Dermatomyositis

WHAT IS DERMATOMYOSITIS?

Juvenile dermatomyositis (JDM) is an inflammatory disease of the muscle (myositis), skin and blood vessels that affects about 3 in 1 million children each year. The cause is unknown. The symptoms of JDM include muscle weakness and skin rash.

FAST FACTS

- Patients with JDM 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.
- Patients with JDM have varying symptoms. They range from minimal muscle weakness, including falling when running and having to turn over to out of bed, to not being able to swallow and changes in the voice. Patients can also have skin rashes that range from mild redness to severe ulcers.
- Other forms of myositis in children include polymyositis, focal myositis and other rare forms of myositis.

WHO GETS IT?

All age and ethnic groups are affected. Most cases occur in children ages 5 -10 and adults ages 40-50. Women are affected about twice as often as men.

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 abnormalities in the body's immune system may lead to inflammation and damage to muscle cells or the blood vessels that are in the muscle and skin.

HOW IS IT DIAGNOSED?

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, knuckles, elbows, knees, chest and back. The rash, which can be patchy with bluish-purple discolorations, is often the first sign of 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, and tends to gradually worsen.

Other JDM signs and symptoms that may occur include:

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

TESTS

- **Magnetic resonance imaging (MRI).** 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.
- **Electromyography.** A doctor with specialized training inserts a thin needle through the skin into the muscle to be tested. Electrical activity is measured as the patient relaxes or tightens the muscle, and changes in the pattern of electrical activity can confirm a muscle disease. The doctor can determine where the disease is by testing different muscles.
- **Muscle biopsy.** A 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 surround and damage the capillary blood vessels in the muscle.
- **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 blood vessels, which suggest active disease, can be seen by magnifying the capillaries in the fingernail bed and looking with a special light.

TREATMENT

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

Medications

Corticosteroids (Prednisone). These medications suppress 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 2-4 weeks, but therapy often is needed for years. The doctor also may prescribe topical corticosteroids for the skin.

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 2-year period (at least). Most patients develop treatment-related side effects with this regimen.

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 eye exams.

Corticosteroid-sparing agents. Other medications work slower, but have fewer side effects than prednisone, and allow the patient to wean off steroids sooner ("spare" the steroids).

- **Methotrexate** is considered best initial treatment for most JDM subjects.
- **Intravenous immunoglobulin (IVIg).** Immunoglobulin contains healthy antibodies from blood donors. High doses can block the harmful antibodies that attack mu**s**cle and skin.

Other steroid-sparing agents, including cyclosporine, azathioprine, tacrolimus, hydroxychloroquine, or anti-TNF therapy. Cyclophosphamide and rituximab may be used in severe disease with IVIg, steroids and methotrexate.

Other aspects of care for myositis:

- **Skin protection.** Protection from ultraviolet A and B (UVA and UVB) light is thought to help control skin disease and potentially muscle disease. Use sunscreen or sunblock that decreases exposure to UVA and UVB light. Wear wide-brimmed hats and photoprotective clothing. Avoid sun exposure during peak day-light hours.
- **Physical therapy.** A physical therapist can teach exercises to maintain and improve strength and flexibility and advise an appropriate level of activity. Physical activity is thought to be important in JDM. Physical therapy is directed at preventing muscle wasting and stiffness, and is particularly necessary in patients with calcium deposits (calcinosis) and muscle involvement. Therapy should focus initially on stretching and splinting and include more aggressive strength-building therapy once inflammation is controlled.
- **Speech therapy.** If the swallowing muscles are weakened by dermatomyositis, speech therapy can help the patient learn how to compensate for those changes.
- **Dietetic assessment.** In JDM, chewing and swallowing can become more difficult. A registered dietitian can teach how to prepare foods that are safe to eat.

HEALTH IMPACTS

Other secondary conditions have been seen with JDM including such as diabetes, celiac disease and arthritis.

Living with JDM

Since the myopathies can be chronic diseases, it is important for patients to have good general health practices. These include eating a well-balanced, nutritious diet, maintaining a healthy weight and managing any other chronic illnesses. Regular exercise is important to regain and maintain strength. It is important for employers, teachers and family members to understand the limitations imposed by muscle weakness, particularly since patients may look entirely normal.

POINTS TO REMEMBER

- Myositis almost always causes loss of muscle strength.
- Some patients develop rashes, swallowing, or breathing problems.
- Early diagnosis and sticking to the treatment plan are important.
- Children with myositis can experience remission.

TO FIND A RHEUMATOLOGIST

For a listing of pediatric rheumatologists in your area, **click here**. Learn more about **rheumatologists** and **rheumatology health professionals**.

FOR MORE INFORMATION

The American College of Rheumatology has compiled this list to give you a starting point for your own additional research. The ACR does not endorse or maintain these websites, and is not responsible for any information or claims provided on them.

http://www.printo.it/pediatric-rheumatology/information/UK/3.htm
http://www.arthritis.org/disease-center.php?disease_id=37
http://www.curejm.com/

REFERENCES

Myositis and You. Ed. Lisa Rider, MD, Lauren Pachman, MD, Frederick Miller, MD, Harriet Bollar, The Myositis Association, 2007.

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