

Juvenile Dermatomyositis

A. **Definition.** This inflammatory condition of children results in myopathy, progressive muscle weakness, and characteristic skin findings.

B. **Epidemiology**

1. JDM is the **most common idiopathic inflammatory myopathy of childhood** (85% of all myopathies).
2. Individuals are usually **5–10 years** of age. The mean age of onset is **6 years**.
3. **Females** are more likely to be affected.

- the most common of the pediatric inflammatory myopathies
- have characteristic rash and proximal symmetric muscle weakness that is often responsive to the immunosuppressive therapy.

ETIOLOGY

- The etiology of JDM is unknown, but it is characterized by activation of T and B lymphocytes, leading to vasculitis affecting small vessels of skeletal muscle, with immune complex deposition and subsequent inflammation of blood vessels and muscle.
- JDM has been documented to follow:
 1. infections (infection 3 mo before disease onset is obtained in most affected children)
 2. allergic reactions
 3. sun exposure

CLINICAL MANIFESTATIONS

WHERE IN THE BODY:

- Affects the proximal muscles, particularly the hip and shoulder girdles, and the abdominal and neck muscles.

COURSE OF ILLNESS:

- Dermatomyositis tends to present in a slow, progressive fashion with insidious onset
- Some children can present in an acute fashion.

SYMPTOMS:

- ✓ low-grade fevers
- ✓ fatigue and malaise
- ✓ rash
- ✓ progressive muscle weakness
- ✓ Difficulty climbing steps, getting out of chairs, and getting off the floor.
- ✓ difficulty swallowing
- ✓ joint pain

SIGNS:

- ✓ positive **GOWER SIGN**: needing to lean on legs while getting up from the ground.
- ✓ In severe cases, the patient is not able to sit up from a supine position or even lift the head off the examination table.
- ✓ **VOICE SOUND NASAL AND DIFFICULTY SWALLOWING**: If muscles of the upper airway and pharynx are involved
- ✓ skin rash is the first symptom in 50% of cases, and concomitantly with weakness in 25% of cases.
- ✓ The classic **JDM rash** (**SHAWL SIGN**) hyperpigmentation and telangiectasia of the skin, followed by atrophy occur on exposed skin, such as the extensor surfaces of the arm, the V of the neck, or the upper part of the back
- ✓ **HELIOTROPE DISCOLORATION** of the eyelids. may cross the bridge of the nose, in a mask-like distribution, and involve the ears as well
- ✓ **GOTTRON PAPULES** Scaly, red plaques, classically are found across the knuckles, but can be found on the extensor surfaces of any joint.
- ✓ **PERIUNGUAL ERYTHEMA AND DILATED NAIL-FOLD CAPILLARIES**.
- ✓ Less commonly, cutaneous vasculitis, with inflammation, erythema, and skin breakdown.
- ✓ At some point in their illness, 15% of patients with JDM develop arthritis. The arthritis commonly affects small joints, but can occur in any joint.
- ✓ Raynaud phenomenon
- ✓ hepatomegaly, and splenomegaly.



LABORATORY AND IMAGING STUDIES

- JDM has no evidence of systemic inflammation, with a normal blood count and ESR, indicating inflammation isolated to the muscle and skin.
- Evidence of myositis can be identified in 98% of children with active JDM by elevated serum muscle enzymes, including: (Serum levels are in the normal range for the 1st 4-5 mo after disease onset):
 - aspartate aminotransferase,
 - alanine aminotransferase,
 - creatine phosphokinase,
 - aldolase,
 - lactate dehydrogenase.
- There may be a Coombs-negative anemia
- Antinuclear antibody (ANA) with a speckled pattern (unknown specificity) is present in >80% of children.
- Pm/Scl Antibodies identify a subgroup of myopathies with a protracted disease course, often complicated by pulmonary interstitial fibrosis, tachycardia, conduction abnormalities, and elevated troponin levels.
- Electromyography(EMG) and muscle biopsy can be used to document the myositis.
- MRI is a noninvasive means of showing muscle inflammation.

DIFFERENTIAL DIAGNOSIS

- Diagnosis of JDM is based on the presence of documented muscle inflammation in the setting of classic rash.
- polymyositis is rare in children, they should have a muscle biopsy to exclude other causes of muscle weakness, such as muscular dystrophy, particularly boys.
 - postinfectious myositis
 - other myopathies.

Criteria for Diagnosis of Juvenile Dermatomyositis* (4of 5)

1. Rash typical of dermatomyositis
2. Symmetric proximal muscle weakness
3. Elevated muscle enzymes (SGOT, SGPT, LDH, CPK, and aldolase)
4. EMG abnormalities typical of dermatomyositis (fasciculations, needle insertion irritability, and high-frequency discharges)
5. Positive muscle biopsy specimen with chronic inflammation
6.

TREATMENT

- Systemic corticosteroids are the cornerstone of therapy for JDM.
- In severe or refractory cases, it may be necessary to use cyclosporine (Immunosuppressives)^{Rx} or cyclophosphamide (Disease modifying antirheumatic drugs- cytotoxic)^{Rx}.
- IVIg as adjunctive therapy.
- Hydroxychloroquine or dapsone^{Rx} (is bactericidal as well as bacteriostatic against *Mycobacterium leprae*/ primary treatment for Dermatitis herpetiformis) has been used for the skin manifestations; they **do not** significantly affect the muscle disease.
- Exposure to the sun worsens the cutaneous manifestations, but also exacerbates the muscle disease; sunlight may lead to flare.

COMPLICATIONS

- calcinosis :The most serious complication of JDM .
 - ✓ consequence of chronic inflammation, Dystrophic calcification occur in the skin and soft tissues in any area of the body
 - ✓ Range from mild to extensive (calcinosis universalis).

- ✓ Are associated with increased morbidity and mortality. They are present at diagnosis in about 25% of children with JDM.
- ✓ They may drain a white, cheesy material and resolve, or serve as a nidus for infection, *most frequently staphylococcal*, progress to septicemia and death.
- ✓ Aggressive immunosuppressive therapy at the time of diagnosis decrease frequency of calcinosis.
- ✓ it seems to occur more commonly in children who have had:
 - A. cutaneous vasculitis,
 - B. prolonged disease activity,
 - C. Delays in onset of therapy.

2. **Gastrointestinal perforation and gastrointestinal bleeding** :JDM patients who develop vasculitis are at risk for it.

3. **lipoatrophy and insulin resistance:**

- ✓ in >10% of chronic JDM cases. It
- ✓ is characterized by:
 1. loss of subcutaneous fat on the extremities
 2. hypermuscular appearance
 3. acanthosis nigricans
 4. weakened abdominal muscles resulting in a potbelly appearance
 5. Abnormal glucose and lipid metabolism. which can progress to frank type 2 diabetes, control of insulin resistance leads to improvement in muscle disease.
 6. Girls may lose their menses; sterility may result if the onset of JDM is before puberty.. In these patients,
- 4. Aspiration pneumonia is a frequent major complication associated with unrecognized impairment in swallowing fluids.

PROGNOSIS

- The outcomes in patients with JDM depend greatly on:
 - a. the extent of muscle disease
 - b. the time between disease onset and initiation of therapy.
- JDM follows one of three clinical courses:
 1. uniphasic course, in which patients are treated and improve without significant sequelae.
 2. chronic recurrent course: poor response to therapy+ loss of function.
 3. chronic progressive course, poor response to therapy + loss of function.
- Patients who ultimately develop calcinosis also are at risk for chronic loss of mobility depending on the extent of calcium deposition.

NOTE:

The association of polymyositis with malignancy is seen in adults and not in children.