An international consensus survey of the diagnostic criteria for juvenile dermatomyositis (JDM).

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Objective. To develop revised criteria for the diagnosis of juvenile dermatomyositis (JDM) using an international consensus process.

Methods. An initial survey was circulated to members of the Network for JDM and the Paediatric Rheumatology International Trials Organisation (PRINTO). Each individual was asked to identify those criteria that were felt to be most helpful in the diagnosis of classical JDM. A second survey was derived from these results and used to rank these proposed criteria in order of their importance and usefulness in clinical practice.

Results. The first survey had a response rate of 49.8% (118 individuals) from 92 centres in 32 countries. All responders routinely used proximal muscle weakness and characteristic skin rash in the diagnosis of JDM, while 86.8% used elevated muscle enzymes. Muscle biopsy, magnetic resonance imaging (MRI) and changes on the electromyogram (EMG) were deemed important diagnostic criteria. Other criteria, including myositis-specific or -related antibodies, nailfold capillaroscopy, factor VIII-related antigen, muscle ultrasound, calcinosis and neopterin, were used by 35.3% of respondents. Seventy-eight respondents to the first survey (66%) responded to the second survey. Typical MRI and muscle biopsy changes were rated by all to be the most useful clinically relevant diagnostic criteria after proximal muscle weakness, characteristic skin rash and elevated muscle enzymes. These were followed by myopathic changes on EMG, calcinosis, dysphonia and nailfold capillaroscopy, which were ranked equally.

Conclusion. This process identified nine criteria that clinicians felt to be helpful or important in the diagnosis of JDM. A further process of refinement and validation is necessary to agree an internationally acceptable, clinically usable set of diagnostic criteria.