PATIENT FACT SHEET

Juvenile Dermatomyositis

CONDITION DESCRIPTION

Juvenile dermatomyositis (JDM) is an inflammatory disease that affects children's muscles, skin or blood vessels. JDM's cause is unknown, but one theory is that the immune system directs inflammation at muscle and skin tissues by mistake. JDM often affects large muscles around the neck, shoulders and hips. Muscle weakness makes it hard for children with JDM to climb stairs, get in and out of a car or chair, brush their hair or stand up from the floor. They may have little or no pain. JDM affects three in one million children each year, usually between ages 5-10. JDM affects girls twice as often as boys, and all ethnic groups equally.

SIGNS/ SYMPTOMS

The most common symptoms are muscle weakness and skin rash. Muscles in the hips, thighs, shoulders, upper arms or neck weaken and get worse over time. Both sides of the body are affected equally. Children may struggle to do basic movements like getting up from a chair or the floor. Rashes are violet or dusky red and may be seen in the face or eyelids, and around the nails, elbows, knees, chest and back. Knuckle rashes may be mistaken for eczema. Other signs of JDM may include difficulty swallowing, voice changes, tender or painful muscles, fatigue, fever, weight loss, hard calcium deposits under the skin, stomach ulcers, intestinal tears and lung problems. Blood tests often show evidence of muscle damage and JDM-related autoantibodies, and a nailfold capillaroscopy can show swollen blood vessels around the cuticles. Diagnosis is supported by MRI and muscle biopsy evidence of muscle inflammation.

COMMON TREATMENTS

Children should be treated early to control inflammation, improve function and prevent disability. High-dose intravenous (injection into vein) and/or oral corticosteriods are the standard treatments. Symptoms and inflammation measures may start to improve in 2-4 weeks. However, long-term corticosteroid use can cause osteoporosis, weight gain, cataracts and other side effects, so methotrexate is often added and used in combination with steroids. Alternative treatment options include intravenous immunoglobulin, azathioprine, cyclosporine, tacrolimus, hydroxychloroquine, mycophenolate mofetil and anti-TNF biologics. Rituximab may be used in very severe disease. JDM is unique among most pediatric rheumatic diseases in that it can often be cured if treated aggressively at the beginning.

CARE/ MANAGEMENT TIPS

Physical therapy can help children with JDM build their strength, and prevent muscle wasting and stiffness. The therapist may start with stretches and gradually increase the child's activity. Children with JDM need to protect themselves from the sun by wearing sunscreen, should wear hats with brims and protective clothing when playing outdoors. If neck or throat muscles are affected, the child may need speech therapy. A dietitian can create a diet for children with chewing or swallowing problems. They should eat a healthy and balanced diet, and stay active. JDM symptoms are not always obvious, so parents should speak to their child's educators about the disease and how to prevent injury.

Updated March 2019 by Jonathan Hausmann, MD, and reviewed by the American College of Rheumatology Committee on Communications and Marketing. This information is provided for general education only. Individuals should consult a qualified health care provider for professional medical advice, diagnosis and treatment of a medical or health condition.

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