Editorial

The classification of myositis: setting the stage for a universal terminology

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Idiopathic inflammatory myopathies are a group of systemic autoimmune diseases characterised by skeletal muscle weakness often accompanied by extramuscular organ involvement, affecting skin, joints, lungs, gastrointestinal tract, and the heart (1). Even though these inflammatory myopathies are usually described as "idiopathic", specific causes have been identified in many cases. Because of the autoimmune-mediated pathophysiology, the term "autoimmune myositis (AIM)" has been proposed but has not been generally accepted. As such, we will use "myositis" as an umbrella term in this Editorial.

In contrast with the International Classification of Diseases (ICD), ICD-10 and ICD-11, identifying "Dermatopolymyositis" (2) and "Dermatomyositis/Inclusion body myositis/polymyositis" (3), respectively, myositis constitutes a heterogeneous group that encompasses several subtypes, which show substantial differences in clinical features, pathogenesis, disease course, prognosis, association with cancer, and therapeutic management.

Classification of myositis has been a source of considerable debate in the field, with increased recognition of these subtypes over the last decades. This is not simply due to semantics, but the identification of homogeneous disease subtypes with distinct phenotypes and pathophysiology can have important implications for diagnosis, treatment approaches, clinical trial design, and outcome measures.

This editorial aims to provide an overview of the terminology of various classifications and diagnostic criteria for myositis, summarise contemporary issues, and discuss the need for future design of globally unified and clinically relevant criteria.

Historical context

Since the Bohan and Peter Criteria, which described five subtypes of myositis (primary idiopathic polymyositis [PM], primary idiopathic dermatomyositis [DM], DM/PM associated with neoplasia, childhood DM/PM associated with vasculitis, and DM/PM with associated collagen-vascular disease) (4, 5) in 1975, much progress has been made in the understanding of myositis heterogeneity. These criteria led to an overdiagnosis of PM for many years due to initial difficulties distinguishing PM from other neuromuscular disorders (*i.e.* muscular dystrophy, metabolic myopathies) (6); secondly, the term PM included subgroups that were later identified as distinct conditions (7, 8). Notably, a proportion of the patients who were diagnosed as PM, based on the Bohan and Peter criteria, were later found to be suffering from a different myositis subgroup, namely inclusion body myositis (IBM) (7, 8). IBM has distinctive clinical and histopathological features compared with PM and DM, consisting of a slowly progressive course, failure to respond to corticosteroid treatment, a specific pattern of muscle involvement, and muscle histopathology (9).

The discovery of several myositis-specific (MSA) and myositis-associated (MAA) autoantibodies associated with distinct clinical features (10) led to a different approach to myositis classification (11). Following such an approach, a separate subclassification of overlap myositis (OM) was proposed by Troyanov *et al.* (11) and defined as the association of myositis with over-

lapping features of another connective tissue disease (*i.e.* polyarthritis, interstitial lung disease [ILD], Raynaud's phenomenon, sclerodactyly, scleroderma proximal to MCP joints, lower oesophageal or small-bowel hypomotility) or autoantibodies associated with clinical overlap phenotypes (*i.e.* anti-PM/Scl, -Ku, -U1-RNP). This entity, accounting for up to 50% of myositis patients (12), is a subtype distinct from PM and DM in terms of clinical features, course, prognosis, association with cancer, and therapeutic response (13-15). OM occurs as myositis in combination with systemic sclerosis (SSc) (16), systemic lupus erythematosus (SLE) (17), rheumatoid arthritis (RA) (18), or Sjögren's syndrome (SS) (19, 20). There is no generally accepted consensus or guidance on diagnosis and care of patients with these overlapping conditions.

On the other hand, the recognition of distinct histopathological patterns in myositis led to the publication of criteria that delineated seven myositis subsets: IBM, PM, DM, amyopathic DM (ADM), possible DM sine dermatitis, nonspecific myositis, and immune-mediated necrotising myopathy (IMNM) (21), according to a worldwide initiative of multidisciplinary researchers and clinicians attending a European Neuromuscular Centre (ENMC) workshop in 2003.

The first of these new approaches to myositis classification was based on the close autoantibody-phenotype association (11); the second was the enhanced understanding of the nuances of muscle histopathology (21, 22).

Later, combining both approaches led to a clinical-sero-pathological classification of myositis that contributed to a more accurate diagnosis (23). Based on this combined approach, ENMC produced classification criteria for IBM (24, 25), DM (26), IMNM (27), and more recently for the anti-synthetase syndrome (ASyS) (28). However, the diagnostic performance of these expert opinion-based criteria is often unknown and needs further validation as demonstrated for the previous ENMC 2013 criteria on IBM (29).

Moving to the latest classification system, a large international multidisciplinary collaborative group of myositis experts, including adult and pediatric rheumatologists, neurologists, dermatologists, epidemiologists, and biostatisticians, was assembled in 2004 to develop a data-driven myositis classification criteria for the first time. With the endorsement of the European League of Associations for Rheumatology (EULAR) and the American College of Rheumatology (ACR), the myositis classification criteria were named EULAR/ACR myositis classification criteria and published in 2017 (30). The criteria aimed to distinguish myositis from mimickers and categorise myositis, with a minimum of clinical and histopathological features, into the following major subgroups: PM, IBM, DM, ADM, juvenile DM, and other juvenile myositis. These data-driven criteria, already partially validated at the time of publication, represented a significant step forward in the myositis classification (31) (sensitivity and specificity: 87%/82% without biopsies, 93%/88% with biopsies, respectively) (30).

The historical evolution of the different myositis classifications is summarised in Table I.

Contemporary issues

Given the scarce availability of the other MSA at the time of the criteria development, the EULAR/ACR criteria only included the anti-Jo autoantibody as MSA, resulting in significant shortcomings in the classification of several more recently recognised subgroups, including ASyS and IMNM (30). Moreover, the criteria mainly focused on muscle and skin involvement, while extramuscular signs (*i.e.* arthritis, ILD, Raynaud's phenomenon) of myositis were not included. These two points are of utmost importance if we consider that i) anti-tRNA-synthetase antibodies (ARS) are characteristic of ASyS (32, 33), and ii) the main three clinical features of ASyS (arthritis, ILD, and myositis) can remain isolated throughout follow-up (34). ASyS can potentially be included in the separately defined overlap myositis group (11), but histological (35) and molecular (36, 37) evidence indicate that ASyS is a distinct entity in the myositis spectrum. In a separate study, hierarchical cluster analysis of phenotypic, biological, and immunologic variables also suggested a classification with four subgroups: DM, IBM, IMNM, and ASyS (23).

Similarly to anti-ARS, several autoantibodies not considered by the 2017 EULAR/ACR myositis classification criteria, such as anti-PM/Scl, -Ku, -U1-RNP, -U3-RNP, -RuvBL1/2, -SMN, have been specifically associated with myositis and clinical features of SSc or certain disease features, such as ILD. Indeed, the respective ACR/EULAR criteria for myositis (30) and SSc (38) lack sensitivity for these overlap patients (39). These patients, included in overlap myositis (23), show specific myopathological abnormalities and molecular signatures, response to treatment, and prognosis (40-44). This has led to the increased use of the term "scleromyositis", although it has not been universally accepted as of yet (16). In summary, the 2017 EULAR/ACR myositis classification criteria do not recognise ASyS or "scleromyositis" as distinct entities (45), leading not only to an underestimation of myositis incidence by more than one-quarter (46) but also limiting the access for these groups of patients to randomised clinical trials dedicated to myositis. To overcome this unmet need, an EULAR/ACR initiative (CLASS project) (47) aimed to develop and validate new ASyS classification criteria.

Despite the overall good performance of the EULAR/ACR criteria in the classification of myositis, it also showed several shortcomings, most of all in that IMNM was not contained as individual subtype (30). When the performance of the EULAR/ACR criteria was assessed, IMNM was the most common myositis type to be classified as non-myositis (45), and even when the patients with IMNM were classified as myositis, they were most commonly misclassified as PM, rarely as DM or IBM (in cases with severe IMNM affecting distal muscles), as this subgroup had only recently been recognised when the classification criteria project had started in 2004 (45). Indeed, IMNM is now established as a distinct entity (48) and evidence of an autoantibody-driven pathogenesis (49).

Table I. The historical evolution of classification criteria in myositis.

Myositis subgroups	Bohan and Peter 1975 (4, 5)	Dalakas 1991 (9)	Tanimoto 1995 (70)	ENMC 2004 (21)	Troyanov 2005 (11)	EULAR/ ACR 2017 (30)
Dermatomyositis (DM)	+	+	+	+	+	+
Clinically amyopathic DM	-	-	-	+	-	+
Juvenile DM (JDM)	+	-	-	-	-	+
Polymyositis (PM)	+	+	+	+	+	+
Juvenile myositis other than JDM	+	-	-	-	-	+
Inclusion body myositis	-	+	-	(+)	(+)	+
Immune-mediated necrotising myopathy	-	-	-	+	-	-
Anti-synthetase syndrome	-	-	-	-	-	-
Overlap myositis	+	-	-	-	+	-
Non-specific myositis	-	-	-	+	-	-

+: the subgroup is included in the classification. (+): the subgroup is mentioned in the paper.
-: the subgroup is not included in the classification.

Whereas some patients with IMNM may be seronegative, the diagnostic value of anti-SRP or anti-HMGCR autoantibodies is high in typical forms characterised by a rapidly progressive and severe muscle disability with high serum creatine kinase levels (48). Including an adequate number of cases with anti-SRP or anti-HMGCR positive IMNM or histopathological features of necrotising myopathy in the muscle biopsy item could improve the accuracy of classification of these patients (45, 50).

By recognising all these conditions (IMNM, ASyS, etc.) as distinct myositis subtypes (50), the number of patients classified as pure PM has continued to decrease, and PM can now be considered a rare myositis subgroup (51). Indeed, some have questioned the existence of pure PM at all (7, 23), and some consider it a diagnosis of exclusion because of its nonspecific phenotype (subacute proximal weakness without overlap features or autoantibodies), which is at high risk for mimickers (12).

While a strength of the EULAR/ACR criteria is the ability to subclassify clinically amyopathic DM (CADM) (30), external validation studies showed a variable sensitivity for CADM between 33% to 100% (45). This wide variability is due to the fact that patients with 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 listed in the EULAR/ACR criteria, do not meet the minimum threshold to be classified as DM (45). Thus, to facilitate the diagnosis of CADM and include these patients in clinical trials, an extensive Delphi process involving 50 global dermatology and rheumatology experts generated a list of 25 items for the classification of skin-predominant DM (52).

Adult-onset myositis is associated with an increased cancer risk within the three years preceding and following myositis onset (53), and a cancer-associated myositis has been described (4, 5, 11). Indeed, high-risk factors for cancer in myositis have been identified, including a diagnosis of DM, anti-TIF1-γ and anti-NXP2 antibodies positivity, age >40 years at the time of myositis onset, persistent high disease activity despite immunosuppressive therapy (including relapse of previously controlled disease), moderate to severe dysphagia, cutaneous necrosis or ulceration (13). However, other myositis subgroups such as DM with anti-SAE1, -Mi2, and -MDA5 antibodies and IMNM with anti-HMGCR antibodies also have an intermediate cancer risk according to the International Guideline for myositis-associated Cancer Screening (13). This indicates that cancer-associated myositis is not a distinct myositis subgroup in the myositis spectrum, but the disease subtype is one of the risk factors for cancer.

The 2017 EULAR/ACR criteria identify JDM and juvenile myositis other than DM (30). However, several reports show that MSA found in adult myositis can also be found in juvenile myositis, indicating that a similar phenotypic spectrum may exist in children.

Other types of myositis which are clinically and histopathologically distinct from the myositis subgroups discussed above are immune checkpoint inhibitor-induced myositis (54), myositis associated with systemic vasculitis (55), idiopathic orbital myositis (which can also be observed in the context of other autoimmune diseases, *i.e.* RA, SLE, SS) (56), eosinophilic myositis (57), giant cell or granulomatous myositis (58), focal myositis (59), myositis associated with graft *versus* host disease (60), myositis in association with inflammatory bowel diseases (61), and monogenic autoinflammatory diseases (62). Conditions with disparate findings on serology, clinical presentation and histology should be termed non-specific (21) and closely monitored during follow-up if a more precise diagnosis can be reached over time. The terminology of IBM has recently been revised in that hIBM or sIBM should be avoided; the respective gene should be used instead for hereditary cases (24, 25).

Due to these critical issues, a new global interdisciplinary collaboration of clinicians is currently revising the 2017 EULAR/ACR criteria to provide a more accurate classification of myositis based on new findings (50).

Proposals for the use of a unified terminology

Given the evolution of the myositis classification above, we here suggest using the terminology provided in Table II. In addition, we recommend avoiding the word "idiopathic" in the context of myositis terminology. The terms "dermatopolymyositis" or "polydermatomyositis" should no longer be used. Diagnosis of the subform polymyositis (PM) should be restricted only to patients who fulfil the most recent criteria. The anti-synthetase syndrome has been described with several acronyms such as ASS, ASyS, ASynS, ASSD, ARSE, etc. To provide a more universally consistent abbreviation, we propose to use ASyS.

However, some points remain to be discussed. Recently, transcriptomic data have revealed unique gene expression

Table II. Current terminology of myositis.

Proposed myositis subgroups*	Current acronym	Other acronyms
Paediatric		
Juvenile dermatomyositis	JDM	-
Other juvenile myositis	JM	JPM
Adult		
Dermatomyositis	DM	-
Clinically amyopathic dermatomyositis	- CADM	ADM
Polymyositis	PM	-
Inclusion body myositis	IBM	-**
Immune-mediated necrotising myopathy	IMNM	NAM, NM
Anti-synthetase syndrome	ASyS	ASS, ASynS, ASSD, ARSE
Overlap myositis (*e.g.* associated with SSc, SLE, RA, etc.)	OM	-
Non-specific myositis#	-	-

NAM: necrotising autoimmune myopathy; NM: necrotising myopathy; RA: rheumatoid arthritis; SLE: systemic lupus erythematosus; SSc: systemic sclerosis.
*This list is not exhaustive and other myositis subgroups exist; however, only major myositis subgroups defined in international diagnostic criteria were included. Distinct, rare subforms exist, such as focal myositis, granulomatous myositis, eosinophilic myositis, orbital myositis, etc.
**A recent international ENMC workshop consented to avoid previous terms such as hIBM or sIBM (25).
#Non-specific myositis should be used in cases with myositis that do not (yet) fulfil diagnostic criteria, *e.g.* when serological, histological and clinical criteria are inconclusive.

profiles in muscle biopsies from patients with MSA-defined subtypes of myositis (30). Thus, any future myositis classifications might take advantage of biomarkers identified by bioinformatics, machine-learning tools (63) and multi-omics approaches.

A notable downside of identifying further subtypes of patients in major myositis subgroups would be that it could potentially hamper the recruitment of patients into clinical trials and increase the risk that these trials will take too much time and resources to conduct due to the rarity of the disease (64). A more pragmatic approach has been proposed by the investigators that led to the approval of the first treatment for DM, intravenous immunoglobulins (65). In this study, all patients showed a DM rash. However, in accordance with previously reported data (66), 20% of the study population tested positive for anti-ARS and would be classified as ASyS, and 13% as "scleromyositis" according to myositis classification (12, 67). Indeed, the term "scleromyositis" should be avoided since no generally accepted definitions are available. Cases with an overlap of SSc and myositis should be defined as OM.

In conclusion, classifying myositis is a work in progress. MSA and MAA have been found in up to 80% of people suffering from myositis (68). Moreover, autoantibodies are linked to clinical phenotypes and recently, their critical role in myositis pathogenesis has been further elucidated (49, 69). Thus, any new myositis classification should aim to integrate autoantibody findings, and consider extramuscular disease manifestations. Nevertheless, we propose here to define a universal terminology, based on most up to date understanding. It should be in all our interest to utilise a harmonised, universal terminology in order to improve diagnostic accuracy, standardise treatment approaches, and progress clinical trial design.

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