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Juvenile Dermatomyositis: A Review of the Literature

ABSTRACT

Juvenile dermatomyositis (JDM) is the most common idiopathic inflammatory myopathy of childhood, characterized by proximal muscle weakness, pathognomonic cutaneous findings, and multisystem involvement. Over the past decade, significant advances have been made in pathogenesis and treatment of JDM. This review provides a comprehensive update on the etiopathogenesis, clinical features, autoantibody profiles, and treatment of JDM. Type I interferon signaling, vasculopathy, and mitochondrial dysfunction have emerged as interconnected pathogenic mechanisms, and their identification has informed the development of targeted therapeutic strategies. In particular, elucidation of the type I interferon pathway has contributed to disease diagnosis, monitoring of disease activity through interferon-related biomarkers, and the development of novel targeted therapies. Myositis-specific autoantibodies (MSA), detected in approximately 70% of patients, define clinically distinct subgroups; for example, anti-NXP2 is associated with calcinosis, and anti-MDA5 positivity carries a risk of interstitial lung disease. Importantly, MSA subtype influences not only clinical phenotype but also treatment response, underscoring the value of systematic autoantibody testing in guiding therapeutic decisions. A treat-to-target approach and close monitoring of disease activity are recommended. In refractory disease, conventional immunosuppressants and biologic agents are escalated sequentially, with Janus kinase (JAK) inhibitors targeting the type I interferon pathway. More selective interferon blockade with anifrolumab and next-generation B-cell depletion with chimeric antigen receptor T-cell (CAR-T) cell therapy represent emerging options for patients with refractory disease. Calcinosis remains without a standardized treatment procedure, and evidence-based management is limited by the lack of pediatric randomized controlled trials. Prospective, multicenter studies are needed to establish treatment algorithms and improve long-term outcomes in this rare disease.

Keywords: Autoantibodies, calcinosis, interferon type I, interstitial lung disease, janus kinase inhibitors, juvenile dermatomyositis

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Introduction

Juvenile idiopathic inflammatory myopathies (JIIMs) are heterogeneous, systemic autoimmune diseases characterized by inflammation of the skeletal muscle and skin, with underlying capillary vasculopathy.[1] Hallmark features include symmetrical proximal muscle weakness and pathognomonic cutaneous findings, such as Gottron papules and heliotrope rash.[2]

Juvenile dermatomyositis (JDM) is the most common JIIM; other JIIMs include clinically amyopathic juvenile dermatomyositis (CAJDM), antisynthetase syndrome (AsyS), immune-mediated necrotizing myopathy (IMNM), and overlap myositis.[3] Juvenile polymyositis, historically defined as a JIIM

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subtype characterized by the absence of typical cutaneous findings and more prominent muscle involvement, is now considered rare and has largely been reclassified into IMNM or AsyS. Inclusion body myositis (IBM) and cancer-associated myositis are mostly seen in adulthood[4] (Table 1).

This review focuses on updates in JDM, in terms of etiopathogenesis, clinical features, myositis antibodies, and treatment strategies.[5]

Epidemiology

The disease occurs worldwide, with an incidence range from 2.5 to 4.1 cases per million children per year.[6,7] As in many autoimmune diseases, girls are more affected, with a girl-to-boy ratio of 2-2.3:1.[8,9] The disease can occur at any age after early childhood, with a mean age at symptom onset ranging from 5.9 to 8.1 years.[10,11] Despite increasing awareness, diagnostic delay remains a significant concern, with reported delays ranging from 2.7 to 9.6 months.[10,12]

Etiopathogenesis

Although the exact etiology of JDM remains unclear, it is considered a "type I interferon-driven autoimmune microangiopathy" triggered by environmental factors in genetically susceptible individuals.[13] Genetic susceptibility has been strongly associated with HLA-DRB1, DRB5, and DRB6. Non-HLA variants, including PTPN22 and TYK2, are known as JDM associated variants, while more recent studies have implicated IRF4 and STAT4 loci.[14] Environmental exposures, such as infections, ultraviolet radiation (UV), medications, vaccinations, and psychosocial stress, have been documented as potential triggers for the disease onset or flares.[15-17]

MAIN POINTS

- *Juvenile dermatomyositis (JDM) is a rare autoimmune myopathy of childhood characterized by proximal muscle weakness and pathognomonic skin findings, with multisystem involvement that substantially impacts long-term outcomes.*
- *The myositis-specific autoantibody profile defines clinical phenotype, prognosis, and treatment decisions in JDM.*
- *Type I interferon signaling, vasculopathy, and mitochondrial dysfunction are interconnected pathogenic mechanisms offering potential therapeutic targets.*
- *Established pathogenic mechanisms are being targeted by more selective agents, including anifrolumab as an alternative to JAK inhibitors for interferon pathway blockade, and CAR-T cell therapy as a next-generation approach to B-cell depletion beyond rituximab.*
- *Calcinosis and lipodystrophy remain major contributors to long-term morbidity in JDM; both lack standardized treatment protocols, and early, aggressive disease control represents the most effective preventive strategy.*

Activated plasmacytoid dendritic cells (pDCs) initiate multiple immunological mechanisms, including adaptive immunity, interferon signaling, complement system, and neutrophil extracellular trap formation (NETosis).[13,18,19] Activated pDCs promote T-cell activation, leading to the expansion of $CD4^+$ T cells, including Th1 and Th17 populations, which produce pro-inflammatory cytokines that amplify inflammation and recruit immune cells to affected tissues. The resulting inflammatory milieu involves multiple immune cell populations, including regulatory T cells, B lymphocytes, macrophages, and plasma cells.[20,21] B-cell activation leads to the production of myositis-specific autoantibodies (MSAs). These autoantibodies may form immune complexes, which further stimulate innate immune pathways and promote activation of the complement system.[21]

Interferon Pathway

Overactivation of the interferon pathway is one of the best-characterized pathogenic mechanisms in JDM. Pattern recognition receptors, such as TLR7 and TLR9, detect endogenous nucleic acids released from damaged cells, while the cGAS-STING pathway is activated by cytosolic DNA.[22,23] Through these signals, pDCs produce type I interferons, which activate the JAK-STAT signaling pathway in target tissues and induce the expression of interferon-stimulated genes (ISGs).[22] Upregulation of ISGs leads to increased expression of MHC class I molecules in affected tissues, particularly on muscle fibers. This may cause endoplasmic reticulum stress and contribute to immune-mediated muscle injury.

Increased ISG expression, also described as interferon signature, correlates with disease activity in JDM.[23,24] Certain ISGs demonstrate tissue-specific patterns of expression; for example, CXCL10, CXCL9, and IFI44L have been associated with cutaneous disease activity,[25] whereas ISG15 is specifically associated with muscle involvement.[26] Interferon-related proteins, such as CXCL10, galectin-9, MxA, and SIGLEC1, are elevated during active disease and represent potential biomarkers for predicting disease flares.[27-29]

Mitochondrial Dysfunction

Mitochondrial dysfunction contributes to JDM pathogenesis independently of interferon signaling. In treatment-naïve patients, CD14+ monocytes exhibit dysregulation of mitochondrial gene expression that persists despite improvement of the interferon (IFN) signature. Oxidative stress-driven oxidized mitochondrial DNA (oxmtDNA) stimulates ISG expression via cGAS-STING and TLR9 pathways; this effect can be suppressed by N-acetylcysteine (NAC) and TLR9 antagonism.[30]

Spatial transcriptomic analysis of treatment-naive JDM muscle reveals downregulation of electron transport chain (ETC) pathways in muscle fibers and CD68+

Table 1. Inflammatory Myopathies

	Frequency (%)	Antibodies	Features
Juvenile dermatomyositis	80-90	anti-TIF1γ, anti-NXP2, anti-Mi2, anti-MDA5	Classic form is characterized by symmetrical proximal muscle weakness accompanied by Gottron papules and heliotrope rash.
Clinically amyopathic juvenile dermatomyositis	5-10	anti-MDA5, anti-TIF1γ	Characteristic skin manifestations without clinically evident muscle weakness. Elevated muscle enzymes indicate hypomyopathic dermatomyositis, whereas normal enzymes indicate amyopathic dermatomyositis. Muscle involvement may develop during follow-up.
Overlap myositis	5-10	anti-Pm/Scl, anti-U1RNP	Coexistence of myositis with another systemic rheumatic disease, such as systemic sclerosis, systemic lupus erythematosus, or inflammatory arthritis.
Immune-mediated necrotizing myopathy	1-5	anti-SRP, anti-HMGCR	Severe muscle weakness and markedly elevated muscle enzymes are seen. Muscle biopsy shows prominent myofiber necrosis with minimal inflammatory infiltrate. May be associated with statin use.
Antisynthetase syndrome	<5	anti-Jo-1, anti-PL7, anti-PL12, anti-EJ, anti-OJ, anti-KS, anti-Zo, and anti-Ha.	Characterized by myositis, inflammatory arthritis, interstitial lung disease, Raynaud phenomenon, fever, and mechanic's hands. Associated with anti-tRNA synthetase antibodies, most commonly anti-Jo-1. More commonly seen in adults.

From references (1, 3, 4).

macrophage infiltrates.[31] Circulating mitochondrial markers, including mtDNA, MT-ND6 protein, and anti-mitochondrial antibody, correlate with calcinosis, while electron microscopy provides direct evidence of mitochondrial calcification.[32]

In experimental myositis models, IFNγ-induced mitochondrial dysfunction and oxidative stress are attenuated by NAC, supporting the use of mitochondrial antioxidants as potential therapeutic targets.[33]

Vasculopathy

Vasculopathy is considered a central component of JDM pathogenesis. Vascular involvement follows a biphasic course: an early inflammatory small-vessel vasculitis followed by a non-inflammatory occlusive vasculopathy with capillary dropout.[34,35]

In the early phase, upregulation of adhesion molecules, such as VCAM-1 and ICAM-1, increases vascular permeability and facilitates perivascular inflammatory cell recruitment while promoting the release of chemokines including monocyte chemoattractant protein-1 (MCP-1) and macrophage inflammatory proteins (MIP).[36] Activation of the complement system, particularly deposition of the membrane attack complex (C5b-9) on endothelial cells, contributes to microvascular damage. Together, these mechanisms result in capillary loss, impaired angiogenesis, and subsequent tissue ischemia.[34] NETosis, driven by calcium crystal-mediated frustrated phagocytosis, contributes to vascular injury through myeloperoxidase-driven oxidative modification of HDL and endothelial damage. Impaired NET clearance further sustains this proatherogenic milieu.[19] Histopathological findings of perivascular Th1 and Th17 infiltrates implicate the vessel wall as an active site of immune-mediated injury.[20]

As the disease progresses, inflammatory vasculitis gives way to a non-inflammatory occlusive vasculopathy marked by endothelial swelling, necrosis, and luminal occlusion, leading to capillary dropout and tissue ischemia that underlie severe complications such as cutaneous ulceration, intestinal ischemia, and calcinosis.[34] Microparticle-mediated thrombin generation contributes to a prothrombotic state, while increased arterial stiffness suggests long-term progression toward accelerated atherosclerosis.[35,37]

Classification

Although the Bohan and Peter criteria[38,39] have been widely used for the classification of JIIM since 1975, they were not specifically developed for pediatric populations (Table 2). The EULAR/ACR 2017 classification criteria introduced a probability-based scoring system incorporating muscle biopsy findings and serological features.[2] These criteria demonstrated improved sensitivity (96.5%) and specificity (85%).[40] However, they include only anti-Jo1 among autoantibodies and do not fully account for distinct histopathological patterns such as IMNM.[2] Therefore, there is an ongoing effort to develop updated classification criteria, including patterns of muscle weakness, biopsy-defined myonecrosis, additional rashes, skin biopsy, capillaroscopy, interstitial lung disease (ILD), arthritis, Raynaud's phenomenon, and a broader spectrum of antibodies.[41]

Table 2. Bohan and Peter Criteria for Dermatomyositis and Polymyositis[38,39]

	Criteria	Definition
A	Symmetrical proximal muscle weakness	Symmetrical weakness of the limb-girdle muscles and anterior neck flexors, progressing over weeks to months, with or without dysphagia or respiratory muscle involvement.
B	Muscle biopsy	Muscle biopsy evidence of necrosis of type I and II fibers, phagocytosis, regeneration, perifascicular atrophy, variation in fiber size, and perivascular inflammatory exudate.
C	Elevated serum muscle enzymes	Elevation of skeletal-muscle enzymes: creatine phosphokinase, aspartate aminotransferase, lactate dehydrogenase, and aldolase.
D	Electromyography	Electromyographic triad of (1) short, small, polyphasic motor units, (2) fibrillations, positive sharp waves, and insertional irritability, and (3) high frequency repetitive discharges.
E	Dermatologic features	Heliotropic rash (lilac discoloration of eyelids) with periorbital edema, Gottron's sign (a scaly, erythematous dermatitis over the dorsum of the metacarpophalangeal and proximal interphalangeal joints
Dermatomyositis Definite- 3 of A-D and E Probable- 2 of A-D and E Possible- 1 of A-D and E		Polymyositis Definite- all of A-D Probable- 3 of A-D Possible- 2 of A-D

Clinical Features

Juvenile dermatomyositis may follow a monocyclic course with sustained remission after a single episode, a polycyclic relapsing–remitting course, or a chronic continuous course. Approximately one-third of patients achieve monocyclic disease, 5-15% follow a polycyclic course, and 30-50% follow a chronic continuous course requiring prolonged immunosuppression. A chronic continuous course is the strongest predictor of poor long-term outcome.[10,11,42,43]

Muscle Involvement

Myopathy, presenting as acute or subacute symmetric proximal muscle weakness, occurs in 85% of patients.[12,42] Pelvic and shoulder girdle involvement typically presents with difficulty getting out of bed, climbing stairs, and raising arms overhead. Neck flexors are among the earliest affected muscle groups. Pharyngeal muscle involvement occurs in one-quarter of patients, manifesting as dysphagia and dysphonia, with aspiration risk.[42,44]

Needle electromyography (EMG) typically shows a myopathic triad of short, low-amplitude, polyphasic motor unit potentials, fibrillations with positive sharp waves and insertional irritability, and high-frequency repetitive discharges.[38,39] Given its invasive nature, EMG is no longer routinely performed and is currently recommended only when neuropathy cannot be excluded on clinical grounds.[45]

Muscle biopsy is likewise recommended for atypical presentations or diagnostic uncertainty.[45] When performed, findings may include perifascicular atrophy, fiber size variability, perivascular inflammatory infiltration, regeneration, and necrosis of muscle fibers.[38,39] Biopsy findings assessed with standardized scoring systems, combined with MSAs, have shown value in predicting long-term outcome.[46]

Magnetic resonance imaging (MRI) has largely replaced EMG and muscle biopsy owing to its non-invasive nature.[45,47] In one cohort, muscle MRI findings supported the diagnosis in 80% of evaluated cases.[20] High signal intensity on T2-weighted or STIR sequences, reflecting muscle edema, is considered a reliable marker of disease activity.[45]

Nearly 25% of patients with JDM present without clinically detectable muscle weakness, a subgroup defined as clinically amyopathic JDM (CAJDM). Clinically amyopathic juvenile dermatomyositis is characterized by cutaneous findings for at least 6 months without clinical muscle weakness or elevated muscle enzymes, whereas hypomyopathic JDM denotes subclinical myositis evidenced by laboratory, imaging, or electrophysiological evaluation.[48] Diagnostic delay is significantly longer in CAJDM compared to classical JDM. Anti-TIF1γ and anti-MDA5 are the most common MSAs associated with CAJDM.[49] Although CAJDM represents a milder phenotype, calcinosis and cutaneous ulceration may develop in inadequately treated patients, necessitating close follow-up.[50,51]

Cutaneous Features

Pathognomonic cutaneous features, such as heliotrope rash and Gottron's papules, occur in nearly all patients. Heliotrope rash is a symmetrical violaceous rash of the upper eyelids, often with periorbital edema (85-100%). Gottron's papules are erythematous scaly papules overlying the extensor surfaces of the joints (90-95%).[8,42]

Both nasolabial sparing "malar rash" (65-70%) and erythema crossing nasolabial folds, called "facial erythema" (50-55%) are seen in JDM. V-sign and shawl sign (3-10%) are photosensitive lesions. The V-sign involves the neck and anterior chest; the shawl sign affects the shoulders and upper back. Both are more prominent in

anti-TIF1γ.[9,12,42] Mechanic's hands, presenting as hyperkeratotic fissuring of the fingertips and lateral digits (5%), are rare in JDM and associated with anti-synthetase syndrome. Cutaneous ulceration (5-40%) occurs more frequently in anti-NXP2 and anti-MDA5. Raynaud's phenomenon, more common in overlap myositis, may also occur[52] (Figure 1) .

Calcinosis and Lipodystrophy: Calcinosis is defined as the dystrophic deposition of insoluble calcium salts in the skin and subcutaneous tissue. As calcinosis may cause ulceration, joint contractures, and recurrent infections, it is one of the most significant complications. It affects up to 40% of patients and is present at diagnosis in 3.4-8.6% of cases.[8,9,11,53]

Risk factors for calcinosis include early disease onset, especially before the age of 6 years, delayed diagnosis, chronic disease course, and anti-NXP2 and anti-MDA5 positivity.[11,54-56] Early nailfold capillaroscopy changes[57] and high inflammation scores in muscle biopsies[11] additionally increase the risk of calcinosis. Calcinosis arises from mitochondrial calcium deposition and correlates with anti-mitochondrial antibody positivity, with levels rising prior to clinical detection, suggesting a role as an early predictive biomarker.[32]

Calcinosis is classified into 4 subtypes based on distribution and depth: circumscripta, tumoral, universalis, and exoskeleton type; different subtypes may coexist in the same[58] (Figure 2).

Plain radiography remains the most accessible and widely used modality for detection and monitoring of calcinosis, although it may miss early, subtle calcifications and deeper fascial deposits. Low-dose whole-body computed tomography (CT), which provides volumetric quantification, has been validated. The use of magnetic resonance imaging (MRI) is limited due to its cost, time demand, and lower sensitivity for calcific deposits. No standardized imaging protocol currently exists for calcinosis.[59] Durometry has been proposed as a quantitative tool for the assessment of calcinosis firmness and monitoring treatment response.[60]

Lipodystrophy is a less frequent but clinically significant complication, affecting 10-40% of patients. It typically emerges at an average of 2.5-6 years after disease onset, following a cephalocaudal distribution from the face to the trunk and extremities. A polycyclic or chronic disease course, panniculitis, delayed diagnosis, and prolonged, uncontrolled disease activity are the principal risk factors for lipodystrophy.[61,62]

Nailfold Capillary Changes: Nailfold capillaroscopy (NFC) is a non-invasive technique reflecting this microvascular injury. At diagnosis, capillary end-row loop density is markedly reduced in treatment-naive patients.[63,64] Capillary loss correlates primarily with cutaneous disease activity, whereas its association with muscle involvement remains weaker.[63-65] Cumulative capillary loss is associated with chronic disease and frequently fails to normalize despite treatment.[66]

Nailfold capillaroscopy findings overlap with the scleroderma pattern, with capillary loss, giant capillaries, microhemorrhages, and avascular areas observed in both conditions. However, abnormal capillary morphology and bushy (ramified) capillaries, reflecting an underlying neoangiogenic process, are more specific to JDM (Figure 3).[67,68] Anti-TIF1-γ is associated with the lowest capillary density and an active scleroderma pattern.[63,64,69]

Nailfold capillaroscopy abnormalities carry prognostic value across multiple outcomes: baseline NFC abnormality predicts calcinosis,[57] cumulative capillary loss is associated with lipodystrophy,[66] periungual hemorrhages correlate with dysphagia,[63] and low capillary density is associated with impaired pulmonary function.[70]

Arthritis

Arthritis in JDM is nonerosive, presenting in an oligoarticular or polyarticular pattern; the knees, wrists, elbows, and small joints of the hands are the most commonly affected joints.[71] Contractures in JDM do not result from chronic arthritis but from muscle inflammation and fibrosis. Although arthralgia and arthritis are reported in 14-28% of patients at disease onset,[9,12] the prevalence approaches 50% during long-term follow-up.[10,42]



Figure 1.Cutaneous findings in juvenile dermatomyositis. (A) Gottron papules, (B) heliotrope rash, (C) malar rash, (D) Raynaud phenomenon.

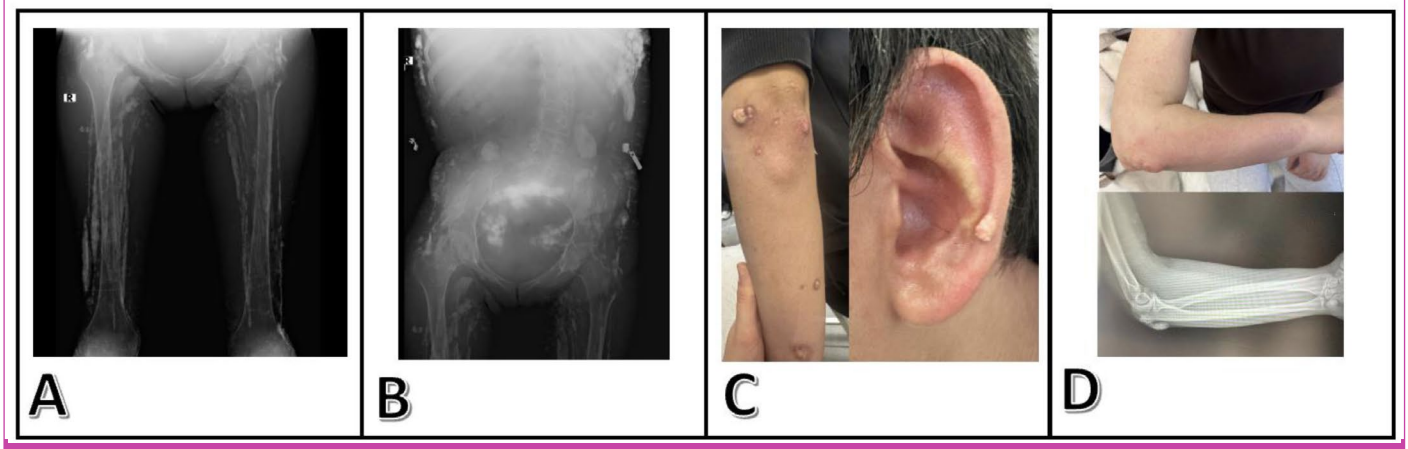


Figure 2.Spectrum of calcinosis phenotypes in juvenile dermatomyositis. (A) *Calcinosis universalis*, characterized by extensive linear calcium deposition along fascial planes and muscle groups, (B) *exoskeleton calcinosis*, with diffuse sheet-like calcification covering a large body surface area, (C) *calcinosis circumscripta*, presenting as localized superficial plaques and nodules in the skin and subcutaneous tissue, and (D) *tumoral calcinosis*, appearing as large focal calcified masses around the joints.

Pulmonary Involvement

Pulmonary involvement occurs in 15% of patients across cohorts,[10,20,42] with a twofold higher prevalence observed in Asian cohorts.[72] Anti-MDA5 positivity is complicated by ILD in up to 80% of cases, with a strong association with rapidly progressive ILD (RP-ILD).[73] Although rare in the pediatric population, anti-Ro52 and anti-ARS antibodies represent additional risk factors for ILD.[73,74] Anti-MDA5 positivity, elevated erythrocyte sedimentation rate (ESR), and elevated IL-10 are independent predictors of ILD.[75]

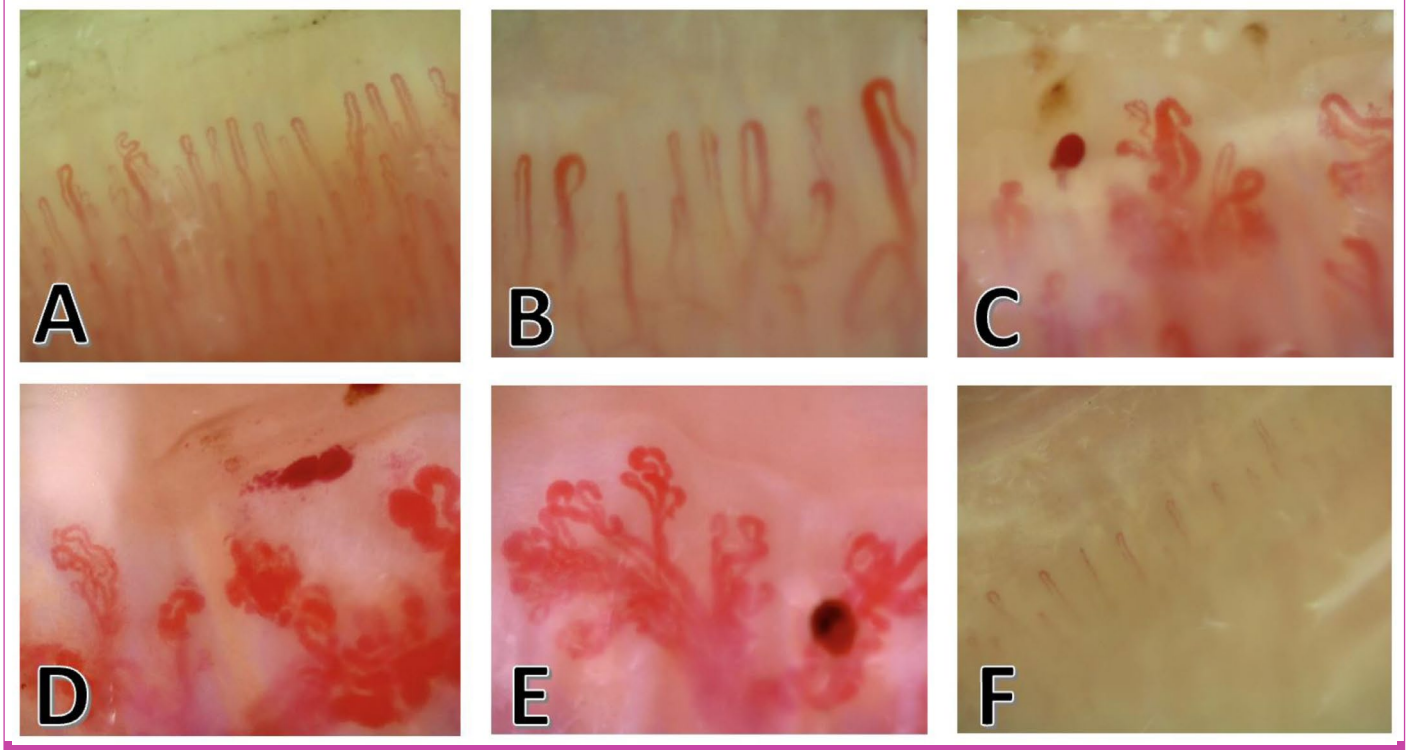


Figure 3.Nailfold capillaroscopy findings in patients with juvenile dermatomyositis. (A) Normal capillary pattern. (B) Early stage: presence of dilated capillaries. (C) Active stage: giant capillaries and neoangiogenesis with active and chronic hemorrhage. (D-E) Active to late stage: neoangiogenesis, bushy capillary structures with hemorrhage. (F) Late stage: reduced capillary density and avascular areas.

In CAJDM, ILD may represent the initial and only manifestation. In classic JDM, ILD frequently follows an asymptomatic early course; by the time respiratory symptoms emerge, fibrosis may have already developed.[75,76] Rapid deterioration can occur even in initially asymptomatic patients with minimal imaging findings.[77] Pulmonary function tests (PFTs), which reveal a restrictive pattern with reduced total lung capacity (TLC) and forced vital capacity (FVC), often reflect respiratory muscle involvement. A reduced diffusing capacity of the lungs for carbon monoxide (DLCO) is more specific for alveolar involvement and may indicate underlying ILD even in the absence of respiratory symptoms.[73,78]

The predominant pattern in high-resolution computed tomography (HRCT) is nonspecific interstitial pneumonia (NSIP), characterized by bilateral ground-glass opacity (GGO) and reticulation with lower lobe predominance. Organizing pneumonia (OP) pattern, manifesting as consolidation with or without GGO, may coexist. Ground-glass opacity predominance reflects active inflammation and is associated with better treatment response, whereas reticulation and traction bronchiectasis indicate advancing fibrosis. In RP-ILD, diffuse or random subpleural GGO with consolidation is characteristic and portends a poor prognosis.[73,78-80]

Serum KL-6 is a useful biomarker for detecting ILD and monitoring disease severity, correlating with CT severity scores and aids in monitoring treatment response.[81,82] Elevated ferritin and IL-18 reflect alveolar macrophage activation in RP-ILD.[77]

Gastrointestinal Involvement

Gastrointestinal involvement occurs in 22-37% of JDM cases,[1,83] with higher rates in anti-NXP2-positive cases. Presentations include bowel perforation, ulceration, obstruction, or wall thickening. The colon and duodenum are the most frequently affected sites.[83] Vasculopathy is the central pathogenic mechanism, and all complications occur during active disease.[83,84] Risk factors include severe disease at onset, malnutrition, skin ulceration, edema, lymphopenia, and hypoalbuminemia.[83] Acute pancreatitis should be included in the differential diagnosis. Serum lipase measurement is recommended in patients with active JDM presenting with abdominal pain.[85] Given the insidious onset and mortality reaching 26%, early recognition is essential.[83]

Cardiac Involvement

Cardiac involvement is uncommon in JDM, and available data remain limited. Conventional echocardiography is normal in most patients, although subclinical myocardial dysfunction may be detected by speckle-tracking echocardiography as reduced left ventricular global longitudinal strain.[86] Cardiac and pulmonary involvement do not appear to correlate.[87] Carotid-radial pulse wave velocity is increased in JDM patients compared with healthy controls, indicating enhanced peripheral arterial stiffness, potentially reflecting ongoing microvascular injury and raising concern for future accelerated cardiovascular morbidity.[35,37]

Autoantibodies

Autoantibodies are identified in approximately 70% of JDM patients, defining clinically and prognostically distinct homogeneous subgroups. The clinical phenotypes associated with the major autoantibodies are summarized in Figure 4. Autoantibody status has a direct implication for treatment stratification, with differential responses observed across MSA subgroups.[4,88]

Myositis-Specific Antibodies

Anti-TIF1 γ: Anti-TIF1γ is identified in 18-30% of patie nts.[4,49,89] Age at disease onset is approximately 7 years; notably, anti-TIF1γ positivity is strongly associated with early-onset disease, especially before the age of 3 years.[90,91]

The associated phenotype features predominant cutaneous involvement. Widespread photosensitive rashes, malar rash, V-sign and shawl-sign, linear extensor erythema, Gottron papules, cutaneous ulceration, and erythroderma are characteristic.[49,89,92] Increased UV radiation exposure in the 30 days preceding symptom onset has been associated with anti-TIF1γ positivity, suggesting a role for UV-triggered interferon activation.[93] Anti-TIF1γ positivity is associated with greater nailfold capillary loss than other MSA subgroups.[69]

This subgroup follows a predominantly chronic disease course, with a monophasic disease less frequently observed than in other subgroups.[69,89] Disease flares after achieving a complete clinical response are common.[90] In adult DM, anti-TIF1γ is a strong marker for cancer-associated myositis. In children, this association is markedly weaker, and routine malignancy screening is not recommended in JDM.[52,91]

Anti-NXP2: Anti-NXP2 is the second most common MSA in JDM, detected in 12-23% of patients,[4,56,89] and is considerably rarer in adults. Median age at disease onset ranges from 6 to 6.5 years.[56,89] Muscle involvement is more severe than in other MSA subgroups, with prominent weakness, muscle cramps, atrophy, joint contractures, and dysphonia.[4,89,90,94] Creatine kinase (CK) levels are elevated accordingly.[56,95] Gottron papules and heliotrope rash are present in nearly all patients, whereas truncal rashes are not typically expected.[56,89,95] Interstitial lung disease is rarely observed.[91]

Due to the more severe and prominent vasculopathic disease course, skin ulcers, lipodystrophy, calcinosis, and gastrointestinal involvement are frequent.[83,96] Calcinosis is the most strongly associated complication, reported in 42% of patients, with the risk increasing substantially in those diagnosed before age 5 years.[56] Gastrointestinal involvement, while uncommon in JDM overall, occurs most frequently in anti-NXP2-positive patients,

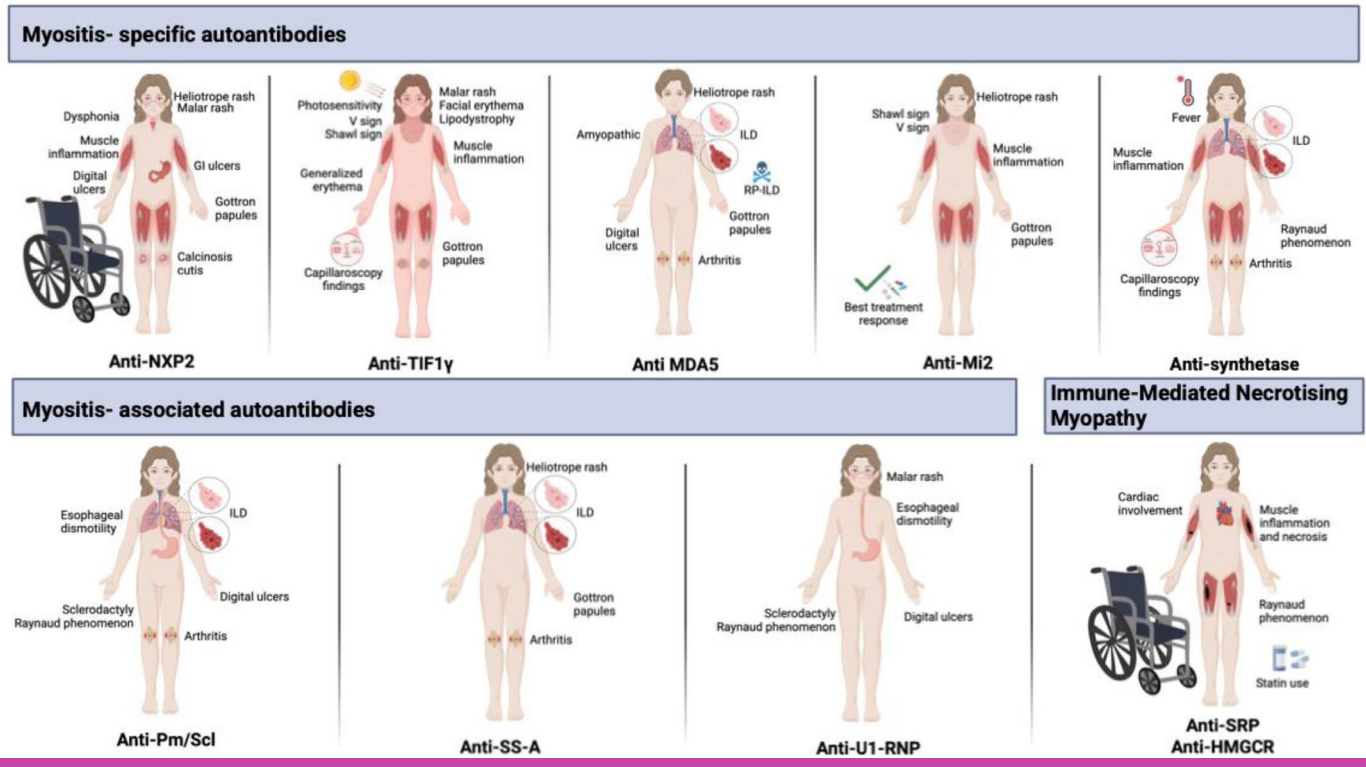


Figure 4. Clinical phenotypes associated with autoantibodies in juvenile dermatomyositis. Myositis-specific autoantibodies (MSAs): Anti-NXP2, Anti-TIF1γ, Anti-MDA5, Anti-Mi-2, Anti-synthetase. Myositis-associated autoantibodies (MAAs): Anti-PM/Scl, Anti-SS-A, Anti-U1-RNP. Immune-mediated necrotizing myopathy (IMNM)-associated autoantibodies: Anti-SRP, Anti-HMG-CoA reductase. Each panel illustrates the predominant clinical features associated with the respective autoantibody. Created with BioRender.com.

exclusively during active disease, with preceding infection as a key trigger.[83]

Disease course is typically chronic and severe, with a low drug-free remission rate.[4,91] Three-year relapse rates reach 60%, significantly exceeding those of other MSA subgroups.[95] Edema, skin ulceration, severe muscle weakness, and lymphopenia predict poor outcomes and gastrointestinal complications.[83,94]

Anti-MDA5: Anti-MDA5 is identified in 6-15% of JDM patients.[49,97,98] Median age at disease onset is 8.7-9.6 years in different series, which is older than in the other major MSA subgroups, with a male predominance.[97,98] Interstitial lung disease is the defining and most consequential feature, occurring in 20-60% of anti-MDA5-positive patients.[97-99] Rapidly progressive interstitial lung disease, the principal cause of mortality in adult anti-MDA5 disease, is uncommon in children, occurring in fewer than 10%, although it carries a high fatality risk when present.[97,100] Concomitant anti-Ro52 antibodies further increase the risk of RP-ILD.[98] Anti-MDA5 JDM exhibits the highest type I interferon score among all MSA subgroups. Its expression pattern most closely overlaps with STING-associated vasculopathy with onset in infancy (SAVI), a monogenic interferonopathy.[101] This overlap reflects a shared pathogenic mechanism driven by STING-IFN-β signaling, which is responsible for both vasculopathy and ILD.[98,101]

Cutaneous involvement extends beyond classic rashes to include skin and oral ulceration, inverse Gottron papules, palmar papules, chilblain-like lesions, livedo reticularis, and peripheral digital gangrene.[97,98,100] Muscle involvement is characteristically mild, with lower creatine kinase levels and lower muscle biopsy scores than other MSA groups; clinically amyopathic presentation, while hallmark in adults, is less common in children.[95,97,100]

Arthritis and arthralgia are present in up to 90% of anti-MDA5 positive patients.[97,100] Small joint polyarthritis is particularly characteristic.[99,102] Fever, weight loss, and lymphadenopathy further distinguish this subgroup.[97] Macrophage activation syndrome may be the initial presentation of anti-MDA5 JDM and should be considered in patients with hyperferritinemia, refractory systemic inflammation, and ILD.[103]

Although monocyclic disease is more frequent and overall prognosis is more favorable than anti-TIF1γ and anti-synthetase subgroups, pulmonary surveillance is warranted regardless of apparent remission, given the risk of subclinical or delayed-onset ILD.[97,102]

Anti-Mi-2: Anti-Mi-2 is detected in 3-10% of JDM patients.[4,89] The phenotype is characterized by classical dermatomyositis features with Gottron papules, heliotrope rash, malar rash, and elevated CK levels.[4,89] Calcinosis is not a prominent feature.[99] Muscle biopsy demonstrates the most severe histopathological changes among all MSA subgroups, yet anti-Mi-2 positivity is associated with a 7-fold lower probability of remaining on treatment over time.[46] Anti-Mi-2 JDM carries a very low mortality rate and the most favorable treatment response among the MSA subgroups.[46,89]

Anti-ARS: Anti-aminoacyl tRNA synthetase (anti-ARS) autoantibodies target cytoplasmic enzymes involved in protein synthesis and include anti-Jo-1, anti-PL-7, anti-PL-12, anti-EJ, and anti-OJ, of which anti-Jo-1 is the most common.[89] Anti-ARS comprises fewer than 5% of JDM patients.[89,91] The phenotype is characterized by ILD, arthritis, Raynaud's phenomenon, fever, and mechanic's hands.[89,97] Interstitial lung disease occurs in approximately 60% of patients, which is the highest frequency among all MSA subgroups, and is the primary driver of mortality.[89,97] Disease course is predominantly chronic, with no drug-free remission reported.[91,97]

Anti-HMGCR and anti-SRP: Juvenile IMNM is defined by 2 autoantibodies, anti-SRP and anti-HMGCR, each identified in 1-2% of JIIM patients.[89,104] Both subgroups present with severe proximal muscle weakness, markedly elevated CK, and minimal or absent skin rash, mimicking muscular dystrophy and causing diagnostic delay.[104-106]

While anti-HMGCR in adults is strongly associated with statin exposure, pediatric cases occur exclusively in statin-naïve children. A distinct HLA profile (DRB1*07:01) suggests a different pathogenic mechanism in the pediatric population.[104,107] Anti-SRP is more frequently associated with neck weakness, dysphagia, cardiac involvement, and ILD compared to anti-HMGCR.[105] Muscle biopsy in both subgroups demonstrates a necrotizing pattern with fiber size variation, regenerating fibers, and sarcolemmal membrane attack complex (MAC) deposition.[104,105] Both subgroups follow a severe and refractory course; virtually all anti-HMGCR–positive children ultimately require biological therapy.[104,106,107]

Myositis-Associated Antibodies

Anti-Ro52: Anti-Ro52 is the most frequently detected MAA in JDM, present in 14% of patients.[74,98] It commonly co-occurs with MSAs, especially anti-ARS and anti-MDA5.[74] The most clinically significant association of anti-Ro52 is with ILD. Anti-Ro52 positivity is an independent predictor of ILD development, regardless of concurrent MSA status. Co-positivity with anti-MDA5 further amplifies this risk and has been associated with RP-ILD.[74,98] Raynaud's phenomenon is also independently associated with anti-Ro52 positivity. Anti-Ro52-positive patients follow a more severe and chronic course of disease.[74]

Anti-U1-RNP: Anti-U1-RNP is detected in 4-5% of JIIM and is strongly associated with overlap myositis. Patients present at an older age with fewer classic JDM skin findings, whereas Raynaud's phenomenon, gastrointestinal dysmotility, and cardiac abnormalities are more frequent. Critically, anti-U1-RNP positivity is associated with mortality in JDM.[49,89,108]

Anti-Pm/Scl: Anti-Pm/Scl is present in 3-5% of JIIM and is characteristically associated with overlap syndrome, most frequently systemic sclerosis. Unlike most MAAs, it rarely co-occurs with an MSA, and its clinical profile includes mechanic's hands, Raynaud's phenomenon, and ILD.[49,108] Sclerodactyly and digital infarcts are additional distinguishing features, and ILD risk is elevated across cohorts.[98,108] Calcinosis and lipoatrophy have also been reported in anti-Pm/Scl positive cases.[49]

Differential Diagnosis

Several conditions may mimic JDM, particularly when pathognomonic cutaneous features are absent. Among muscular dystrophies (MD), Becker MD and symptomatic female carriers may present with proximal weakness indistinguishable from JDM; however, a slowly progressive course, a positive family history, absence of nailfold capillaroscopy changes, and absence of MSAs favor a dystrophic etiology.[109] Dysferlinopathy (LGMDR2) presents with subacute proximal weakness and prominent inflammatory infiltrates, which mimic myositis; however, absent dysferlin expression on immunohistochemistry is the key distinguishing feature. Importantly, unlike in JIIM, inflammatory infiltrates in muscular dystrophies do not typically invade non-necrotic myofibers.[110]

Metabolic myopathies should also be considered, particularly when exercise intolerance or episodic rhabdomyolysis is reported. Riboflavin-responsive late-onset form of multiple acyl-CoA dehydrogenase deficiency, presenting with subacute, fluctuating proximal weakness, and elevated CK, may closely mimic JDM.[111] Late-onset Pompe disease, mitochondrial myopathies, and endocrine myopathies should also be considered. Steroid-induced myopathy may complicate interpretation of disease course in patients already receiving glucocorticoids and is distinguished by normal CK. A systematic approach integrating clinical history, nailfold capillaroscopy, MSA testing, and muscle biopsy is essential to avoid misdiagnosis.[109-111]

Treatment

General Principles: The main goal of this treatment is to achieve inactive disease, defined as the complete elimination of inflammatory activity in muscles, skin, and other organ systems. In refractory cases, minimal disease activity may serve as an acceptable interim target.[112] Management is informed by guidelines and recommendations from the Childhood Arthritis and Rheumatology Research Alliance (CARRA),[113,114] the Single Hub and Access point for pediatric Rheumatology in

Europe (SHARE),[45] and the British Society for Rheumatology (BSR).[115]

A treat-to-target approach is recommended, with milestones of minimal clinical improvement by week 6, moderate improvement and normalized muscle enzymes by month 3, normalized muscle strength by month 6, and inactive disease by month 12.[112] If targets are not met, immunomodulatory therapy should be intensified rather than escalating glucocorticoids.[112] Disease-modifying antirheumatic drug (DMARD) discontinuation may be considered after at least 1 year of inactive disease off corticosteroids.[45] All treatment decisions should be individualized within a multidisciplinary team.[45,112,115]

Initial Treatment: Glucocorticoids are the cornerstone of initial therapy.[45,115] Induction with high-dose intravenous methylprednisolone (30 mg/kg/day, max 1 g) for 3 consecutive days, followed by oral prednisolone 1-2 mg/kg/day, is recommended; the intravenous route is preferred when gastrointestinal absorption is a concern.[115,116] Glucocorticoid dose should be tapered with clinical improvement and discontinued within 12 months where possible.[112]

Combination of glucocorticoids with methotrexate (MTX) is recommended as standard induction therapy.[45,115] In the PRINTO randomized controlled trial, the combination of glucocorticoids with either MTX or cyclosporine (CSA) was superior to prednisolone monotherapy. However, MTX demonstrated a more favorable safety profile and better long-term outcomes.[116] Cyclosporine remains an alternative for patients intolerant of or unresponsive to MTX.[45,116] Real-world data show that only one-third of patients achieve clinically inactive disease with glucocorticoid plus MTX.[117]

Hydroxychloroquine (HCQ) is recommended as an adjunct in all patients by CARRA consensus treatment plans, with particular benefit to those with mild or skin-predominant disease.[113,114]

Conventional Therapies in Refractory Disease: In cases of insufficient response to MTX/CSA, mycophenolate mofetil (MMF) is an effective alternative for both muscle and skin disease, including calcinosis.[45]

In severe disease manifestations such as major organ involvement or extensive ulcerative skin disease, cyclophosphamide (CYC) should be considered. A marginal structural model analysis of a UK cohort demonstrated significant reductions in skin and muscle disease activity with CYC, which sustained beyond 12 months.[45,118]

Intravenous immunoglobulin (IVIG) at 1-2 g/kg/dose monthly has demonstrated improvements in muscle strength, skin activity, global assessment scores, and laboratory parameters.[119] Intravenous immunoglobulin remains the most frequently used immunomodulatory agent for calcinosis.[55]

Biologic Agents: Biologic DMARDs (bDMARD) should be considered when there is an inadequate response to at least one conventional DMARD (cDMARD) and IVIG, each for a minimum of 12 weeks. The CARRA consensus treatment plans include 4 bDMARD options: TNFα inhibitors (adalimumab or infliximab), abatacept, rituximab, and tocilizumab.[114]

Rituximab depletes B cells via CD20-targeted depletion. In the "Rituximab in Myositis" (RIM) trial, 83% of patients with refractory myositis showed improvement with a significant steroid-sparing effect.[120] Onset of action may be delayed up to 26 weeks.[45] Rituximab is the most frequently preferred bDMARD.[114]

Abatacept is a recombinant CTLA4 fusion protein that inhibits T cell activation by blocking CD80/CD86-CD28 binding. In an open-label pilot trial of refractory JDM, 9 of 10 patients achieved improvement at week 24 with steroid-sparing effect, and reductions in IFN gene score correlated with clinical and MRI improvement.[121]

Among anti-TNF agents, infliximab and adalimumab have been associated with significant improvements in skin, muscle, and global disease activity, with calcinosis reduction in half of patients and a steroid-sparing effect.[45,122]

JAK Inhibitors: The prominent type I IFN pathway activation in JDM provides a strong rationale for JAK inhibitors (JAKi). Tofacitinib inhibits JAK1/3, whereas baricitinib and ruxolitinib inhibit JAK1/2; all are currently used off-label in JDM.[123] In a systematic literature review of 195 patients with JDM , tofacitinib was the most frequently used agent (57.4%), with improvement rates of 89.7%, 69.2%, and 92.7% for tofacitinib, ruxolitinib, and baricitinib, respectively.[124] Tofacitinib has demonstrated improvement in pulmonary findings beyond skin and muscle response.[125] Baricitinib has shown sustained benefit over long-term follow-up, with shorter disease duration prior to initiation predicting better outcomes.[126] The absence of anti-TIF1γ antibodies was the main predictor of complete or partial response to JAKi.[88] The safety profile of these inhibitors is acceptable, with upper respiratory tract infections being the most frequently reported adverse event.[124,126]

Emerging Therapies: Anifrolumab, a humanized monoclonal antibody blocking IFNAR1, has shown promising responses in refractory JDM, including in patients who failed prior JAK inhibitor treatment. Multiple case reports describe rapid improvement in skin disease, muscle activity, and MRI edema.[127,128]

Dazukibart, an anti-IFN-β monoclonal antibody, demonstrated rapid and clinically meaningful improvement in skin and muscle disease in adults with refractory dermatomyositis in a phase II randomized controlled trial, with a favorable safety profile;[129] a pediatric case of refractory JDM also showed dramatic multisystem improvement.[130]

Anti-CD19 CAR-T cell therapy enables a deep immune reset through the depletion of CD19-expressing B cells and plasmablasts, potentially eliminating autoreactive clones and achieving drug-free remission. Initial evidence in adults demonstrated major clinical response and sustained drug-free remission.[131] The first reported case of JDM showed normalization of muscle strength, near-complete resolution of skin disease, and reduction in IFN-related biomarkers after a single infusion.[132] A subsequent case series of 8 children with severe refractory autoimmune disease, including 3 with JDM, confirmed clinically substantial improvement and drug-free remission across all patients over a median follow-up of 16.5 months.[133] Formal clinical trials in pediatric rheumatology are needed to establish the role of this therapy.

Adjunctive Therapies: Sun exposure may trigger and exacerbate cutaneous manifestations through UV-mediated activation of the type I IFN pathway; therefore, broad-spectrum sunscreen and sun-protective measures are recommended for all patients. Persistent skin activity should prompt escalation of systemic immunosuppression, with topical tacrolimus 0.1% or topical corticosteroids for local control. Active physiotherapy within a multidisciplinary team, including a supervised exercise program, is integral to management. Calcium and vitamin D supplementation is recommended given the cumulative impact of corticosteroid use, systemic inflammation, and reduced mobility on bone health. Psychosocial support for patients and families should not be overlooked as part of comprehensive JDM management.[45,114,115]

Treatment of Calcinosis: No standardized treatment protocol exists for calcinosis in JDM, and evidence remains limited to case reports and small series.[45,134] Early disease control is the most established preventive strategy; when calcinosis develops, improvement is slow and may require years of sustained treatment.[55] Intravenous immunoglobulin is the only treatment linked to improvement of calcinosis, though not a major improvement, indicating that IVIG alone may not achieve complete resolution.[55,119] Anti-TNF therapy resulted in lesion reduction in half of the patients and complete resolution in one-third of the patients.[122] JAK inhibitors have shown promising results in refractory cases, although non-response has been documented and controlled data are lacking.[124,125,134] Calcium-modifying agents, including bisphosphonates (most commonly pamidronate), calcium channel blockers, and sodium thiosulfate, are used in selected cases; however, their efficacy remains uncertain and the level of evidence low.[45,134]

Discussion

Over the past decade, significant advances have been made in both the pathogenesis and treatment of JDM. This review highlights the increasingly central role of autoantibody profiling in defining clinical phenotype, prognosis, and treatment decisions. The identification of type I interferon signaling, vasculopathy, and mitochondrial dysfunction as interconnected pathogenic mechanisms has deepened the understanding of disease biology and provided a rationale for targeted therapeutic development.

Despite these advances, there are certain limitations. Calcinosis and lipodystrophy lack standardized treatment protocols, and the evidence base for managing these complications remains largely limited. Concurrently, the therapeutic landscape continues to evolve; anifrolumab and dazukibart offer more selective interferon pathway blockade beyond JAK inhibitors, while CAR-T cell therapy is gaining attention as a next-generation approach to B-cell depletion beyond rituximab. Further prospective studies are needed to establish evidence-based treatment algorithms for this rare disease.

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