Juvenile Idiopathic Inflammatory Myopathies

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KEYWORDS

• Juvenile idiopathic inflammatory myopathy • Juvenile dermatomyositis
• Juvenile polymyositis

KEY POINTS

- The juvenile idiopathic inflammatory myopathies (JIIM) are several acquired, autoimmune disorders that affect muscle and, to a lesser extent, skin.
- JIIM should be considered in children presenting with either signs of muscle weakness or typical rash.
- The diagnosis of juvenile polymyositis should be made cautiously, with careful consideration and evaluation of other differentials.
- Consultation and referral to a provider with expertise in diagnosis and management of JIIM should occur before initiation of treatment.

INTRODUCTION

Juvenile dermatomyositis (JDM) is a rare disease but is the most common and recognizable of the systemic inflammatory myopathies. The term juvenile idiopathic inflammatory myopathy (JIIM) is used to emphasize that there are several acquired autoimmune disorders that affect muscle and, to a lesser extent, skin. In addition to JDM, this group includes juvenile polymyositis (JPM), immune-mediated necrotizing myositis (a disorder recently distinguished from JPM and characterized by severe weakness marked increase in muscle enzyme levels, poor response to therapy, and specific autoantibody associations[1]), and myositis associated with another connective tissue disease. A more complete listing of other disorders can be found in **Box 1**.[2]

All forms of JIIM have muscle involvement as a common feature, which presents as weakness, poor endurance, and reductions in physical function. Skin manifestations are important in some forms, particularly JDM, and children may have several pathognomonic and typical skin lesions. Involvement of organ systems outside the muscle and skin is possible and may have a major impact on both morbidity and mortality.

Historically, the JIIMs were severe, chronic illnesses with mortality in excess of 30%.[3] However, with current therapy, mortality is uncommon but morbidity remains a concern.

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Box 1
Forms of juvenile idiopathic inflammatory myopathy

JDM

JPM

Immune-mediated necrotizing myositis

Focal/nodular myositis

Orbital/ocular myositis

Granulomatous myositis

Eosinophilic myositis

Macrophagic myofasciitis

Myositis associated with another rheumatic illness[a]

[a] Myositis when seen with another rheumatic illness, such as lupus, is considered to be a form of JIIM by some clinicians, whereas others view this as a potential mimic of JIIM.

Data from Rider LG, Nistala K. The juvenile idiopathic inflammatory myopathies: pathogenesis, clinical and autoantibody phenotypes, and outcomes. J Intern Med 2016;280(1):24–38.

New diagnostic approaches hold promise to identify patients at higher risk of poor outcomes, leading to more tailored therapy. Given the rarity and complexity of these disorders, early referral to providers with expertise in the management of the JIIMs is necessary.

INCIDENCE OF JUVENILE IDIOPATHIC INFLAMMATORY MYOPATHIES

All of the JIIMs are rare. The most common is JDM, with an incidence of approximately 2.5 per million per year[4] and prevalence of approximately 2.5 per 100,000.[5] Data are more limited for other forms of JIIM, although JPM is estimated to be about one-tenth as common as JDM, with the other forms rarer still.[4]

DEMOGRAPHICS OF JUVENILE IDIOPATHIC INFLAMMATORY MYOPATHIES

Age

For JDM[6–10]

- Median age at onset 5.7 to 6.9 years
- Median age at diagnosis 7.4 to 7.7 years

For JPM[9]

- Median age at onset 11 years
- Median age at diagnosis 12.1 years

Race and Ethnicity

The impact of race or ethnicity has not been adequately studied, but patients from all races and ethnicities have been reported. However, a study conducted in the United States suggests a similar incidence of JDM for white, non-Hispanic and African American, non-Hispanic children, and possibly slightly lower incidences for Hispanic children.[10]

DELAYS IN DIAGNOSIS ARE COMMON

Most studies report delays in diagnosis, averaging approximately 6 months from disease onset to diagnosis.[6,11] However, much longer delays are common. It is important to make the diagnosis of JIIM in a timely fashion, because delays in diagnosis are