Review

Performance of the 2017 EULAR/ACR Classification Criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups: a scoping review

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ABSTRACT

The 2017 EULAR/ACR classification criteria for adult/juvenile idiopathic inflammatory myopathies (IIM) were established using a data-driven approach by an international group of myositis experts to allow classification of IIM and its major subtypes. Since their publication, the performance of the criteria has been tested in multiple cohorts worldwide and significant limitations have been identified. Moreover, the understanding and classification of IIM have evolved since 2017. This scoping review was undertaken as part of a large international project to revise the EULAR/ACR criteria and aims to i) summarise the evidence from the current literature on the performance characteristics of the 2017 EULAR/ACR classification criteria in various cohorts and IIM subtypes, and ii) delineate the factors that need to be considered in the revision of the classification criteria. A systematic search of Medline (via PubMed), Cumulative Index to Nursing and Allied Health Literature, and conference abstract archives was conducted independently by three investigators for studies on the EULAR/ACR criteria published between October 2017 and January 2023. This scoping review of 19 articles and 13 abstracts revealed overall good performance characteristics of the EULAR/ACR criteria for IIM, yet deficiencies in lack of inclusion of certain IIM subtypes, such as immune mediated necrotising myopathy, amyopathic dermatomyositis, anti-synthetase syndrome and overlap myositis. Published modifications that may improve the performance characteristics of the criteria for classification of IIM subtypes were also summarised. The results of this review suggest that a revision of the EULAR/ACR criteria is warranted.

Introduction

Idiopathic inflammatory myopathies (IIM) are a rare, heterogenous group of systemic autoimmune diseases that include a number of subgroups: dermatomyositis (DM), juvenile dermatomyositis (JDM), polymyositis (PM), inclusion body myositis (IBM), immune-mediated necrotising myopathy (IMNM), antisynthetase syndrome (ASSD), overlap myositis (OM), and non-specific myositis, among others (1). Even though amyopathic forms also exist (clinically amyopathic dermatomyositis [CADM]), IIM are primarily characterised by muscle weakness. Extra-muscular manifestations include rash, interstitial lung disease, arthritis, cardiac involvement, and Raynaud's phenomenon. For classification of this heterogeneous group of diseases, no single clinical or diagnostic test suffices, and diagnostic evaluation often involves history, clinical examination, laboratory analyses, neurophysiological tests, imaging, skin, and muscle biopsy.

The Bohan and Peter (Bohan-Peter) criteria laid the groundwork as the first classification criteria for IIM and was developed from a single-institution case series in 1975 (2, 3). In the following years, several additional IIM classification criteria were developed. The first multicentre and interdisciplinary classification criteria were the Tanimo-

to criteria for PM and DM in 1995 (4). However, these criteria did not include IBM or JDM. Other published criteria, such as those by Targoff *et al*. in 1997, supplemented the Bohan-Peter criteria with the myositis-specific autoantibodies (MSA) that were known at the time as well as magnetic resonance imaging (MRI) findings of the muscle (5). In 2004, experts within the European Neuromuscular Centre (ENMC) and Muscle Study Group published additional criteria with an emphasis on histopathological features specific to different subtypes of IIM, combined with clinical criteria including MRI, age of onset, MSA, and electromyography (EMG) findings (6).

Given the lack of universally accepted and well-validated criteria, Lundberg *et al*. formed a large international multidisciplinary collaborative group to establish the European Alliance of Associations for Rheumatology (EULAR) - American College of Rheumatology (ACR) classification criteria for adult and juvenile IIM and their major subgroups (EULAR/ACR Criteria) (7). The 2017 EULAR/ACR criteria are data driven and include 16 items from six categories with different weights assigned for each item: age of symptom onset, pattern of muscle weakness, DM skin manifestations (heliotrope rash, Gottron's papule or sign), dysphagia or gastrointestinal dysmotility, laboratory measurements (elevation of muscle enzymes and anti-Jo1 autoantibody positivity), muscle biopsy findings (endomysial mononuclear cell infiltration of myofibres, perimysial/perivascular mononuclear cell infiltration, perifascicular atrophy, rimmed vacuoles) that were optional for cases with DM rash. The criteria provide a probability score for IIM allowing categorisation of the patients as possible (50-<55%), probable (55-<90%), and definite IIM (≥90%) based on different thresholds. Additionally, the criteria allow for IIM subclassification for DM, PM, CADM, IBM, and JDM subtypes. As part of the criteria development process, the EULAR/ACR criteria were both internally and externally validated using retrospective data; however, the external validation did not include any controls and included only few patients with CADM (8).

Even though it has been only six years since the publication of the EULAR-ACR criteria, the criteria development process took almost 10 years to enrol a sufficient number of participants reflecting the body of knowledge from almost two decades ago. Since then, several critical developments have occurred in the understanding and classification of IIM including the increased number and availability of MSA in routine clinical practice and recognition of IMNM and ASSD as newer subtypes. This scoping review was performed as part of a large international project to revise the EULAR-ACR criteria and aims to summarise the current literature on the performance characteristics of the criteria and systematically delineate the factors that need to be considered in the revision.

Methods

Eligibility criteria and search strategy

A scoping review format was chosen to best address the identification and analysis of knowledge gaps, as well as identification and discussion of certain characteristics in studies (9). Three authors (D.S., R.Z., and S.G.) independently identified studies published after the publication of the EULAR-ACR Criteria, on the performance of the criteria, by systematically searching Medline (via PubMed), Cumulative Index to Nursing and Allied Health Literature (CINAHL), and the online or in-print abstract archives of the relevant conferences. The inclusion criterion was any original study assessing the performance of the EULAR/ACR Criteria. Case reports/series, review articles, book chapters, editorials/opinion pieces, and non-English articles were excluded. In addition, the authors screened the reference lists of the articles identified in accordance with the Preferred Reporting Items for Systematic reviews and Meta-analyses (PRISMA) reporting guidelines for scoping reviews (10). A protocol was not developed for this scoping review.

The relevant conferences were identified based on the input from the Steering Committee members of the project, which included 25 participants from seven different specialties and 12 countries spanning six continents. These conferences included the ACR, EULAR, and American Academy of Neurology (AAN) annual meetings, American Academy of Dermatology (AAD) and Society for Investigative Dermatology (SID) conferences, European Academy of Dermatology & Venereology (EADV) conference, World Congress of Dermatology (WCD), International Congress on Neuromuscular Diseases (ICNMD), Global Conference on Myositis (GCOM), World Muscle Society (WMS) conference, and Muscle Study Group. The authors searched through the conference agendas, online abstract archives, and/or abstracts published in the respective journals from these conferences. If an original article of a conference abstract was published on Medline, the original article was used for data extraction instead of the conference abstract.

The terms used in the database search were a combination of terms for "myositis", "classification", and "criteria" as following: ("myositis"[MeSH Terms] OR "myositis"[Text Word]) AND ("classification"[MeSH Terms] OR "classification"[Text Word]) AND ("standards"[MeSH Subheading] OR "criteria"[Text Word]). The specific search terms used in different abstract archives along with the weblinks of each abstract archive are detailed in Supplementary Table S1. A critical appraisal of individual sources of evidence was not performed. The authors of the articles and abstracts were not reached to confirm the data obtained from the investigators.

Data extraction and synthesis

Three authors (D.S., R.Z., and S.G.) extracted the following variables from each article and conference abstract using a pre-determined standardised approach: authors, year, geographic location, study design (number of participating centres [single/two/multiple centres] and prospective/retrospective), number of participants, characteristics of the study population (average age, sex and race/ethnicity distribution), gold standard(s) used for diagnosis,

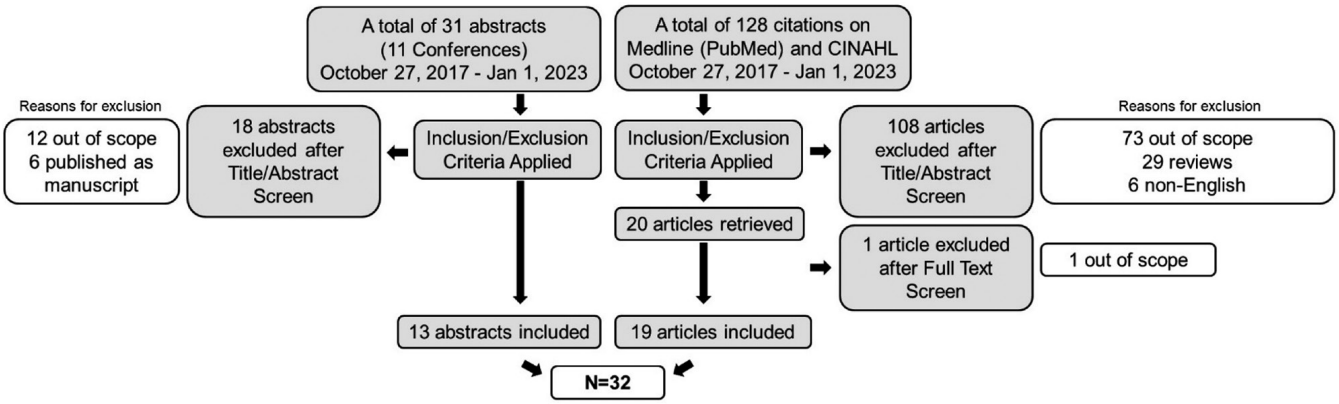

Fig. 1. Flow diagram of study identification and selection.

performance characteristics of the EULAR/ACR criteria reported (sensitivity, specificity, and number of true positive, true negative, false positive and false negative subjects), performance characteristics of any other classification criteria reported, any variables tested that were not included in the EULAR/ACR criteria, proportion of participants who had EMG and MRI, and any suggestions in the discussion section of the manuscripts or abstracts for future consideration in revised classification criteria. All the included studies were summarised descriptively by the same three authors (Suppl. Tables S2 and S3). If reported, the number of true positive, true negative, false positive, and false negative subjects was used to calculate the confidence interval for the sensitivity and specificity results. Studies that used the Bohan-Peter criteria as the gold standard for diagnosis, did not report whether cases fulfilled the criteria for possible, probable, or definite IIM, or did not report sensitivity and/or specificity results were not included in the analyses. Forest plots were generated to display the sensitivity/specificity results of the criteria across different studies. Study selection, data extraction, data synthesis and analysis were performed in accordance with the PRISMA guidelines for scoping reviews (10) (Suppl. Table S4).

Results

Search results

The search revealed 128 citations on Medline (via PubMed) and 15 citations on CINAHL between October 27, 2017 (publication date of the EULAR/ACR criteria) and January 1, 2023 (Fig. 1). All the articles identified via CINAHL search overlapped with the Medline search. After applying the inclusion and exclusion criteria detailed above to the titles and abstracts, a total of 113 articles were excluded due to being outside the scope (n=75), publication type (n=32), or non-English language (n=6). After this initial title/abstract review, 20 articles were retrieved, and the full texts were reviewed according to the inclusion and exclusion criteria. Of these, one article was excluded due to being outside the scope, yielding 19 articles for final review (Supplementary Table S2). The search of the abstract archives of the relevant conferences with the same inclusion and exclusion criteria yielded 31 conference abstracts. Six of these abstracts were published as articles and 12 were beyond scope (Fig. 1). Therefore, 13 conference abstracts were included in the final review (Suppl. Table S3).

Characteristics of the published articles and abstracts

The total number of patients with IIM included in these studies was 7,007. The mean age of patients was 50.9 years (SD 6.1) in adults with IIM and 7.1 years (SD 1.5) in children with JDM, respectively. Overall, 70% of the patients with IIM were female. The largest sample size per study was 1,024 in an original article from the U.S. (11) and 1,370 in a conference abstract from China (12). The smallest sample size in a study was 26 in an original article from India (13). The studies were performed in Asia (n=11), Europe (n=11), South America (n=5), North America (n=4), and Australia (n=1). Studies from the African continent were lacking. Of the 32 articles and abstracts included, study population consisted of patients with IIM in 24 studies, DM in four studies (12; 14-16), juvenile IIM in two studies (17; 18), anti-MDA5 autoantibody positive IIM in one study (19), and scleromyositis in one study (20).

The subclassification performance of criteria was assessed for DM in eight studies (11; 21-27), for PM in eight studies (11, 21-27), for IBM in six studies (11, 21-24, 27), for CADM in six studies (11, 15, 22, 24, 25, 27), and for JDM in five studies (17, 18, 23, 25, 27). All studies were retrospective except one abstract which had an ambispective design with a prospective component (28) and one retrospective study of a prospectively collected data (15). The majority of the studies were single-centre studies (n=19; 59.4%), while data were collected from multiple centres in 10 studies (11, 18-20, 23, 29-33). Three studies did not specify the number of centres (34-36). The gold standard used for IIM diagnosis was based on physician assessment or expert consensus in the majority of the studies (24 out of 30 studies with reported gold standard, 80%). Remaining studies used Bohan-Peter, ENMC or Sontheimer criteria as the gold standard for IIM diagnosis (2, 3, 28, 29, 35-38).

Classification performance of the EULAR-ACR myositis classification criteria

After excluding the studies that used

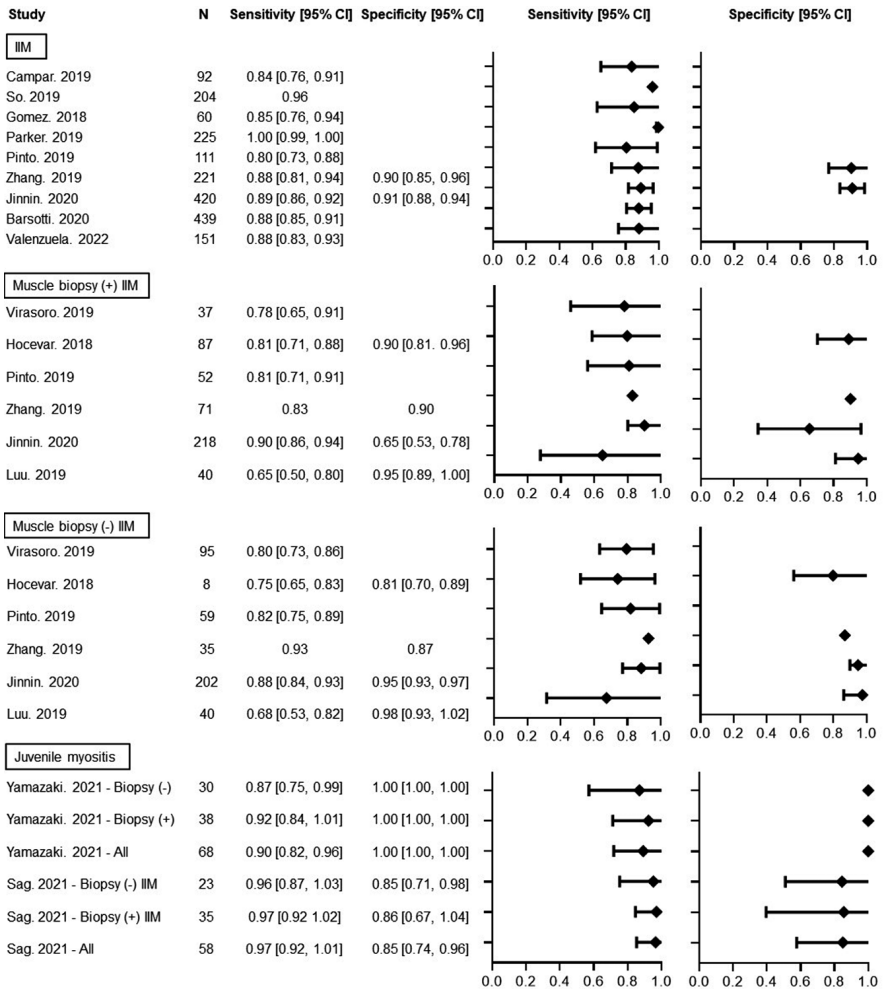

Fig. 2. Forest plots of sensitivity and specificity of the EULAR-ACR myositis classification criteria for patients with IIM, those with and without available muscle biopsy results, and patients with juvenile myositis.
IIM: idiopathic inflammatory myopathies.

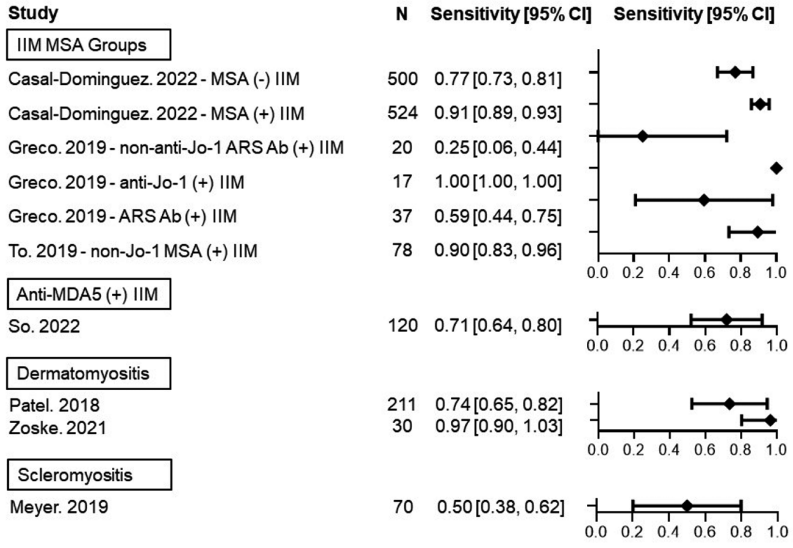

Fig. 3. Forest plots of sensitivity and specificity of the EULAR-ACR myositis classification criteria for specific IIM subtypes.
IIM: idiopathic inflammatory myopathies; MSA: myositis-specific autoantibodies; ARS: anti-tRNA synthetase autoantibodies.

Bohan-Peter criteria as a gold standard (28, 29, 35-38), did not specify definite/probable/possible case classification (12-14, 39) or did not report sensitivity/specificity information (32), 21 studies were identified that reported the performance of the EULAR-ACR criteria for IIM classification. The majority of the studies (n=15) assessed the performance of the criteria in patients with IIM with one study specifically focusing on anti-aminoacyl-tRNA synthetase (ARS) autoantibody positive IIM (31), one study on non-Jo-1-MSA positive IIM cases (40) and one study on MSA positive and negative IIM (11). The performance of these criteria was also assessed in specific IIM subgroups including DM (15, 16), juvenile myositis (17, 18), anti-MDA-5 positive IIM (19), and scleromyositis (20).

The sensitivity of the criteria for IIM ranged from 80% to 100%, while the specificity was over 90% (Fig. 2). Sensitivity of the criteria was 100% in patients with anti-Jo1 positivity, 25% in patients with non-anti-Jo1 ARS positivity, 90% in patients with non-anti-Jo-1 autoantibody positive MSA, 91% in patients with positive MSA and 77% in patients with negative MSA (11, 31, 40) (Fig. 3). The sensitivity of the criteria was 74–97% in DM patients, 50% in scleromyositis, and 71% in anti-MDA-5 autoantibody positive IIM. The criteria had 87–97% sensitivity and 85–100% specificity in patients with juvenile myositis. There were no studies focusing solely on IMNM. IMNM was the most common myositis type to be classified as non-myositis (11, 24, 41). In two studies, 35–64% of the patients with IMNM were misclassified as non-myositis (24, 41). One study noted a substantial increase in the sensitivity of the criteria for IMNM when biopsy results were not included (24). This result was attributed to the non-specific muscle biopsy findings that may be observed in patients with IMNM. The patients with IMNM who were classified as IIM were most commonly subclassified as PM, followed by IBM, and DM (11, 23, 24, 27).

Six studies additionally reported the sensitivity and specificity of the criteria for IIM based on the availability of

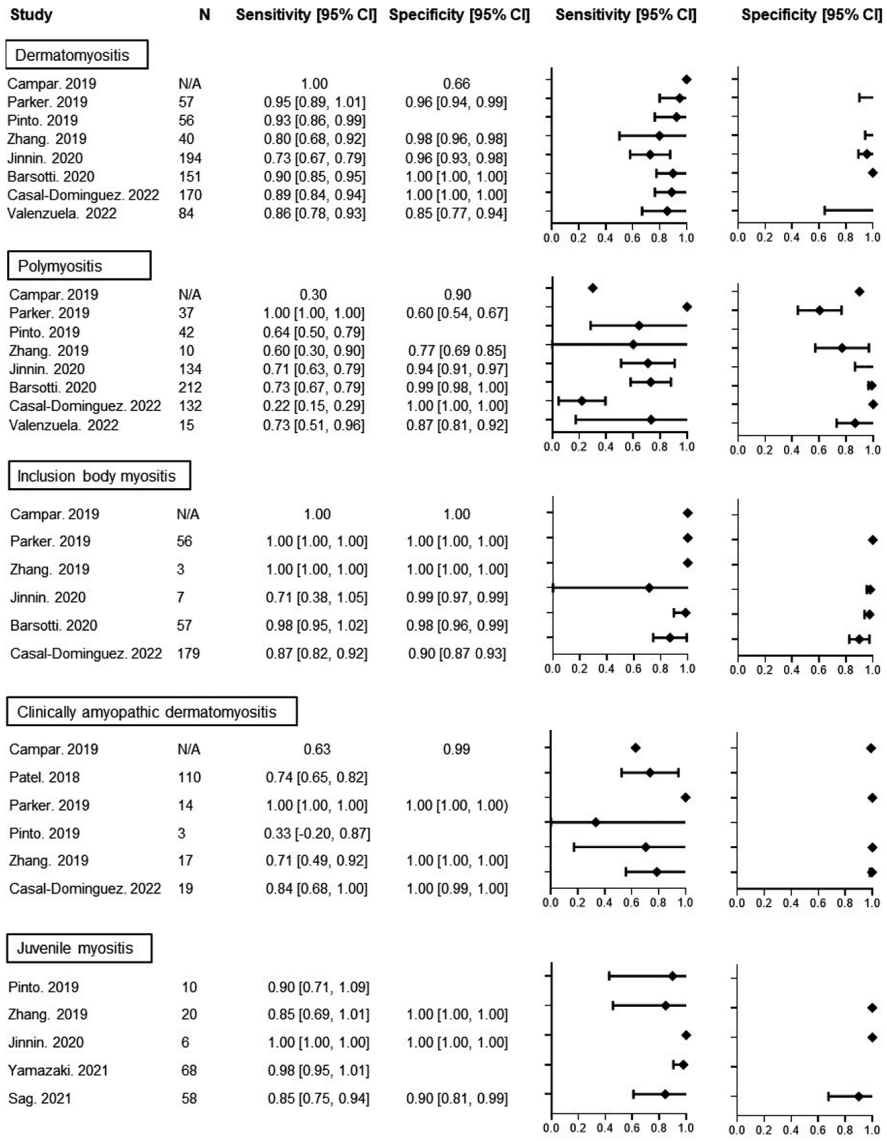

Fig. 4. Forest plots of sensitivity and specificity of the EULAR-ACR myositis classification criteria for the subclassification of major IIM subtypes.

muscle biopsy findings with variable results (23, 25, 27, 30, 34, 41). While one study showed an increase in sensitivity with the presence of muscle biopsy from 75% to 81% (41), two other studies showed a decrease in sensitivity when adding muscle biopsy findings (27, 30). Jinnin *et al*. found no significant difference in sensitivity based on the availability of muscle biopsy findings (23). Similarly with regard to specificity, both improvement (41) and worsening (23) were reported based on the availability of muscle biopsy findings. Additionally, two studies showed little difference in specificity for IIM depending on muscle biopsy findings (27, 30).

Subclassification performance of the EULAR-ACR myositis classification criteria

- Dermatomyositis

Eight studies reported the performance of the criteria for subclassification of DM (11; 21-27). The sensitivity of the criteria for DM subclassification ranged from 73% to 100% with the majority of the studies showing sensitivity over 80% (Fig. 4). The specificity ranged from 66% to 100%, and was over 85% in all studies except one study (22). For CADM, six studies reported the performance of the criteria for subclassification of CADM (11, 15, 22, 24, 25, 27). The sensitivity of the criteria for CADM subclassification ranged from 33% to 100%, while the specificity was between 99% and 100% (Fig. 4).

- Polymyositis. Eight studies reported the performance of the criteria for subclassification of PM (11, 21-27). The sensitivity of the criteria for PM subclassification varied widely between studies ranging from 22% to 100% (Fig. 4). The specificity ranged from 60% to 100%.

- Inclusion body myositis. Six studies reported the performance of the criteria for subclassification of IBM (11, 21-24, 27). The sensitivity of the criteria for IBM subclassification ranged from 71% to 100% with all studies showing a sensitivity over 85% except for one study (23) (Fig. 4). The specificity ranged from 90% to 100%.

- Juvenile myositis. Five studies reported the performance of the criteria for subclassification of JDM (17, 18, 23, 25, 27). The sensitivity of the criteria for JDM subclassification ranged from 85% to 100%, while the specificity was between 90% to 100% (Fig. 4).

Comparison of the performance of the EULAR-ACR myositis classification criteria and other myositis classification criteria

In 18 studies, the EULAR/ACR criteria were compared with other myositis classification criteria, including Bohan-Peter, ENMC, Tanimoto, Targoff, Solomon and Senecal criteria.

- Bohan-Peter criteria. For IIM, the EULAR/ACR criteria had a similar or higher sensitivity and higher specificity in all studies compared to the Bohan-Peter criteria (23, 25, 27, 30, 33, 42) (n=6). For DM, the EULAR/ACR criteria had a comparable sensitivity of to the Bohan-Peter criteria in two studies (21, 26), lower sensitivity in one study (12) and higher sensitivity of in two studies (14, 42). The specificity of the EULAR/ACR for DM was higher in two studies, comparable in one study, and lower in one study (12, 14, 21, 26). For PM, the EULAR/ACR criteria achieved a modestly lower sensitivity but a higher specificity than the Bohan-Peter criteria (21, 26, 42). Concordance rates between the EULAR/ACR and Bohan-Peter criteria were 54–89.5% for IIM, 42–83.7% for DM and 44– 95.8% for PM (32, 33,

35, 36, 38). For JDM, the EULAR-ACR criteria had a higher sensitivity and similar specificity to the Bohan-Peter criteria (17, 18). For MDA-5 autoantibody positive patients with IIM, the EULAR/ACR criteria had remarkably higher sensitivity than the Bohan-Peter criteria (19).

- *ENMC criteria*. The EULAR/ACR criteria showed similar sensitivity to the ENMC criteria for DM (12, 16, 29), and PM (29) except in one abstract. This abstract from India showed that the EULAR-ACR criteria classified fewer patients with DM than the ENMC criteria (35% *vs*. 83%) which was suggested to be due to difficulty in identifying heliotrope rash in dark-skinned individuals (28).

- *Tanimoto criteria*. In two studies with JDM patients, the EULAR/ACR criteria had a higher sensitivity than the Tanimoto criteria (17, 18). One study showed a 100% specificity for both criteria (18), while the other showed a higher specificity for the Tanimoto criteria than the EULAR/ACR criteria (17). The EULAR-ACR criteria also showed a higher sensitivity in IIM, DM, ASSD, and overlap myositis, and a higher specificity (87.8%) than the Tanimoto criteria in adults (23, 42).

- *Targoff criteria*. The EULAR/ACR criteria was compared with the Targoff criteria in one study and showed higher sensitivity and similar specificity (30).

- *Solomon criteria*. In one study with patients with clinicoserological diagnosis of ASSD, a larger number of patients met the EULAR/ACR criteria compared to Solomon criteria for IIM (59.5 *vs*. 45.9%) (31).

- *Senecal criteria*. In one study, the EULAR-ACR criteria classified 53.2% of patients as DM and the rest of the cases as PM, while Senecal criteria classified 75.9% of patients as overlap myositis, 12.7% as PM and 10.1% as DM (29).

Proposals for modifications of the EULAR-ACR myositis classification criteria

- *Myositis-specific autoantibodies*. Overall, there was a strong support for adding further myositis-associated autoantibodies (MAA) and MSA beyond anti-Jo1 to the classification criteria (11, 19, 20, 23, 30, 31, 33, 40). The most commonly misclassified patients as not having myositis by the EULAR-ACR criteria were those with anti-HMG-CoA reductase, anti-SRP, anti-MDA-5, anti-PL-7 and anti-PL-12 autoantibodies (11, 41). Eight studies examined the impact of adding MSA and/or MAA to the current criteria.
Including non-Jo1 anti-ARS autoantibodies in the criteria improved the sensitivity of the criteria from 25% to 95% for patients with ASSD (31). In a cohort of patients with MDA-5 (+) IIM, replacing the anti-Jo-1 autoantibody item with anti-MDA-5 increased the sensitivity of the EULAR/ACR criteria from 71.7% to 98.3% (19). Including MAA (anti-SS-A, anti-SS-B, anti-RNP, anti-Ku, anti-PM-Scl) and non-Jo-1 MSA (anti-Mi-2, anti-PL-7, anti-PL-12, anti-EJ, anti-OJ, anti-SRP, anti-HMG CoA reductase antibody) slightly increased the accuracy of the criteria by increasing the area under the curve (AUC) from 0.79 to 0.83 and 0.81, respectively (30). Another study showed an increase in definite IIM cases from 73.1% to 96.2% and a decrease in probable IIM from 16.7% to 3.8% and non-IIM from 10.3% to none by including MSA (40). These autoantibodies included anti-PL-7, PL-12, EJ, OJ, Mi-2, MDA-5, SAE1, TIF-1γ, NXP-2, SRP and HMG CoA reductase autoantibodies and were given the same weight as anti-Jo-1 in the criteria (40). Inclusion of MSA increased the agreement between physician diagnosis and EULAR/ACR criteria from a kappa coefficient of 0.43 to 0.77 in one study (13) and the sensitivity of the EULAR/ACR criteria by 1.4% in another study (33). Another study showed that adding MSA increased the sensitivity of the EULAR/ACR criteria from 95.6% to 98.3%, but decreased the specificity from 44.8% to 15.2% and agreement from 88.5% to 85.2% (14). However, the types of autoantibodies included were not reported in these studies. Lastly, in a group of patients with physician-diagnosed scleromyositis, including anti-Ku, SMN1, PM-Scl, U1-RNP, and RuvBL1/2 autoantibodies improved the sensitivity of the criteria from 56% to 84% (20).

- *Muscle biopsy*. Two studies examined the impact of expanding the muscle biopsy findings on the performance of the criteria. The first study showed that including invasion of myofibres by endomysial infiltrates on muscle histopathology improved the sensitivity of the criteria slightly from 65% to 68% without reducing the specificity at 95% for IIM diagnosis (30). The second study showed that including perifascicular atrophy (along with MSA) increased the agreement between physician diagnosis and the criteria for DM (13).

- *Muscle MRI*. The frequency of MRI availability was reported in 4 studies with a mean of 47% (0.2 SD) of patients with IIM having available muscle MRI results. Addition of positive MRI findings to the criteria has been recommended in several studies (17, 23, 27, 30). However, only one study has examined the effect of adding MRI findings to these criteria (30). This study showed that including MRI evidence of myositis improved the accuracy of the criteria with increase in AUC from 0.79 to 0.91 for IIM diagnosis (30).

- *Cutaneous findings*. Expanding the skin findings for DM in the classification criteria was suggested in several studies (13, 15, 25, 27, 28). Patients with mechanics hand, shawl sign, V neck sign, and erythroderma were misclassified as PM using the current criteria (27). In another study, 26% of patients with amyopathic DM did not meet the criteria for IIM (15). These patients were most commonly presented with V neck sign, malar rash, rash in other areas of face, Gottron's sign on areas other than hands, scalp, and arms (15). This study also suggested that skin biopsy findings could be included in the criteria but did not report the effect of including these findings on the criteria performance. Importantly, in three studies from India, the sensitivity of Bohan-Peter and ENMC criteria were higher than the EULAR-ACR criteria for DM which was attributed to a wider range of DM rashes allowed in the ENMC criteria and dif-

ficulty in identifying heliotrope rash in individuals with dark skin (13, 25, 28). Only one study examined the effect of including other DM rashes (shawl sign, Vneck, Holster sign, malar rash not sparing nasolabial fold, mechanic hands and periorbital oedema) on the criteria performance (14). This study showed an increase in the sensitivity of the EULAR-ACR criteria from 95.6% to 97.7% and a decrease in specificity from 44.8% to 13.3% and agreement with physician diagnosis from 88.5% to 85.7% (14).

- *Interstitial lung disease (ILD)*. The patients with ILD were one of the most commonly misclassified groups (11, 41). One study examined the impact of adding ILD on the performance of the criteria and showed that the sensitivity of the criteria increased by 1.3% when ILD was added as an interchangeable item with dysphagia (21).

- *EMG*. The frequency of EMG availability was reported in eight studies with an average of 64% (SD 0.2) of patients with IIM with available EMG results. No studies have examined the impact of adding EMG findings to the performance of the criteria.

Discussion

This scoping review of 19 articles and 13 abstracts from 16 countries demonstrated the performance characteristics of the EULAR/ACR myositis classification criteria for adult and juvenile IIM, with estimates of sensitivity and specificity ranging from 65% to 99.6% and 77.4% and 98%, respectively. The EULAR/ACR criteria generally had better performance characteristics than the previously published criteria including the Bohan-Peter, ENMC, Tanimoto and Targoff criteria for IIM. Importantly, this review also demonstrated the critical deficiencies of the current EULAR/ACR criteria in subclassification of the existing and emerging IIM subtypes such as IMNM, CADM, and ASSD. Several published modifications to the original criteria which may improve the performance of the criteria for these IIM subtypes were also summarised including broadening the MSA in the criteria.

The EULAR/ACR myositis classification criteria were developed and externally validated in a predominantly Caucasian population who had to have an IIM diagnosis for at least six months (8). In this review, the performance of the criteria in non-Caucasian populations were seen to be overall favourable in studies from Argentina, Chile, China, India, Japan, and South Korea while the performance was slightly lower in Australia (19, 23, 25-27, 30). This result may not be due to differences in the Australian cohort, but the availability of datapoints. The performance of these criteria in an inception cohort of patients with newly diagnosed IIM also showed good results (24). These external validation studies showed acceptable to good performance of the criteria in non-Caucasian patients and those with newly diagnosed IIM.

Unlike the majority of classification criteria in rheumatology in which the primary goal is to accurately classify and distinguish the disease of interest from its mimickers, the IIM classification criteria also aim to subclassify IIM subsets. This additional task in a tremendously heterogenous disease such as IIM is important but challenging because of evolving understanding of the disease with discovery of new clinico-serological phenotypes over the last several years. This review showed favourable performance characteristics for IBM and JDM subtypes with over 90% sensitivity and specificity (21, 23, 24, 27). However, the results were more variable for PM and CADM subtypes. With the discovery of new autoantibodies after the data collection in the classification criteria project, PM is now recognised to constitute only a minority of IIM subtypes (43). The majority of the cases who were classified as PM had other physician diagnoses including ASSD, OM, and IMNM which suggests that the criteria likely lead to overestimation for PM (24, 25, 27). This conflict between the current practice and the criteria decreases the generalisability of the study results and hampers studies to be conducted in these IIM subtypes.

Not surprisingly, IMNM was the most commonly misclassified subtype which was often classified as PM or rarely as DM or IBM (in cases with severe IMNM affecting distal muscles) by the EULAR/ACR criteria, as this subgroup had hardly been recognised when the classification criteria project had started in 2004 (11, 24, 41). These results show that the IMNM subclassification should be one of the prioritised goals in consideration of the revision of the EULAR-ACR criteria. Including adequate number of cases with IMNM, anti-HMG-CoA reductase and anti-SRP auto-antibody information, and/or histopathological features of necrotising myopathy in the muscle biopsy item may help with the accurate classification of these patients.

Even though the EULAR/ACR criteria showed good sensitivity for classifying ASSD cases with anti-Jo-1 autoantibodies as IIM (21), the sensitivity was much lower in ASSD patients with non-anti-Jo-1 autoantibodies (31). Furthermore, the majority of patients with ASSD were classified as either PM or DM based on the EULAR/ACR criteria (11, 24). This represents a classic dilemma between a pure clinical versus a serological classification. Including non-anti-Jo1 autoantibodies in the criteria may improve the sensitivity for the classification of ASSD as IIM; however, the problem of misclassification can only be resolved by recognition of ASSD as a distinct subtype in the criteria (31). There are currently ongoing efforts for development and validation of classification criteria for ASSD by the ACR/EULAR (44). Unlike the revised EULAR/ACR criteria which will subclassify ASSD as a distinct IIM subtype, the ASSD criteria aim to classify ASSD patients who present with a whole spectrum of extramuscular manifestations.

A criterion by Casal-Dominguez *et al*. which propose to classify patients with IIM according to their MSA outperformed the EULAR/ACR criteria with perfect sensitivity and specificity in two independent cohorts (11). While MSA are extremely helpful in phenotypic characterisation of patients, the reliability of immunoassays used for MSA testing are variable and not standardised across different regions of the world

which is evident by several reports on high rates of false positive MSA results (45). Furthermore, MSA negative patients represent approximately 30% of patients with IIM (1). Recognising the importance of MSA as pointed out by Casal-Dominquez *et al*. and others, the revised ACR/EULAR criteria will attempt to include a whole spectrum of MSA despite the limitations in MSA testing.

One of the major strengths of the EULAR/ACR criteria is their ability to subclassify CADM as an IIM subtype which is different than the previous criteria such as the Bohan-Peter, Tanimoto and Targoff criteria. However, owing to the small number of patients with CADM, the criteria were not externally validated in those with CADM and did not include cases with hypomyopathic DM, further necessitating external studies (8). External studies showed variable sensitivity of the EULAR/ACR criteria for CADM between 33% to 100% (15, 23, 24, 42). In one study, one third of the patients with CADM did not meet the minimum threshold to be classified as DM (15). These patients had skin manifestations including V neck sign, malar rash, erythema overlying the rest of the face, and scalp rash, which are not part of the skin variables in the EULAR/ACR criteria (15). Therefore, inclusion of other skin manifestations, MSA information, and skin biopsy results in the revised criteria may improve the sensitivity of the EULAR/ACR criteria for CADM. Prospective data collection across different specialties would allow for systematic evaluation of all the pertinent cutaneous findings in the revised criteria; thus, should be considered.

In the EULAR/ACR criteria, EMG and muscle MRI were only available in 29% and 38% of the cases, respectively. Therefore, these diagnostic modalities were thought to not be commonly used and were ultimately not included in the criteria. However, subsequent studies showed available EMG in 64% of patients with IIM with some centres having EMG in over 90% of their cases (41), while MRI was available in about half of the patients with IIM. These results highlight the variation in the diagnostic tools used for IIM across different geographical regions and including these tests may improve the sensitivity and specificity of the criteria for IIM (30). With better recognition of the specificity and distinct phenotypes associated with MSA, myositis diagnosis will likely rely more on MSA and other ancillary tools such as MRI and EMG for diagnosis in the future with muscle biopsy reserved for specific cases. In order to adapt to upcoming changes in the field, besides including a more extended MSA, MRI and EMG could also be considered for inclusion in the revision of the EULAR/ACR criteria.

This scoping review has several limitations. First, the manuscripts and conference abstracts in non-English languages were excluded which may result in disproportionate representation from English-speaking countries. However, the majority of the publications included in this scoping review were from Asia and Europe. Second, only two data sources were used for study selection. Lastly, there were a limited number of studies that reported specificity of the criteria, which could be due to requiring a control group in the study. Nevertheless, this scoping review was the first to systematically assess the performance of the EULAR/ACR criteria and the findings will inform efforts to revise the EULAR/ACR criteria.

Conclusion

This scoping review, conducted as part of the project aiming to revise the 2017 EULAR/ACR myositis classification criteria, highlights the overall good performance characteristics of the criteria in the classification of IIM as well as critical deficiencies in the subclassification of certain IIM subtypes. Revision of the EULAR/ACR criteria should enable classification of the new subtypes to allow for studies to be conducted in these subtypes which will ultimately facilitate the access of these patients to new therapeutics. Harmonisation of various criteria development efforts will accelerate progress in optimal classification of these patients with IIM.

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