Criteria Would Ease JSLE Remission Definition

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BRUGES, BELGIUM – A normal physical examination may be all that it takes to confirm inactive disease in a child with juvenile-onset systemic lupus erythematosus, according to new criteria.

Dr. Rina Mina of Cincinnati Children's Medical Center said that she and her associates developed the criteria to determine clinical remission and clinically inactive disease in juvenile-onset systemic lupus erythematosus (JSLE), a complex disease. They used Delphi surveys of 210 pediatric rheumatologists. The senior author on the paper (Pediatr. Rheum. 2011;9[suppl. 1]:O17) was Dr. Hermine I. Brunner, also of Cincinnati Children's.

The surveys achieved consensus on four descriptors: clinically inactive disease – which is defined without regard to medications used – and clinical remission on medication (including steroids and immunosuppressants), clinical remission on preventive medication, and clinical remission off medication. Consensus was lowest – only 86% – on what defined clinical remission off medication, while consensus for the other categories was between 94% and 96%.

According to the criteria, a JSLE patient with either inactive disease or clinical remission "should have no signs of disease activity on physical examination," Dr. Mina explained, adding that physical examination may ultimately prove to be enough to define inactivity. A patient with inactive disease or remission can have at most two nonlimiting symptoms such as headaches or fatigue.

Some select laboratory abnormalities can persist under either definition, she said, including a persistently positive antinuclear antibody test result, proteinuria from lupus-related kidney damage, or low levels of C4, for example. Members of the conference challenged Dr. Mina on the proteinuria, suggesting that it was dangerous to accept a patient with proteinuria as in remission; however, Dr. Mina argued that proteinuria was defined as a stable proteinuria as the result of damage, not a changing condition.

Use of preventive medication other than a steroid or immunosuppressant was defined as any medication to thwart disease progression or the development of disease-related damage. This category includes angiotensin-converting enzyme inhibitors, vitamin D, omega-3 acids, statins, and/or bisphosphonates. There was no consensus among the surveyed pediatric rheumatologists on whether nonsteroidal anti-inflammatory agents used daily could be considered preventive, Dr. Mina said at the meeting. Also lacking was any consensus on whether antimalarial drugs were to be considered preventive.

One of the key areas of consensus achieved by the surveys was that "inactive disease and clinical remission are distinct and should be differentiated from cure as well as from minimal disease activity," Dr. Mina said. What was "crucial" in differentiating between inactive disease and clinical remission was the time frame. Clinical remission on medication or with preventive medication is defined as requiring 6 continuous months. The criteria, she noted, are similar to current American College of Rheumatology/European League Against Rheumatism criteria for defining clinically inactive disease and remission in juvenile idiopathic arthritis.

Remission off medication was defined as disease inactivity for at least 12 continuous months. Patients could be considered "off medication" if they were not being treated with steroids, immunosuppressants, or preventive medications; however, a patient could be treated with insulin to treat steroid-induced diabetes, for example, and still be included.

Dr. Mina reported that her group had validated the consensus definitions with a cohort of JSLE patients and had found the accuracy to be "outstanding" compared with scores from widely used disease indexes for JSLE. The new criteria surpassed both the SLE Disease Activity Index and the British Isles Lupus Assessment Group score in establishing inactive disease and remission. "All core parameters support the content validity of the consensus process," she said.

Dr. Mina said she had no relevant financial disclosures. ■