PG Textbook of PEDIATRICS

VOLUME 3

Systemic Pediatrics, Pediatric Subspecialties, and Social Pediatrics

Editor-in-Chief

Piyush Gupta
MD FIAP FNNF FAMS
Professor and Head
Department of Pediatrics
University College of Medical Sciences
New Delhi, India
President, Indian Academy of Pediatrics (2021)

Editors

PSN Menon
MD MNAMS FIAP
Consultant and Head
Department of Pediatrics
Jaber-Al-Ahmed Armed Forces Hospital
Kuwait City, Kuwait

Siddarth Ramji
MD FNNF
Former Director-Professor
Department of Neonatology
Maulana Azad Medical College and
Associated Lok Nayak Hospital
New Delhi, India

Rakesh Lodha
MD
Professor
Department of Pediatrics
All India Institute of Medical Sciences
New Delhi, India

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46.7 Juvenile Dermatomyositis

Priyankar Pal

Juvenile dermatomyositis (JDM) is an uncommon chronic inflammatory multisystem disease affecting predominantly the skin and striated muscles. It is the most common inflammatory myopathy in children. It is characterized by symmetrical proximal muscle weakness, heliotrope rash, and Gottron papules **(Fig. 1)** over the knuckles and elbows. In the presteroid era, one-third children died, one-third spontaneously recovered and one-third survived with significant residual contractures and muscle atrophy. Over the last few decades, outcome has improved considerably with aggressive immunosuppressive therapies. Mortality rate in children in the West is now reported to be <3%. However, JDM is still associated with significant morbidity and mortality in our country because of late diagnosis and even later initiation of immunosuppression.

EPIDEMIOLOGY

Incidence is reported to be 3.2 per million children per year (US National Registry) and girls are more commonly affected than the boys. Onset is usually between 4 and 10 years, with an average age of 7 years. However, 25% of children have onset before 4 years of age. There is no racial predilection.

ETIOLOGY

Environmental Risk Factors

Like most autoimmune diseases, JDM is thought to be the result of environmental triggers in genetically susceptible individuals, leading to immune dysfunction.

Familial Dermatomyositis

There are several reports of rare occurrence of familial JDM and an increased frequency of other autoimmune diseases in families of children with JDM.

Human Leukocyte Antigen Relationships

Human leukocyte antigen (HLA)-B*08, DRB1*0301, and DQA1*0501 confer risk of myositis. The DQA1*0301 allele is an additional risk factor for JDM.

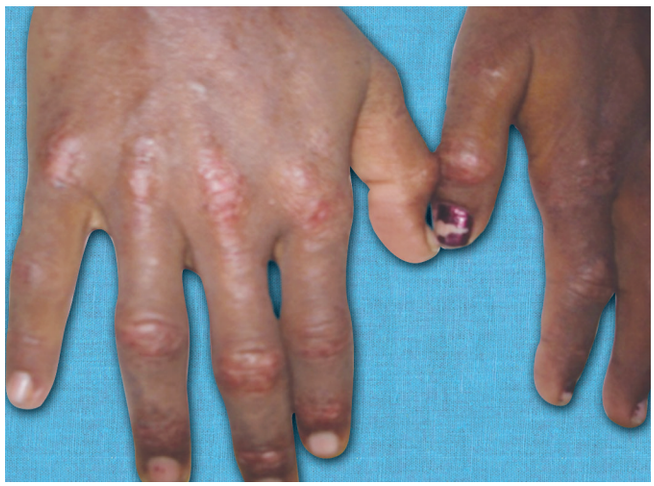


Fig. 1: Gottron papules.

PATHOGENESIS

It is characterized by perivascular inflammation early in its course. Although the exact sequence in pathogenesis is uncertain, the initial event is an immune attack on muscle capillary endothelium, followed by infiltration of dendritic cells with a resulting interferon response, and upregulation of major histocompatibility (MHC) class I expression on the surface of myofibers.

CLINICAL MANIFESTATIONS

Onset is usually insidious, with development of progressive muscle weakness and pain. A more acute onset occurs in approximately one-third of children. Easy fatigability, weakness, and low-grade fever may precede actual muscle weakness by 3–6 months.

Musculoskeletal Disease

At onset, muscle weakness is predominantly proximal, and lower extremity involvement is more common. Weakness of the anterior neck flexors, back, and abdominal muscles leads to inability to hold the head upright, and maintain a sitting posture with protrusion of the abdomen. The child may stop walking and be unable to dress or climb stairs. There may be associated muscle pain or stiffness. Physical examination demonstrates symmetrical weakness that is most pronounced in the proximal muscles of the shoulders and hips. There may be occasional edema and induration of the overlying subcutaneous tissue. The muscles may be tender and Gower sign is often present. Later in the course of disease, the distal muscles may show varying weakness especially in patients with more severe disease. Even with severe muscle weakness, the tendon reflexes are well-preserved. Pharyngeal and palatal muscles are frequently affected resulting in difficulty in swallowing and increased risk of aspiration. Esophageal hypomotility may be associated and contributes to this difficulty. Weakness of the voice, nasal speech, and nasal regurgitation are frequent signs.

Muscle strength should be sequentially measured and recorded using a standard scale like childhood myositis assessment scale (CMAS) or disease activity scale (DAS). Some children may have transient arthralgia or nondeforming arthritis. Presence of persistent arthritis in a patient with JDM signifies an overlap syndrome. Children with JDM may develop early flexion contractures.

Mucocutaneous Involvement

In a majority of children, the pathognomic cutaneous abnormalities appear either simultaneously or soon after the onset of muscle weakness. Three most typical manifestations are heliotrope discoloration of the upper eyelids, Gottron papules **(Fig. 1)**, and periungual erythema and capillary loop abnormalities. The heliotrope rash **(Fig. 2)** occurs over the upper eyelids as a violaceous, purple discoloration that is often associated with a malar rash that resembles rash of systemic lupus erythematosus (SLE) in its distribution but is less well demarcated. There may be associated edema of the eyelids and face. The rash varies in intensity and area of distribution and is photosensitive in 50% children **(Fig. 3)**. As already stated, there may be associated edema and induration, but severe edema is unusual and is indicative of a very severe disease **(Fig. 4)**. Later in the disease course, the skin may thin out and there may be atrophy of the subcutaneous structures with hypo-/hyperpigmentation.

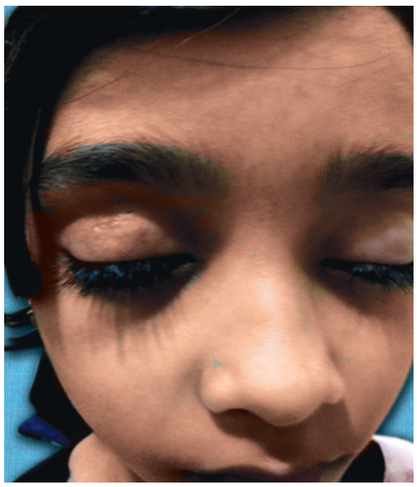


Fig. 2: Heliotrope rash.

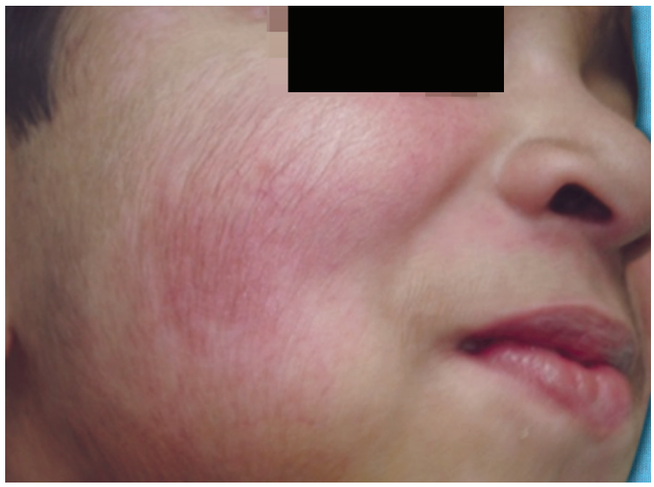


Fig. 3: Facial telangiectatic rash.

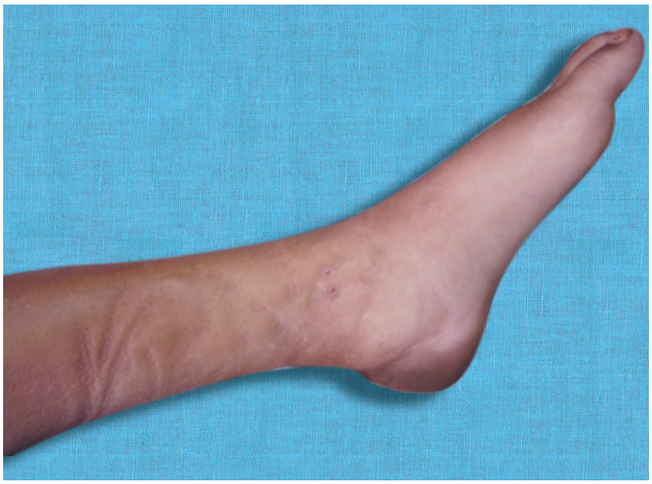


Fig. 4: Gross edema of foot in severe disease.

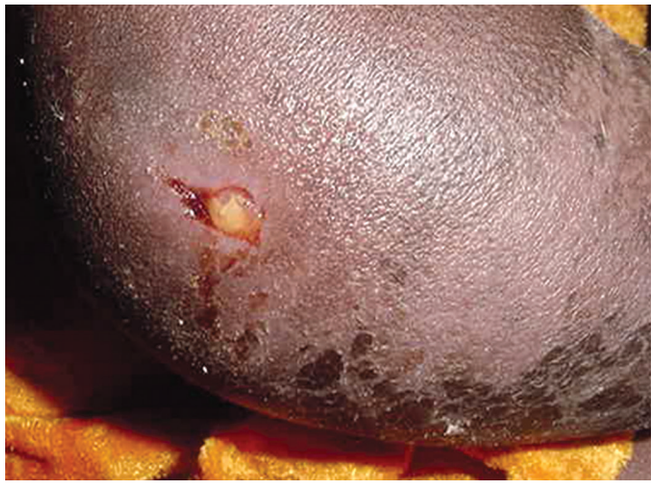


Fig. 5: Calcinosis.

Gottron papules **(Fig. 1)** are shiny, erythematous plaques occurring on the extensor surfaces of the joints. They are common over the proximal interphalangeal joints of the hands but may occasionally appear over the extensor surfaces of the elbows and knees. The periungual skin is often intensely erythematous, and careful examination with naked eye or an ophthalmoscope documents the presence of telangiectasias. Dilatation of isolated loops, thrombosis and hemorrhage, dropout of surrounding vessels, and tortuosity are distinctive, if not pathognomonic. There is often associated marked cuticular overgrowth which is a sign of active disease.

Dermatomyositis sine myositis or *amyopathic dermatomyositis* is rare in children. Some of them never develop myositis but others go on to develop classical JDM.

Calcinosis

Dystrophic calcification may occur in subcutaneous plaques or nodules in 12–43% of children **(Figs. 5 and 6)**. Risk factors include delay in diagnosis, prolonged duration of untreated disease, inadequate therapy, and underlying cardiac or pulmonary disease. These deposits are often painful and disfiguring, may restrict movements and in severe cases, the child may be encased in an exoskeleton of calcium salts.

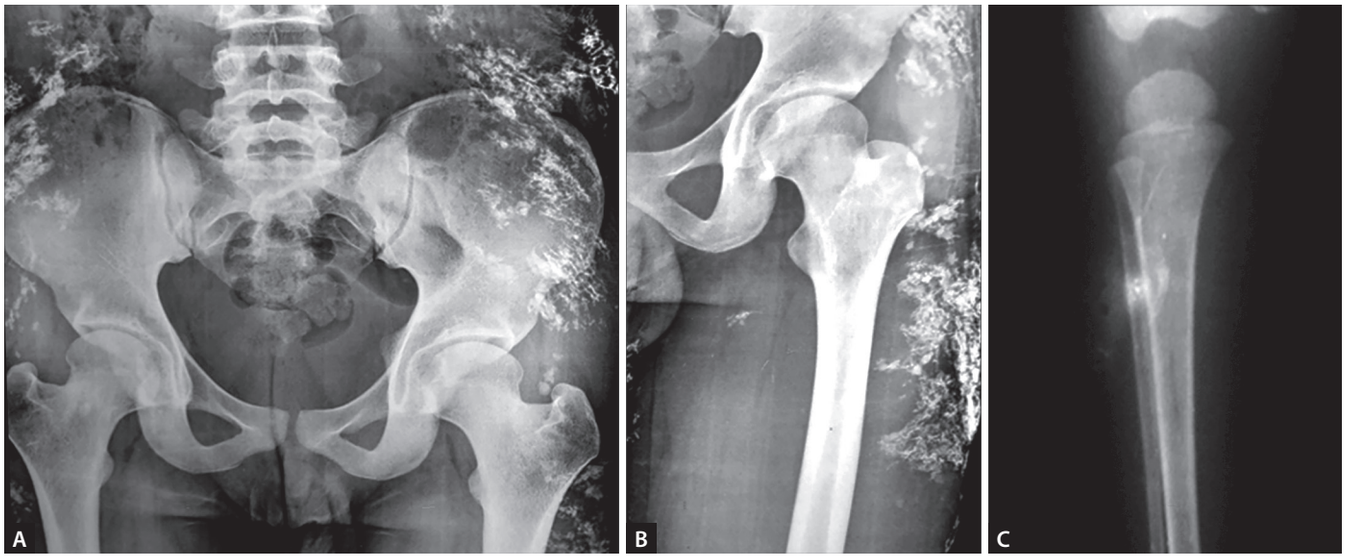
Systemic Involvement

Respiratory muscle weakness may result in symptomatic restrictive pulmonary disease.

Visceral involvement in JDM is due to vasculopathy, which is not a true vasculitis. It is limited to small arterioles and capillaries demonstrating fibrinoid necrosis on biopsy. The vasculopathy can involve the abdominal viscera causing ischemic pain, acute abdomen, hematemesis, melena, and perforation. Cardiac involvement may manifest as sinus tachycardia, innocent murmurs or cardiomegaly. The most severe (though rare) manifestation of vasculopathy is involvement of the central nervous system which can present with varying degrees of encephalopathy and seizures.

Lipodystrophy and Metabolic Abnormalities

Juvenile dermatomyositis is the most common systemic autoimmune disease associated with lipodystrophy which is characterized by slow but progressive loss of subcutaneous fat, best noticed over the upper body and face. This is accompanied frequently by hypertriglyceridemia, insulin resistance, abnormal glucose tolerance, acanthosis nigricans, hypertension, and nonalcoholic steatohepatitis. Lipodystrophy can be focal or generalized and presents usually years after disease onset. It is common in inadequately treated individuals.



Figs. 6A to C: X-ray showing dystrophic calcifications.

DIAGNOSIS

Traditionally, the diagnosis of JDM is based on Bohan and Peter's criteria **(Box 1)**. The diagnosis is essentially clinical. These criteria are likely to undergo revision through the International Myositis Criteria Classification Project.

Investigations

Acute Phase Reactants

Thrombocytosis, elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) correlate with the degree of inflammation and help to differentiate inflammatory myopathies from noninflammatory disorders like muscular dystrophy.

Muscle Enzymes

Serum levels of muscle enzymes are important not only for diagnosis but also for monitoring patients undergoing treatment. Aspartate transaminase (AST), creatinine kinase (CK), lactate dehydrogenase (LDH), and aldolase should be measured at diagnosis. AST or CK may be elevated 20–40 times of normal. However, CK levels may be normal in about 20% children, particularly with a longer duration of untreated disease. LDH appears to correlate best with disease activity. Serum levels of muscle enzymes usually decrease 3–4 weeks before improvement in muscle strength and rise 5–6 weeks before clinical relapse. As a general rule, CK levels return to normal first (usually several weeks after instituting therapy); and aldolase and LDH levels return to normal the last.

Autoantibodies

Antinuclear antibodies (ANA) may be positive in 10–85% children with JDM. Myositis specific antibodies like anti Jo-1 are uncommon in pediatric population and occur only in about 10% of children with JDM. Anti-PM/Scl antibody is associated with overlap syndrome. Anti-synthetase antibodies are associated with increased mortality. Chronic disease course is associated with anti-p 155/140 (TIF1) antibody. Anti-NXP2 antibodies are associated with calcinosis.

Electromyography

Electromyography (EMG) findings that suggest inflammatory myopathy include a combination of changes of myopathy and denervation.

BOX 1: Bohan and Peter criteria for diagnosis of juvenile dermatomyositis (JDM).

- Symmetrical weakness of the proximal musculature
- Characteristic cutaneous changes consisting of heliotrope discoloration of the eyelids, which may be accompanied by periorbital edema and erythematous papules over the extensor surfaces of joints, including the dorsal aspects of metacarpophalangeal and proximal interphalangeal joints, elbows, knees, or ankles (i.e., Gottron papules)
- Elevation of the serum level of one or more of the following skeletal muscle enzymes: creatine kinase (CK), aspartate aminotransferase (AST), lactate dehydrogenase (LDH), and aldolase
- Electromyographic demonstration of the characteristics of myopathy and denervation
- Muscle biopsy documenting histological evidence of necrosis, fiber size variation, degeneration and regeneration, and a mononuclear inflammatory infiltrate, most often in a perivascular distribution

Muscle Biopsy

Muscle biopsy is performed when the diagnosis is in doubt (especially, if there are no skin findings) and sometimes to evaluate the disease activity. Biopsy is performed usually from quadriceps or deltoid muscle, although the best specimen may be chosen based on EMG or magnetic resonance imaging (MRI). Muscle biopsy reveals perifascicular atrophy and variation in fiber size because of ongoing degeneration and regeneration. Areas of focal necrosis may be noted and inflammatory infiltrates are often present.

Magnetic Resonance Imaging

Magnetic resonance imaging has replaced the need for EMG and muscle biopsy for diagnosis. MRI of thigh muscles demonstrates symmetrical muscle edema and inflammatory changes on T2-weighted image or short-tau inversion recovery (STIR) sequences. MRI also helps in selecting a site for muscle biopsy, if needed.

DIFFERENTIAL DIAGNOSIS

- *Idiopathic inflammatory myopathies [juvenile polymyositis (JPM), overlap syndromes, cancer associated myositis, eosinophilic myositis]:* JPM is rare in children and has no dermatological findings. However, absence of dermatological findings in a patient of myositis does not rule out JDM; since they may be absent at onset and can appear months later.

TABLE 1: Differences between juvenile dermatomyositis, juvenile pyomyositis, and juvenile CTD-associated myositis.

Characteristic	*JDM*	*JPM*	*Juvenile CTD-associated myositis*
Frequency, %	81.2	7.6	11.2
Median age at diagnosis (years)	7.4	12.1	10.2
Frequency of severe disease, %	30	58	25
Clinical features			
Musculoskeletal:			
Proximal muscle weakness	++	++	++
Arthralgia/arthritis	+	++	++
Cutaneous:			
Gottron papules	++	–	+
Heliotrope rash	++	–	+
Periungual capillary abnormalities	++	+	+
Malar rash	++	–	+
Photosensitivity	+	–	+
Skin ulcer	++	–	++
Raynaud phenomenon	+	++	+++
Gastrointestinal (GI)*:*			
GI bleed	+	+	++
Pulmonary:			
Dyspnea on exertion	++	+	++
Interstitial lung disease	+	++	+++
Cardiac:			
Abnormal ECG/ECHO	+	++	+
Constitutional:			
Fever	++	++	++
Fatigue	+++	+++	+++
Weight loss	+	++	++
Autoantibodies:			
ANA titers	Intermediate	Intermediate	Highest
Other antibodies	Anti-p155/140	Anti-SRP Anti-Jo-1	Anti-U1-RNP Anti-PM-Scl Anti-Ro/La
Muscle enzymes:	Lowest	Highest	Intermediate
Outcome			
Mortality, %	2.4	6.3	14.6
Complications	Calcinosis (34%)		

(ANA: antinuclear antibody; CTD: connective tissue disease; ECHO: echocardiogram; ECG: electrocardiogram; JDM: juvenile dermatomyositis; JPM: juvenile polymyositis; +++: very frequent; ++: frequent; +: less common; –: does not occur)
Source: Modified from Shah M, Mamyrova G, Targolf IN, et al. The clinical phenotypes of the juvenile idiopathic inflammatory myopathies. Medicine (Baltimore). 2013;92:25-41.

- *Infectious myopathies:* Viral (enterovirus, influenza, Coxsackie, echovirus, parvovirus, hepatitis B, human T lymphotropic virus I), bacterial (*Staphylococcus*, *Streptococcus*, Lyme borreliosis) and parasitic *(Toxoplasma, Trichinella)*
- *Noninflammatory myopathies:* Muscular dystrophies, congenital myopathies, myotonic disorders, metabolic myopathies (glycogen storage diseases, lipid myopathies, periodic paralyses, mitochondrial myopathies), endocrinopathies
- *Systemic rheumatic diseases:* SLE, scleroderma, juvenile idiopathic arthritis, mixed connective tissue disease (MCTD), vasculitis
- *Others:* Trauma, toxins, drug-induced myopathies, disorders of neuromuscular transmission

The differences between JDM, JPM, and juvenile CTD-associated myositis are enumerated in **Table 1.**

TREATMENT Juvenile Dermatomyositis

Pharmacotherapy, along with early individualized physiotherapy, remains the mainstay of therapy. All patients should be advised sun protection and use of sunscreen lotions.

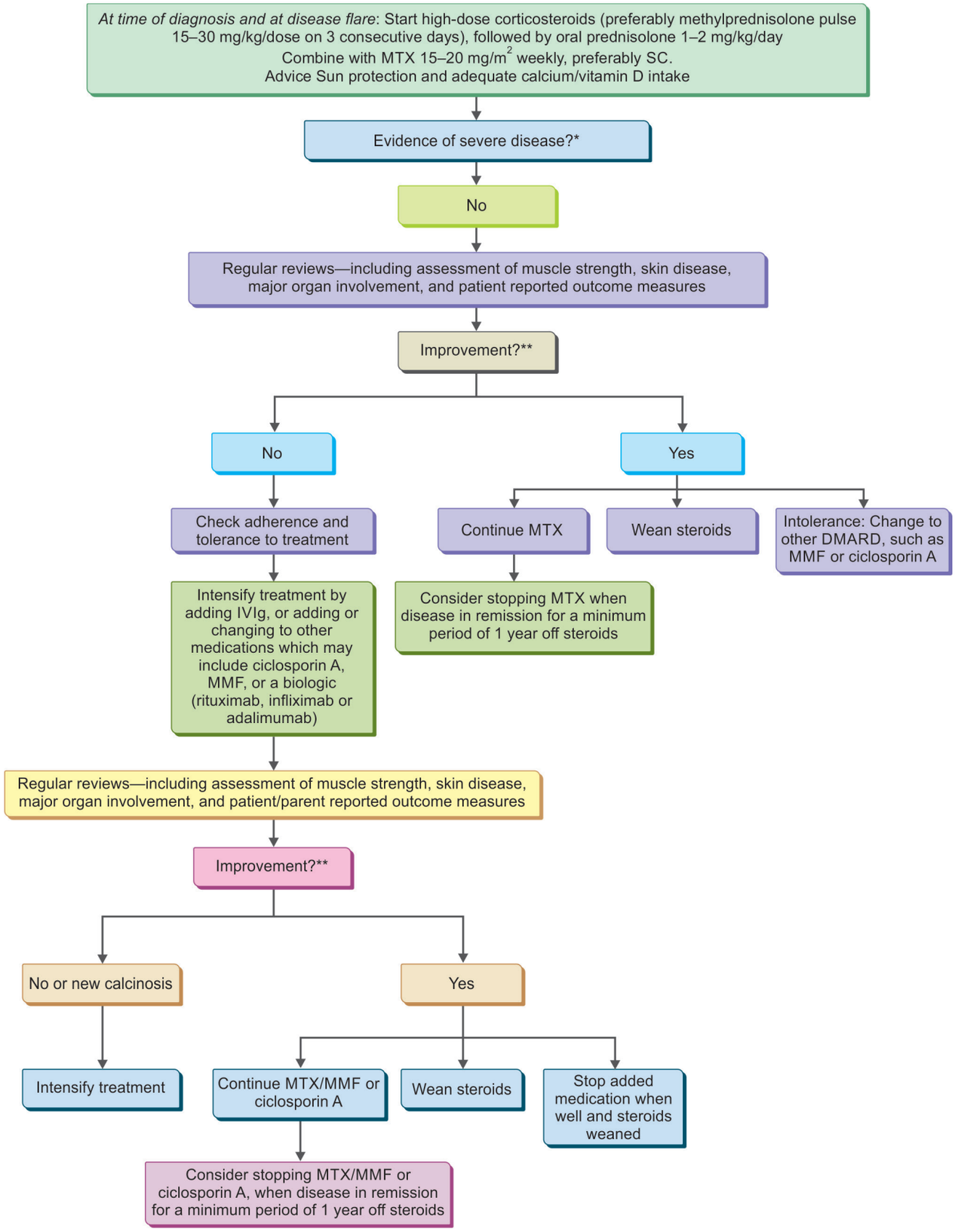
Remission Induction

This is usually achieved by intravenous pulse methylprednisolone (30 mg/kg/day for 3–5 days) or oral prednisolone (2 mg/kg/day for 4 weeks) followed by gradual tapering. Methotrexate (15–20 mg/m^2/week) is started concomitantly, orally or subcutaneously. Combined initiation of corticosteroids and methotrexate has synergistic action and gives the liberty of tapering steroids early without risking a disease flare.

Maintenance

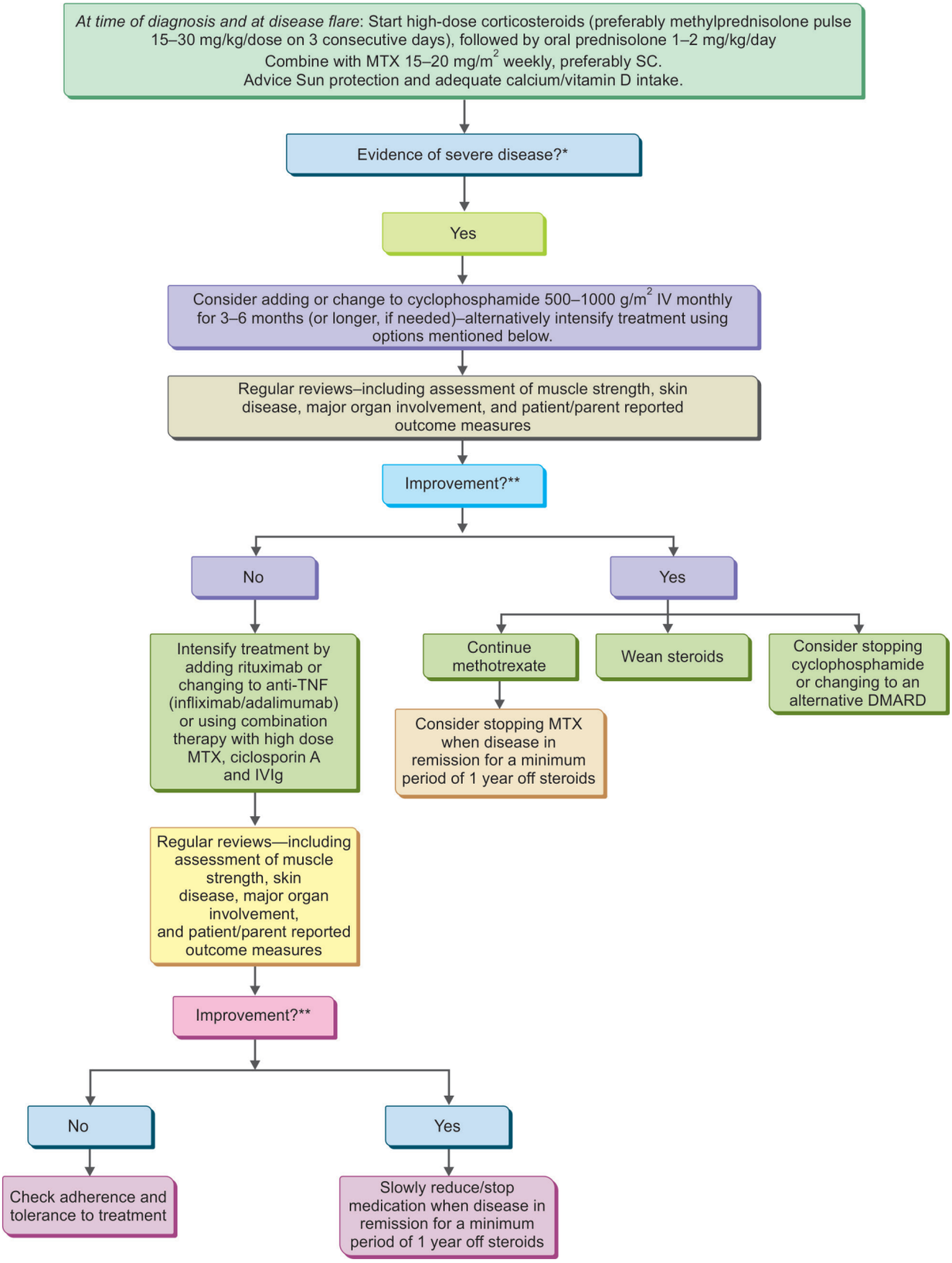
- Therapy during this phase consists of oral prednisolone which is initially given at a dose of 1 mg/kg/day and then gradually tapered off over a period of 2 years depending on the clinical response. In case of disease exacerbation on tapering, low-dose maintenance steroids may be continued for years.
- Methotrexate is continued during this phase and helps in reducing the dose of steroids.
- Oral hydroxychloroquine at a dose of 3–6 mg/kg/day may be added along with oral prednisolone and methotrexate. This is particularly useful for skin disease in JDM.
- Photoprotective measures like full sleeved cotton clothing and sunscreen lotion and calcium and vitamin D supplementation for bone protection are given to most patients.

Flowchart 1: Treatment of mild/moderate juvenile dermatomyositis.



*Such as major organ involvement/extensive ulcerative skin disease.
**Improvement based on clinician opinion.
(MTX: methotrexate; MMF: mycophenolate mofetil; IVIg: intravenous immunoglobulins; DMARD: disease-modifying antirheumatic drug; SC: subcutaneously)
Source: Adapted from Bellutti Enders F, Bader-Meunier B, Baildam E, et al. Consensus-based recommendations for the management of juvenile dermatomyositis. Ann Rheum Dis. 2017;76:329-40.

Flowchart 2: Treatment of severe juvenile dermatomyositis (JDM).



*Such as major organ involvement/extensive ulcerative skin disease.
**Improvement based on clinician opinion.
(MTX: methotrexate; MMF: mycophenolate mofetil; IVIg: intravenous immunoglobulins; DMARD: disease-modifying antirheumatic drug; SC: subcutaneously)
Source: Adapted from Bellutti Enders F, Bader-Meunier B, Baildam E, et al. Consensus-based recommendations for the management of juvenile dermatomyositis. Ann Rheum Dis. 2017;76:329-340.

Second-line Therapies
Whereas majority of patients will show good response to steroids with methotrexate, some patients do need second-line treatment. These consist of intravenous immunoglobulin at 2 g/kg/month, cyclosporine at 2.5–7.5 mg/kg/day, azathioprine at 1–3 mg/kg/day or combinations of these. Third-line drugs are less frequently needed and consist of monthly pulses of intravenous cyclophosphamide, mycophenolate mofetil, tacrolimus, or biologicals. Second- and third-line therapies are reserved for refractory patients or those with unacceptable toxicities to first-line drugs. The anti-CD20 biologic rituximab is beneficial in refractory cases. **Flowcharts 1 and 2** depict consensus-based recommendations for management of mild/moderate and severe JDM, respectively.

Management of Complications
Patients with palatal involvement may require prolonged nasogastric tube feeding.

Calcinosis
There is no accepted effective therapy but early aggressive therapy with corticosteroids and other medicines results in decreased frequency and severity of calcinosis. Medications like colchicine, aluminum hydroxide, bisphosphonates, and tumor necrosis factor blockade have shown variable benefits.

Lipodystrophy
Recent data have suggested leptin deficiency as an important causative factor and trials of recombinant human leptin have shown promising results.

IN A NUTSHELL

1. Juvenile dermatomyositis is the most common idiopathic inflammatory myopathy of childhood.
2. Clinical features, if present, are characteristic and help in diagnosis. Skin changes include Gottron papules and heliotrope rash. Muscle changes include features of proximal muscle weakness with inflammation.
3. MRI of muscles showing inflammation and edema helps in diagnosis. Muscle biopsy is usually reserved for children where clinical features are not characteristic.
4. Lipodystrophy and calcinosis are two most important long-term complications and may progress despite therapy.
5. Immunosuppression with steroids and an additional steroid sparing agent is the mainstay of therapy. Early diagnosis and effective therapeutic regimen help in reducing the risk of calcinosis.

Monitoring
Success of therapy is evaluated using the following parameters:
- Abatement of systemic signs and symptoms: Systemic symptoms and fever usually abate within a few days.
- Regression of acute inflammatory markers
- Improvement in muscle strength (preferably tested by single observer and by a standardized tool): Improvement in muscle strength and dermatitis occurs over months.
- Improvement in muscle enzymes: Muscle enzymes decrease appreciably by 1–2 weeks of initiation of therapy.
- Sometimes by imaging modalities like MRI or muscle ultrasound.

PROGNOSIS

Majority of patients have a uniphasic disease course with good functional outcome. About 30% patients survive with minimal atrophy or contractures. A few develop lipoatrophy and insulin resistance and some evolve into MCTD. Mortality is about 1–2% and usually occurs within the first 2 years of onset due to respiratory insufficiency, acute gastrointestinal hemorrhage, myocarditis, or interstitial lung disease.

MORE ON THIS TOPIC

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46.8 Large Vessel Vasculitis: Takayasu Arteritis

Anju Gupta

Takayasu arteritis (TA) is a chronic idiopathic granulomatous large vessel vasculitis associated with a chronic continuous or waxing-waning course. It involves elastic arteries, i.e., aorta and its major branches and pulmonary arteries. Inflammation involves all the three layers of vessel wall producing a typical pathology characterized by stenosis, occlusions, dilation, aneurysms, and rarely rupture. Most clinical features occur due to systemic inflammation and distal ischemia.

Initial description of this systemic vasculitis was given by Japanese ophthalmologists, who identified "arteriovenous anastomosis in retina" (now known to be due to ischemic retinopathy) and its association with "absence of radial pulsations". Autopsy studies described occurrence of panarteritis involving aorta and its major branches and pulmonary arteries. Even after passage of more than 120 years after initial description, not much is known about the etiopathogenesis of this disease.