Inflammatory Myopathies

What is myositis? Myositis is the general term used to describe swelling of the muscles. Injury, infection, and even exercise can cause muscle swelling. The swelling will go away once the injury or infection is treated, or once you rest your muscles from exercise. Certain medicines can also cause some muscle swelling that goes away once you stop taking the medicine.

Often people who have temporary myositis from one of these causes become concerned when they find the TMA web site and read about the serious, chronic form of myositis. When we talk about myositis, we mean the inflammatory myopathies.

What are inflammatory myopathies? Myositis is the popular term used to describe a number of inflammatory myopathies: dermatomyositis (DM) inclusion-body myositis (IBM) juvenile forms of myositis (JM) polymyositis (PM)

These diseases cause swelling and loss of muscle. They are also called idiopathic inflammatory myopathies. “Idiopathic” means the causes are unknown.

Inflammatory myopathies are thought to be autoimmune diseases, meaning the body's immune system, which normally fights infections and viruses, is misdirected and attacks the body's own normal, healthy tissue through inflammation, or swelling. All of these diseases can cause muscle weakness, but each type is different.

**Signs and symptoms:** Muscle weakness, sometimes with muscle pain General tiredness and fatigue Trouble climbing stairs, standing from a seated position, or reaching up Difficulty swallowing

Dermatomyositis Dermatomyositis (DM) affects people of any age or sex, but is found in more women than men.

DM is the easiest type of myositis to diagnose because of the skin rash which is often seen before any muscle weakness is felt. The DM rash looks patchy, dusky, and reddish or purple. It is found on the eyelids, cheeks, nose, back, upper chest, elbows, knees and knuckles. Some people also have hardened bumps under the skin, called calcinosis. The skin rash and weak muscles are caused by inflammation, or swelling, in the blood vessels under the skin and in the muscles, also called vasculitis. Patients who have the skin rash but feel no muscle weakness have
amyopathic DM, or DM sine myositis.

The weakness begins with muscles that are closest to and within the trunk of the body. Neck, hip, back and shoulder muscles are examples. Some DM patients have muscle pain.

**Signs**  Appearance of a rash on the eyelids, cheeks, nose, back, upper chest, elbows, knees and knuckles  Scaly, dry or rough skin  Trouble rising from a seated position, or getting up after a fall  General tiredness

**Symptoms**  Painful and/or itchy rash caused by inflammation of blood vessels under the skin and in the muscles  Sudden or progressive weakness in muscles in neck, hip, back and shoulder muscles  Difficulty swallowing (dysphagia), a feeling of choking  Hardened lumps or sheets of calcium, called calcinosis, under the skin  Changes in voice (dysphonia), especially hoarseness

Juvenile dermatomyositis  Juvenile Myositis (JM) is found in children under the age of 18 and affects 3,000 to 5,000 children in the United States. Juvenile dermatomyositis, or JDM, is a disease marked by muscle weakness and skin rash. The other form of myositis that can occur in children – juvenile polymyositis – is extremely rare.

The first sign of JM is usually a skin rash. The rash may be red and patchy, like dry skin; a red or purplish color on the eyelids or cheeks that may look more like allergies; or both. Children with juvenile polymyositis do not experience skin symptoms.

JM patients can have weak muscles at the same time they see the skin rash, or the weak muscles may come after the rash over days, weeks or months. The weaker muscles are usually closer to the body (for example, neck, shoulders, back, and stomach), and you may notice your child having trouble climbing or standing from a seated position. The skin rash and weak muscles are caused by inflammation or swelling in the blood vessels under the skin and in the muscles.

Other signs may include falling, weaker voice (dysphonia), or problems swallowing (dysphagia). About half of the children with JM have pain in their muscles.

Some children may have calcinosis, hardened lumps or sheets of calcium under the skin, or contractures, in which the muscle shortens and causes
the joint to stay bent. Exercising the muscles can prevent contractures.

**Signs** Visible, reddish-purple rash over the eyelids or joints General tiredness Moodiness or irritability Complaints of tummy aches &nbsp Trouble climbing stairs or onto a bus; standing from a seated position; getting dressed Difficulty reaching up, like to shampoo or comb hair Trouble lifting the head

**Symptoms** Characteristic reddish-purple rashes of JM—Gottron’s papules (bumps found over the knuckles, elbows and knees) and heliotrope rash (purplish rash around the eyes) Gradual muscle weakness, most often of those closest to the body like neck, stomach, upper arms and legs Hardened lumps or sheets of calcium (calcinosis) under the skin &nbsp Trouble swallowing (dysphagia) Hoarse-sounding voice Stomach problems

Inclusion-body myositis Inclusion-body myositis (IBM) is found in more men than women with onset usually occurring after age 50. A small number of IBM cases may be hereditary (h-IBM) but most are “sporadic” (s-IBM) meaning there is not a direct genetic link. In most cases, IBM progresses slowly over months or years. There is currently no effective treatment for IBM.

**Signs** Frequent falling episodes Trouble climbing stairs or standing from a seated position A foot that seems to drop when walking, causing tripping Weakened hand grip Difficulty swallowing

**Symptoms** Weakness and noticeable shrinking of the quadriceps (main muscle of the thighs), causing falls Weakness in the forearm muscles Weakness of muscles below the knees, causing the foot to drop and toes to catch when walking Weakness of flexor muscles of the fingers used for gripping Weakness of throat muscles, causing trouble swallowing (dysphagia) and possibly choking Pain or discomfort as muscles weaken

Polymyositis Polymyositis (PM) is found mostly in people over the age of 20 and affects more women than men. Muscle weakness usually happens over days, weeks or months. The weakness begins with muscles closest to and within the trunk of the body. Neck, hip, back and shoulder muscles are examples. Some patients also have weakness in muscles farther from the trunk, like hands and fingers. Some PM patients experience muscle pain, breathing problems, and trouble swallowing.
Researchers are finding that each case of PM is quite different from others. Sometimes, cases originally diagnosed as PM and not responding to treatment are later found to be inclusion-body myositis (IBM). Patients with certain types of PM may have one or more other autoimmune diseases.

**Signs**
- Sudden or gradual weakness in the muscles
- Difficulty swallowing (dysphagia)
- Falling and difficulty getting up from a fall
- General feelings of tiredness

**Symptoms**
- Marked weakness in the muscles closest to the center of the body, like the forearms, thighs, hips, shoulders, neck and back
- Sometimes, weakness in the fingers and toes
- Thickening of the skin on the hands (mechanic’s hands)

**Diagnosis**
Diagnosing myositis is often a complicated and lengthy process. Different tests and examinations help doctors establish an accurate diagnosis so you can begin treatment as soon as possible.

During your clinical exam, your doctor will ask questions about your health in general, including detailed personal and family health histories. The doctor will want to know when you first saw signs of the skin rash or muscle weakness, and will examine your muscle strength and any skin symptoms.

The doctor may use one or more of the following tests to help confirm a specific diagnosis: Conventional blood tests: Doctors look for elevated levels of muscle enzymes in patients’ blood samples. Muscle and skin biopsy: Small samples of muscle tissue show abnormalities in muscles, including inflammation, damage, and abnormal proteins. For those with skin symptoms, doctors often biopsy a piece of skin to study. Electro-diagnostic tests: Muscle resonance imaging scans (MRI) reveal inflammation in muscles; electromyograms (EMG) detect changes in muscles’ electrical patterns that indicate muscle disease and which muscles are affected. Antibody testing: Since the discovery of certain myositis-specific and myositis-associated antibodies, more detailed blood testing confirms a diagnosis and provides insight into the possible course of the disease as well as potential complications.

Other tests may rule out another type of disease or condition that has similar symptoms to myositis. If you have questions about any test, talk with your doctor or lab technician.
Treatment Myositis varies tremendously from patient to patient, and no one treatment works for everyone. Your physician may use a combination of drugs to treat you, or change medications over time. It is extremely important that you communicate well with your doctor about your treatment and any side effects.

An important part of your treatment will be the regular practice of an exercise program that works for you, no matter what your level of ability or weakness. Talk with your physician about supplements that are sometimes recommended for muscle strength.

Standard medications for myositis patients  Physical therapy and a regular exercise routine  Supplements recommended for myositis patients

Frequently asked questions I’ve been diagnosed. Now what? Patients with polymyositis and dermatomyositis generally respond to treatment in a month or two. The active period of the disease is usually 2-3 years in both children and adults, but patients with cardiac or pulmonary complications have a longer active period than others. IBM patients have a slow-progressing disease, with no effective treatment.

Is there a cure? A review of many dermatomyositis and polymyositis cases found that 20% of the patients recover completely, although we don’t say they are “cured.” These patients will never have another active period, or flare. Often doctors will say that people are “in remission” when there is no sign of disease activity. Others will have flares for a long time before the disease is controlled, and some will have periodic flares all their lives. These outcomes depend in part on the speed of diagnosis and treatment. Patients diagnosed with polymyositis who don’t respond to treatment should ask to be tested for inclusion-body myositis.

Do people die from myositis? Research shows that the mortality rate after several years of the disease is approximately 15%, which reflects the higher rate of mortality in patients with connective tissue diseases, cardiac involvement or cancer.

Do IBM patients have flares? There is no effective treatment for inclusion-body myositis. This form of myositis causes people to get slowly weaker rather than experience flares. Sometimes doctors give patients a trial course of prednisone followed by methotrexate or azathioprine. If there is no improvement in a month or two, the drugs are discontinued. Patients with IBM need to anticipate the imminent limitations in their strength and mobility and prepare for each change.
How soon will I respond to medication? Although every case varies, blood muscle enzymes usually return to normal in about 4 to 6 weeks and polymyositis and dermatomyositis patients gradually regain strength in 2 to 3 months. Prednisone is the most common treatment. Methotrexate or azathioprine is usually added for better long-term control of the disease and to avoid prednisone’s long-term side effects. In cases where people either don’t respond to prednisone or have serious side effects, doctors may prescribe intravenous immune globulin (IVIG) and other immunosuppressive medications including cyclosporine (Neoral, Sandimmune), tacrolimus (Prograf) or mycophenolate (Cellcept). Other, newer drugs are being tested for myositis. Ask your doctor if you are eligible for the clinical trials now being conducted.

What is autoimmunity, and what causes an autoimmune disorder? The body’s immune system normally fights infections, viruses and other harmful things by producing antibodies and white blood cells called lymphocytes, a process known as the immune response. In someone with an autoimmune disorder, the immune response is overactive and doesn’t properly “turn off.” The immune system is misdirected and essentially turns on itself, attacking the body’s own normal, healthy tissue. In the inflammatory myopathies, these overactive immune cells target the muscles, causing inflammation.

How will pregnancy and nursing affect and be affected by myositis? Some people with dermatomyositis (DM) first notice the signs (for example, rash and weakness) during pregnancy. For patients who already have myositis and become pregnant, the weakness and other symptoms can become worse, or flare, during pregnancy. For children with juvenile myositis, it’s difficult to predict how myositis will affect them later in terms of fertility, but there have been enough studies with positive pregnancy outcomes for these children and their families to remain optimistic about the possibility of having a healthy pregnancy.

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