



PATIENT FACT SHEET

Dermatomyositis (Juvenile)



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

JDM's 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. Early rashes may be patchy. Common sites are the face or eyelids, and around the nails, elbows, knees, chest and back. Knuckle rashes may be mistaken for eczema.

Other possible signs 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.

Diagnosis includes MRI and muscle biopsy to look for tissue inflammation, blood tests for levels of inflammatory markers or JDM-related autoantibodies, and nailfold capillaroscopy to show swollen blood vessels in the cuticles.



COMMON TREATMENTS

Children should be treated early to control inflammation, improve function and prevent disability. High-dose corticosteroids, or up to 2mg/kg/day with the dose tapering down for up to two years, is the standard treatment. Symptoms and inflammation measures may start to improve in 2-4 weeks.

Long-term corticosteroid use can cause osteoporosis, cataracts and other side effects, so other options include methotrexate in combination with steroids, as well as intravenous immunoglobulin, azathioprine, cyclosporine, tacrolimus, hydroxychloroquine, mycophenolate mofetil and anti-TNF biologics. Rituximab may be used in very severe disease in combination with steroids, methotrexate and immunoglobulin.



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. Therapy is necessary for children with calcium deposits. Some children need splints.

Children with JDM should wear hats with brims and protective clothing when playing outdoors to protect their skin from sun damage. 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.

Children with JDM may also have diabetes, celiac disease or arthritis. 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 2017 by James Udell, MD, and reviewed by the American College of Rheumatology Communications and Marketing Committee. 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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