

Review Article

# Juvenile Dermatomyositis: A Review

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## Abstract

Juvenile dermatomyositis is a systemic vasculopathy, affecting mainly the skin and muscle. In the United States, it is seen in more than three per million children per year. It is diagnosed on the basis of the criteria set by Bohan and Peter. The following review describes the characteristic clinical manifestations, the pathophysiology and immunology of the disease. The various treatment modalities are discussed. *Int Pediatr.* 2000;15(1):21-25.

*Key words:* juvenile dermatomyositis, vasculopathy

## Introduction

Juvenile dermatomyositis is a systemic vasculopathy, affecting primarily the skin and muscle, causing symmetric proximal weakness and characteristic skin rash. It differs from the adult form of dermatomyositis by the presence of vasculitis of the small blood vessels, which can involve the gastrointestinal tract and myocardium, besides skin and muscle. Calcinosis is an additional feature that is present in juvenile dermatomyositis, but not in the adult form of dermatomyositis. Juvenile dermatomyositis is not associated with development of malignancies, unlike adult dermatomyositis.<sup>1</sup>

## Epidemiology

Juvenile dermatomyositis is the most common of the inflammatory myopathies of childhood, affecting about three per million children per year.<sup>1</sup>

Dermatomyositis has a bimodal distribution in the age of onset, occurring in two peaks, one at 5 to 14 years and the other at 45 to 64 years of life.

Juvenile dermatomyositis is 10 – 20 times more common than polymyositis in children, and tends to have a more

acute and severe onset. It appears to have a seasonal predilection, occurring more frequently in the spring and summer months. A history of antecedent illness is often obtained in newly diagnosed juvenile dermatomyositis. Studies implicate Coxsackie virus on the basis of viral antibody findings or viral isolation from patients.

Juvenile dermatomyositis has a strong association with the HLA antigens B8/DR3 and DQA1\*0501 allele.<sup>1</sup>

## Diagnostic Criteria

Bohan and Peter set forth criteria for the diagnosis of juvenile dermatomyositis and polymyositis in childhood. These criteria assume that the child has the characteristic rash, after which three of the four criteria must be fulfilled for definite disease, two of four for probable disease and one of four for possible disease.<sup>2</sup> (Table 1)

**Table 1.—Diagnostic Criteria For Juvenile Dermatomyositis<sup>2</sup>**

	Juvenile dermatomyositis	Polymyositis
Characteristic rash	+	-
Symmetric proximal muscle weakness	+	+
Elevated muscle derived enzymes	+	+
Muscle histopathology	+	+
Electromyographic changes: Inflammatory myopathy	+	+

## Clinical Features

The onset of juvenile dermatomyositis is acute in 50% of patients, with rapid development of weakness and rash. Children who have a subacute onset may present a skin rash, a gradually progressive weakness of muscles, joint contractures or difficulty using hands because of tendon involvement. The usual time period between onset of disease symptoms and diagnosis is approximately three months. In those children who had only weakness as the presenting complaint, diagnosis could be delayed to 12 months or greater.<sup>3</sup>

Besides the characteristic manifestations described below, children often have low-grade fever, malaise, weight loss, and poor appetite.

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### Cutaneous Manifestations

The rash may precede or follow the onset of proximal muscle weakness. The characteristic rash is violaceous or heliotropic, occurring most prominently on the eyelids (Figure 1). Periorbital edema can be seen. Eyelid telangiectasia accompanies the periorbital edema in 50 – 90% of children. Exposure to sunlight can cause exacerbation of the skin inflammation or may precipitate activation of myositis. Erythema can occur over the upper parts of the body (shawl sign) and extensor surfaces of arms and legs.

The skin over the knuckles may be hypertrophic or pale red and evolve into atrophic bands. Gottron's papules are flat-topped red papules on the knuckles (Figure 2). Erythematous plaques with fine scales are seen on the elbows, knees, and medial malleoli of the ankles. A livedo reticularis pattern may be seen on the extremities. Diffuse vasculitis may be manifested by nailbed telangiectasia, infarction of oral epithelium, skin folds or digital ulceration.<sup>1</sup>

The scalp may be involved with diffuse, scaly dermatosis and often nonscarring alopecia<sup>4</sup> (Figure 3). This is often misdiagnosed as seborrheic dermatitis or psoriasis.

Panniculitis is a rare finding, where indurated plaques and nodules are found mainly on the arms, thighs and buttocks. These can be erythematous and painful.<sup>5</sup>

Hypertrichosis is another unusual feature of juvenile dermatomyositis, the pathogenesis of which is not known. Hair growth may be more prominent on the forehead, cheeks, forearms and legs. Hypertrichosis may respond to the steroid therapy given for the dermatomyositis.<sup>6</sup>



Fig 1.—Heliotrope, erythema on the face of a 10 year old with a one year history of dermatomyositis

### Calcinosis

Soft tissue calcifications occur in up to 40% of the patients in the late stages. Skin calcinosis is seen as crusted papules or plaques around joints or as nonhealing sores. Sometimes, the calcific material is extruded through the skin as a white cheesy exudate, leaving behind a dry pitted scar. Muscle calcification results in contractures or severe muscular pain. Four patterns of calcification are seen: superficial and deep calcarial masses, deep linear deposits (Figure 4) and lacy reticular subcutaneous deposition encasing the torso.<sup>7</sup>

### Musculoskeletal Features

Proximal muscle weakness is the major feature. This is manifested by difficulty in raising the arms above the shoulders, inability to comb the hair, and difficulty in climbing stairs. Neck flexor weakness is an especially sensitive indicator of muscle weakness. Muscle pain is not a frequent symptom. The child usually keeps the limbs in a flexed position, which promotes development of flexion contractures and soft tissue calcification.

There may be difficulty in swallowing due to palatal muscle weakness, with regurgitation and a nasal voice. Pooling of barium in a wide atonic pyriform fossa may be seen. Esophageal dysfunction may be present.

The myopathy may be focal, especially early in disease onset. Therefore a normal area may be erroneously targeted in EMG or biopsy. MRI of the muscles can help detect the involved areas.

Decreased bone density may be present, leading to increased rate of bone fracture.



Fig 2.—Gottron's papules (same patient as in Figure 1)



Fig 3.—Occipital alopecia in a child with dermatomyositis



Fig 4.—Calcinosis cutis in dermatomyositis (courtesy of Guinter Kahn, MD, Miami, Florida)

#### Gastrointestinal Symptoms

Decreased esophageal motility leads to difficulty in handling secretions. Involvement of the masseter can lead to difficulty in chewing food. Vasculitis leading to mucosal ulcerations may result in frank bowel perforation, which can be life threatening.

#### Cardiac Abnormalities

Abnormalities in EKG are seen in over half the children with juvenile dermatomyositis. Myocarditis most com-

monly leads to asymptomatic conduction abnormalities. These resolve when disease activity subsides.

#### Pulmonary Findings

There is a decrease in ventilatory capacity in the absence of respiratory complaints. Pulmonary fibrosis can occur, but is more common with children who have antibodies to Jo-1.

#### Ophthalmic Findings

Thrombosis of vessels at the eyelid margin may be seen. "Cotton-wool" spots on the retina result from small vessel occlusion. Intraretinal edema can cause injury to the retinal nerve fibers and lead to optic atrophy and visual loss.

#### Pathophysiology

Vascular lesions without a prominent inflammatory component can be seen in juvenile dermatomyositis. Capillaries, venules and small arteries are damaged with deposition of IgM, C3D and fibrin, with loss of muscle capillary network and structural changes in the nailfold capillary bed. The primary lesion occurs in the endothelial cell, which contains reticulotubular inclusions that are the site of thrombosis and vessel obliteration.<sup>1</sup>

The muscle pathology reflects vascular compromise and capillary dropout, with perivascular atrophy of both type I and type II muscle fibers, and inflammatory infiltrates of mononuclear cells and plasma cells. CD4+ cells predominate in the infiltrate, which is primarily around the blood vessels.

Soft tissue calcification is accompanied by urinary excretion of gamma-carboxyglutamic acid, which is a component of the vitamin K dependent coagulation pathway. However, no clear mechanism has been identified for the occurrence of calcinosis.

#### Immunology

Antinuclear antibodies, mainly of the speckled variety, are seen in over 60% of the patients. Myositis-specific autoantibodies are seen in about 10% of children with juvenile dermatomyositis, the most common being anti-Mi2 antibody (Table 2). Von Willebrand factor (vWF) released from the damaged endothelial cells was noted to be increased in active juvenile dermatomyositis in various studies.

Serum levels of neopterin, a pteridine derived from activated macrophages, is elevated in about 60% of patients and correlates with clinically active disease.

Studies have also shown that absolute lymphocyte counts were low in active juvenile dermatomyositis, but the percentage of B lymphocyte counts were significantly increased, with an increase in CD4/CD8 ratio, and this reverted only after starting cytotoxic therapy.<sup>8</sup>

**Table 2.—Myositis Associated Antibodies<sup>3</sup>**

<b>Anti-synthetase autoantibodies</b>	
Anti-Jo-1	[HisRS]
Anti-PL-7	[ThrRS]
Anti-PL-12 (1)	[AlaRS]
Anti-PL-12 (2)	[tRNA-Ala]
Anti-OJ	[IleRS]
Anti-EJ	[GlyRS]
<b>Anti-Mi-2 autoantibodies</b>	
Anti-Mi-2	[five proteins]
<b>Anti-signal recognition particle autoantibodies</b>	
Anti-SRP	[SRP54]
<b>Other myositis-specific autoantibodies</b>	
Anti-FER	[Elongation Factor 1a]
Anti-KJ	[unidentified protein]
Anti-MAS	[unidentified RNA]
Myositis without any of the above autoantibodies	

**MRI Studies**

MRI is useful in identifying areas of involvement, which is detected by positive T2 images. Studies have shown that MRI detects areas of involved muscle in those children with normal muscle enzymes. MR spectroscopy using P31 can be used to monitor response to therapy, when other indicators have normalized.

One must however remember that MRI images are not specific to juvenile dermatomyositis. Similar changes can be seen in inflammatory myopathies, strenuous exercise, a prolonged muscle cramp or local trauma.<sup>1</sup>

**Electromyography**

EMG shows changes suggestive of myopathy. However, it can be negative in up to 10% of new onset juvenile dermatomyositis despite elevated muscle enzymes, due to improper electrode placement into normal areas of muscle.

**Prognosis**

Prognosis is related to the degree of vasculitis. Death can occur in the acute phase due to myocarditis, progressive unresponsive myositis or occasionally due to lung involvement or bowel perforation secondary to ulceration. Infection during the course of intensive therapy may also result in death.

The clinical course can be monocyclic, chronic polycyclic and chronic continuous. The overall prognosis for survival is improved following better use of corticosteroids.

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**Table 3.—Laboratory Features<sup>2</sup>**

Antinuclear antibodies	Positive speckled
DNA	Normal or negative
Smith antigen	Normal or negative
Ribonucleoprotein	Normal or negative
SSA-Ro / SSB-La	Normal or negative
VonWillebrand factor related antigen	Elevated
Creatine kinase	Elevated
Aspartate aminotransferase	Elevated
Lactic acid dehydrogenase	Elevated
Aldolase	Elevated
Erythrocyte sedimentation rate	Normal or elevated
White blood cell count	Normal or elevated
Hemoglobin	Normal or elevated
D-xylose	Impaired

Residual disability is related mainly to calcification and flexion contractures.

**Treatment**

Corticosteroids are the mainstay of therapy for children with juvenile dermatomyositis.<sup>7</sup>

With severe vasculitis, IV methylprednisolone at the dose of 30 mg/kg (max. 1 gm) can be given every other day until the muscle enzymes and vWF have returned to normal. Then oral therapy with prednisone is started at a dose of 2 mg/kg/day. In most cases, improvement is seen over the first 4 weeks of steroid treatment. After this, the dose of oral steroids is gradually reduced and changed to an alternate day regimen. Monitoring the patient for the adverse effects of steroids is very important.<sup>9</sup>

If the skin rash is very prominent, hydroxychloroquine (up to 7 mg/kg/day) is started, along with topical steroids and lubricants. Use of sunscreen (SPF 16 or more) will reduce exposure to UV light, so as to decrease disease activation.

Presence of myocarditis, persistent dysphagia, diplopia and dyspnea, especially with weakness of intercostal muscles suggests bad prognostic features. In these children, early therapy with cyclosporine or cytotoxic drugs is desirable. Cyclosporine A can be given in at a dose of 6 – 8 mg/kg daily.<sup>10</sup>

Cytotoxic therapy is considered in those children who cannot be maintained on reasonable doses of steroids, with or without cyclosporine, or who have late vasculitis. Azathioprine, 2.3 mg/kg/day, given orally or methotrexate given orally or parenterally may be tried. Short courses of cyclophosphamide with mesna (2-mercaptoethanesulfonic acid)

may be used in presence of severe vasculitis.<sup>7</sup>

IV gammaglobulin has been given with some success in anecdotal cases. IVIG was used in the form of monthly infusions of 2 gm/kg/month in those patients who did not respond to the traditional line of treatment.<sup>11</sup>

Once inflammation subsides, it is important to start muscle-training exercises. Foot drop must be prevented by using appropriate splints.

Acute pain due to calcinosis may be treated with indomethacin or colchicine. As the disease becomes inactive and the patient is mobilized, it tends to improve. In some patients, however, it may be intractable. Surgical removal may be done in solitary subcutaneous lesions. Some reports have suggested that aluminium hydroxide may be effective in calcinosis.<sup>12</sup>

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