at any age; older children may develop seropositive arthritis indistinguishable from adult rheumatoid arthritis. Systemic JIA (sJIA) is characterized by fevers and rash at onset of disease, though it may evolve into an afebrile chronic polyarthritis that can be resistant to therapy. Patients with sJIA, like those with adult onset Still's disease (AOSD), are susceptible to macrophage activation syndrome, a "cytokine storm" characterized by fever, disseminated intravascular coagulation, and end organ dysfunction. Other forms of arthritis in children include psoriatic JIA and so-called "enthesitis related arthritis," encompassing the non-psoriatic spondyloarthropathies. Approximately 50% of JIA patients will have active disease into adulthood.

2. **JIA can be accompanied by destructive chronic uveitis.** In addition to joints, JIA can involve the eyes, resulting in a form of chronic scarring uveitis not seen in adult arthritis. Patients at particularly high risk are those with oligoarticular or polyarticular arthritis beginning before the age of 6 years, especially if accompanied by positive ANA at any titer. In thehighest risk group, up to 30% of children may be affected. Patients who did not develop uveitis in childhood are very unlikely to do so as adults. However, if a JIA patient had uveitis as a child, ongoing ophthalmological surveillance remains important because uveitis can recur with minimal or no symptoms. Young adults with persistently active uveitis may require ongoing, systemic immunosuppression, even if the arthritis is quiescent. Acute anterior uveitis associated with juvenile-onset spondyloarthritis can still begin in adulthood, but (as in adults) this condition is overtly symptomatic and screening is not required.

3. **Pain in the adult JIA patient may be complex.** Joint pain accompanying longstanding JIA may arise from multiple mechanisms. Active arthritis remains a primary concern, and is not always reflected by inflammatory markers. Pain can arise from structural derangements resulting from altered growth, including leg length discrepancy, fusion of cervical vertebrae, temporomandibular joint disease or atlantoaxial instability. Cartilage injury can lead to early osteoarthritis. Psychosocial factors and pain amplification play a role in some patients and should be considered.

4. **Disease modifying anti-rheumatic therapies are used similarly in JIA as in adult arthritis.** The same treatments are used in JIA as in adult-onset arthritis, including joint injections, methotrexate, and biologics. As in adults, immunosuppression can be tapered in the absence of active disease, though many patients who have required methotrexate or biologic therapy will flare if treatment is stopped altogether. Some patients may be accustomed to receiving joint injections under sedation, and will need assistance to adjust to a different practice style. Systemic JIA, like AOSD, can respond well to blockade of IL-1 or IL-6, even if the febrile phase has resolved. In general, initiation of therapy during childhood is not associated with cumulative toxicity. The exception is glucocorticoid-induced osteoporosis, because failure to attain normal skeletal mass may be irreversible.

5. **Teens and young adults are still learning to care for themselves.** Teens and young adults often lack well-developed self-management and self-advocacy skills. This can translate into missed appointments and non-adherence to treatment, requiring steady patience and support on the part of the health care team. Particular guidance may be required with respect to pregnancy, substance use (including interaction between alcohol and medications), and lifestyle choices to minimize the effect of longstanding arthritis on cardiovascular risk.