Performance of the 2017 European League Against Rheumatism/American College of Rheumatology classification criteria for adult and juvenile idiopathic inflammatory myopathies in clinical practice

The 2017 European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) classification criteria for idiopathic inflammatory myopathies (IIM) are the first validated classification criteria for IIM. The criteria provide a score with a corresponding probability of having IIM. The cut-offs for classifying a patient with IIM were set at ≥5.5 and ≥6.7 in the probability models without and with available muscle biopsy results, respectively. At these cut-offs, the criteria had an 87% and 93% sensitivity, and 82% and 88% specificity, in the model without and with the muscle biopsy results. The criteria were tested for sensitivity, but not for specificity in two independent cohorts.[1]

We evaluated the performance of these criteria in a retrospective cohort of consecutively diagnosed IIM cases administered at our secondary/tertiary rheumatology centre between January 2010 and October 2017. The controls were the consecutive patients, without proof of IIM, who had an extensive diagnostic work-up for IIM, including muscle biopsy, during the same time period.

During the 94-month observation period we diagnosed IIM in 95 patients (72.6% female, age range 28–87 years). A muscle biopsy was performed in 87 (91.6%) patients, and was consistent with IIM in 97.7% of them. The control group consisted of 72 subjects (55.6% female, age range 18–88 years). The baseline clinical features of the cases and controls, and the clinical diagnoses of the cases are presented in table 1.

We assessed the sensitivity and specificity of the EULAR/ACR IIM criteria using the model that predicted the probability of IIM without muscle biopsy data in all patients, the model that predicted the probability of IIM with muscle biopsy data in all patients that had biopsy results available, and the model with or without biopsy data depending on the availability of the muscle biopsy results, respectively. The results are presented in table 2. The cases that were misclassified were predominantly those with an immune-mediated necrotising myopathy (35%), and those with a mild myopathy or without a myopathy, who had an interstitial lung disease, and myositis-specific antibodies other than a-Jo-1 (40%). In the control group, the cases presenting with severe toxic rhabdomyolysis were most commonly false positive (71.4%).

To summarise, in our retrospective IIM cohort, the 2017 EULAR/ACR IIM criteria showed a lower sensitivity at a comparable specificity to those originally reported. The lower sensitivity might be at least partially explained by the omission of the histological features of a necrotising myopathy in the *muscle biopsy* item, and the restriction of the *antibodies*' item solely to the presence of anti-Jo-1. These limitations had already been raised by the authors.

Table 1 Clinical characteristics of patients with an inflammatory myopathy and controls

Characteristics	IIM	Controls
n	95	72
Age (years)*	62.3 (52.3–71.3)	54.5 (45.0–72.4)
Female:male ratio	2.7	1.3
Symptom duration time (months)*	4.0 (2.0–10.0)	7.0 (1.5–24.0)
Proximal muscle weakness	76.8%	47.2%
Dysphagia	26.3%	9.7%
Elevated muscle enzymes	83.2%	69.4%
Myopathic EMG	67.8% (61/90)	36.5% (23/63)
Skin involvement	53.7%	1.4%
Interstitial lung disease	36.8%	4.2%
Arthritis	24.2%	2.8%
Raynaud phenomenon	21.1%	4.2%
MSA and/or MAA†	58.9%	0%
Anti-Jo-1	16.8%	0%
HEP-2 titre ≥1:80	61.1%	9.7%
Muscle biopsy done	91.6%	100%
Muscle biopsy positive	97.7%	0%
Clinical diagnosis		
Dermatomyositis	27.1%	
Antisynthetase syndrome	22.1%	
Immune-mediated necrotising myopathy	14.7%	
Myositis overlap syndrome	14.7%	
Polymyositis	10.5%	
Cancer-associated myositis	7.4%	
Inclusion body myositis	1.1%	
Unspecified myositis	2.1%	

*Median (IQR).
†MSA (myositis- specific antibodies) or MAA (myositis-associated antibodies): anti-Jo-1, anti-PL7, anti-PL12, anti-Mi-2, anti-SRP, anti-HMGCR, anti-MDA5; anti-TIF1γ; anti-NXP-2; anti-SAE; anti-Ku; anti-PM-Scl; anti-Ro; anti-U1RNP; anti-Scl-70.
EMG, electromyography; IIM, idiopathic inflammatory myopathies.

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Contributors All authors met the authorship requirements. AH and MT designed the study question. AH, SC and MK collected the data. AH and ZR analysed the data. AH prepared the first draft of the manuscript. AH, ZR, MT, MK and SC significantly contributed to the final manuscript.

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Table 2 The sensitivity and specificity of the new EULAR/ACR classification criteria for idiopathic inflammatory myopathies

	Sensitivity % (95% CI)	Specificity % (95% CI)	Positive predictive value % (95% CI)	Negative predictive value % (95% CI)
Using the probability model *without* muscle biopsy results n_{cases}=95; $n_{controls}$=72	74.7 (64.8 to 83.1)	80.6 (69.5 to 88.9)	83.5 (75.8 to 89.2)	70.7 (62.7 to 77.7)
Using the probability model *with* muscle biopsy results n_{cases}=87; $n_{controls}$=72	80.5 (70.6 to 88.2)	90.3 (81.0 to 96.0)	90.9 (83.1 to 95.3)	79.3 (71.3 to 85.5)
Using the probability model depending on the availability of the muscle biopsy results n_{cases}=95; $n_{controls}$=72	78.9 (69.4 to 86.6)	90.3 (81.0 to 96.0)	91.5 (84.0 to 95.6)	76.5 (68.6 to 82.8)

ACR, American College of Rheumatology; EULAR, European League Against Rheumatism.

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