Validation of the EULAR/ACR 2017 idiopathic inflammatory myopathy classification criteria in juvenile dermatomyositis patients

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Abstract

Objective

In 2017, a new set of criteria was proposed by EULAR/ACR to classify idiopathic inflammatory myopathies. Our aim was to validate the EULAR/ACR 2017 classification criteria in juvenile dermatomyositis (JDM) patients.

Methods

This study was carried out at Hacettepe University Children's Hospital Department of Paediatrics, Divisions of Rheumatology, Neurology and Paediatric Pathology Unit. Control patients included inborn errors of metabolism presenting with myopathy and/or rhabdomyolysis, idiopathic rhabdomyolysis, dystrophinopathies, neuromyotonia and systemic rheumatic disorders. Patients' data were collected retrospectively from patient files.

Results

Fifty-eight JDM patients (60.3% female) and 40 controls (32.5% female) were included in this study. When the probability cut-off was set at 55% as recommended, the sensitivity/specificity of the new criteria to diagnose JDM were 96.5%/85% in the total cohort, 95.8%/84.6% without the muscle biopsy data and 97%/85.7% with biopsy data. With the ROC curve analysis, the optimal probability cut-off for the whole cohort was found to be >62%; providing a sensitivity/specificity of 96.6% (95% CI: 88.1% to 99.6)/90% (95% CI: 76.3% to 97.2%), and >68.5% for the patients with muscle biopsy providing sensitivity/specificity of 97% (84.7–99.9%)/100% (76.8–100%), respectively. The new EULAR/ACR criteria were the most sensitive, however, the least specific compared to the Tanimoto criteria (sensitivity/specificity 64%/97.5%) and Bohan-Peter criteria (sensitivity/ specificity 74.1%/92.5%).

Conclusion

The new EULAR/ACR criteria performed favourably in our JDM cohort especially with the probability cut-off of >62%.

Key words

juvenile dermatomyositis, EULAR/ACR criteria, Bohan-Peter criteria, Tanimoto criteria, myositis specific antibodies, muscle biopsy

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Introduction

Juvenile dermatomyositis (JDM) is the most common idiopathic inflammatory myopathy (IIM) affecting 2/1,000,000 children per year (1). It is characterised by the classical skin rash, symmetrical muscle weakness and elevated muscle enzymes due to inflammation in the muscle tissue. Several classification criteria have been proposed (Table I). The Bohan-Peter criteria, published at 1975, is the most widely used one (2, 3). It has five parameters including classical skin rash, symmetrical muscle weakness, muscle biopsy features, electromyography (EMG) findings, and elevated muscle enzymes. The skin rash was not defined in detail but it is a mandatory criterion. An important drawback of this criteria is EMG which has been removed from the routine diagnostic work-up in many centres, especially in younger children, as it is an invasive tool providing very little data specific for JDM. In 1995, Tanimoto published his criteria with nine variables.(4). Tanimoto *et al*. have suggested skin rash as a mandatory criterion, and included a more detailed muscle involvement evaluation and systemic features; however, it fails to diagnose amyotrophic dermatomyositis. Other than Jo-1 antibody, no other autoantibodies were included, which was another important disadvantage of this criteria.

In 2017, EULAR/ACR classification criteria for IIM was proposed and validated in a large cohort including 976 IIM patients and 624 controls (5, 6). These criteria set has two options depending on the presence of the muscle biopsy data. Each variable has a weighted value; at the end, it provides a total score as well as the probability of IIM diagnosis calculated with a web-based calculator or a formula using the total score. A diagnostic tree is available to subclassify the patients. The new criteria include age, detailed muscle weakness and skin rash evaluation, elevation of the muscle enzymes, muscle biopsy features (inflammation, perifascicular atrophy and rimmed vacuole) and anti Jo-1 positivity. There are external validation studies in different adult cohorts however our study is the first external validation study of the new criteria in a paediatric JDM cohort after the original paper.

Methods

Study population

This study was held at Hacettepe University Children's Hospital Department of Paediatrics, Divisions of Rheumatology, Neurology and Paediatric Pathology Unit. In our centre, each JDM patient is evaluated together with the Paediatric Rheumatology and Neurology departments. Thus, the judgment was based on the conjunct opinion of these two groups of experts. The paediatric pathologist contributes if a muscle biopsy is deemed necessary. Definitive diagnosis of JDM and non-JDM patients were made by expert opinion of paediatric rheumatologist (YB >20 years of experience; SO >25 years of experience), paediatric neurologist (GH >20 years of experience) and paediatric pathologist (BT >20 years of experience). JDM patients followed at Hacettepe University Divisions of Paediatric Rheumatology and Neurology between 2000–2020 were included. To test the specificity, patients manifesting with myalgia, muscle weakness, myositis and skin features in whom JDM is one the most possible differential diagnosis were selected as control group. Patients' data were collected retrospectively from patient files.

Methodology

The new criteria have three sets of information; muscle features, skin features and laboratory results including muscle biopsy findings. Each parameter has a weighted score depending on the presence of muscle biopsy. The sum of the scores is then converted into a probability of IIM using a web-based calculator. It can also be calculated according to following formulas (5, 6).

Probability of IIM = 1/ (1 + exponential (6.49 – score)) when muscle biopsy data are present.

Probability of IIM = 1/ (1 + exponential (5.33 – score)) when muscle biopsy data are absent.

The missing data were defined as 'score 0'. A patient was classified as JDM if the patient's probability is above a specified cut-off value. The cut-off probabil-

Table I. Different JDM classification criteria sets.

Bohan-Peter criteria (2, 3)	Tanimoto criteria 4)	EULAR/ACR criteria (5, 6)
1. Symmetric proximal muscle weakness 2. Elevation of the serum levels of skeletal muscle enzymes 3. Electromyography features of myopathy 4. Muscle biopsy evidence 5. Typical skin rash of JDM (Heliotrop rash, Gottron's sign) Definite: Skin rash + 3 out of 4 criteria Probable: Skin rash + 2 out of 4 criteria Possible: Skin rash + 1 out of 4 criteria	1. Skin lesions a. Heliotrop rash b. Gottron's sign c. Erythema on the extensor surface of extremity joints 2. Proximal muscle weakness 3. Elevated serum CK or aldolase levels 4. Muscle pain on grasping or spontaneous pain 5. Myogenic changes on electromyography 6. Positive anti-Jo-1 antibody 7. Non-destructive arthritis or arthralgia 8. Systemic inflammatory signs (fever, elevated CRP or ESR) 9. Pathological findings compatible with inflammatory myositis At least 1 item from criterion 1 and at least 4 criteria from criteria 2-9	1. Age at onset 2. Muscle weakness a. Objective symmetric weakness, usually progressive, of the proximal upper extremities b. Objective symmetric weakness, usually progressive, of the proximal lower extremities c. Neck flexors are relatively weaker than neck extensors d. In the legs, proximal muscles are relatively weaker than distal muscles 3. Skin manifestations a. Heliotrop rash b. Gottron's papule c. Gottron's sign 4. Other clinical manifestations a. Dysphagia or oesophageal dysmotility 5. Laboratory measurements a. Anti-Jo-1 positivity b. Elevated CK or LDH or AST or ALT 6. Muscle biopsy features a. Endomysial infiltration b. Perimysial infiltration c. Perifascicular atrophy d. Rimmed vacuole

CK: creatine kinase; CRP: C-reactive protein; ESR: erythrocyte sedimentation rate; ALT: alanine aminotransferase; AST: aspartate aminotransferase; LDH: lactate dehydrogenase.

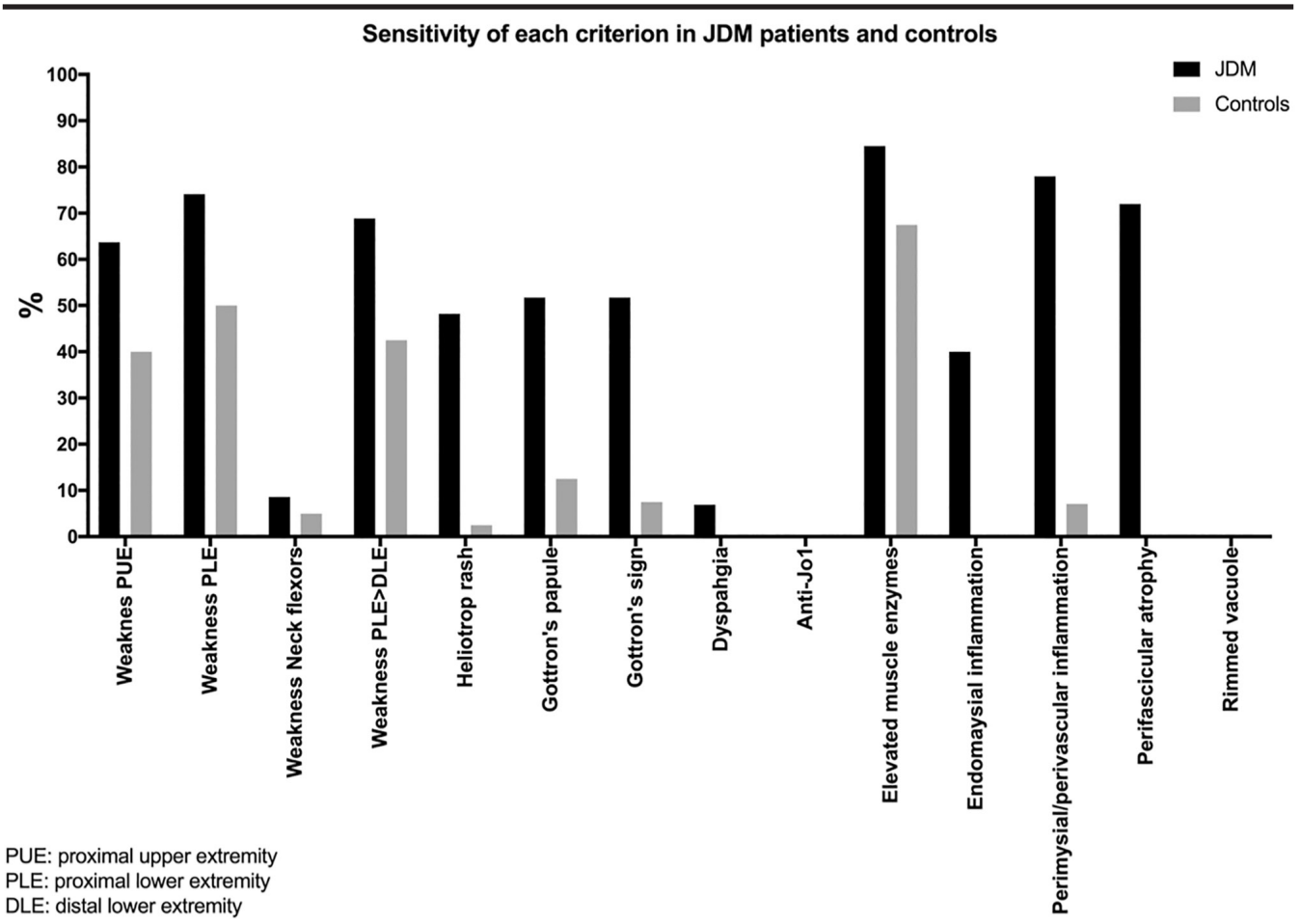

Fig. 1. Sensitivity of each criterion in JDM patients and controls.

Table II. Clinical and laboratory features of the patients.

Clinical features	JDM patients (n=58) *	Control group (n=40)
Female n (%)	35 (60.3%)	13 (32.5%)
Age at disease onset (mean±SD years)	8.1 ± 4.3	8.7 ± 5.4
Age at the time of diagnosis (mean±SD years)	8.7 ± 4.4	9.9 ± 5.3
Classical rash n (%)	53 (91%)	9 (22.5%)
Gottron's papule	43 (74%)	8 (20%)
Heliotrop rash	28 (48%)	1 (2.5%)
Symmetrical proximal muscle weakness n (%)	44 (76%)	20 (50%)
Upper extremity	37 (64%)	16 (40%)
Lower extremity	43 (74%)	20 (50%)
Gowers sign n (%)	40 (69%)	23 (58%)
MRC score of quadriceps femoris (mean ± SD)	3.71 ± 1.02	3.9 ± 1.1
Other skin features (malar rash, V sign, facial erythema) n (%)	19 (33%)	15 (38%)
Calcinosis n (%)	5 (9%)	4 (10%)
Dysphagia, swallowing dysfunction	4 (7%)	-
Skin ulceration n (%)	2 (3%)	4 (10%)
Laboratory investigations	JDM patients	Control group
Haemoglobin gr/dL (median/IQR)	12.2 (11.4-13.1)	12.3 (11.4-14.0)
White blood cells x10^3/mm^3 (median/IQR)	7950 (6275-9525)	7150 (4875-9500)
Platelets x10^3/mm^3 (median/IQR)	274500 (233500-371500)	274000 (225500-320750)
Erythrocyte sedimentation rate mm/hr (median/IQR)	13 (6-21)	17 (5-38)
C-reactive protein mg/dL (median/IQR)	0.18 (0.10-0.31)	0.5 (0.2-1.9)
Creatine kinase U/L (median/IQR)	337 (87-1913)	1568 (163-13876)
Lactate dehydrogenase U/L (median/IQR)	629 (434-865)	605 (359-1277)
ANA positivity n (%)	28/36 (77%)	14/21 (66.6%)
ENA positivity n (%)	4/30 (13%)	6/14 (42.8%)
MSA/MAA positivity n (%)	34/46 (75.9%)	NA
NXP2	10 (21.7%)	
TIF-1g	8 (17.4%)	
MDA5	4 (8.7%)	
Mi-2	4 (8.7%)	
Others	8 (17.4%) (3 PM-Scl, 2 Ku, 1 OJ, 1 PL12, 1 HMGCR)	
Negative	12/46 (24.1%)	
EMG showing myopathy	23/25 (92%)	5/8 (62.5%)
MRI showing myositis	12/15 (80%)	-
Muscle biopsy performed	35/58 (60.3%)	14/40 (35%)

MRC: Medical Research Council; ANA: anti-nuclear antibody; ENA: extractable nuclear antigen; MSA: myositis-specific antibody; MAA: myositis-associated antibody; EMG: electromyography; MRI: magnetic resonance imaging; IQR: interquartile range; SD: standard deviation; NA: not available.
*The features of our JDM patients have also been presented as our single-centre experience as well (submitted work).

ity was set at 55%, as recommended by the new EULAR/ACR criteria.

Each patient's score was also calculated for the Bohan-Peter criteria (2, 3) and Tanimoto criteria (4). The patients were then dichotomised into none-low/possible *versus* probable/definite JDM. Sensitivity, specificity, positive, and negative predictive values of the EULAR/ACR scoring criteria with/without muscle biopsy against the gold standard clinician's diagnosis were calculated and used to assess the diagnostic accuracy of the EULAR/ACR classification criteria, and compared with those of the Bohan-Peter and Tanimoto criteria.

Receiver operator characteristic (ROC) analysis was used to determine a suggested cut-off probability for the EULAR/ACR classification criteria in our paediatric cohort. The Youden method was considered when determining the most appropriate cut-off values.

Results

Demographic features of the patients

Fifty-eight (60.3% female) JDM patients and 40 non-JDM (32.5% female) control patients were included in this study. The control group was composed of inborn errors of metabolism presenting with myopathy and/or rhabdomyolysis [multiple acyl-CoA dehydrogenase (MADD) deficiency (n=8), carnitine-palmitoyl transferase II deficiency (n=2), long-chain 3-hydroxy/acyl-CoA dehydrogenase deficiency (n=1)], idiopathic rhabdomyolysis (n=3), dystrophinopathies [Duchenne/Becker muscular dystrophy (n=10)], neuromyotonia (n=1) and systemic rheumatological disorders [systemic lupus erythematosus (n=5), mixed connective tissue disorders (n=4), interferonopathies (n=4), polyarteritis nodosa (n=2)].

The mean age at disease onset was 8.1±4.3 years in the JDM patients and 8.7±5.4 years in the control group. The mean age at diagnosis was 8.7±4.4 years in the JDM patients and 9.9±5.3 years in the control group. The classical skin rash (91%) was the most common finding in the JDM group and elevated muscle enzymes (67.5%) was the most common finding in the control group. The sensitivities of each criterion are presented in Figure 1. Electromyogra-

phy was performed in 43% of JDM patients (92% of them showing myopathy) and 20% (62.5% of them showing myopathy) of the control group. Muscle biopsy was performed in 60.3% (n=35) of the JDM patients and 35% (n=14) of the control group. All of the muscle biopsies of the JDM patients had the classical JDM histopathology. 34 out of 46 (76%) JDM patients were myositis-specific or myositis-associated antibody positive. Clinical and laboratory features compared with the control group are summarised in Table II.

Sensitivity and specificity of the new EULAR/ACR criteria in the paediatric JDM population

The new EULAR/ACR criteria recommends the optimal probability as 55% for both patients with and without muscle biopsy, providing the optimal balance between sensitivity and specificity. We have analysed our cohort with receiver operator curve (ROC) analysis, and calculated different probability cut-off values with Youden's index (Fig. 2). The analysis was done separately in patients with and without muscle biopsy data as well.

When the probability cut-off was set at 55% as recommended in the original article (5, 6), the sensitivity/specificity of the new criteria to diagnose JDM were 96.5%/85% in the total cohort, 95.8%/84.6% without muscle biopsy data and 97%/85.7% with biopsy data. Adding subclassification tree analysis decreased the sensitivity of the EULAR/ACR criteria from 96.5% to 84.5% however increased the specificity from 85% to 90%.

The optimal probability cut-off for the whole cohort was calculated as 62% giving a sensitivity of 96.5% (95% CI: 88.1–99.6%) and a specificity of 90% (76.3–97.2%). The optimal cut-offs were found at 53% in patients without muscle biopsy data; sensitivity and specificity were 95.6% (78.9–99.9%) and 84.6% (65.1–95.6%), respectively, and at 68.5% in patients with muscle biopsy data; sensitivity and specificity were 97% (84.7–99.9%) and 100% (76.8–100%), respectively. ROC curve analysis indicated that the area under the curve for all cases, the cases without muscle biopsy data and the cases with muscle biopsy data were 0.97, 0.95 and 0.99, respectively.

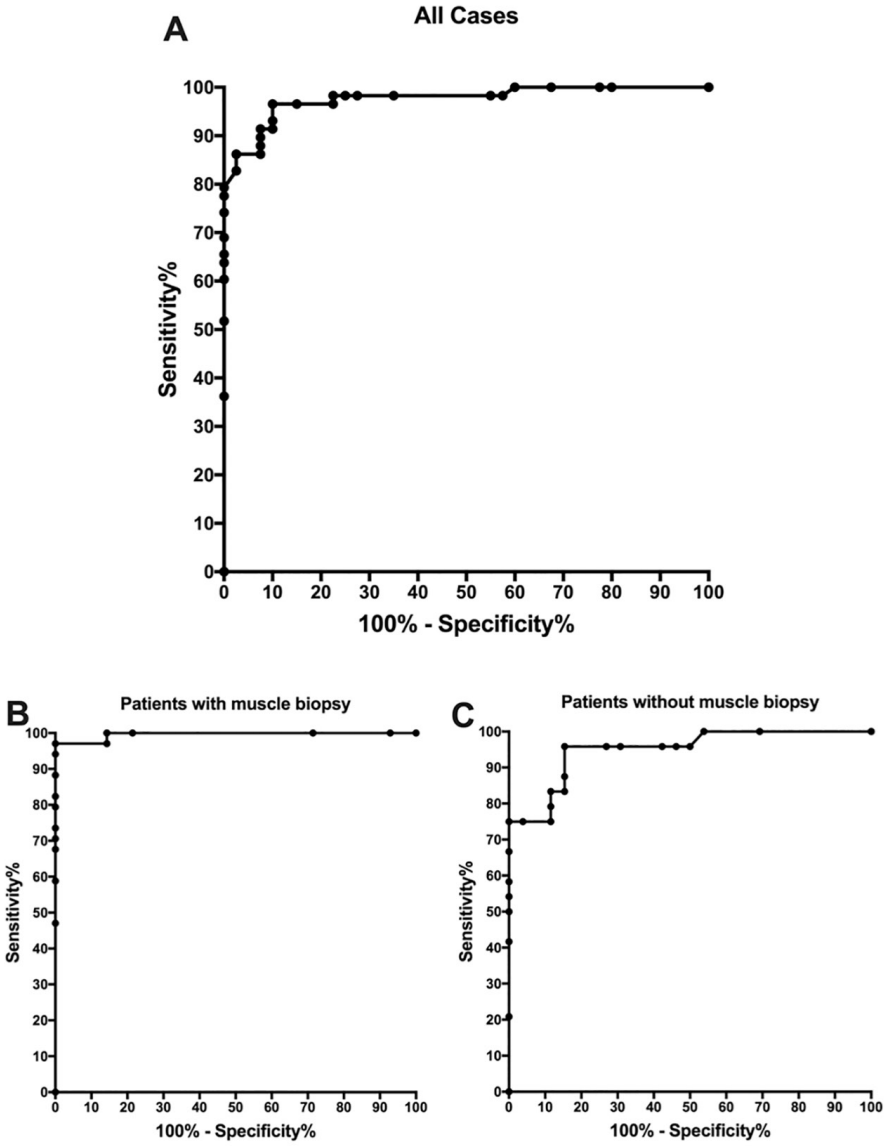

Fig. 2. ROC analysis of the new EULAR/ACR criteria in JDM patients. Sensitivity/1-specificity of the EULAR/ACR classification criteria for JDM in all cases (**A**), the cases with muscle biopsy data (**B**) and the cases without muscle biopsy data (**C**). The probability cut-off was calculated with Youden's index (ROC: receiver operator curve; JDM juvenile dermatomyositis)

Comparison of the new EULAR/ACR criteria with other classification criteria

We compared the sensitivity and specificity of the new EULAR/ACR criteria using the recommended cut-off of 55% with the previously published criteria (Bohan-Peter and Tanimoto criteria). The new EULAR/ACR criteria (sensitivity/specificity 96.5%/85%) was the most sensitive, however, the least specific compared to the Tanimoto criteria (sensitivity/specificity 64%/97.5%) and Bohan-Peter criteria (sensitivity/ specificity 74.1%/92.5%). The new EULAR/ACR criteria had the lowest (90.1%) positive predictive value (Bohan-Peter criteria 93.4%, Tanimoto criteria 97.3%) and the highest (94.4%) negative predictive value (Bohan-Peter criteria 71.1%, Tanimoto criteria 65%) (Table III).

Discussion

This study is the first external validation of the new EULAR/ACR criteria in a pure paediatric JDM population after the original study.

Table III. Sensitivity and specificity of different criteria for classification of JDM.

	Sensitivity (n=58)	Specificity (n=40)	Positive predictive value	Negative predictive value
EULAR/ACR criteria	96.5% (56/58)	85% (34/40)	90.1%	94.4%
without biopsy	95.8% (22/23)	84.6% (22/26)	85.1%	95.6%
with biopsy	97% (34/35)	85.7% (12/14)	94.3%	92.3%
subclassification tree analysis	84.5% (49/58)	90% (36/40)	92.5%	83.7%
Bohan-Peter criteria	74.1% (43/58)	92.5% (37/40)	93.4%	71.1%
Tanimoto criteria	64% (37/58)	97.5% (39/40)	97.3%	65%

Bohan-Peter classification criteria have been the most widely used criteria since 1975 (2, 3). In 1995 Tanimoto *et al*., proposed a new set of criteria as well (4). In this study, we have also compared the sensitivity and specificity of these criteria with the new EULAR/ACR criteria. The new criteria have the highest sensitivity, but on the other hand the lowest specificity. The classical rash is a mandatory criterion in both Bohan-Peter and Tanimoto criteria, and is also included in the subclassification tree of the new criteria. The muscle biopsy data slightly increased the sensitivity and specificity of the new criteria as well. The adult IIM cohorts testing the new criteria revealed a sensitivity between 80.2-92.7% and the specificity between 87–98% (7-11). However, the paediatric JDM population was not adequately represented in any of these studies. The original EULAR&ACR paper reported the sensitivity and specificity of the new criteria as 87–93% and 82–88%, respectively in adult IIM patients. This paper also included JDM patients, yielding a good sensitivity, however despite including 115 juvenile comparators in the criteria development cohort, no childhood controls were included in the validation cohort thus specificity was not available (6).

The shift of the probability cut-off from 55% to 62% in the whole cohort provided only very subtle (5%) change in the specificity, thus the cut-off of 55% could be acceptable for JDM patients as well, validating the performance of the new criteria in JDM patients. However, the probability cut-off of 68.5% in patients with muscle biopsy data remarkably improved the specificity in this group, thus this value might be useful when the muscle biopsy data is available.

In our control group we have mainly included challenging patients whom the caring physician listed JDM on the top of the differential. This selection bias might influence the specificity, however, classification criteria should help to differentiate especially these patients in the routine daily clinic. In our control group, 37.5% of the patients had systemic inflammation, 27.5% had metabolic myopathies and 25% had dystrophinopathies; which was similar to the original validation study (6). Six (15%) control patients were misclassified as JDM with the new criteria. Muscle weakness parameters lowered the specificity and led to misclassification for three patients with inborn errors of metabolism. Of note, 6 out of 8 patients with a diagnosis of MADD deficiency within the metabolic diseases group had an initial diagnosis of inflammatory myopathy, representing challenges on clinical grounds. Two patients with interferonopathy and one with mixed connective tissue disorder presented with skin features resulting in misclassification as well. Adding subclassification tree analysis increased the specificity of the new criteria for the sake of the sensitivity.

The recent advances in the field of myositis-specific/myositis-associated antibody (MSA/MAA) improved our insight into JDM. These antibodies provided valuable information to predict the probable comorbidities and the disease course. The most common MSA/MAAs in JDM were NXP2, TIF-1g, Mi-2 and MDA5 in different cohorts (12-15). In the new criteria only Jo-1 positivity has a definitive value, but only 2–5% of the JDM patients are Jo-1 positive (1, 16) ; in fact, we have no anti-Jo1 positive patients. We then included all the MSAs in the criterion of Jo-1 autoantibody, which increased the sensitivity to 98.3% without changing the specificity Having MAA/MSA as a distinct criterion in the classification criteria might increase the sensitivity and specificity but should include other MSA/MAAs, at least the most common ones (NXP2, TIF-1g, Mi-2 and MDA5) especially in the JDM population.

There has been a shift towards use of non-invasive diagnostic tools in parallel to the advancing technology. Electromyography was substituted with muscle MRI as it can provide more specific data, especially in patients with patchy involvement. There are some reports showing that either whole-body MRI or thigh MRI are helpful diagnostic tools, and their findings correlated with the extent of the muscle pathology (17, 18). There is an imaging scoring system for JDM based on muscle MRI findings too (19) , In our cohort 12 out of 15 JDM patients had myositis in muscle MRI of the upper leg. In contrast to Bohan-Peter and Tanimoto criteria, the new EULAR/ACR criteria excluded EMG findings from the variable list, which is in accordance with our daily practice. MRI may well be considered and discussed in future revisions of the criteria.

Despite being invasive, muscle biopsy still stands as the gold standard tool providing valuable data on the type, degree and extent of the muscle pathology and even some prognostic features. There is a muscle biopsy scoring tool to evaluate muscle histopathology, validated in JDM patients (20, 21). This score tool is composed of four different domains; inflammatory domain, muscle fibre domain, connective tissue domain and vascular domain. In the new criteria only a part of the inflammatory domain and perifascicular atrophy was included. In our cohort only 78% of the patients had perifascicular atrophy and 72% of them had inflammation on their muscle biopsy samples. On the other hand, 94% of our patients had MHC (major histocompatibility complex) class-1 overexpression which is highly suggestive for JDM and was not included in the new criteria. Instead, the presence of rimmed vacuole which is an important feature of inclusion body myositis was included although it is not a feature of JDM (16, 22). Tak-

ing into account the muscle biopsy data increased the positive predictive value of the new criteria, but the sensitivity and specificity might be increased if the biopsy parameters for the criteria are revised according to the JDM biopsy score tool features suggested by Wedderburn *et al*. (17).

This study has some limitations. There might be selection bias in purpose while choosing the control group patients which may lower the specificity of the new criteria. Retrospective evaluation of the skin features may not be reliable, thus further studies including prospective cohorts may be useful. Unknown or missing data were regarded as "score 0" as recommended in the original paper (6).

Conclusion

The new EULAR/ACR criteria performed favourably in our JDM cohort especially with the probability cut-off of >68.5% in patients with muscle biopsy data. One can speculate that the yield of the criteria in childhood presentations may be improved by including the recently identified myositis-specific antibodies and validated muscle biopsy score tool parameters.

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