



Inflammatory Myopathies

The prospect of a muscle disease worries some people because they are afraid of not being able to walk. Yet in many cases, treatment exists for myopathy (muscle disease). Proper diagnosis and treatment raise the chance of living life fully despite this illness.

What are myopathies?

Myopathy is the medical term for muscle disease. Some muscle diseases occur when the body's immune system attacks muscles. The result is misdirected inflammation, hence the name inflammatory myopathies. This damages muscle tissue and makes muscles weak.

People with inflammatory myopathies may have these features:

- Weakness in the large muscles around the neck, shoulders and hips
- Trouble climbing stairs, getting up from a seat, or reaching for objects overhead
- Little, if any, pain in the muscles
- Choking while eating or aspiration (intake) of food into the lungs
- Shortness of breath and cough

The inflammatory myopathies include **polymyositis** and **dermatomyositis**. Muscle inflammation and weakness occur in both conditions, and patients with dermatomyositis also have a rash. This rash most often appears as purple or red spots on the upper eyelids. Scaly, red bumps over the back of the fingers may also occur on the knuckles or the elbows or knees. Children with the disease also may have white calcium deposits in the skin called calcinosis.

Sometimes patients can have the rash with no sign of muscle disease. Doctors call this form of the disease amyopathic dermatomyositis.

People with dermatomyositis may also have lung inflammation (pneumonitis). Children with the disease may have an inflammation of the blood vessels ([vasculitis](#)).



Some doctors group a health problem called **inclusion body myositis** with the inflammatory myopathies. Yet, it differs from them. Men get it more often than women, and the patients tend to be older. Most of these patients do not respond to standard treatment. Therefore, this fact sheet will not discuss this disease.

Who gets myopathies?

Inflammatory myopathies are rare. Polymyositis and dermatomyositis occur in about one person per 100,000.

All ages can get these diseases. The peak time to get them, though, is ages 5 to 10 in children, and 40 to 50 in adults. Women get inflammatory myopathies about twice as often as men. These diseases affect all ethnic groups. No one can predict who will get an inflammatory myopathy.

What causes myopathies?

There are many causes of muscle disease. They include infection, muscle injury due to medicine, inherited diseases that affect muscle function, disorders of electrolyte levels, and thyroid disease.

We do not know what causes inflammatory myopathies. A top theory is that something goes wrong in the immune system that leads to inflammation. This damages muscle cells or the blood vessels that are in the muscle.



Fingers of a person with dermatomyositis show red bumps on the knuckles (referred to as Gottron's papules), redness around the nails and thick, ragged cuticles.

How are myopathies diagnosed?

A doctor suspects myopathy when patients complain of trouble doing tasks that require muscle strength, or when they get certain rashes or breathing problems.

Most people with myopathy have little or no pain in their muscles. This differs from other health problems. Examples are other forms of muscle disease, joint pain due to arthritis and nerve problems that cause numbness or tingling in the hands and feet. Many patients with other illnesses think they feel weak. In fact, they are tired, short of breath or depressed, rather than having true muscle weakness.

A doctor will do a muscle strength exam to find if true muscle weakness is present. These tests likely will follow:

- A blood test to measure the level of various muscle enzymes
- An electromyogram — often referred to as an EMG— to gauge electrical activity in muscle
- A biopsy of a weak muscle (a small piece of muscle tissue is removed for testing)
- Magnetic resonance imaging — more often called MRI — to try to show abnormal muscle



Some patients may have a blood test for myositis-specific antibodies (immune proteins). This test helps detect myopathy. It also gives some information about prognosis (how serious the disease is).

How are myopathies treated?

Doctors most often treat these diseases with medications.

Corticosteroids. Often, the first treatment is an oral (by mouth) corticosteroid, such as prednisone (Deltasone and other brands), at a high dose. This reduces inflammation. Blood muscle enzymes tend to return to normal about 4–6 weeks after treatment starts. Most patients regain muscle strength in 2–3 months.

Combination treatment. Your doctor likely will add another drug to your treatment plan: [methotrexate](#) (Rheumatrex and Folex) or [azathioprine](#) (Imuran). This gives better long-range control of the disease and helps avoid long-term side effects of corticosteroids. These side effects include weight gain and redistribution of body fat, thinning of the skin, [osteoporosis](#) and cataracts. Even muscle weakness can be a side effect.

Because patients taking prednisone are at risk for osteoporosis, they should receive proper treatment to prevent it.

Immune treatments. Patients whose disease is severe or who do not respond to standard treatment have other options. They include intravenous immunoglobulin — sometimes referred to as IVIg — or medicines that suppress the immune system. These immunosuppressive drugs include [cyclosporine](#) (Neoral, Sandimmune), tacrolimus (Prograf), [mycophenolate mofetil](#) (CellCept) and [rituximab](#) (Rituxan).

Physical therapy. Physical therapy and exercise are important in the treatment of muscle disease. Very weak patients who do not walk should receive range of motion exercises. These exercises prevent joint contractures (deformity of the joint). Patients with medium weakness should start an exercise program to strengthen their muscles. The goal is to slowly increase in intensity as the patient regains strength. Mildly weak patients should take part in their normal activities.



Broader health impacts of myopathies

In adults, dermatomyositis and, to a much lesser extent, polymyositis at times may be linked to cancer. Therefore, all adults with these diseases should have tests to rule out cancer.

Living with myopathy

Myopathies are chronic (long-term) diseases. To help control your illness, it is important to practice good health measures. Eat a healthy, well-balanced diet, exercise and try to keep a healthy weight.

If you have a dermatomyositis rash, protect yourself from the sun. This is because the rash gets worse after sun exposure, for reasons that are not clear. Therefore, limit time outdoors, and put on sunscreen when you go outside.

If you have trouble swallowing, eat soft or semisolid foods, or puree your solid foods. To prevent choking if you are bedbound, you should be seated up in bed to eat.

People with myopathy may look healthy and normal. It is important for employers, teachers and family members to understand the limits that muscle weakness causes in people with myopathy.

Points to remember

- Myopathy almost always causes loss of muscle strength.
- Some patients also have rashes or breathing problems, or both.
- While there is no cure for myopathies, treatment is most often effective. Following your doctor's treatment plan is key to having a good outcome.

The role of a rheumatologist in the treatment of myopathies

You need a correct diagnosis of myopathy to have the best possible outcome. Rheumatologists are doctors who are experts in diagnosing and treating diseases of the muscles, joints and bones. Therefore, they are more likely to make a proper diagnosis of myopathy. They can also advise patients about the best treatment options available.

To find a rheumatologist

For a listing of rheumatologists in your area, [click here](#).

Learn more about [rheumatologists](#) and [rheumatology health professionals](#).



Physical therapy can help patients with muscle disease go on leading active lives.



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 Web sites, and is not responsible for any information or claims provided on them. It is always best to talk with your rheumatologist for more information and before making any decisions about your care.

The Arthritis Foundation

www.arthritis.org

National Institute of Arthritis and Musculoskeletal and Skin Diseases Information Clearinghouse

www.niams.nih.gov

The Myositis Association of America

www.myositis.org

Information about specific myopathies:

National Institute of Neurological Disorders and Stroke

www.ninds.nih.gov/health_and_medical/disorders/polymyos_doc.htm

www.ninds.nih.gov/disorders/dermatomyositis/dermatomyositis.htm

www.ninds.nih.gov/health_and_medical/disorders/inclusion_doc.htm

American College of Rheumatology Research and Education Foundation

Learn how the ACR Research and Education Foundation advances research and training to improve the health of people with rheumatic diseases.

www.rheumatology.org/REF

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This information was written by Marc Miller, MD, and reviewed by the American College of Rheumatology Communications and Marketing Committee.

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