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Myositis

WHAT IS MYOSITIS?

Myositis (my-oh-SIGHT-iss) is a disease that causes inflammation of the muscles (myo means muscle and itis means inflammation). There are several different forms of myositis, including:

- polymyositis;
- dermatomyositis;
- inclusion body myositis;
- myositis in children (juvenile myositis); and
- myositis that occurs in people with other autoimmune diseases, such as rheumatoid arthritis (RA), lupus and scleroderma.

The various forms of myositis differ in signs and symptoms, course and treatment.

Myositis is a rare disease. It is estimated that each year five to 10 out of every one million adults in the United States gets one of the forms of myositis.

Myositis can affect people of any age. Children who get the disease usually are between five and 15 years of age. Adults usually are between 30 and 60 years old. Like many other autoimmune diseases, most forms of myositis affect more women than men.

All forms of myositis involve inflammation that develops within muscle tissues, most commonly the hips or shoulders. Rarely, muscle inflammation occurs in a single part of the body, such as one arm, one leg or just muscles that move the eye. The muscle inflammation almost always causes weakness. Sometimes warmth, swelling and pain of the muscles can occur. Myositis may be associated with inflammation in other organs, such as the:

- joints;
- heart;
- lungs;
- intestines; and
- skin.

The most common form of myositis is polymyositis (poly means many). Muscle inflammation develops in both arms and legs. The form of myositis called dermatomyositis (dermato means skin) is associated with a skin rash. This rash usually is found over the knuckles in the hands and around the eyes. However, in some people, rashes occur on other parts of the body.

WHAT CAUSES MYOSITIS?

Scientists don't know yet what causes myositis. Because myositis has many forms, it probably has many different causes. Some scientists think that myositis develops after a person with a certain genetic background is exposed to particular drugs, chemicals, viruses or other infectious agents. Other environmental exposures such as ultraviolet light or stressful life events may trigger the development of myositis.

It is important for people to tell their doctors about any prescription or over-the-counter medications they may be taking, infections or other environmental causes that may have occurred before the onset of myositis. In cases resulting from drug exposure or infection, the myositis goes away when the drug is stopped or the infection is treated.

Whatever triggers the disease also may result in abnormalities in the immune system. The immune system is made up of groups of cells called lymphocytes that move throughout the body. In healthy people, lymphocytes defend the body by making substances that attack viruses, bacteria and other disease-causing agents. But in people with myositis, lymphocytes attack muscle.

In addition, some people have an abnormality in the immune system that produces proteins called autoantibodies. Autoantibodies and some of the lymphocytes turn against the body's own tissues and cause tissue damage. This process is called autoimmunity (auto means self). Therefore, myositis is called an autoimmune disease.

SYMPTOMS OF MYOSITIS

The many forms of myositis begin and progress in different ways. In most people, the illness develops slowly over a period of months or even years. In some people, problems can

occur suddenly. Typically, myositis develops so slowly that people aren't aware that their muscles are weak for a long time. This makes it hard for doctors to diagnose at first.

Once the disease is diagnosed, it can appear to come (disease flare) and go (disease remission) without warning, and the myositis can change over time. Myositis flares usually are recognized by increasing symptoms of:

- muscle weakness;
- fatigue;
- skin changes; and
- worsening of arthritis, lung disease or other organ systems affected by myositis.

Muscle Weakness

The major symptom of myositis is muscle weakness. Hip and shoulder muscles are most likely to be involved. The weakness causes difficulty doing daily activities, such as:

- combing hair;
- putting on clothes;
- getting in and out of the bathtub, bed, chair or car; and
- climbing stairs.

Other muscles sometimes affected are those in the front of the neck and throat, making it hard to swallow, sing or even speak clearly.

Skin Changes

Dermatomyositis causes several types of rashes. A bumpy, patchy, reddish or purplish rash on the face, around the eyes and on the eyelids is called a heliotrope rash. A rash on the knuckles, elbows or knees is called Gottron's papules. If the rash is over the upper back, back

of the neck and along the sides of the arms, it is known as shawl sign. Some people have puffy eyelids, wrists or hands as a result of myositis. If you have myositis you also may develop:

- redness or irritation around the base of your fingernails;
- overgrown cuticles;
- cracked and bleeding skin on the sides and tips of your fingers; or
- small, whitish lumps of calcium deposits called calcinosis, usually on the fingers.

Other Symptoms

Other symptoms of myositis include:

- fatigue;
- fever;
- poor appetite;
- weight loss;
- pain;
- shortness of breath; or
- tenderness or pain in the muscles or joints.

Sometimes it's very difficult to tell whether the joint or the muscles between the joints are causing pain and other problems. If your chest muscles or lungs are affected, you may have trouble breathing or may develop a cough.

Some people become sensitive and react to the cold resulting in white, blue and red color changes in the fingers. This is referred to as Raynaud's (RAY-nodes) phenomenon and is caused by the narrowing of the blood vessels in the fingers. This reduces blood flow and turns the fingers white and then gradually blue or red.

Occasionally, the heart can become damaged by myositis. This damage can result in an irregular heartbeat or an enlarged heart. Swelling in the legs

and ankles (called edema) and shortness of breath also can occur as a result of heart damage.

DISEASE COURSE

How myositis begins and progresses differs from person to person. Some people first develop rashes, shortness of breath and joint inflammation. New symptoms may develop over time and the initial symptoms go away.

Rarely, myositis or dermatomyositis may be a sign of cancer. Generally, the myositis improves or entirely goes away once the tumor is removed or the cancer is treated.

DIAGNOSIS

Because myositis is similar to many other diseases and differs from person to person, it can be difficult to diagnose. Your doctor will often need to perform many tests and see you several times before diagnosing myositis. Generally, specialists will be involved in the evaluation of people with suspected myositis and in providing treatment. These specialists include:

- a rheumatologist (roo-ma-TALL-o-jist), a doctor who specializes in arthritis and related diseases, such as myositis;
- a pediatric rheumatologist (pee-dee-AAH-trik roo-ma-TALL-o-jist), a doctor who specializes in treating children with arthritis and related diseases; or
- a neurologist (nur-ALL-o-jist), a doctor who specializes in treating problems of the central nervous system.

You will be asked questions about your symptoms and a physical examination will be performed. Blood tests will be done, including some that test for autoantibodies and muscle enzymes.

One of the most useful blood tests performed is to determine muscle enzyme levels called creatine kinase (CK). The CK level in the blood is unusually high in people with active myositis. The CK level can go up with increasing myositis disease activity and go down as the myositis improves.

Another procedure that is useful in diagnosing myositis is an electromyogram (EMG). An EMG measures the electrical pattern of the muscles. Sometimes, magnetic resonance imaging (MRI) of the weak muscles is done. An MRI produces a picture of the muscle that can show areas of inflammation.

A muscle biopsy also may be performed. In this procedure, a small piece of muscle is removed and the tissue is examined under a microscope. The muscle biopsy shows the amount and type of muscle tissue damage. It helps determine what type of myositis you have.

TREATMENT

There are many types of treatment for myositis. These include medications, rest, exercise and physical therapy. Treatment will vary from person to person and will change over time for each person. The treatment recommended will depend upon the severity and type of problems, other medical conditions, and any bad reactions you may have to previous therapy.

Medication

GLUCOCORTICOIDS

The most effective and most commonly prescribed drugs in treating myositis are glucocorticoids (such as prednisone and cortisone). Glucocorticoids are powerful drugs that help reduce inflammation and relieve stiffness and pain. However, they can cause side effects, which include:

- weight gain;
- stunted growth (in children);
- thinning of the bones (osteoporosis);
- depression and mood swings;
- cataracts;
- glaucoma;
- worsening of diabetes or onset of diabetes in someone who has never had it;
- rounding of the face;
- sleep problems; and
- high blood pressure.

Prednisone is the glucocorticoid medication most often used for myositis. If your myositis is severe, high doses of glucocorticoids may be prescribed. Prednisone may be given orally (by mouth) in the form of pills, or intravenously (into a vein) in a liquid form. Several weeks or months may pass before you see obvious improvement, but you may be getting better sooner without realizing it. Throughout the treatment of myositis you will need to get blood tests to check myositis disease activity.

After your strength improves, your doctor can begin to slowly reduce the dose of prednisone. If your myositis remains under control, your doctor may eventually be able to take you off prednisone completely. This will probably not occur for at least six months after starting prednisone. More treatment with prednisone may become necessary if your myositis flares again.

Special Instructions for Glucocorticoids

Your body naturally produces small amounts of hormones that are forms of glucocorticoids. They help your body perform many daily functions, such as maintaining blood pressure and keeping the salts (electrolytes) in your body in balance. When you take glucocorticoid drugs

for more than a few days, your body may stop producing some of these hormones. This is not permanent. Your body will start making the hormones again as the dose of your medication is lowered.

It is important to pay attention to your diet when taking glucocorticoids. Reducing your calories and avoiding added salt and high-sodium foods lessen the chance of weight gain and high blood pressure. A dietitian can help you change your diet.

Taking glucocorticoids may put you at risk for osteoporosis (ah-stee-oh-po-RO-sis). This is a disease that causes bones to weaken and have an increased risk of fracture. Bone-density tests should be done before you begin taking glucocorticoids. A bone-density test measures your bone mass. It is painless and accurate. This test should be repeated each year while taking glucocorticoids in order to monitor your bone health. Calcium and vitamin D supplements, hormone replacement and/or bone-preserving medications are regularly prescribed to help prevent osteoporosis.

Remember:

- Do not stop taking glucocorticoids without talking to your doctor first.
- Do not try to increase or decrease this medication on your own. You and your doctor will need to work together to gradually wean you off of this medicine.
- Tell your other health-care providers that you are taking this medicine. Your medication may need to be increased for short periods of time if your body is under added stress – for example, if you are having surgery or if you have a secondary illness.
- Wear a medical identification bracelet or necklace to let people know that you take these medications in case a medical emergency occurs.

- If you become sick or vomiting prevents you from taking this medication, let your doctor know right away. You may need to increase your usual dose for several days while you are sick.
- Talk to your doctor about ways to prevent osteoporosis.
- Talk to a dietitian to help you with your diet.

Be Aware

If you suddenly stop taking glucocorticoids, your body may not be able to make enough hormones on its own on short notice. This can make you very sick. Slowly reducing the dose is required to prevent you from getting sick.

IMMUNOSUPPRESSIVE DRUGS AND OTHER MEDICATIONS

Your myositis may not respond to glucocorticoid therapy alone. If this is the case, both prednisone and a drug that slows the immune system may be prescribed. These drugs are called immunosuppressive drugs. The most common ones used in myositis are methotrexate and azathioprine. Immunosuppressive drugs slow down the immune system. This reduces the immune system's attacks on healthy tissue such as muscle, but also decreases your body's ability to fight infection.

Immunosuppressive drugs can cause side effects. If you are taking these medications, you will need to keep regular follow-up appointments in order to monitor any side effects and medication dose.

Call your doctor if you have any of the following side effects: loss of appetite; excessive weight gain; mouth sores; skin rash; fever; chills; or shortness of breath.

Other treatments that may be prescribed for your myositis include:

- other immunosuppressive drugs (such as cyclosporine and chlorambucil);
- tumor necrosis factor (TNF) inhibitors etanercept or infliximab;
- intravenous immunoglobulin therapy (IVIG);
- hydroxychloroquine.

Each of these treatments has different possible side effects. Talk to your doctor before taking these and all other medications.

Caution

Methotrexate may cause birth defects to unborn children. You should use birth control while on methotrexate. Women taking methotrexate must go off their medication during pregnancy and for several months prior to a planned pregnancy. People who have serious kidney or liver disease, or who drink alcohol should not take methotrexate.

Rest

Getting enough rest is another important part of managing myositis. During times of increased muscle weakness, take frequent rests during the day and limit your activity to an acceptable level. It is more helpful and efficient to schedule regular rest breaks rather than waiting until you are too tired or fatigued.

If you are employed, discuss your situation with co-workers, supervisors and your doctors to develop a work plan to accommodate your disease. The Americans with Disabilities Act (ADA) gives you the right to ask for work accommodations. To learn about the federal laws that can help you in your place of employment, contact the Arthritis Foundation to request a free copy of the booklet *Arthritis in the Workplace*.

When your myositis is active, you should limit strong physical activity, and you may not be able to work. If you are able to work, your day should include regularly scheduled rest breaks. You can do more activity and take fewer rest breaks as your myositis improves. Avoid overdoing activity at the first sign of improvement. Your doctor and physical therapist can help decide the right balance of rest and activity at each stage of myositis.

Exercise and Physical Therapy

A physical therapy or exercise program is an important part of treating myositis. It can improve muscle strength and endurance. It is important for you to not lose strength in muscles that are not affected by the myositis. Generally, you should not begin vigorous exercise until the drug treatment begins working and your muscle strength improves. A physical examination and laboratory tests will help your doctor and therapist decide the right time to start an exercise program and how hard and how much you should exercise.

Even when strenuous exercise is not recommended, it is important to prevent loss of flexibility in your arms and legs by doing range-of-motion exercise or gentle stretching of the joints and muscles. Other physical therapy activities may include simple exercises at home or sessions with a health-care professional.

As you get better, your physical therapy program will be adjusted. People with myositis can learn to exercise safely to maintain or improve strength and endurance. For more information about exercise, request a copy of the Arthritis Foundation's booklet *Exercise and Your Arthritis*.

RESEARCH

Since 1948, the Arthritis Foundation has invested nearly \$320 million on research to

help prevent, control and cure arthritis and related conditions such as myositis. Currently, researchers funded by the Arthritis Foundation are using mice to examine the role of the immune system in the damage and repair of muscle cells. Other investigators are trying to learn what genes are associated with the onset of these conditions. Such studies could lay the basis for more effective, novel therapies.

THE ARTHRITIS FOUNDATION

The mission of the Arthritis Foundation is to improve lives through leadership in the prevention, control and cure of arthritis and related diseases.

The Arthritis Foundation supports research with the greatest potential for advances and has invested more than \$320 million in these efforts since its inception in 1948. Additionally, the Arthritis Foundation supports key public policy and advocacy efforts at a local and national level in order to make a differ-

ence on behalf of 70 million people living with arthritis.

As your partner in taking greater control of arthritis, the Arthritis Foundation also offers a large number of programs and services nationwide to make life with arthritis easier and less painful and to help you become an active partner in your own health care.

Contact us at (800) 283-7800 or visit us on the Web at www.arthritis.org to become an Arthritis Advocate or to find out how you can become involved.

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For more information: The Arthritis Foundation offers a wide variety of books, brochures and videos about different forms of arthritis, treatment and self-management techniques to help you take control of your arthritis. To order any of these products, become an Arthritis Foundation member or to subscribe to the Arthritis Foundation's award-winning consumer health magazine, *Arthritis Today*, call (800) 283-7800. Call or visit our Web site (www.arthritis.org) to find out how you can take control of your arthritis and start living better today!

MISSION STATEMENT:

The mission of the Arthritis Foundation is to improve lives through leadership in the prevention, control and cure of arthritis and related diseases.

This brochure has been reviewed by the AMERICAN COLLEGE OF RHEUMATOLOGY.



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