AMERICAN COLLEGE OF RHEUMATOLOGY
EDUCATION • TREATMENT • RESEARCH

Specialists in Arthritis Care & Research

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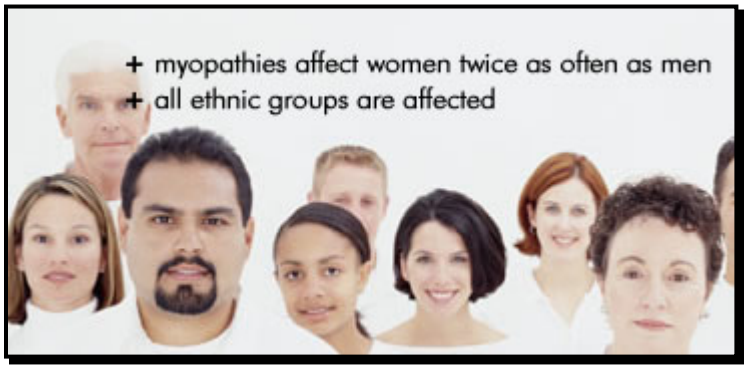

Myopathies

The prospect of a muscle disease is worrisome to many people because they are afraid of becoming immobile. Yet in many cases, treatment exists for myopathy (muscle disease). Proper diagnosis and therapy increase the chances of living life fully in spite of myopathy.

What are myopathies?

Myopathy is the medical term for muscle disease. There are many causes of muscle disease, such as infection, muscle injury due to medications, inherited diseases affecting muscle function, disorders of electrolyte levels, and thyroid disease. Some of these disorders, such as polymyositis, dermatomyositis and inclusion body myositis, develop when the immune system attacks muscles. This inflammation damages muscle tissue and makes them weak.

Patients with myopathy develop weakness in the large muscles around the neck, shoulders and hips. This causes difficulty in climbing stairs, getting up from a chair or toilet seat, or reaching for objects overhead. Most patients have little if any pain in their muscles, which distinguishes them from patients with other forms of muscle disease, from those who have joint pain due to arthritis, and from those with numbness or tingling in their hands and feet due to neurological problems. Many patients with other conditions complain of weakness, but when questioned closely, they really mean that they are tired, short of breath or depressed rather than suffering from true muscle weakness.

Some patients with myopathy develop weakness of throat muscles involved in swallowing, and this may cause choking or aspiration (intake) of food into the lungs when eating. Others may experience shortness of breath and cough due to inflammation of the lungs.

Polymyositis is an inflammation of the muscle tissue that leads to weakness. Dermatomyositis is characterized by immune inflammation of muscles as well as by the presence of a rash. This rash, which appears as a purple or red discoloration of the upper eyelids, is present in almost all children with inflammatory myopathy. Scaly red lesions over the back of the fingers at the knuckles or over the elbows or knees may also occur. Occasionally patients can develop the rash with no evidence of muscle disease, a condition which is known as “amyopathic dermatomyositis.” People with Dermatomyositis may also have lung inflammation (pneumonitis), and children may have an inflammation of the blood vessels (vasculitis) and calcium deposits in the skin referred to as calcinosis.

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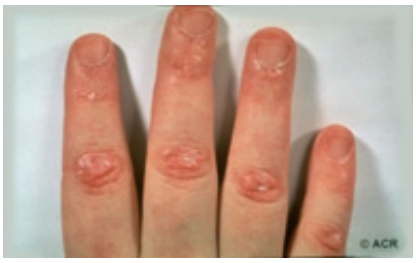
Who gets myopathies?

The inflammatory myopathies are rare diseases. Polymyositis and dermatomyositis occur in approximately 1 person per 100,000. All age groups are affected with peak incidence between the ages of 5 and 10 in children, and between 40 and 50 in adults. Women are affected about twice as often as men. All ethnic groups are affected. It is not possible to predict who will develop an inflammatory myopathy.

Inclusion body myositis is also a very rare disease, but differs from the other inflammatory myopathies in that men are affected more commonly than women, and the patients tend to be older.

What causes myopathies?

It is not known what causes the idiopathic inflammatory myopathies. A leading theory is that abnormalities in the body's immune system may lead to the development of inflammation and subsequent damage to muscle cells or the blood vessels that are in the muscle.



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

How are myopathies diagnosed?

A diagnosis of myopathy is suspected when patients complain of difficulty performing tasks that require muscle strength or when they develop certain rashes or respiratory problems. To establish a diagnosis, a muscle strength examination will be performed to determine if true muscle weakness is present. This likely will be followed by a blood test to measure the level of various muscle enzymes, an electromyogram to gauge electrical activity in muscle, and finally a biopsy of a weak muscle. Sometimes MRI scanning can help to establish the presence of abnormal muscle. Blood may also be tested for the presence of myositis-specific antibodies (immune proteins), which help to establish a diagnosis and give some information about prognosis. Inclusion body myositis is diagnosed by the presence of specific changes on the muscle biopsy.

How are myopathies treated?

Treatment depends on the type of myopathy diagnosed.

Polymyositis and dermatomyositis are usually treated with medications. An oral corticosteroid such as prednisone (*Deltasone* and others) is usually administered in high doses once the diagnosis has been established. Blood muscle enzymes usually return to normal in about 4 to 6 weeks and patients gradually regain strength in 2 to 3 months. Methotrexate or azathioprine are usually added to ensure better long term control of the disease and to avoid long term side effects of cortisone such as weight gain and redistribution of body fat, thinning of the skin, osteoporosis, cataracts and even muscle weakness. In severe or treatment-resistant cases additional measures may include intravenous immunoglobulin and other immunosuppressive medications including cyclosporine *(Neoral, Sandimmune)*, tacrolimus (*Prograf)* or mycophenolate (*Cellcept*).

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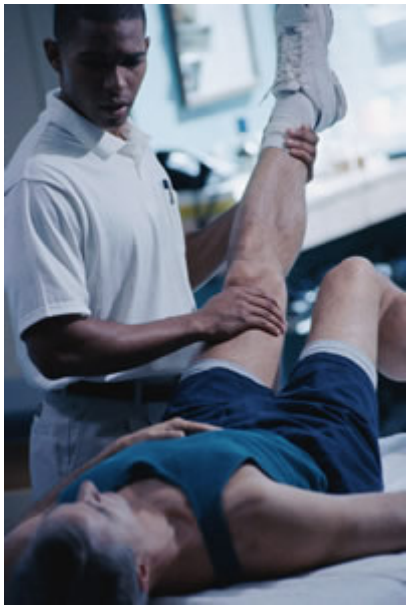
Unfortunately there is no dependably effective treatment for inclusion body myositis. Patients may be given a trial course of prednisone followed by methotrexate or azathioprine. However, if there is no improvement in 2 to 3 months, all drugs should be discontinued.

Patients with dermatomyositis and rash should protect themselves from the sun by limiting time outdoors and using sunscreen when they do go outside. This is because the rash worsens after sun exposure, for reasons that are unclear.

Physical therapy and exercise are important in the treatment of myopathy. Severely weak patients who are confined to their beds should receive range of motion exercises to prevent joint contractures (distortion or deformity of the joint). Patients with moderate weakness should begin a muscle-strengthening program that gradually increases in intensity as strength is regained. Mildly weak patients should be encouraged to participate in normal activities. Patients with swallowing difficulties should receive appropriately prepared food and should be positioned in bed to prevent choking. Patients being treated with prednisone are at risk for the development of osteoporosis and should receive appropriate preventive treatment.

Broader health impacts of myopathies

In adults, dermatomyositis and to a lesser extent polymyositis at times may be related to an underlying cancer. Therefore all adult patients with these disorders should receive appropriate testing to rule out cancer.



Physical therapy can help patients continue to lead active lives.

Living with myopathy

Since the myopathies are chronic diseases, it is important for patients to practice good general health measures including a well-balanced nutritional diet, maintenance of normal weight and proper management of any other chronic illnesses. As discussed above, 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 in myopathy patients, particularly since they may look entirely normal.

Points to remember

Myopathy almost always causes loss of muscle strength. Some patients develop rashes or breathing problems. While the myopathies cannot be cured, most can be effectively treated. Early diagnosis and adherence to the treatment plan are important.

The role of a rheumatologist in the treatment of myopathies

Myopathies should be accurately diagnosed to ensure the best possible outcomes. Rheumatologists are specialists in musculoskeletal disorders and therefore are more likely to make a proper diagnosis. They can also advise patients about the best treatment options available.

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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
http://www.myositis.org/

Specific information about particular myopathies:
http://www.ninds.nih.gov/health_and_medical/disorders/polymyos_doc.htm
http://www.emedicine.com/emerg/topic474.htm
http://www.ninds.nih.gov/health_and_medical/disorders/inclusion_doc.htm

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This information was written by Marc Miller, MD, and reviewed by the American College of Rheumatology Patient Education Task Force.

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