Review

Idiopathic inflammatory myopathies: one year in review 2025

M. Diomedi[1], F. Fattorini[1], S. Aliberti[2], L.M. Bianchessi[3], A. Castellucci[4], J. Schmidt[5], L. Cavagna[3,6], L. Carli[1], S. Barsotti[7]

[1]Rheumatology Unit, University of Pisa, Italy; [2]Dept. of Experimental and Clinical Medicine, University of Florence, Section of Rheumatology, AOU Careggi, Florence, Italy; [3]Rheumatology Unit, Fondazione IRCCS Policlinico San Matteo, Pavia, Italy; [4]Rheumatology Unit, Dept. of Medicine and Surgery, University of Perugia, Italy; [5]Dept. of Neurology, Neuromuscular Center, University Medical Center Göttingen; Dept. of Neurology and Pain Treatment, Immanuel University Hospital Rüdersdorf, Brandenburg Medical School Theodor Fontane, Rüdersdorf; and Faculty of Health Sciences Brandenburg, Brandenburg Medical School Theodor Fontane, Rüdersdorf, Germany; [6]Dept. of Internal Medicine and Therapeutics, Università di Pavia, Italy, [7]Dept. of Rheumatology, ASL Toscana Nord Ovest, Ospedale Versilia, Lido di Camaiore, Italy.

Michele Diomedi, MD
Federico Fattorini, MD
Sabrina Aliberti, MD
Lorenzo Mattia Bianchessi, MD
Andrea Castellucci, MD
Jens Schmidt, MD, FEAN, FAAN
Lorenzo Cavagna, MD, PhD
Linda Carli, MD, PhD
Simone Barsotti, MD, PhD

Please address correspondence to:
Simone Barsotti
Reumatologia, ASL Toscana Nord Ovest, Ospedale Versilia, SS 1 n. 335, 55041 Lido di Camaiore (LU), Italy.
E-mail: simone.barsotti.pisa@gmail.com

Received on January 25, 2026; accepted in revised form on February 5, 2026.

Clin Exp Rheumatol 2026; 44: 167-177.

Key words: review, myositis, antisynthetase, inclusion body, treatment

Competing interests: J. Schmidt has received payments for advisory boards, speaker's honoraria, travel expenses, reasearch projects from Abcuro, Argenx, Biotest, CSL Behring, Johnson & Johnson, Kezar, LFB, Lupin, Takeda and UCB. The other authors have declared no competing interests.

ABSTRACT

Idiopathic inflammatory myopathies (IIMs) are a heterogeneous group of autoimmune diseases characterised by skeletal muscle inflammation and frequently by involvement of other organs, in particular lung and skin, but also joints, heart and gastrointestinal tract.

Although they are rare diseases, the literature on IIMs has been growing rapidly and many studies have been published in order to clarify pathogenesis and to better define diagnosis, clinical manifestations (muscular and extramuscular) and treatment.

The purpose of this review article is to summarise the most relevant contributions published over the last year on this topic.

Introduction

Idiopathic inflammatory myopathies (IIMs) comprise a heterogeneous group of rare autoimmune disorders characterised by diverse clinical presentations and complex pathogenic mechanisms. Although progressive, symmetrical proximal muscle weakness remains the classical hallmark, extra-muscular involvement is frequent and may significantly influence disease course and prognosis. Pulmonary manifestations, particularly interstitial lung disease (ILD), represent one of the most impactful complications, but cutaneous, articular, cardiac and gastrointestinal involvement may also occur. This broad clinical variability highlights the need for a multidisciplinary approach to ensure timely diagnosis, risk stratification and personalised management (1).

Advances in the characterisation of myositis-specific autoantibodies (MSA), combined with distinctive clinical and histopathological features, have led to a more refined subclassification of IIMs. Current categories include dermatomyositis (DM), antisynthetase syndrome (ASyS), immune-mediated necrotising myopathy (IMNM), inclusion body myositis (IBM), polymyositis (PM) and overlap myositis (OM) (2).

In continuity with previous contributions to the "One Year in Review" series (3), the present review summarises original studies published over the past year addressing key advances in the understanding of IIMs. A systematic search of the PubMed database was conducted to identify English-language articles published between 1 July 2024 and 30 June 2025. The following terms were used: "idiopathic inflammatory myopathies", "myositis" (MeSH and semantic search), "pathogenesis", "diagnosis", "clinical manifestations" and "therapy". Titles and abstracts were screened for relevance, and full texts of potentially eligible studies were reviewed. The most pertinent articles were selected and are discussed in this review.

Pathogenesis

Genetic predisposition, immune dysregulation, and muscle-intrinsic factors may converge to drive chronic inflammation and tissue damage in IIM.

A key advancement about genetic predisposition has been proposed, particularly the role of specific HLA alleles across different IIM subtypes. A systematic review and meta-analysis confirmed significant associations between several HLA-DRB1 alleles and IBM while *HLA-DRB1*15:01* emerged as protective. These associations exhibit ethnic variability, being most prominent in Caucasians and differing in East Asian populations (4). In INMN with anti-HMGCR antibodies positivity, the predominance of HLA-DRB1*11:01 suggests that genetic susceptibility is pivotal even in statin-naïve patients, challenging the view that statins are the primary trigger (5).

Epigenetic mechanisms are also gaining attention. In IBM, under-expression of miR-16-5p leads to overexpression of MHC-I molecules on muscle fibres, providing a mechanistic link between microRNA dysregulation and sustained immune activation (6).

At the cellular level, recent studies have refined our understanding of the specific immune profiles that characterise IIM subsets. For instance, activated dendritic cell (DC) subpopulations have been identified as key features in the muscle tissue of IBM patients. These activated DCs correlate with the presence of KLRG1$^+$ and TBX21-expressing CD8$^+$ T lymphocytes, reinforcing the type 1 polarised immune response typical of this subtype and suggesting a role for DCs in sustaining cytotoxic T-cell activation within the muscle microenvironment (7). Moreover, in IBM, activated DCs correlate with CD8$^+$ T cells showing a cytotoxic Th1-polarised phenotype, sustaining ongoing inflammation. In anti-Jo1-positive myositis, B cells with reduced CD73 expression and distinctive cytokine profiles suggest impaired purinergic signalling as a contributor to disease (8).

Checkpoint biology provides further insight: loss of TIGIT expression in CD4$^+$ T cells promotes Th1/Th17 polarisation through metabolic, epigenetic reprogramming, enhancing glycolysis and histone acetylation of inflammatory genes. Clinically, checkpoint inhibitor therapies may unmask severe myocarditis, myositis overlap, underscoring their role in muscle immune tolerance (9).

In addition, recent data from large international registries of immune checkpoint inhibitor-related myocarditis have underscored that checkpoint disruption can precipitate not only cardiac but also generalised skeletal muscle inflammation, often including respiratory muscle involvement. The development and validation of a prognostic score based on troponin levels, cardiomuscular symptoms, ejection fraction, QRS voltage, and thymoma status highlight how therapeutic interference with checkpoint pathways can unmask severe myositis-myocarditis overlap, further emphasising the pivotal role of checkpoint biology in maintaining muscle immune tolerance (10).

Large-scale molecular profiling has also begun to delineate the immunological fingerprints of IIM subtypes. Bulk RNA-seq of 669 muscle biopsies demonstrated that IBM is dominated by a type 1 immune response, with strong expression of IFNG, CXCL9/10/11, and the CCL5-CCR5 axis. Additional analysis pointed to the XCL1-XCR1 pathway as a potentially important recruiter of immune cells into muscle tissue (11).

At the same time, proteomic profiling of plasma samples identified a distinct inflammatory panel in anti-MDA5 dermatomyositis, where CXCL9/10, IL-17C, IL-18R1, and TNF were particularly elevated (12). These results do more than confirm the well-known role of interferon-driven inflammation: they highlight that each autoantibody-defined subgroup is characterised by its own set of dominant cytokines and chemokines.

Additionally, the possible pathogenic role of some autoantibodies is gaining increasing interest. For instance, anti-Mi2 antibodies have been demonstrated to cross-react with the transcriptional regulator AIRE through a shared PHD1-finger epitope. By interfering with the Mi2/NuRD complex, these antibodies are thought to promote transcriptional derepression in muscle cells. This aspect can be also involved in the pathogenesis of lung involvement in patients with anti-Mi-2 positive ILD. This evidence challenges the historical notion that anti-Mi-2 is rarely associated with ILD and suggests that its detection, even in isolation, has significant clinical prognostic value. The findings align with the mechanistic model whereby anti-Mi-2 antibodies could contribute directly to tissue dysfunction, potentially in lung tissue as well as muscle, through mechanisms like transcriptional dysregulation, moving their role beyond that of a simple serologic marker to that of an active participant in disease pathogenesis (13).

The view of IBM as a disorder that combines inflammation with neurodegenerative-like features has been reinforced by the observation that α-synuclein accumulates within muscle fibres. Sarcoplasmic deposits of α-synuclein showed high diagnostic accuracy, and circulating levels correlated with disease duration (14). While previous studies emphasised aggregates of TDP-43 or p62, this new finding draws attention to α-synuclein as both a biomarker and a potential pathogenic factor, underscoring the dual autoimmune-degenerative nature of IBM. This concept of convergent autoimmune and degenerative pathology extends to other antibody subsets. A detailed clinicopathological study of patients with isolated anti-Ku antibodies revealed a highly heterogeneous muscular phenotype, featuring not only classic myositis with necrosis and endomysial inflammation but also prominent vacuolar pathology (rimmed and non-rimmed vacuoles in 50% of cases) and features of neurogenic atrophy. Immunohistochemistry and western blot analysis suggested a significant role for autophagy dysregulation, with elevated p62 protein levels and a tendency for increased LAMP2 and LC3 expression, distinguishing it from both IBM and immune-mediated necrotising myopathy (IMNM). This positions anti-Ku myositis at the intersection of autoimmunity, defective protein clearance, and altered cellular degradation pathways (15).

Laboratory investigations and autoantibodies

Myositis-specific and myositis-associated autoantibodies (MSAs and MAAs) are valuable biomarkers in idiopathic IIMs. Their prevalence and clinical associations vary significantly across different patient cohorts, underscoring the influence of geographic and ethnic factors.

A Vietnamese IIM cohort showed that anti-MDA5 (15.6%) was the most common autoantibody, followed by anti-Jo-1 (10.9%) (16). In a Moroccan dermatomyositis (DM) cohort, 68.5% of patients were positive for a DM-specific antibody. The most frequent antibody was anti-Mi2 (22.2%), followed by anti-TIF1γ (14.8%) and anti-NXP2 (9.2%) (17). An Indian patient cohort identified Ro52 as the most common autoantibody (22.46%), followed by

anti-Mi2B (18.84%) and anti-SRP (14.49%). In the Indian cohort, cutaneous manifestations and joint involvement were more frequent in the anti-Mi2B and MAA-positive subgroups. Histopathological analysis showed that perifascicular atrophy was also more prevalent in these subgroups (18).

A study of a Chinese cohort with anti-SAE-positive DM showed that the disease was associated with a lower mortality rate, with malignancies being the leading cause of death. Surviving patients had significant skin damage, while ILD tended to stabilise (19). In a French cohort of IIM patients, those with cardiac involvement at diagnosis had a significantly higher risk of cardiovascular events (CVEs) and experienced them earlier than patients without cardiac involvement (20).

While classical autoantibodies remain a diagnostic pillar, new research highlights their role in pathogenesis and prognosis. Studies show that anti-Mi-2 autoantibodies target the PHD fingers of SP140L and TIF1γ, while anti-TIF1γ antibodies recognise epitopes outside this region, providing mechanistic insights into clinical differences (21).

Autoantibodies may act as prognostic indicators. Anti-NXP2 positivity is linked to aggressive ILD (22). Long-term data from the MYONET registry confirm that antibodies such as anti-PM/Scl predict severe cumulative damage, while dermatomyositis-specific autoantibodies associate with less tissue damage than seronegativity (23).

Emerging biomarkers complement classical antibodies. Ultra-sensitive interferon quantification may track disease activity (24); adhesion molecules ICAM-1 and VCAM-1 correlate with dermatomyositis-associated ILD; and cytokine/chemokine panels help distinguish antibody-defined subgroups (25).

Take home messages

- Genetic predisposition strongly influences IIM risk and clinical expression, with HLA alleles shaping susceptibility and disease severity across ethnic groups (4, 5).
- Autoantibodies are active mediators, directly disrupting cellular processes, driving inflammation, and influencing organ involvement and prognosis (13, 15, 22, 23).
- Each IIM subtype has a distinct immune and molecular profile, with specific cellular actors, cytokine signatures, and transcriptomic patterns (6-8, 11, 12).
- Integrated biomarker strategies combining autoantibodies, interferon signatures, and cytokines enable personalised risk assessment and tailored management (24, 25).

General and muscular involvement (classification criteria, disease activity criteria, histology) and muscular imaging

General aspects and classification criteria

Over the years, various classification criteria for IIMs have been published, starting in 1970. In 2017, new criteria were approved by the European League Against Rheumatism (EULAR) and the American College of Rheumatology (ACR), which remain in use today. Bi Chen *et al*. evaluated the effectiveness and reliability of the 2017 EULAR/ACR classification criteria in a cohort of 250 anti-MDA5-positive patients with ILD, and compared the results with those of the Bohan/Peter criteria combined with Sontheimer's CADM criteria: 70% of the patients satisfied the EULAR/ACR criteria, whereas only 25.6% of patients were classified as having definite or possible IIM using the Bohan and Peter criteria, reaching 60.4% when adjusted according to Sontheimer's criteria for CADM. All patients who did not meet the EULAR/ACR criteria did meet the IPAF criteria, suggesting that future IIM classification criteria should consider the presence of ILD (26).

In a cohort of 129 patients who met the 2017 EULAR/ACR criteria for IIMs, the Kaplan-Meier survival analysis showed that no significant difference was observed in relapse-free survival duration between DM and PM patients. A longer relapse-free survival was associated with achieving lower CK during a low-dose glucocorticoid phase. Additionally, the use of immunosuppressants was associated with a lower probability of relapse (27).

Muscular imaging

Magnetic resonance imaging (MRI) is a key tool for assessing muscle involvement in IIMs allowing to detect muscle and subcutaneous oedema, fatty infiltration, fasciitis, and fatty replacement. A recent retrospective study outlined the pattern of muscle involvement in different subgroups of IIM based on the modified Goutallier-Lamminen-Mercuri scoring system. It confirmed that severe oedema with minimal fatty replacement was more common in the early stages of the disease, whereas patients with long-standing disease exhibited mild to moderate oedema along with more significant fatty replacement. Notably, thigh adductors and anterior compartment muscles showed higher scores for muscle oedema, while posterior thigh muscles were less affected. Interestingly, 81.2% of DM patients had muscle oedema in the peronei muscles, yet only 28.1% experienced clinically significant distal weakness. Moreover, the authors found that the presence of muscle oedema was correlated with CK levels but not with MMT-8 scores (28).

AS conventional MRI has limited sensitivity in the early stages of IIM, Saran et al hypothesised that heightened diffusion of water molecules across muscle membranes, a phenomenon detectable by Diffusion Tensor Imaging (DTI), would be an early indicator of muscle inflammation at histopathological examination in individuals with IIM. Although both average DTI and DWI values were significant predictors of an effaced fascicular architecture, the Vastus Lateralis oedema signal intensity weighted score proved to be the most reliable parameter for predicting this condition as well as marked lymphocytic inflammation in the endomysium. Similarly, fatty infiltration weighted score was the main predictor of perifascicular atrophy (29).

Muscle ultrasound is a well-established tool in myositis, in particular to identify the involvement of deep finger flexors in IBM. Recent data suggest that this technique may be a useful tool for distinguishing IIM patients from patients with dysferlinopathies. In particular, patients with dysferlinopathies showed a greater degree of atrophy and mostly

moderate to severe Heckmatt scores, suggesting a possible role of muscular US for differential diagnosis or as a screening test (30).

Histology

Muscle biopsy is essential for confirming the diagnosis of IIM, but the most effective muscle selection strategy and sampling technique remain unclear. A 96-item online survey of 116 members of the International Myositis Assessment and Clinical Studies (IMACS) organisation revealed that the most common approach to muscle biopsy was open surgical biopsy (74.5%), followed by needle biopsy (11.3%) and conchotome forceps biopsy (9.4%). Regarding the open surgical approach, local anaesthesia was commonly used (in 54.8% of cases), and most procedures were performed by general surgeons. In contrast, needle or percutaneous biopsies and conchotome forceps biopsies were commonly performed by rheumatologists. The most common complication, regardless of the type of procedure, was postoperative pain lasting more than three days, followed by post-procedural hematoma. As for muscle selection, clinical examination was the main site selection strategy, followed by muscle MRI and electromyography, while "blind" biopsies were uncommon (31). Ultrasound-guided muscle biopsy may represent a valid alternative to the open method and may grant adequate biopsy specimen in up to 90.1% of the cases, similarly to the open procedure (32).

Parallel advances in histopathological assessment have emerged: a 2025 study applied machine learning to quantify $CD31^+$ micro vessels in muscle biopsies, achieving reliable discrimination of altered vascular density across IIM subgroups while reducing analysis time by 90%, underscoring the feasibility of AI-supported vascular profiling in routine diagnostics (33).

Disease activity criteria

In IIM, fatigue is one of the main symptoms that affects patients QOL. George *et al*. studied the associations between fatigue scores (fatigue VAS and SF-36 energy-fatigue domain) and different demographic and clinical parameters, patients-reported outcomes (PROs) and Clinician-reported outcomes (ClinRO) in a cohort of 50 IIM patients. Both patient-reported measures of physical function and clinical measures of disease activity, such as muscle disease activity, physician global disease activity (PhGA), patient global disease activity, and MMT-8, were found to have a moderate to strong correlation with the aforementioned fatigue measures. Fatigue was associated with both, extra-muscular global disease activity and physician global disease activity. Furthermore, muscle enzymes, including CPK, aldolase, AST, and ALT, showed a strong correlation with both fatigue measures. Fatigue served as a useful treatment bioindicator (34).

Furthermore, a cross-sectional multicentre study within the Spanish Myo-Spain registry evaluated disease activity in a cohort of 554 IIM patients using the total 7-domain index and the 6-domain extra-muscular activity of the Myositis Disease Activity Assessment (MYOACT), MMT-8 and PhGA. As expected, the incident group showed significantly higher values for disease activity compared to the prevalent group, whereas the degree of damage wax similar between the two groups. An analysis of the MYOACT organ systems further revealed that skin, constitutional, and muscle involvement were all more frequent in the incident group (35).

Although several PRO measures have been validated for use in dermatomyositis, these instruments were not originally developed specifically for this condition. Consequently, they may not adequately capture crucial, disease-specific symptoms and their functional impacts. Christopher-Stine et al tried to develop a DM-specific PRO tool: the Dermatomyositis Disease Symptom Questionnaire (DM-DSQ). The new DM-DSQ successfully captured symptoms reported by patients, such as fatigue, muscle weakness, muscle/joint pain, and skin issues. It also accounted for the effects of these symptoms on functional, emotional, and cognitive aspects, as well as role functioning (like work) (36).

Take home messages

- MRI is a valuable tool for evaluating muscle involvement in IIMs, showing signs like oedema and fatty infiltration. In early disease stages, severe oedema is common, while long-standing disease shows more fatty replacement (28, 29).
- Despite being crucial for IIM diagnosis, the optimal biopsy procedure and muscle selection strategy remain unclear. While the open surgical biopsy is most common, ultrasound-guided biopsy is a valid alternative. Site selection is primarily based on clinical examination, followed by MRI and electromyography (31, 32).
- Fatigue is a significant symptom that correlates with disease activity and affects quality of life in IIM patients (34).

Extra-muscular involvement

Beyond muscular involvement, IIMs frequently display extra-muscular manifestations, including those of the skin and major internal organs like heart, lungs, and gastrointestinal tract (Table I).

Lung

Interstitial lung disease (ILD) is the most frequent and prognostically relevant extra-muscular manifestation of IIM. Recent studies have advanced new imaging-based assessment and tools for the early recognition of prognostic complications. Nowadays, assessment of ILD still relies mainly on chest HRCT combined with pulmonary function measurements. Recently, quantitative chest CT has gained increasing importance: in anti-synthetase syndrome, semi-automated HU-based quantification correlated closely with both functional decline and radiologist evaluation, proving more sensitivity in capturing early progression, supporting its role as an objective monitoring tool (37). Beyond quantification, novel analytic approaches such as the standardised threshold ratio analysis and distribution (STRAD) have been applied to anti-MDA5 dermatomyositis. When combined with anti-Ro52 antibody status, the resulting STRAD-Ro52 model achieved high accuracy in predicting

Table I. Overview of extra-muscular manifestations reported across the review articles.

Lung involvement	Quantitative chest CT and novel analytic tools (STRAD, AI-based models) improve sensitivity for early ILD progression (38). Pneumomediastinum and pleural effusion are independent predictors of poor outcome (40, 41). Emerging biomarkers (*e.g.* KL-6) support functional correlation and monitoring (39).
Heart involvement	Multiparametric CMR detects subclinical myocarditis beyond standard imaging (42). Left atrial strain analysis identifies early diastolic dysfunction (46). Arrhythmic risk markers and phenotypic subgroups confirm that cardiac disease in IIM is heterogeneous (49, 50).
Skin, microvascular, GI	Calcinosis and panniculitis remain challenging cutaneous complications; anti-SAE panniculitis may represent a distinctive phenotype (51, 52). NVC consistently reveals scleroderma-spectrum abnormalities and microhaemorrhages in DM (55).
Malignancy	Cancer accounts for a major share of mortality in IIM, especially within the first years of disease (59). Beyond classical ovarian and lung cancers, melanoma emerges as a significant risk (58). PET/CT and CA-125 lack sensitivity/specificity, targeted approaches are needed (62).

severe ILD within six months. This framework may allow earlier identification of patients at risk of acute respiratory failure and help refine treatment strategies (38).

Lung ultrasound (LUS) has also emerged as a complementary imaging tool. In a large cohort of patients, ultrasonographic B-lines number closely correlated with ILD severity on HRCT and pulmonary function. When incorporated into a multivariable nomogram together with age, respiratory symptoms, and serum KL-6, predictive performance improved markedly. The availability of a web-based version further enhances its daily clinical applicability: although not replacing HRCT, LUS offers a radiation-free bedside tool suitable for screening and follow-up, particularly useful in high-risk groups of patients (39).

Other than ILD, other lung involvement may also occur that may dramatically reduce the outcome of the patients: pneumomediastinum, although infrequent, was confirmed as an independent predictor of mortality in myositis-ILD, with median survival reduced to 13 months compared with over six years in those without this complication (40). Similarly, pleural effusion, often underrecognised, predicted both rapidly progressive ILD and increased mortality, emphasising the importance of careful imaging evaluation beyond parenchymal changes (41).

Heart

Cardiac involvement in IIM is increasingly recognised as a major driver of morbidity and mortality, and recent studies have refined how we could detect subclinical myocardial inflammation and stratify risk in routine care (42). Multiparametric cardiac MRI (CMRI) has emerged as the cornerstone for non-invasive diagnosis: in anti-synthetase patients, applying the updated Lake Louise Criteria with mapping techniques (late gadolinium enhancement, T1/T2 and extracellular volume) markedly increased diagnostic sensitivity for myocarditis compared to conventional CMRI, with abnormalities present in nearly all clinically suspected cases (43). In patients with clinically stable IIM, cross-sectional CMRI studies demonstrate that up to 10–18% of patients have abnormal native T1/T2 despite preserved function, supporting ongoing surveillance even outside overt flares (44). Prospective observational studies consistently report elevated native T1/T2 values and LGE. These mapping metrics correlate with composite disease activity scores, highlighting their potential as markers for early detection and disease monitoring (45). Beyond ventricular tissue characterisation, feature-tracking CMRI of the left atrium reveals impaired reservoir and conduit function even when conventional ventricular indices are normal, suggesting diastolic involvement and providing an additional window on detecting subclinical disease (46).

From an outcomes standpoint, a contemporary French cohort found that patients with cardiac involvement at myositis diagnosis had a higher incidence of cardiovascular events (CVE) and experienced them earlier during follow-up, highlighting the prognostic weight of early cardiologic abnormalities and the need for intensified monitoring in the first months (47). Moreover, subclinical atherosclerosis deserves attention alongside inflammatory cardiomyopathy: non-gated chest CT analyses in IIM demonstrated excess coronary artery calcification and thoracic aortic dilation compared to matched controls, with aortic diameters correlating with Framingham risk in those without calcification (48). In a large Chinese cohort of 281 dermatomyositis patients, a significant prolongation of QTc, Tp-e interval and Tp-e/QT ratio, compared with matched controls, has been demonstrated. These markers of ventricular repolarisation heterogeneity were positively correlated with inflammatory activity and were particularly elevated in anti-Ro52 positive patients. These findings indicate that repolarisation abnormalities may underlie an increased risk of malignant ventricular arrhythmias and sudden cardiac death in dermatomyositis, even in patients without overt cardiac dysfunction (49).

A cluster analysis of 88 IIM patients with cardiac involvement identified two distinct clinical subgroups. Category I was characterised by marked structural and functional cardiac abnormalities (chamber enlargement, arrhythmias, pulmonary hypertension, elevated NT-proBNP), with heart failure representing the predominant cause of death. In contrast, Category II showed more systemic features (interstitial lung disease, rash, dysphagia) and deaths were mainly attributable to infections, particularly severe pneumonia (50).

Skin, microvascular and gastrointestinal tract

Cutaneous disease in dermatomyositis (DM) remains clinically heterogeneous, ranging from hallmark rashes to complications such as panniculitis and calcinosis. A recent review highlighted that calcinosis affects up to 20% of adults and 75% of children with DM, with pathogenesis increasingly linked to neutrophil activation and mitochondrial dysfunction. New imaging approaches, including CT-based calcium scoring are being explored, as well as emerging therapies such as JAK inhibitors or intralesional sodium thiosulfate, but evidence remains limited (51). In anti-SAE1/2-positive DM, panniculitis may represent a distinctive, though rare, cutaneous manifestation potentially associated with treatment resistance (52).

Vascular abnormalities also represent a key extra-muscular domain. A systematic review of NVC in adult-onset DM synthesised data from 37 studies (346 patients), confirming that over 60% presented with enlarged capillaries and nearly half with a scleroderma-spectrum pattern or microhaemorrhages, especially in patients with anti-MDA5 and anti-TIF1-γ (53).

NVC abnormalities, particularly low capillary density, were present in most (73.7%) DM patients. The presence of NVC abnormalities, may allow to classify patients in two clusters, each one with significant differences in ILD incidence (54). On the other hand, a Mayo Clinic study using NVC in anti-TIF1γ-positive dermatomyositis found more frequent microvascular abnormalities (capillary density loss, microhaemorrhages, disorganisation) in patients with cancer, suggesting that NVC may be used as a non-invasive biomarker for malignancy risk stratification in this subgroup (55).

The gastrointestinal system may also be affected, sometimes with meaningful consequences for patient management. Dysphagia remains the most common manifestation, with a retrospective review of DM patients identifying risk factors including older age, disease duration, and severity of muscle weakness (56). Hepatic dysfunction has also been described, particularly in the context of anti-MDA5-positive DM, where liver enzyme abnormalities may signal poor prognosis (57).

Malignancies

The association between IIM and malignancy is well established and accounts for a substantial part of the disease burden. A recent meta-analysis specifically addressing melanoma found that IIM patients carry a markedly elevated risk of developing this type of malignancy, with a pooled standardised incidence ratio of 6.3 compared to the general population. This highlights melanoma as a distinct cancer type of concern in addition to the more established ovarian, lung, and gastrointestinal tumours (58). Large-scale registry data further quantify the burden: in a Swedish cohort of 1,826 IIM patients, 17% developed cancer after diagnosis, and the five-year probability of death from malignancy was 31%, more than four times compared to patients without subsequent cancer. Risk was the highest in dermatomyositis and when cancer occurred within the first year of IIM onset (59).

Screening approaches remain challenging: serum levels of CA-125 and whole-body PET/CT for occult cancer detection showed limited accuracy, with false positives and false negatives as high as 28.6% for PET/CT and poor discrimination by CA-125 across subgroups (60). Regarding that, a recent international survey of rheumatologists and neurologists revealed striking heterogeneity in clinical practice (61). Complementing this, a recent review summarised clinical risk factors consistently associated with cancer in DM, including older age, male gender, dysphagia, cutaneous necrosis or vasculitis, and the absence of interstitial lung disease. The study emphasised that conventional screening modalities might prove inadequate, and that tailor-made and risk-adapted protocols are needed (62).

Take home messages

- High-resolution CT (HRCT) remains the gold standard for diagnosis of ILD in IIMs, but quantitative CT and novel analytic approaches (*e.g.* STRAD-Ro52 model) enhance early detection and risk stratification (37, 38).
- Rare but severe complications like pneumomediastinum and pleural effusion are strong predictors of mortality (40, 41).
- Multiparametric cardiac MRI (CMRI), including T1/T2 mapping and feature-tracking, improves detection of subclinical myocarditis and diastolic dysfunction (43-46).
- Electrophysiologic markers (*e.g.* QTc prolongation, Tp-e interval) suggest increased risk of malignant arrhythmias even in patients without overt cardiac dysfunction (49).
- Calcinosis and panniculitis may indicate more severe or treatment-resistant disease (51, 52).
- Nailfold capillaroscopy (NVC) detects microvascular abnormalities in >60% of adult DM, serving as a potential non-invasive biomarker (53).
- Liver involvement, including idiopathic portal hypertension, can occur, particularly in anti-MDA5 or anti-synthetase syndromes (57).

Particular subsets of disease

Immune-mediated necrotising myopathy

Immune-mediated necrotising myopathy (IMNM) is characterised by severe proximal muscle weakness, significantly increased serum CK levels and major necrosis of muscle fibres with little or no inflammatory infiltration found on muscle biopsy.

Yang *et al*., using clustering analysis considering clinical, serological and pathological parameters, identified three clusters with distinct phenotypes and prognoses. Cluster 1 had the highest CK levels, the shortest disease course, severe muscle weakness and more inflammation infiltration in muscle biopsy. Cluster 2 had the lowest CK level and moderate inflammation infiltrate. Cluster 3 had the youngest age of onset, the longest disease course and the least frequency of inflammatory infiltration. Positive prognostic factors were age of onset >55 years, more regeneration of muscle fibres, more CD4 T infiltration and membrane attack complex (63). The same group analysed the relation-

ship between anti-HMGCR isotypes and disease outcome: Anti-HMGCR IgM-positive patients had a younger age of onset and more neck weakness than anti-HMGCR IgM negative patients. Interestingly, the levels of anti-HMGCR IgG and IgM are associated with disease activity in anti-HMGCR-positive patients (64).

Inclusion body myositis

Inclusion body myositis (IBM) is the most prevalent inflammatory muscle disease in older adults with no effective therapy available. In contrast to other IIMs, IBM follows a chronic disease course with both inflammatory and degenerative features of pathology (65). Yamashita *et al*. compared patients with IBM according to the autoantibody status: patients with anti-cN1A autoantibodies had a higher frequency of finger flexion weakness than those without (66).

Regarding comorbidities, IBM patients had a significantly increased risk of myocardial infarction (MI) compared to matched population referents without IBM, despite a relatively similar prevalence of baseline traditional cardiovascular risk factors. Corticosteroid and other immunosuppressant use were neither risk factors nor protective for the development of MI in IBM (67).

Anti-synthetase syndrome

Anti-synthetase syndrome (ASyS) is a rare subtype of IIM, characterised by the presence of autoantibodies against aminoacyl-transfer RNA synthetases (ARSs). The clinical manifestations of ASyS include the classic "triad" of arthritis, myositis and ILD, along with other typical clinical features including fever, Raynaud phenomenon and mechanic's hands/hiker's feet.

The Classification Criteria for Anti-Synthetase Syndrome (CLASS) project is an international collaborative study funded by EULAR/ACR to develop and validate data and consensus-driven classification criteria for ASyS. In 2025 CLASS published an analysis of clinical characteristics driven from their database considering patients with mimicking conditions as controls (Table II). In terms of serological variables, Jo-1 and non-Jo-1 anti-synthetase autoantibodies, antinuclear antibodies with cytoplasmic pattern, and anti-Ro52 autoantibodies were associated with ASyS (68).

Each ARSs seemed to represent a different clinico-pathological phenotype. In a retrospective multicentre longitudinal study, Tang *et al*. highlighted that patients with anti-Jo1 had significantly higher muscle enzyme levels and more arthritis, all patients with anti-EJ would develop ILD on follow-up and malignancy was noted in 28.6% of the anti-OJ positive patients (69).

Although anti-Ha (tyrosyl-tRNA synthetase) antibody was identified firstly in 2005, only a few cases with anti-Ha antibody have been reported. In a recent study, anti-Ha antibody seems not to be as rare as previously believed and could coexist with other MSAs. The anti-Ha patients generally display muscle weakness with a typical limb-girdle pattern, while extra-muscular manifestations except for the cutaneous lesions are unusual. The most common pathological picture for anti-Ha patients is a necrotising myopathy with less frequent MHC-II expression (70).

An unmet need in ASyS are the frequent flares that frequently occur when glucocorticoids are tapered close to 10 mg of prednisolone. A Japanese group focused on the risk factor for flares in ASyS, linked to the presence of anti-EJ/Jo-1 antibody and Gottron's sign. An early use of calcineurin inhibitors may prevent flares in ASyS patients (71).

ILD is the most prominent feature and an important prognostic factor associated with poor survival in ASyS. Yamaguchi *et al*. evaluated the long-term clinical prognosis in ASyS with lung involvement: anti-PL-7 antibodies were associated with risk of all-cause mortality, the UIP pattern affected prognosis and pulmonary events within the first 5 years (72). Similar results were found by Shan *et al*.: anti-PL7-ASyS patients with ILD are at significant risk of developing progressive pulmonary fibrosis (PPF) (73).

Myocarditis is an overlooked manifestation of ASyS but it is frequent, even at clinical onset, associated with peculiar clinical features. Patients with ASyS with myocarditis were more frequently males with fever and had higher hs-TnT, NT-proBNP and CRP, compared with those without myocarditis. Interestingly, in most patients this manifestation was the clinical manifestation of disease onset (43).

Table II. Clinical variables included in each CLASS domain.

Specific ASyS domain	Clinical variables
Joint	Arthralgia Arthritis
Muscle	Weakness Myalgia Dysphagia Increase muscle enzymes MRI findings EMG findings Muscle biopsy findings
Lung	HRCT pattern
Skin	Mechanic's hands Hiker's feet Gottron sign/papules Heliotrope rash
Cardiac	Myocarditis Pulmonary hypertension
Other clinical variables	Raynaud phenomenon Unexplained fever Sicca syndrome

Take home messages

- Clustering analysis with combined clinical, serological and pathological parameters could classify IMNM into three clusters with distinct phenotypes and prognoses (63).
- IBM is associated with increased risk of MI compared to population referents; heightened cardiovascular monitoring and prevention strategies are needed in IBM (67).
- The clinical profiles of patients with ASyS are heterogeneous but could be differentiated by specific ARS (69).
- The majority of patients with ASyS experience a flare during tapering of glucocorticoids, that may be prevented by an early use of calcineurin inhibitors (71).

Therapy

Despite the many advances in knowledge in IIMs, clinical practice guidelines (CPGs) are sparse and heterogeneous, particularly due to the paucity of randomised controlled trials (RCTs)

and due to the rarity of the disease (74). Glucocorticoids (GCs) are considered the first-line therapy, but the early use of immunosuppressive drugs is crucial, both for adjunctive efficacy and for steroid sparing effect (75). Table III shows the new results for different therapeutic molecules studied in the last two years.

Intravenous immunoglobulins (IVIg) have been approved by the US Food and Drug Administration (FDA) and the European Medicines Agency (EMA) for IIM. The phase 3 ProDERM study demonstrated that IVIg were safe and effective in patients with DM. Predictors of response to IVIg, analysed by Charles-Schoeman *et al*., were higher cutaneous disease activity and/or anti-TIF1-γ positivity. Instead, pulmonary disease activity predicted a lower IVIg response (76).

Sharf *et al*. compared two different regimes of IVIg administration (before and after six months receiving the diagnosis) in IMNM patients, demonstrating that IVIg may prove to be a valuable addition to an early and aggressive induction regimen. Interestingly, delay in IVIg treatment may lead to the development of permanent residual weakness and long-term disability (77).

Starting from 2005, several case reports, case series, open label trials and RCTs have suggested the efficacy of treatment with RTX in patients with IIMs. In 2025, the group of the University of Padua demonstrated that RTX was effective in most IIMs refractory patients with pulmonary and muscular involvement. Low dose RTX maintenance therapy (1 g every 6 months) seemed able to maintain the remission after induction with standard dose (two infusions of 1 g two weeks apart) (78). Data from a pilot study conducted by Conticini *et al*. showed that the precocious administration of RTX may cover most aspects of ASyS, in particular arthritis and ILD (79). Furthermore, RTX was demonstrated to be effective and safe in both treatment-naïve and refractory IIMs patients. An early use can be associated with faster improvement and better outcomes (80).

Abatacept (ABA) has not displayed superiority over placebo after 24 weeks in a RCT because in this study the response rate for the placebo group was higher than expected in DM patients. However, analysis by IIM subtype suggested there may be a sustained benefit of ABA for patients with non-DM subtypes (PM, IMNM) (81).

Type-I interferon (IFN) dysregulation has a major role in the pathogenic mechanisms of IIMs, especially for DM. Accordingly, blocking IFN signalling through inhibition of the Janus kinase/signal transducer and activator of transcription (JAK/STAT) pathway represents a novel treatment option for DM. The use of JAK Inhibitors (JAKi) in patients with ASyS has produced promising results: Shan *et al*. suggested that JAKi therapy should be considered in the treatment of refractory ASyS patients, especially those with ILD, arthritis and skin rash (82). In a multicentre Indian study, tofacitinib (TOFA) demonstrated a steroid sparing efficacy in IIM, with good clinical improvement, especially in the cutaneous domain, even in patients with refractory disease. Modest benefits were evident also in calcinosis (83). Xing *et al*. demonstrated significant immunologic effectiveness of TOFA in DM and ASyS patients, reducing key immune cell populations (Tfh and Th17 cells) and downregulating immune activation pathways (IL-17 signalling pathway) (84).

Efgartigimod (EFG) is an Fc fragment of human IgG1 that augments its affinity for FcRn, diminishing IgG recirculation and augmenting IgG degradation. Peng *et al*. used EFG for the treatment of anti-SRP-IMNM, demonstrating a substantial decrease in CK values and effectively improved muscle strength (85). Similar positive results were found by a Chinese pilot study in IMNM patients. EFG can expand the therapeutic arsenal for refractory IMNM to shorten duration and minimise chronic myopathic features (86).

Table III. Therapeutic molecules studied in 2025 for the treatment of IIM. The domains or the clinical situations in which the different therapies have been shown to be effective are in the right column.

Therapy	IIM subset	Involvement
IVIg (76, 77)	DM IMNM	Skin rash Muscle
Rituximab (78, 79)	ASyS IIMs	ILD and arthritis Muscle and ILD
Tofacitinib (83)	IIMs	Skin rash and calcinosis
Efgartigimod (85, 86)	IMNM	Muscle

Take home messages

- Higher cutaneous disease activity and anti-TIF1-γ positivity are predictors of response to IVIg in DM (76).
- ABA may be more effective in PM and IMNM patients (81).
- RTX is effective in both treatment-naïve and refractory IIM patients, especially with pulmonary and muscular involvement (80).
- TOFA demonstrated a steroid sparing efficacy in IIM, with good clinical improvement, especially in the cutaneous domain, including calcinosis (83).
- EFG is a promising novel therapy for muscular involvement in IMNM (85, 86).

Conclusions

In 2025, research on IIMs continued to strengthen and expand recent achievements, providing an increasingly integrated and nuanced view of these rare and complex disorders. Advances in the understanding of pathogenetic mechanisms, together with refinements in diagnostic tools and the growing relevance of biomarkers and myositis-specific autoantibodies, have further supported a personalised medicine approach.

Greater attention to clinical phenotypes and extra-muscular involvement remains central to patient management. Although many challenges persist, the 2025 landscape reflects steady progress towards more targeted and effective therapeutic strategies, ultimately aiming to improve patient outcomes and QoL.

Acknowledgements

Some authors are members of the European Reference Network for Rare Neuromuscular Diseases (ERN EURO-NMD) and/or on Rare and Complex Connective Tissue and Musculoskeletal Diseases (ERN ReCONNET).

References

1. CONNOLLY CM, GUPTA L, FUJIMOTO M, MACHADO PM, PAIK JJ: Idiopathic inflammatory myopathies: current insights and future frontiers. *Lancet Rheumatol* 2024; 6(2): e115-e127. https://doi.org/10.1016/s2665-9913(23)00322-3
2. BETTERIDGE Z, MCHUGH N: Myositis-specific autoantibodies: an important tool to support diagnosis of myositis. *J Intern Med* 2016; 280(1): 8-23. https://doi.org/10.1111/joim.12451
3. FATTORINI F, CONTICINI E, DOURADO E *et al*.: Idiopathic inflammatory myopathies: one year in review 2024. *Clin Exp Rheumatol* 2025; 43(2): 167-77. https://doi.org/10.55563/clinexprheumatol/yizkja
4. DHAOUADI T, RIAHI A, BEN ABDALLAH T, GORGI Y, SFAR I: Association of HLA-DR, HLA-DQ, and HLA-B alleles with inclusion body myositis risk: A systematic review, a meta-analysis, a meta-regression and a trial sequential analysis. *Int J Immunopathol Pharmacol* 2025; 39. https://doi.org/10.1177/03946320251321747
5. LLANSÓ L, SEGARRA-CASAS A, DOMÍNGUEZ-GONZÁLEZ C *et al*.: Absence of pathogenic mutations and strong association with HLA-DRB1*11:01 in statin-naive early-onset anti-HMGCR necrotizing myopathy. *Neurol Neuroimmunol Neuroinflamm* 2024; 11(5): e200285. https://doi.org/10.1212/nxi.0000000000200285
6. WIJNBERGEN D, JOHARI M, OZISIK O *et al*.: Multi-omics analysis in inclusion body myositis identifies mir-16 responsible for HLA overexpression. *Orphanet J Rare Dis* 2025; 20(1): 27. https://doi.org/10.1186/s13023-024-03526-x
7. KIROU RA, PINAL-FERNANDEZ I, CASAL-DOMINGUEZ M *et al*.: Activated dendritic cell subsets characterize muscle of inclusion body myositis patients and correlate with KLRG1^{+} and TBX21^{+} CD8^{+} T cells. *medRxiv* 2025; 5. https://doi.org/10.1101/2025.06.04.25328910
8. NAKAZAWA M, HORULUOGLU B, DE VRIES C *et al*.: CD73low B-cell phenotypes and distinct cytokine profiles in patients with active anti-Jo-1 antibody positive idiopathic inflammatory myopathies. *RMD Open* 2025; 11(2): e005401. https://doi.org/10.1136/rmdopen-2024-005401
9. LAI Y, WANG S, REN T *et al*.: TIGIT deficiency promotes autoreactive CD4^{+} T-cell responses through a metabolic-epigenetic mechanism in autoimmune myositis. *Nat Commun* 2025; 16(1): 4502. https://doi.org/10.1038/s41467-025-59786-z
10. POWER JR, DOLLADILLE C, OZBAY B *et al*.: International ICI-Myocarditis Registry. Immune checkpoint inhibitor-associated myocarditis: a novel risk score. *Eur Heart J* 2025. https://doi.org/10.1093/eurheartj/ehaf315. Erratum in: *Eur Heart J* 2025. https://doi.org/10.1093/eurheartj/ehaf529
11. KIROU RA, PINAL-FERNANDEZ I, CASAL-DOMINGUEZ M *et al*.: Distinct cytokine and cytokine receptor expression patterns characterize different forms of myositis. *Rheumatology* (Oxford) 2025; 64(11): 5751-60. https://doi.org/10.1093/rheumatology/keaf346
12. ZHANG Y, HU W, LI T *et al*.: Shared and distinctive inflammation-related protein profiling in idiopathic inflammatory myopathy with/without anti-MDA5 autoantibodies. *J Inflamm Res* 2025; 18: 6009-24. https://doi.org/10.2147/jir.s509777
13. FLASHNER BM, IMAI R, SYNN AJ *et al*.: Anti-Mi-2 positive interstitial lung disease (ILD): a progressive disease comparable to other myositis-ILD. *Respir Med Res* 2025; 88: 101176. https://doi.org/10.1016/j.resmer.2025.101176
14. MAYER T, SCHOLLE L, FOERSTER L *et al*.: Alpha-synuclein as a potential biomarker for inclusion body myositis in blood and muscle. *Neuropathol Appl Neurobiol* 2025; 51(3): e70019. https://doi.org/10.1111/nan.70019
15. QIAO L, LIN Y, LIU M *et al*.: The clinical features, muscle pathology, and role of autophagy in anti-Ku-positive patients. *Front Immunol* 2025; 16: 1608735. https://doi.org/10.3389/fimmu.2025.1608735
16. LUO YB, NAKAZAWA M, MINH NPT *et al*.: Immune characterization of a vietnamese cohort with idiopathic inflammatory myopathies. *Muscle Nerve* 2025; 72(3): 464-74. https://doi.org/10.1002/mus.28455
17. CHIHI M, BARAKAT L, BENHAYOUN FZ *et al*.: Clinical features of dermatomyositis associated with myositis-specific antibodies in Moroccan patients. *Clin Pract* 2025; 15(2): 31. https://doi.org/10.3390/clinpract15020031
18. SANTHAPPAN GIRIJA M, VENGALIL S, KULANTHAIVELU K *et al*.: Autoantibody-based clinicoradiopathologic phenotyping of idiopathic inflammatory myopathies: an Indian cohort. *J Clin Neuromuscul Dis* 2024; 26(2): 70-81. https://doi.org/10.1097/cnd.0000000000000487
19. ZHANG Y, LIU L, DUAN X *et al*.: Longitudinal study of patients with anti-SAE antibody-positive dermatomyositis: a multicenter cohort study in China. *Rheumatology* (Oxford) 2025; 64(3): 1377-85. https://doi.org/10.1093/rheumatology/keae232
20. KASSER C, GUÉDON AF, ALLENBACH Y, FAIN O, COHEN A, MEKINIAN A: Cardiovascular events in patients with myositis: results from a French retrospective cohort. *RMD Open* 2025; 11(1): e005276. https://doi.org/10.1136/rmdopen-2024-005276
21. PINAL-FERNANDEZ I, MUSAI J, CASAL-DOMINGUEZ M *et al*.: Anti-Mi2 autoantibodies target PHD fingers of SP140L and TIF1γ, while anti-TIF1γ autoantibodies primarily bind TIF1γ outside the PHD region. *Rheumatology* (Oxford) 2025; 64(12): 6371-77. https://doi.org/10.1093/rheumatology/keaf426
22. FLASHNER BM, IMAI R, SYNN AJ *et al*.: Progressive course of anti-nuclear matrix protein-2 (NXP-2) positive-interstitial lung disease. *Respir Med Res* 2025; 87: 101170. https://doi.org/10.1016/j.resmer.2025.101170
23. ESPINOSA-ORTEGA S, LODIN K, DASTMALCHI M *et al*.: MYONET Registry Study Group. Autoantibodies and damage in patients with idiopathic inflammatory myopathies: A longitudinal multicenter study from the MYONET international network. *Semin Arthritis Rheum* 2024; 68: 152529. https://doi.org/10.1016/j.semarthrit.2024.152529
24. BOLKO L, ANQUETIL C, LLIBRE A *et al*.: Ultrasensitive interferons quantification reveals different cytokine profile secretion in inflammatory myopathies and can serve as biomarkers of activity in dermatomyositis. *Front Immunol* 2025; 16. https://doi.org/10.3389/fimmu.2025.1529582
25. BAE SS, SHAHBAZIAN A, WANG J *et al*.: Plasma levels of adhesion molecules are elevated in dermatomyositis-interstitial lung disease and associated with low paraoxonase-1 activity. *Arthritis Res Ther* 2025; 27(1): 53. https://doi.org/10.1186/s13075-025-03520-z
26. CHEN B, XI B, XIN H *et al*.: External validation of the 2017 EULAR/ACR classification criteria for idiopathic inflammatory myopathies in anti-MDA5 antibody-positive interstitial lung disease patients: A multicenter retrospective cohort study in China. *Semin Arthritis Rheum* 2025; 72: 152700. https://doi.org/10.1016/j.semarthrit.2025.152700
27. CHOI J, NAM SH, LEE JS *et al*.: Relapse risk factors and clinical characteristics of idiopathic inflammatory myopathies in 105 patients. *Clin Rheumatol* 2024; 43(11): 3379-87. https://doi.org/10.1007/s10067-024-07120-1
28. SRIDHAR S, NASHI S, KULANTHAIVELU K *et al*.: Magnetic resonance imaging in idiopathic inflammatory myopathies: deciphering the pattern of muscle involvement. *Neuromuscul Disord* 2025; 47: 105257. https://doi.org/10.1016/j.nmd.2024.105257
29. SARAN S, NANDOLIA K, BAWEJA A, PAI VS, KUMAR M, BOTCHU R: Diffusion tensor imaging of vastus lateralis in patients with inflammatory myopathies. *Rheumatology* (Oxford) 2025; 64(5): 2961-69. https://doi.org/10.1093/rheumatology/keae560
30. SOLORZANO-FLORES SY, SOTO-FAJARDO C, ÁNGELES-ACUÑA A *et al*.: Can we differentiate patients with dysferlinopathies and inflammatory myopathies by ultrasound? A discriminant analysis study. *Rheumatol Int* 2024; 44(12): 2829-36. https://doi.org/10.1007/s00296-024-05721-2
31. MAXWELL S, ROSS L, OON S, WICKS IP, DAY J: Muscle biopsy practices in the evaluation of idiopathic inflammatory myopathies: an international survey of expert clinicians. *Semin Arthritis Rheum* 2024; 68: 152519. https://doi.org/10.1016/j.semarthrit.2024.152519
32. COSTA F, CAMPANILHO-MARQUES R, DOURADO E *et al*.: Efficacy and safety of ultrasound-guided needle muscle biopsy in the diagnosis of idiopathic inflammatory myopathies. *Rheumatology* (Oxford) 2025;

64(9): 5123-31. https://doi.org/10.1093/rheumatology/keaf241
33. MASSARO A, CAZZATO G, INGRAVALLO G *et al*.: Pre-screening of endomysial microvessel density by fast random forest image processing machine learning algorithm accelerates recognition of a modified vascular network in idiopathic inflammatory myopathies. *Diagn Pathol* 2025; 20(1): 13. https://doi.org/10.1186/s13000-025-01608-3
34. GEORGE TB, KERET S, PILLAI AC *et al*.: Fatigue is common in myositis and is associated with disease activity. *Semin Arthritis Rheum* 2025; 73: 152730. https://doi.org/10.1016/j.semarthrit.2025.152730
35. COBO-IBÁÑEZ T, CASTELLVÍ I, PROS A *et al*.: Disease activity in patients with idiopathic inflammatory myopathy according to time since diagnosis and positivity to antisynthetase autoantibodies: data from the Myo-Spain registry. *Arthritis Res Ther* 2025; 27(1): 5. https://doi.org/10.1186/s13075-024-03471-x
36. CHRISTOPHER-STINE L, CIESLUK A, CHINOY H *et al*.: The Dermatomyositis Disease Symptom Questionnaire (DM-DSQ): a measure to assess the patient experience of dermatomyositis symptoms. *J Rheumatol* 2024; 51(12): 1198-207. https://doi.org/10.3899/jrheum.2023-1137
37. JAMAL F, SHASHI K, VAZ N, DOYLE T, DELLARIPA P, HAMMER M: Quantitative chest computed tomography for progression of interstitial lung disease in antisynthetase patients. *J Thorac Imaging* 2024; 39(5): 281-84. https://doi.org/10.1097/rti.0000000000000770
38. XIAO F, CHEN F, LI D *et al*.: Severe interstitial lung disease risk prediction in anti-melanoma differentiation-associated protein 5 positive dermatomyositis: the STRAD-Ro52 model. *Ann Med* 2025; 57(1): 2440621. https://doi.org/10.1080/07853890.2024.2440621
39. ZHANG W, HUANG G, ZHENG S *et al*.: Risk prediction modelling in idiopathic inflammatory myositis-associated interstitial lung disease based on seven factors including serum KL-6 and lung ultrasound B-lines. *Clin Exp Rheumatol* 2025; 43(2): 260-68. https://doi.org/10.55563/clinexprheumatol/ylf0oe
40. JUNG J, KIM MJ, YOO B *et al*.: Clinical impact of pneumomediastinum in patients with myositis-associated interstitial lung disease. *PLoS One* 2025; 20(7): e0328043. https://doi.org/10.1371/journal.pone.0328043
41. LIANG X, REN H, XIAO F *et al*.: Pleural effusion as a predictor of rapidly progressive interstitial lung disease and mortality in idiopathic inflammatory myopathies. *Clin Exp Rheumatol* 2025; 43(2): 221-29. https://doi.org/10.55563/clinexprheumatol/ve77nv
42. SEN G, SCULLY P, GORDON P, SADO D: Advances in the diagnosis of myocarditis in idiopathic inflammatory myopathies: an overview of diagnostic tests. *Rheumatology* (Oxford) 2024; 63(7): 1825-36. https://doi.org/10.1093/rheumatology/keae029
43. DE LUCA G, CAMPOCHIARO C, PALMISANO A *et al*.: Myocarditis in anti-synthetase syndrome: clinical features and diagnostic modalities. *Rheumatology* (Oxford) 2024; 63(7): 1902-10. https://doi.org/10.1093/rheumatology/kead541
44. YURCHENKO K, HØJGAARD P, PECINI R *et al*.: Cardiac involvement in established idiopathic inflammatory myopathy assessed by cardiac magnetic resonance mapping. *Clin Rheumatol* 2025; 44(7): 2941-50. https://doi.org/10.1007/s10067-025-07530-9
45. SAMIM MM, BARTHUR A, VENGALIL S *et al*.: Cardiac magnetic resonance imaging markers in idiopathic inflammatory myopathy - a prospective observational study. *Ann Indian Acad Neurol* 2025; 28(3): 392-99. https://doi.org/10.4103/aian.aian_1001_24
46. YU LZ, LIN YW, SHI RY *et al*.: Quantification of left atrial strain in patients with idiopathic inflammatory myopathy using cardiovascular magnetic resonance feature tracking. *Clin Radiol* 2024; 79(7): 544-52. https://doi.org/10.1016/j.crad.2024.03.005
47. KASSER C, GUÉDON AF, ALLENBACH Y, FAIN O, COHEN A, MEKINIAN A: Cardiovascular events in patients with myositis: results from a French retrospective cohort. *RMD Open* 2025; 11(1): e005276. https://doi.org/10.1136/rmdopen-2024-005276
48. YANG T, QIU Y, ZHANG Y *et al*.: The association of cardiovascular disease risk with coronary artery calcification and thoracic aortic dilation: a study in idiopathic inflammatory myopathies and systemic lupus erythematosus. *Clin Rheumatol* 2024; 43(10): 3117-25. https://doi.org/10.1007/s10067-024-07115-y
49. WANG H, LIN P: Evaluation of ventricular repolarization in dermatomyositis and relationship with inflammation and autoimmunity. *Heart Vessels* 2024; 39(11): 979-87. https://doi.org/10.1007/s00380-024-02413-6
50. DONG J, MENG X, XU H *et al*.: Cluster analysis of clinical phenotypes in idiopathic inflammatory myopathy patients complicated with cardiac involvement. *Clin Rheumatol* 2024; 43(7): 2237-44. https://doi.org/10.1007/s10067-024-06986-5
51. DAVULURI S, CHUNG L, LOOD C: Calcinosis in dermatomyositis. *Curr Opin Rheumatol* 2024; 36(6): 453-58. https://doi.org/10.1097/bor.0000000000001036
52. TAGO M, KOIZUMI H, KAMIYA S *et al*.: Panniculitis on the trunk as a possible characteristic feature of anti-SAE1/2 antibody-positive dermatomyositis: A possible cutaneous manifestation of treatment resistance. *J Dermatol* 2024; 51(7): e227-e228. https://doi.org/10.1111/1346-8138.17128
53. FLATLEY EM, COLLINS D, LUKOWIAK TM, MILLER JH: Nailfold microscopy in adult-onset dermatomyositis in association with myositis antibodies. *Arch Dermatol Res* 2024; 317(1): 34. https://doi.org/10.1007/s00403-024-03521-z
54. XU H, QIAN J: The role of nailfold video-capillaroscopy in the assessment of dermatomyositis. *Rheumatology* (Oxford) 2025; 64(5): 2987-94. https://doi.org/10.1093/rheumatology/keae677
55. MUMTAZ S, PHILLIPPS J, SULLIVAN MM *et al*.: Microvascular abnormalities between anti-TIF1-γ-associated dermatomyositis with and without malignancy. *BMC Rheumatol* 2025; 9(1): 50. https://doi.org/10.1186/s41927-025-00504-z
56. CHENG I, WONG CSM, CHAN HHL: A retrospective review of clinical characteristics and risk factors of dysphagia in patients with dermatomyositis. *Dysphagia* 2025; 40(3): 626-36. https://doi.org/10.1007/s00455-024-10763-6
57. MO W, XING X, ZHAI S *et al*.: Anti-melanoma differentiation-associated gene-5 antibody-positive dermatomyositis with liver dysfunction: a warning sign of higher death risk. *Clin Rheumatol* 2024; 43(11): 3389-97. https://doi.org/10.1007/s10067-024-07093-1
58. LUO R, XIA D, YU S: A meta-analysis of melanoma risk in idiopathic inflammatory myopathy patients. *Z Rheumatol* 2024; 83 (Suppl 3): 299-304. https://doi.org/101007/s00393-024-01473-3
59. CHE WI, KUJA-HALKOLA R, HELLGREN K *et al*.: Impact of cancer on the mortality of patients with idiopathic inflammatory myopathies by flexible parametric multistate modelling. *J Intern Med* 2024; 296(4): 336-49. https://doi.org/10.1111/joim.20003
60. WANG X, ALBAYDA J, PAIK JJ *et al*.: Evaluating CA-125 and PET/CT for cancer detection in idiopathic inflammatory myopathies. *Rheumatology* (Oxford) 2025; 64(4): 2115-22. https://doi.org/10.1093/rheumatology/keae470
61. JEKIELEK M, NISENBAUM R, VINIK O, KASSARDJIAN CD: Idiopathic inflammatory myopathies and malignancy screening: a survey of current practices amongst Canadian neurologists and rheumatologists. *Muscle Nerve* 2025; 72(3): 502-8. https://doi.org/10.1002/mus.28463
62. LIU S, ZHANG Z, YAN S *et al*.: Risk, risk factors, and screening of malignancies in dermatomyositis: current status and future perspectives. *Front Oncol* 2025; 15: 1503140. https://doi.org/10.3389/fonc.2025.1503140
63. YANG H, ZHANG L, TIAN X *et al*.: Distinct phenotype and prognosis of immune-mediated necrotizing myopathy based on clinical-serological-pathological classification. *Rheumatology* (Oxford) 2025; 64(4): 2252-64. https://doi.org/10.1093/rheumatology/keae361
64. YANG H, SUN C, YE L *et al*.: Association of anti-HMGCR antibodies of the IgM isotype with refractory immune-mediated necrotizing myopathy. *Arthritis Res Ther* 2024; 26(1): 158. https://doi.org/10.1186/s13075-024-03387-6
65. WISCHNEWSKI S, THÄWEL T, IKENAGA C *et al*.: Cell type mapping of inflammatory muscle diseases highlights selective myofiber vulnerability in inclusion body myositis. *Nat Aging* 2024; 4(7): 969-83. https://doi.org/10.1038/s43587-024-00645-9
66. YAMASHITA S, TAWARA N, SUGIE K, SUZUKI N, NISHINO I, AOKI M: Impact of sex, age at onset, and anti-cN1A antibodies on sporadic inclusion body myositis. *J Neurol Sci* 2024; 464: 123164. https://doi.org/10.1016/j.jns.2024.123164
67. BEECHER G, MUHAMMAD S, SHAMMAS I *et al*.: Increased risk of myocardial infarction in inclusion body myositis: a non-concurrent cohort study. *Eur J Neurol* 2025; 32(5): e70177. https://doi.org/10.1111/ene.70177
68. FAGHIHI-KASHANI S, YOSHIDA A, BOZAN F *et al*.: Classification Criteria for Anti-Synthetase

Syndrome Project participating investigators. Clinical Characteristics of Anti-Synthetase Syndrome: Analysis from the Classification Criteria for Anti-Synthetase Syndrome Project. *Arthritis Rheumatol* 2025; 77(4): 477-89. https://doi.org/10.1002/art.43038
69. TANG HS, TANG IYK, HO RTC *et al*.: Clinical heterogeneity and prognostic factors of anti-synthetase syndrome: a multi-centred retrospective cohort study. *Rheumatology* (Oxford) 2025; 64(1): 212-20. https://doi.org/10.1093/rheumatology/kead671
70. ZHAO B, HOU Y, SHAO K *et al*.: Clinico-seropathological characteristics of anti-Ha antisynthetase syndrome. *Brain Pathol* 2025; 35(3): e13319. https://doi.org/10.1111/bpa.13319
71. HASEGAWA A, KURASAWA K, KOIKE R *et al*.: Clinical features and risk factors of flare in anti-synthetase syndrome. *Clin Rheumatol* 2025; 44(6): 2431-38. https://doi.org/10.1007/s10067-025-07398-9
72. YAMAGUCHI K, SULLIVAN DI, KHUSHBOO S *et al*.: Long-term clinical prognosis of anti-aminoacyl-tRNA synthetase antibodies and interstitial lung disease. *Clin Rheumatol* 2025; 44(8): 3341-52. https://doi.org/10.1007/s10067-025-07521-w
73. SHAN X, HUANG Z, WANG G, GE Y: Predictive factors and clinical outcomes of progressive pulmonary fibrosis in anti-threonyl (PL7) positive anti-synthetase syndrome. *Rheumatology* (Oxford) 2025; 64(10): 5379-87. https://doi.org/10.1093/rheumatology/keaf306
74. BARSOTTI S, LUNDBERG IE: Current treatment for myositis. *Curr Treatm Opt Rheumatol* 2018; 4(4): 299-315. https://doi.org/10.1007/s40674-018-0106-2
75. NEVES A, VIVEIROS L, VENTURELLI V, ISENBERG DA: Where are we now in biologic drugs for myositis? *Rheumatology* (Oxford) 2024; 63(11): 2938-47. https://doi.org/10.1093/rheumatology/keae096
76. CHARLES-SCHOEMAN C, SCHESSL J, BATA-CSÖRGŐ Z *et al*.: Predictors of response to intravenous immunoglobulin in patients with dermatomyositis: the ProDERM study. *Rheumatology* (Oxford) 2025; 64(6): 3767-76. https://doi.org/10.1093/rheumatology/keaf070
77. SHARF K, DO T, GHETIE D, CHOI D, CHAHIN N: Benefits of early versus late initiation of intravenous immunoglobulin in the treatment of patients with anti-3-hydroxy-3-methylglutaryl-coenzyme a reductase immune-mediated necrotizing myopathy. *Arthritis Care Res* (Hoboken) 2024; 76(11): 1584-92. https://doi.org/10.1002/acr.25406. Erratum in: *Arthritis Care Res* (Hoboken) 2025; 77(3): 419. https://doi.org/10.1002/acr.25496
78. GAMBA A, DEPASCALE R, ZANATTA E *et al*.: Effectiveness and safety of low dose rituximab as remission-maintenance treatment for patients with refractory idiopathic inflammatory myopathies: results of a retrospective study from a monocentric cohort. *Clin Rheumatol* 2024; 43(10): 3167-74. https://doi.org/10.1007/s10067-024-07079-z
79. CONTICINI E, CAMELI P, GRAZZINI S *et al*.: Efficacy and safety of a step-down regimen of low dosage of glucocorticoids combined with early administration of synthetic or biologic immunosuppressants in anti-synthetase syndrome: a pilot study. *Semin Arthritis Rheum* 2024; 69: 152560. https://doi.org/10.1016/j.semarthrit.2024.152560
80. MANWATKAR A, NARESH K, MATHEW J *et al*.: Comparison of rituximab efficacy in treatment-naïve and refractory inflammatory myopathies: experiences from a tertiary care centre. *Rheumatology* (Oxford) 2025; 64(4): 2091-98. https://doi.org/10.1093/rheumatology/keae307
81. AGGARWAL R, LUNDBERG IE, SONG YW, SHAIBANI A, WERTH VP, MALDONADO MA: Efficacy and safety of subcutaneous abatacept plus standard treatment for active idiopathic inflammatory myopathy: phase 3 randomized controlled trial. *Arthritis Rheumatol* 2025; 77(6): 765-76. https://doi.org/10.1002/art.43066
82. SHAN X, WU S, CHEN X, GE Y: Janus kinase inhibition (JAKi) therapy in refractory anti-synthetase syndrome: A retrospective cohort study. *Semin Arthritis Rheum* 2024; 68: 152474. https://doi.org/10.1016/j.semarthrit.2024.152474
83. SHOBHA V, KODALI RS, AMIN SN *et al*.: Effectiveness of generic tofacitinib in idiopathic inflammatory myositis (IIM)-a retrospective analysis from Indian Myositis Registry (MyoIN). *Clin Rheumatol* 2024; 43(7): 2245-52. https://doi.org/10.1007/s10067-024-07019-x
84. XING X, LI Y, LIU Q *et al*.: Tofacitinib treatment for active dermatomyositis and anti-synthetase syndrome: a prospective cohort pilot study. *Rheumatology* (Oxford) 2025; 64(6): 3756-66. https://doi.org/10.1093/rheumatology/keaf046
85. PENG Q, YAO X, CHEN S, LI X, LIN F, ZOU Z: Efgartigimod combined with steroids as a fast-acting therapy for anti-SRP immune-mediated necrotizing myopathy. *Front Neurol* 2025; 16: 1560483. https://doi.org/10.3389/fneur.2025.1560483
86. YANG MT, YUAN JC, WANG YK *et al*.: Treatment of refractory immune-mediated necrotizing myopathy with efgartigimod. *Front Immunol* 2024; 15: 1447182. https://doi.org/10.3389/fimmu.2024.1447182