Pediatric Neurology Part III: Chapter 149. Juvenile dermatomyositis (Handbook of Clinical Neurology)

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Juvenile dermatomyositis (JDM) is a systemic, inflammatory, idiopathic disease, mainly affecting the skin and the muscles, starting before the age of 16, with an incidence around one case per 1 million children. Some patients display typical features of JDM without skin involvement, or even without muscle involvement; however, both tissues are affected over time in most cases. Diagnosis criteria were established by Bohan and Peter 35 years ago, based on the presence of typical skin rash and proximal muscle involvement. Other conditions have to be ruled out before making a diagnosis of JDM, such as other connective tissue diseases, polymyositis, infectious/postinfectious myositis, genetic diseases, or metabolic or drug-induced myopathies. Unlike adult-onset dermatomyositis, JDM is exceptionally associated with a malignant disease. JDM may also affect several organs, including the lungs and the digestive tract. In a subset of patients, glucose intolerance, lipodystrophia and/or calcinosis develop. Delay in treatment initiation or inadequate treatment may favor diffuse, debilitating calcinosis. JDM patients have to be referred to reference pediatric centers to properly assess disease activity and disease-related damage (including low bone density in most cases), and to define the best treatment. Long-lasting corticosteroid therapy remains the gold standard, together with physiotherapy. Ongoing clinical trials are assessing the effect of several immunosuppressive and immunomodulatory drugs, which may help to control the disease and possibly demonstrate a corticosteroid-sparing effect. Most patients respond to treatment; relapses are frequent but a complete disease remission is achieved in most cases before adulthood.

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