New Criteria for Idiopathic Inflammatory Myopathies

The European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) has developed new classification criteria for idiopathic inflammatory myopathies (IIMs) that generally perform better than existing criteria.

The most common subgroups of IIMs in adults are dermatomyositis (DM), polymyositis (PM), and inclusion body myositis (IBM). In children, juvenile dermatomyositis (JDM) is most common.

The International Myositis Assessment and Clinical Studies group (IMACS), led by Ingrid E. Lundberg, MD, PhD, of the Karolinska University Hospital and Karolinska Institutet in Stockholm, has developed consensus on outcome measures and definitions of improvement for myositis.

The objective of this international project was to develop classification criteria for adult and juvenile IIM and their major subgroups. The researchers used a probability-score model because of its superior discriminating performance.

Based on statistical models, 16 variables from six categories best distinguished IIM cases from comparators. Each variable was assigned a weight (score) based on its influence to discriminate IIM from non-IIM, and a total score was computed by adding score points corresponding to each criterion being present. As pediatric experts generally use fewer muscle biopsies for classification of JDM in clinical practice than adult rheumatologists, a second model not including biopsy variables was also developed.

With or without muscle biopsy, the classification criteria provide a score and a corresponding probability of having IIM. To facilitate use of the new criteria, a web-based calculator

has also been created.

A patient classified as having IIM by the new EULAR/ACR classification criteria can be further subclassified using a classification tree developed by IMACS. Adult IIM is distinguished from juvenile IIM by age at onset of first symptom. Based on this data set, juvenile patients with skin rash can be classified into JDM. For adults, clinical findings and muscle biopsy features can separate PM, IBM, DM, or amyopathic DM (ADM).

Because of small sample sizes, these criteria are unable to separate three additional subgroups: juvenile PM, immune-mediated necrotizing myopathy (IMNM), and hypomyopathic DM.

“The EULAR/ACR classification criteria are the first myositis criteria to be validated and tested for sensitivity in other cohorts and revealed no misclassification,” the researchers wrote. Compared with most previous criteria, the new criteria are superior in sensitivity, specificity, and classification accuracy. The criteria including muscle biopsy displayed high sensitivity (93%) and specificity (88%). Performance was slightly lower without biopsy variables (sensitivity of 87% and specificity of 82%).

While the previous Targoff criteria showed the highest sensitivity (93%) and specificity (89%), it was not able to capture all subgroups of IIM, as ADM patients were not included. Additionally, the variables of the Targoff criteria are not defined clearly enough and it requires testing more variables, including electromyography, which is not always easily accessible and may be painful for patients. By comparison, the EULAR/ACR criteria use a limited number of accessible, defined clinical and laboratory variables, and can be used for both adults and children.

Other criteria had either high sensitivity and low specificity (Bohan and Peter; Tanimoto critiera) or low sensitivity and

high specificity (Dalakas and Hohlfeld; European Neuromuscular Center criteria).

Potential Limitation

Despite utilizing data from centers in Europe, America, and Asia, a potential limitation of the new EULAR/ACR criteria is that a majority of the patients were Caucasian. Thus, the new criteria must still be validated in Asian and African populations.

Revision of the criteria will also be important in the future when additional validated myositis autoantibody tests, imaging, and other tests become available. “Importantly, these criteria are proposed as classification criteria in research and clinical trials, not as diagnostic criteria,” the researchers concluded.

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