Juvenile Dermatomyositis

Juvenile Dermatomyositis (JDM) is a disease involving **myositis,** inflammation of the muscles, that classically presents as proximal weakness, as well as **characteristic rashes.** JDM affects 2-4 children per million in one year and is more common in females. The cause of JDM is unknown, but it is believed to be an autoimmune condition causing the muscle and skin inflammation.

PRESENTATION

- Proximal, usually symmetrical, muscle weakness
- Dysphonia
- Dysphagia
- Dyspnea
- Rashes: extensor surfaces of fingers, elbows, and knees; around eyes
- Skin ulcerations
- Arthritis
- Fatigue
- Rarely: cardiac and GI involvement

PHYSICAL EXAM

- Vitals, height and weight
- Assess muscle strength, looking for findings such as Trendelenburg sign, neck flexor weakness, and Gower's sign
- Joint exam
- Cardio exam: pericardial friction rub
- Pulm exam: chest expansion
- Abdo exam: distension, rigidity
- Skin exam
  - Gottron's papules
  - Heliotrope rash
  - Malar rash on face
  - Capillary changes at nail folds

DIAGNOSIS

- Diagnosis is usually made using the **Bohan and Peter criteria**:
  1. Symmetrical proximal muscle weakness
  2. Muscle biopsy evidence of myositis
  3. Elevation in serum skeletal muscle enzymes
  4. Characteristic electromyography pattern of myositis
  5. Typical rash of dermatomyositis
- In current clinical practice, however:
  - MRI is now routinely used in diagnosis
  - EMG is rarely used
  - Muscle biopsy is rarely used, unless the diagnosis is uncertain, or the patient is not responding to therapy

Definitive JDM	Typical rash plus any **three** of 1-4
Probable JDM	Typical rash plus any **two** of 1-4
Possible JDM	Typical rash plus any three of 1-4

INVESTIGATIONS

Serum muscle enzymes, CRP, CBC, Creatine, liver enzymes
Consider autoantibodies: ANA, Myositis antibodies
Evaluating myositis: MRI +/- EMG, muscle biopsy
Consider PFTs, ECG, and swallowing study

MANAGEMENT

- Prompt consultation with a **pediatric rheumatologist** when JDM is suspected
- **Admission** if: severe weakness, aspiration pneumonia, severe major organ system involvement, or secondary infection
- **Corticosteroids** and **methotrexate**
  - Consider adding **IVIG**
  - If no response/contraindications for use, consider other **DMARDs, biologics,** and **cytotoxic agents**
- Physiotherapy, OT, dietician, social work support

PROGNOSIS

- About 1/3 will have a **monocyclic** course – meaning they will recover from their disease.
- About 1/3 have a **polycyclic** course – disease free periods followed by relapses.
- About 1/3 will have a **chronic continuous** course.
- Mortality is low with greater than **95% long-term survival** since the introduction of corticosteroids.
- The main long-term complications include calcinosis, lipodystrophy, contracture, and metabolic problems such as diabetes, insulin resistance, and hyperlipidemia.

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