THE INTERNATIONAL MYOSITIS CLASSIFICATION CRITERIA PROJECT

BACKGROUND

The EULAR/ACR classification criteria for idiopathic inflammatory myopathies (IIM) were developed within an international and multidisciplinary collaboration, the International Myositis Classification Criteria Project (IMCCP), comprising adult and pediatric rheumatologists, dermatologists, neurologists, epidemiologists and biostatisticians. The project involves 47 clinics from Europe (n=23), America (North America n=17, and South America n=1) and Asia (n=6) (figure 1).

Figure 1. Geographical location of participating clinics in the International Myositis Classification Criteria Project.

Demographic, clinical and laboratory data on 1600 patients (976 myositis and 624 comparators) were collected. The myositis patients (74.5% adults; 25.5% children) included juvenile dermatomyositis (DM) (n=248), polymyositis (PM) (n=245), DM (n=239), inclusion body myositis (IBM) (n=176), amyopathic DM (ADM) (n=44), hypomyopathic DM (n=12),

immune-mediated necrotizing myopathy (IMNM) (n=11) and juvenile PM (n=1). The comparators (81.6% adults; 18.4% children) embodied a broad spectrum of mimicking conditions, including muscle dystrophies, systemic autoimmune diseases and non-inflammatory myopathies.

The developed classification criteria (see web-calculator) can be used with or without muscle biopsy data and provide a probability of having IIM. Each obtained probability corresponds to unique sensitivity and specificity measures (figure 2A-D).

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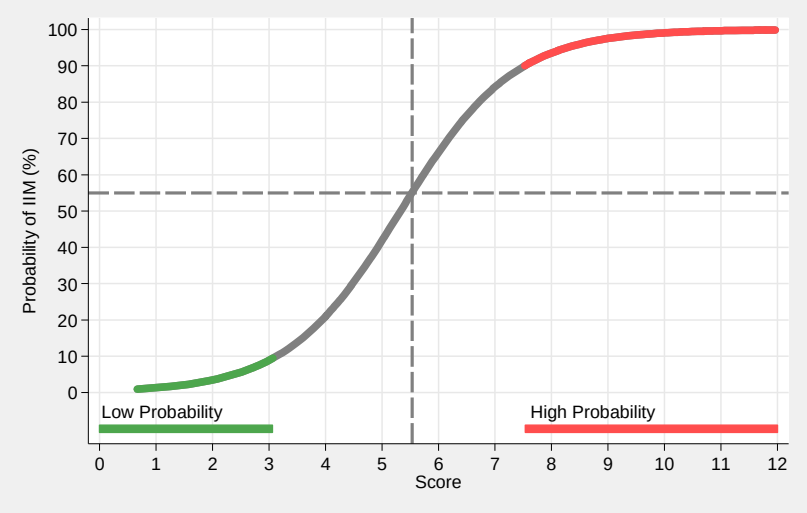

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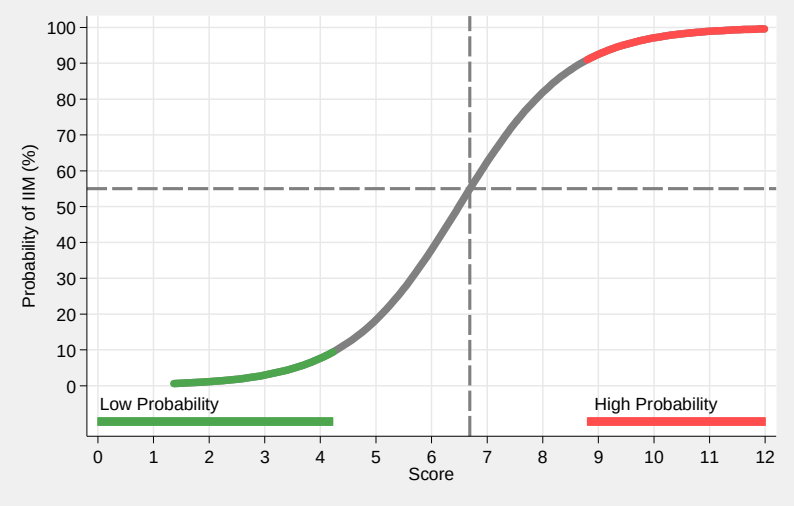

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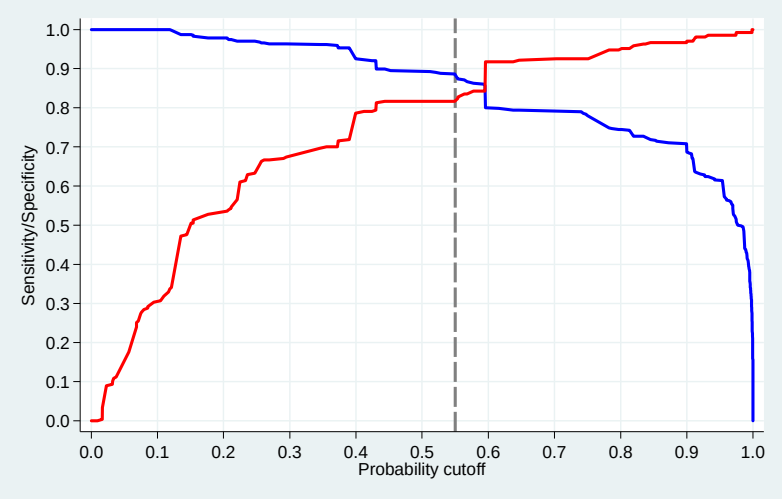

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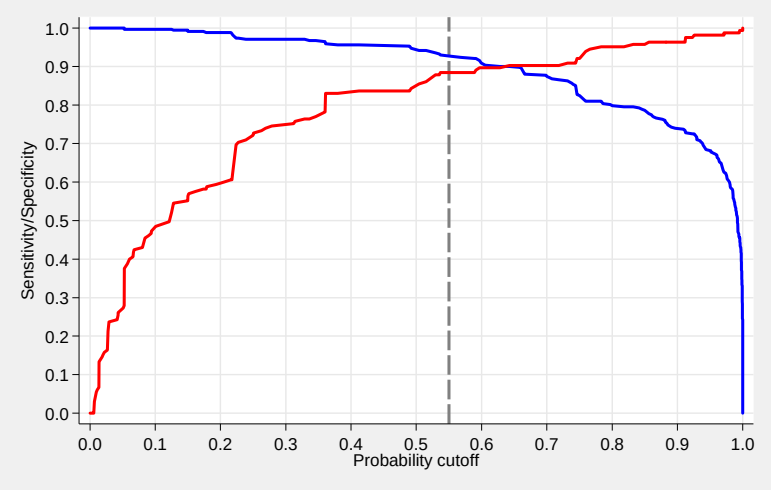

Figure 2. Probability of having idiopathic inflammatory myopathies (IIM) based on the EULAR/ACR classification criteria for IIM. Each score obtained from the classification criteria corresponds to a probability of having the disease, without muscle biopsy data (A), or with muscle biopsy data (B). Each score and probability of disease display a unique set of sensitivity (blue line) and specificity (red line) measurements for the classification criteria not including muscle biopsy data (C) or including muscle biopsy data (D). The most optimal point of accuracy should be stated in publications and be appropriate to the intended purpose, with the recommendation of using a minimum of 55% probability (score of 5.5 without biopsies; 6.7 with biopsies) for classifying a case as IIM ("probable IIM") (dotted line). "Definite IIM" corresponds to a probability of at least 90% (score of 7.5 without biopsies; 8.7 with biopsies).

Recommendations

- Patients with pathognomonic skin rashes (heliotrope rash, Gottron's papules and/or Gottron's sign) of JDM or DM are accurately classified with the EULAR/ACR classification criteria without including muscle biopsy data. For patients without these skin manifestations muscle biopsy is recommended. For DM patients without muscle involvement a skin biopsy is recommended.
- The EULAR/ACR classification criteria provide a score and a corresponding probability of having IIM. Each probability displays a unique sensitivity and specificity. The best balance between sensitivity and specificity can be found for a probability of 55-60% (total aggregated score of ≥5.5 and ≤ 5.7) for the criteria not including muscle biopsy data, and 55-75% (total aggregated score ≥6.7 and ≤ 7.6) when including muscle biopsies. These cases are designated "probable IIM". The recommended cutoff needed for classifying a patient as IIM is ≥55%.
- "Definite IIM" corresponds to a probability of ≥90% or a total aggregate score of 7.5 or more without muscle biopsy and 8.7 with muscle biopsy, and is recommended in studies where a high specificity is required.
- A patient is termed "possible IIM" if the probability is ≥50% and <55% (a minimum score of 5.3 without biopsies and 6.5 with biopsies).

For clarity and transparency, both the descriptive term ("possible", "probable" or "definite") and the probability and the aggregated score should be reported in studies.

Sub-classification of IIM

Sub-classification of IIM can be made after a patient has been classified as having IIM using the EULAR/ACR classification criteria, and is based on a classification tree (figure 3). Sub-classification of IBM is established either on clinical features (finger flexor weakness and response to treatment: not improved), or muscle biopsy data (rimmed vacuoles). Patients with IMNM cannot be distinguished from patients with PM in the classification tree. The probability range (minimum to maximum) of having IIM, the range of score and the sub-classification of IIM can be obtained with the web-calculator. Sub-classification of IIM can only be obtained after sufficient information, according to the classification tree, is included.

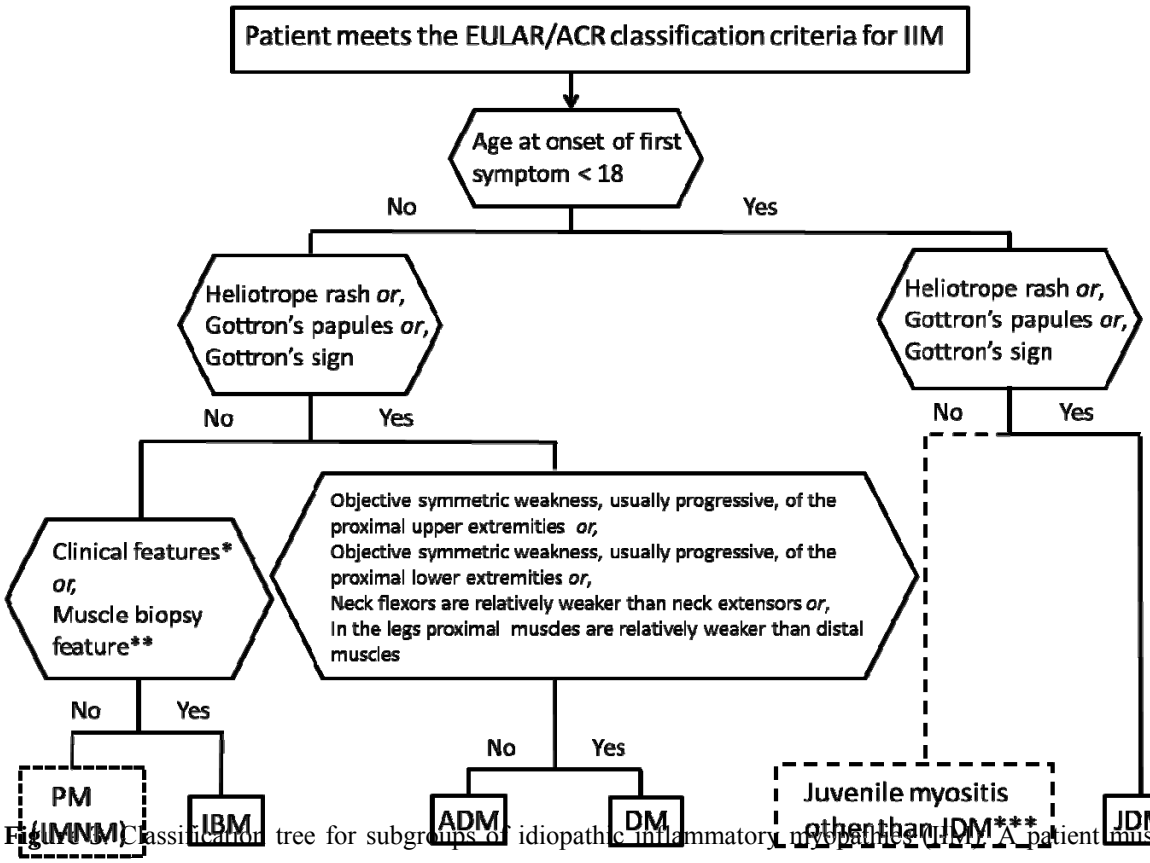

Figure 1. Classification tree for subgroups of idiopathic inflammatory myopathies (IIM). A patient must first meet the EULAR/ACR classification criteria for IIM. The patient can then be sub-classified using the classification tree. The subgroup of PM patients includes patients with immune-mediated necrotizing myopathy (IMNM). For IBM classification one of the following, *Finger flexor weakness and response to treatment: not improved, or **Muscle biopsy: rimmed vacuoles, is required for diagnosis. ***Juvenile myositis other than JDM was developed based on expert opinion. IMNM and hypomyopathic DM were too few to allow sub-classification.

PM, polymyositis; IMNM, immune-mediated necrotizing myopathy; IBM, inclusion body myositis; ADM, amyopathic dermatomyositis; DM, dermatomyositis; JDM, juvenile dermatomyositis.

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