AMERICAN COLLEGE OF RHEUMATOLOGY
EDUCATION • TREATMENT • RESEARCH

Dermatomyositis (Juvenile)

Fast Facts

- Patients with JDM have varying symptoms. They range from mild muscle weakness like difficulty getting out of a chair or difficulty turning over in bed to severe symptoms including profound weakness or difficulty swallowing. Patients can also develop rash or skin changes ranging from mild redness to more severe ulcer formation.
- Other forms of myositis in children include polymyositis, focal myositis, and other rare forms of myositis.
- Myositis almost always causes loss of muscle strength and most all patients also have a rash.
- Early diagnosis and sticking to the treatment plan are important to prevent permanent muscle weakness.
- Children experience JDM differently. While remission is often possible, a minority of children with JDM may have a more chronic course that is less responsive to therapy.

Juvenile dermatomyositis is an inflammatory disease of the muscle (myositis), skin, and blood vessels that affects about three in one million children each year. The cause is unknown. The primary symptoms of JDM include muscle weakness and skin rash. All age and ethnic groups are affected. Most cases start in children ages 5 -10 and adults ages 40 - 50. Girls are affected about twice as often as boys.

Patients with juvenile dermatomyositis (JMD) develop weakness in the large muscles around the neck, shoulders, and hips. This causes difficulty in climbing stairs, getting into cars, getting up from a chair or off the floor, or brushing hair. Most patients have little, if any, pain in their muscles, which distinguishes them from patients with other forms of muscle disease. Many patients with other conditions complain of weakness; however, when questioned closely, they really mean that they are tired, short of breath, or depressed rather than suffering from true muscle weakness.

What causes it?

Dermatomyositis is in a group of diseases or disorders of the muscles called inflammatory myopathies. The cause of most inflammatory myopathies is unknown; however, the leading theory is that the body's immune system mistakenly directs inflammation against muscle cells and blood vessels in the skin and muscles causing damage, rash, and weakness.

What are common signs and symptoms of juvenile dermatomyositis?

The most common signs and symptoms of JDM include:

- **A violet-colored or dusky red rash**, most commonly on the face, eyelids, and areas around the nails, elbows, knees, chest, and back. The rash, which can be patchy with bluish-purple discolorations, is often the first sign of dermatomyositis. A rash on the knuckles occasionally can be misdiagnosed as eczema when on fact it is dermatomyositis.
- **Progressive muscle weakness**, particularly in the muscles closest to the trunk (such as those in the hips, thighs, shoulders, upper arms, and neck). This can affect the ability to get out of a chair, off the floor or into the car and leads to falls. The weakness affects both the left and right sides of the body equally, and tends to gradually worsen.

Other JDM signs and symptoms that may occur include:

- Difficulty swallowing
- Voice changes
- Muscle pain or tenderness
- Fatigue, fever, and weight loss
- Hardened deposits of calcium under the skin
- Stomach ulcers and intestinal tears
- Lung problems

How is juvenile dermatomyositis diagnosed?

There are a number of tests doctors may use to help diagnose juvenile dermatomyositis. These tests include:

- **Magnetic resonance imaging:** A scanner creates images of the muscles from data generated by a powerful magnetic field and radio waves. It does not involve any radiation exposure. As MRI has become more sensitive, doctors have been using it more to diagnose myositis. MRI can detect subtle muscle inflammation and swelling early in the disease. A benefit of MRI is that it allows us to view whole muscles to look for patterns or patches of muscle inflammation, instead of taking a small sample from a single muscle.
- **Muscle biopsy:** Minor surgery is done to remove a small piece of muscle tissue to look at under the microscope. A muscle biopsy may reveal inflammation in the muscles or other problems, like damage, muscular dystrophy, or infection. In dermatomyositis, inflammatory cells are seen surrounding and damaging the tiny blood vessels within the muscles.
- **Blood tests:** A blood test will let the doctor know if enzymes from inflamed muscle are elevated. A blood test also can detect specific autoantibodies associated with JDM, which can help in determining the best medication, treatment, and prognosis.
- **Nailfold capillaroscopy:** Abnormal swelling and distortion of the blood vessels can be seen with the cuticles of the nails. This finding suggests active disease. Your doctor can examine the nailbeds by using a lighted magnifying tool.

Treatment & medications for juvenile dermatomyositis

The goal of treatments for juvenile dermatomyositis is to minimize inflammation, improve function, and prevent disability. The treatment should be early and requires a team approach between the rheumatologist, physical therapist, dermatologist, and primary care doctor.

Corticosteroids

Corticosteroids alter the immune system, limiting the production of antibodies and reducing skin and muscle inflammation, as well as improving muscle strength and function. Corticosteroids, especially prednisone, are usually the first choice in treating inflammatory myopathies such as dermatomyositis, because they work fast.

The doctor may start with a very high dose, and then decrease it as signs and symptoms improve. Signs of improvement may be seen in about two - four weeks as the inflammation is diminished, but the muscles still need to be rebuilt and this may require weeks to months. Often, physical therapy is required for strengthening and retraining the muscles that were damaged.

Standard treatment for JDM has been high-dose daily oral glucocorticoids (e.g., up to 2 mg/kg/day of prednisone, at times in divided doses), which is continued until clinical and laboratory improvements are evident and then reduced slowly over a two-year period (at least). Most patients develop treatment-related side effects with this regimen. In many cases, however, prednisone is introduced early as a treatment option and may be discontinued before the two-year period is completed.

Prolonged use of corticosteroids can have serious and wide-ranging side effects, like osteoporosis and cataracts, so the doctor may recommend supplements like calcium and vitamin D to strengthen bone and regular eye exams to detect cataracts.

Corticosteroid-sparing agents

Other medications work slower, but have fewer side effects than corticosteroids like prednisone, and allow the patient to wean off steroids sooner (“spare” the steroids).

- Methotrexate is considered best initial treatment for most JDM subjects and is now often started along with corticosteroids.
- Immunoglobulin may be administered through an IV, because it contains healthy antibodies from blood donors. High doses can block the harmful antibodies that attack muscle and skin.
- Other steroid-sparing agents include cyclosporine, azathioprine, tacrolimus, hydroxychloroquine, mycophenalate mofetil or Anti-TNF drugs. and rituximab may be used in very severe disease along with immunoglobulin, steroids, and methotrexate.

Other aspects of treating juvenile dermatomyositis include:

- **Skin protection:** Protection from ultraviolet A and B (UVA and UVB) light is thought to help control the rash and potentially muscle disease. Use sunscreen or sunblock that decreases exposure to UVA and UVB light. Wear wide-brimmed hats and photo-protective clothing. Avoid sun exposure during peak daylight hours.