Idiopathic Inflammatory Myopathies in Childhood: Current Concepts

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KEYWORDS

• Juvenile idiopathic myopathies • Juvenile dermatomyositis
• Juvenile polymyositis • Connective tissue myositis

The juvenile idiopathic inflammatory myopathies (JIIM) are a group of rare, chronic autoimmune diseases that have muscle involvement as a primary feature. Muscle inflammation results in muscle weakness (proximal greater than distal), reductions in endurance, and impairment of physical function. Juvenile dermatomyositis (JDM), the most common member of this group, also manifests a variety of cutaneous features. Other forms of JIIM include juvenile polymyositis (JPM) and myositis associated with another connective tissue disease (JCTM) (also called overlap myositis). Less commonly, JIIM can be associated with other organ involvement, including lung, heart, and brain.

The underlying pathogenesis of JIIM is unknown. The current state of this knowledge has been recently reviewed.[1]

Although JIIM are rare, affecting 2 to 4 per 1 million children,[2,3] they occupy an important proportion of time in pediatric rheumatology clinics because of their complexity, chronicity, and risk of mortality and morbidity. This rarity also makes it unlikely that most pediatricians or family physicians will have seen or cared for a child with JIIM, making recognition and early management challenging.

This review focuses on practical issues of relevance to the practicing pediatrician, including clinical presentation, differential diagnosis, investigation, therapy, and prognosis. The goal is to facilitate early recognition and referral for appropriate specialist care and to assist the reader in comanagement of these patients with a pediatric rheumatologist.

CLINICAL PRESENTATION AND EVALUATION

Presenting features of children with JIIM have been reported by numerous investigators. Two of the largest descriptions have included a total of 641 patients, mostly with JDM.[4,5] Their findings are summarized in **Table 1**. As expected, most patients presented with weakness and rashes. However, it should be noted that a significant

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Table 1
Typical presenting features of children with JIIM

	McCann et al,[5] 2006	Guseinova et al,[4] 2011
Population	151 children with JIIM 120 JDM 4 JPM 27 other JIIM (most overlap)	490 JDM
Gender (F:M)	104:47	321:169
Age at diagnosis (years) (median)	7.0 (range 1–16)	7.0 (172 were <5, 211 were 5–10, 103 were 10–18)
Characteristic or Gottron rash	88%	n/a
Gottron rash	n/a	72%
Heliotrope rash	n/a	61%
Malar rash	n/a	56%
Proximal muscle weakness	82%	84%
Systemic features	81%	n/a
Myalgias	68%	n/a
Arthralgias or arthritis	66%	n/a
Arthritis	36%	35%
Edema	32%	n/a
Dysphagia	29%	18%
Contractures	27%	n/a
Skin ulcers	23%	6%
Dyspnea	18%	n/a
Dysphonia	17%	11%
Lipoatrophy	10%	n/a
Raynaud phenomenon	n/a	5%
Calcinosis	6%	4%

Abbreviation: n/a, no data available.

proportion of children were not found to be clinically weak. Many children also present with systemic features (>80% in one series)[5] including fever, fatigue, and weight loss. Other common symptoms include muscle and joint pain, subcutaneous edema, and symptoms of pharyngeal weakness (dysphonia or dysphagia).

Given that muscle involvement is common to all forms of JIIM, many children will present with complaints of weakness or changes in physical function. However, the extent of these changes may vary from nearly imperceptible to profound. Careful assessment of muscle strength is crucial, and the possibility of JIIM should not be discounted if weakness is not immediately obvious. Weakness primarily affects the proximal (shoulder and hip girdle) and axial (mainly paraspinal and abdominal) musculature. Distal muscle weakness may be present, but it should be less prominent than the proximal muscle weakness. Otherwise, alternative diagnoses should be considered.

In the clinic, muscle strength is most commonly evaluated using confrontational manual muscle testing (MMT). An abbreviated form of MMT has recently been validated in children using 8 muscle groups.[6] However, MMT can be challenging, due to a variety of factors. MMT requires cooperation, and may be difficult for young