A New Guideline for Juvenile Idiopathic Arthritis

It places a greater emphasis on early disease-modifying treatments, shared decision-making and immunizations.

THE LARGEST-EVER clinical practice guideline for the treatment of infants to teenagers with juvenile idiopathic arthritis (JIA) was published in September by the American College of Rheumatology (ACR).

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JIA is the most common rheumatic disease in children, causing significant short- and long-term disability for about 1 to 20 per 100,000 children. There are several types of JIA: oligoarticular affecting four or fewer joints, polyarticular affecting five or more joints, and psoriatic arthritis, which is associated with psoriasis. Joint inflammation, pain and stiffness can be chronic and continue into adulthood.

"The updated guideline was necessary to incorporate recently approved medications and shifting treatment paradigms," says HSS pediatric rheumatologist Karen Onel, MD, chief of the Division of Pediatric Rheumatology and primary investigator for the development of the ACR's 2021 Guideline for the Treatment of Juvenile Idiopathic Arthritis. "Given that about half of children and teens with JIA are cared for by adult, not pediatric, rheumatologists, we hope the new guideline will be helpful for them, too."

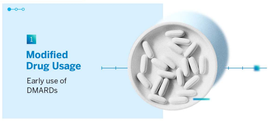
50%
of children and teens with JIA are cared for by adult, not pediatric rheumatologists

HERE ARE DR. ONEL'S ANSWERS TO SOME KEY QUESTIONS ON THE NEW JIA GUIDELINE.

Q: What were the top three major updates from the previous JIA guideline published in 2019?

A: The 2021 JIA guideline: (1) recommends early use of disease-modifying antirheumatic drugs, both conventional synthetic and biologic drugs, and a decreased use of nonsteroidal anti-inflammatory drugs and oral corticosteroids; (2) stresses the importance of shared decision-making with parents, caregivers and patients; and (3), for the first time, recommends children and teens with JIA should receive regularly scheduled immunizations for infectious diseases.

GIF, see storyboard doc



Q: How did you form teams of contributors?

A: It is essential that guideline contributors are as free from potential bias as possible. I am chair of the American College of Rheumatology's Committee on Ethics and Conflicts of Interest and unconflicted. I invited a mix of experts for the core team, the voting panel and the literature review team and put out a call for interested others to apply, intending to limit those with conflicts to less than 50% of the total, in line with best practices for guideline development as suggested by the Institute of Medicine. Overall, only one-third of our contributors were conflicted.

Q: How did including patient and parent panels benefit guideline development?

A: Patient and parent panels shared valuable perspectives. Some of their opinions were the same and some were different from ours. All parents agreed that when a child or teen has JIA, it affects the whole family, including navigating finances and co-pays, infusion units only open during school and work hours and repeat appointments for monitoring. This insight kept us laser-focused on the fact that there are real people at the other end of our recommendations.

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Q: How was the methodology different from the previous guidelines published in 2019?

A: This is the first JIA Guideline that used the Grading of Recommendations Assessment, Development and Evaluation (GRADE) approach to evaluate the strength of the available evidence in answering PICO (population, intervention, control and outcomes) questions. As a result, our recommendations reflect varying levels of evidence quality with caveats noted to assist clinicians in determining what's right for their patients.

Q: What was your biggest challenge?

A: The literature is not as voluminous for JIA as it is for other diseases, and most evidence was very low or low quality, and none was moderate or high quality. However, JIA is complex, with many phenotypes and there are many medications to consider, including recently approved tofacitinib (Xeljanz®) and golimumab (Simponi Aria®). In some areas, we did not follow the manufacturers' package inserts. For example, screening for hepatitis infection makes sense for adults, but we did not recommend it for children since there have been no cases of hepatitis B in immunized children in the past 20 years and there is no hepatitis C in the U.S. In other areas, we harmonized with adult recommendations, such as reducing the monitoring schedule for methotrexate, where appropriate.

Q: Did you intentionally leave anything out of the new treatment recommendations?

A: We removed time frames and quantifications of abnormalities, which have been in every previous version of the guideline. We did not want insurers and other groups to insist patients wait three months before starting treatment, because that's too long to wait for children who get sicker by the minute.

The evidence shows that aggressive, early treatment helps many children improve and do incredibly well. Rheumatologists should treat patients according to their symptoms and risk factors, in a shared decision-making process that takes their preferences into account. Insurers are not part of that decision.

Q: Why the shift to recommending immunizations and does that include the COVID-19 vaccine?

A: There is voluminous data showing that vaccines do not cause disease flares in children or teens with JIA. As trusted partners with our patients, we need to make sure they get their immunizations, including influenza and COVID-19 vaccines. The Centers for Disease Control and Prevention's guidelines for immunizations for immunosuppressed patients are clear and the ACR also categorically says that all children should be immunized against infectious diseases.

Q: How often will the guideline be updated?

A: It takes years to conduct a review process and formulate a guideline update, but it's essential to keep pace as new medications are approved and treatment paradigms evolve. The 2021 JIA Guideline should be considered a work-in-process road map that will continue to be filled in over the years, as required.