Adults With Juvenile-Onset RA Need Special Care

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BOSTON — Adults with juvenile-onset rheumatoid arthritis have greater accumulated morbidity than do those with adult-onset rheumatoid arthritis, and failing to take that into account can lead to suboptimal outcomes, Dr. Peter A. Nigrovic reported at a rheumatology conference sponsored by Harvard Medical School, Boston.

"This may include structural bony deformity related to disturbed growth, premature osteoporosis, uveitis, and rarely amyloidosis," said Dr. Nigrovic, director of the Center for Adults with Pediatric Rheumatic Illness at Brigham and Women's Hospital in Boston. In addition, impaired development of life goals, excess anxiety, and impaired social and vocational functioning can lead to psychosocial morbidity.

In terms of the medical management of JRA in adulthood, "a substantial proportion of these patients will have ongoing synovitis. Therefore, aggressive treatment to prevent further injury is a major priority," Dr. Nigrovic said. Appropriate anti-inflammatory management can be challenging, however, as it is relatively difficult to assess disease activity using conventional measures. Inflammatory markers, such as erythrocyte sedimentation rate and C-reactive protein level, are often unreliable indicators of active disease in this population, and biomechanical joint pain is common despite clinical remission.

Drug therapy for adult JRA patients is similar to that for adult RA, with some exceptions, Dr. Nigrovic said. Medications used to control inflammation include NSAIDs, intra-articular and/or oral steroids, disease-modifying antirheumatic drugs, anti–tumor necrosis factor–α (anti-TNF-α) agents, and occasionally anakinra and thalidomide.

Two drugs—sulfasalazine and gold injections—are usually avoided in patients with systemic-onset JRA because of evidence linking their use for this condition with the rare but potentially fatal macrophage activation syndrome, he said.

Because inflammation in JRA affects the growing skeleton, adults in whom the disease persists often have a range of unique orthopedic complications that can be the source of pain and disability, including shortened upper extremities, small femoral heads, and deformed ankles and feet.

The temporomandibular joint (TMJ) and the cervical spine are both sites of hallmark growth abnormalities, according to Dr. Nigrovic. Involvement of the [TMJ] correlates strongly with cervical spine disease, and as such, clinicians should pay close attention to adult JRA patients with micrognathia for signs of spinal problems.

Because of the unique orthopedic considerations of adult JRA patients, joint replacement surgery can involve numerous obstacles, including the need for custom prosthetics to compensate for growth-related contractures, osteopenia, and abnormal bone morphology.

For optimal patient care, physicians also need to be cognizant of the extra-articular complications associated with JRA, because they may have an impact on clinical management. For example, patients with a history of uveitis should receive continued specialist follow-up into adulthood, and those without a history of the eye condition should be watched carefully for symptoms of the disease.

Peak bone mass attained during adolescence is lower in affected teens than in their healthy peers. Given their increased risk for osteoporosis, patient education on nutritional and lifestyle issues pertinent to building and maintaining bone strength, along with drug therapy as warranted, should be a priority in adults, he said.

Ongoing systemic inflammation associated with JRA can lead to amyloidosis, and potentially renal failure, in a small percentage of patients. Symptoms of the condition, which is diagnosed by fat pad biopsy, include proteinuria with or without hypertension, splenomegaly, hepatomegaly, and abdominal pain. Treatment of this condition has focused on suppressing the underlying inflammation with drugs such as cyclophosphamide and chlorambucil. More recently, anti-TNF-α therapy has proved to be an effective treatment option.

The psychosocial consequences of the disease, including low self-esteem, difficulty with social interactions, and increased risk of depression, can lead to diminished quality of life and social/vocational failure, said Dr. Nigrovic. The transition from juvenile to adult care should be facilitated in stages to best meet the patients' developmental needs. ■

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