



MYOSITIS 101

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Your guide to understanding myositis

Patients who are informed, who seek out other patients, and who develop helpful ways of communicating with their doctors have better outcomes. Because the disease is so rare, TMA seeks to provide as much information as possible to myositis patients so they can understand the challenges of their disease as well as the options for treating it.

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MYOSITIS BASICS

“Myositis” means general inflammation or swelling of the muscle. There are many causes: infection, muscle injury from medications, inherited diseases, disorders of electrolyte levels, and thyroid disease. Exercise can cause temporary muscle inflammation that improves after rest.

A more specific use of the word is to describe a chronic inflammatory muscle disorder, also called myopathy, or disease of the muscle. Dermatomyositis (DM), polymyositis (PM), inclusion-body myositis (IBM), and juvenile forms of myositis (JM) are all inflammatory myopathies, or diseases where there is swelling and loss of muscle. It often appears gradually. Long before you were diagnosed, you may have had trouble getting up from a chair, climbing stairs, or grasping objects with your hands.

Inflammatory myopathies are 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. Inflammatory myopathies are rare diseases. All forms combined affect an estimated 50,000 to 75,000 people in the United States. The causes of DM, PM, IBM and JM are not known, but some doctors believe there is an environmental exposure to infection, virus, toxin or sunlight that triggers the disease in someone who has an inherited tendency for it. There is no cure for any of the forms of myositis.

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DERMATOMYOSITIS (DM) affects people of any age and sex but is more common in women. It's the easiest type of myositis to recognize because there's usually a visible skin rash caused by inflammation of blood vessels under the skin (vasculitis). The rash is patchy and reddish or purple, and it can be seen on eyelids, cheeks, nose, back, upper chest, elbows, knees or knuckles. Some patients with DM usually report gradual muscle weakness and sometimes pain, and they often notice the rash well before the muscle weakness. Two sub-types of DM are amyopathic DM, where the skin is affected but muscles are not involved; and cancer-associated DM, where cancer and dermatomyositis are diagnosed within two or three years of one another. The juvenile form of DM, which has both similarities and difference, is described on the following page.

POLYMYOSITIS (PM) affects mostly adults and is more common in women than men. Patients experience muscle weakness gradually, usually beginning with muscles closest to the body's core, like neck, hip, back and shoulder muscles, although some patients also have weakness in their hands and fingers. It affects both sides of the body equally. Some patients have trouble swallowing, called dysphagia; or difficulty breathing, which can be a sign of an inflammation in the lining of the lung, called interstitial lung disease. Many patients feel pain as well as weakness in their muscles. Like DM, PM may be associated with a malignancy or with other autoimmune diseases. When someone has more than one autoimmune disease, it is called "overlap syndrome," which is further described later.

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INCLUSION-BODY MYOSITIS (IBM) affects more men than women and is rarely seen in people younger than 50 years old. IBM progresses more slowly than the other types of myositis, and weakness happens gradually, sometimes over years. Some of the first signs of IBM are falling, difficulty getting up from a chair, and weakening grip. Muscles most often affected are those at the front of the thighs, those that elevate the feet, and those in the hips, fingers, wrists, upper arms, shoulders, neck, back, and, less often, in the face. Many IBM patients notice shrinking, or atrophy, in the arms and thighs as the muscles become weaker. Trouble swallowing (dysphagia) is a common problem for IBM patients.

JUVENILE MYOSITIS (JM) occurs in children younger than 18 and affects more girls than boys. Juvenile dermatomyositis (JDM) is the most common form, affecting an estimated 3,000 to 5,000 children in the United States. Unlike the adult forms, JM is not associated with cancer. Polymyositis in children is very rare. Signs of JM include skin rashes, the visible, reddish-purple rash over the eyelids or over joints; trouble climbing or lifting the head; a weak voice (called dysphonia); or problems swallowing (dysphagia). The muscles most often affected are those closest to the center of the body-neck, stomach, upper arms and legs. About half the children with JM report pain in their muscles. Some children have calcinosis (hardened lumps under the skin) or contractures (when the muscle shortens and causes the joint to stay bent). Like adults, children may have more than one autoimmune disease, or overlapping diseases.



DIAGNOSIS

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If you have read any of the Sherlock Holmes mysteries, you have had a glimpse into the way physicians make diagnoses. Dr. Watson, the narrator of the stories, often expresses awe at the seeming brilliance of Holmes' observations, but the great detective explains frequently that his conclusions are based on simple observations and deductive reasoning.

Medical diagnoses are made by this same process, and it is no coincidence that Sherlock Holmes employed this method. Sir Arthur Conan Doyle, Holmes' creator, was a physician, and he modeled the detective's character after that of one of his medical school professors who was noted for his ability to make brilliant diagnoses based on simple observations in his patients.

Physicians who care for patients with myositis use the process of deductive reasoning to arrive at their diagnoses as well. Although very few of us are as consistently brilliant as Sherlock Holmes, the process of making a diagnosis most often begins with the collection of clues from the history of the illness and observations from the physical examination. Although our muscles make up a substantial part of our body mass, there are only a limited number of ways in which they can produce symptoms when they malfunction. The neuromuscular physician is trained to recognize the signs and symptoms of muscle malfunction.

PHYSICAL EXAMINATION

The main symptom of muscle disease is, of course, weakness. In patients with myositis, we have learned that not all muscles in the body are affected equally. For example, certain groups of muscles, such as the ones that we use to bend our fingers to make a fist, seem to be disproportionately weak in inclusion-body myositis (IBM). Observation of weakness in finger flexor muscles is thus an important clue to the possibility that a patient may have IBM. The neuromuscular physician assembles such observations (“findings” in medical speak, “clues” in Holmes’ terms) to make a tentative preliminary diagnosis.

BLOOD TESTS

Once the preliminary diagnosis is made, a search for supporting evidence begins. Such evidence can come from the results of a number of tests. Blood tests, in particular “muscle enzymes,” provide important clues. The muscle enzymes consist of proteins that exist in high concentrations in muscles. The main one is creatine kinase, or CK. When muscle is damaged or degenerates, muscles leak CK into the bloodstream. In someone with myositis, the CK level in the blood may be elevated, thus when an elevated CK level is found, it is an important clue that something is wrong with the muscle. A number of other blood tests may be done to determine whether or not other diseases, which may produce secondary damage to muscles, are present.

ELECTROMYOGRAM (EMG)

Although few who have it remember it fondly, the electromyogram (EMG) study can provide particularly helpful information. This study consists of two parts. The first is the nerve conduction study (NCS), where recordings are made from nerves and muscles while small electrical shocks are applied to various points along a nerve. The NCS is then followed by the EMG study, where a fine needle electrode is inserted into various muscles. Those who have had this test will recall hearing a sound like static on a radio receiver during this test. There are patterns of electrical abnormalities in nerves and muscles that can indicate the presence of an inflammatory disease, and the physician who does the test is trained to interpret them.

MAGNETIC RESONANCE IMAGING (MRI)

In recent years, physicians have learned that the appearance of the muscles on magnetic resonance imaging (MRI) scans of a limb can provide evidence about muscle disease. Increasingly, neuromuscular physicians are performing MRI scans on patients who are suspected to have myositis. This study, too, can provide evidence about selective involvement of muscles.

MUSCLE BIOPSY

Ultimately, a biopsy of one of the muscles is usually needed to make a confident diagnosis. The information obtained from physical examination, blood tests, EMG, and MRI scans can suggest that the most likely diagnosis is a muscle disease, but there are other diseases of the nervous system that might produce similar abnormalities. Since decisions about treatment hinge on having an accurate diagnosis, the neuromuscular physician will usually feel most confident when the evidence from a muscle biopsy is available.

Although the biopsy procedure requires removal of a piece of muscle, it is really a rather minimal procedure. A small piece of muscle is obtained by one of two methods. The most common method is through a small incision (usually an inch or so) in the skin over a muscle in the arm or leg. This is done through a patch of skin that has been numbed by local anesthetic, so there is little or no discomfort. As an alternative to the "open" procedure, some physicians will use a needle to obtain muscle tissue. This has the advantage of only requiring a small nick in the skin, but the disadvantage is that the amount of muscle obtained is sometimes insufficient to allow a confident diagnosis.

However the muscle sample is obtained, it is examined by a physician who has special training in interpretation of muscle biopsies. The muscle sample must be processed and stained with various chemical compounds before it can be interpreted under the microscope. The processing can take a week or more, so the final diagnosis from a muscle biopsy can be delayed.

Occasionally, the information obtained on the biopsy is inconclusive, and a second biopsy is sometimes necessary.

The neuromuscular physician will make the most accurate diagnosis possible based on information from all of the sources described above. The process described is used by neuromuscular physicians, to make diagnoses in other muscle diseases as well, but it is particularly important in the evaluation of a patient who may have some form of myositis.

Specific blood tests are further discussed on page 12.



BLOOD TESTS

Your doctor will order blood tests when trying to diagnose myositis or monitor the progress of your disease (see Diagnosis, page 5). One or more of the following tests will help give an accurate picture of your disease:

ALANINE AMINOTRANSFERASE

(ALT, also called serum glutamate pyruvate transaminase [SGPT]); enzyme found in many tissues, including muscle. When muscle tissue is damaged, muscle cells leak the enzyme into the blood so that a blood sample will show increased ALT levels. Normal ranges vary by age, gender and other factors.

ALDOLASE

This is an enzyme found in high concentrations in muscle tissue. When the muscle is damaged, the contents of the muscle cells (including aldolase) are released into the blood. Aldolase testing is an indicator of muscle damage. Normal values range between 1.0 and 7.5 units per liter (U/L), but check your own laboratory's normal value range.

ANTINUCLEAR ANTIBODY

(ANA): protein produced by the immune system that attacks the body's own tissue rather than foreign antigens. There is typically no ANA detectable in the blood, so presence of ANA may indicate a type of autoimmune disease, like myositis.

FLOW CYTOMETRY

This is a blood test looking at a specific group of white blood cells that is used to learn more about the extent and severity of juvenile myositis (JM).

LACTATE DEHYDROGENASE

This is an enzyme found in skeletal muscle tissue. When muscle tissue is damaged, levels of the enzyme in the blood increase. Normal values vary, but typical values range from 105 to 333 IU/L (international units per liter).

MYOSITIS-SPECIFIC ANTIBODIES

In addition to the conventional blood tests above, many patients are now tested for “myositis-specific antibodies” (MSAs). These antibodies, and others called “myositis-associated antibodies” (MAAs) were identified several years ago, and are present in about 50 percent of PM and DM patients. Since they almost never occur unless a patient has one of these diseases, they assist in confirming the diagnosis. Because those with the same antibodies follow certain clinical patterns, the presence of MSAs and MAAs offers some insight into the possible course of the disease. The MSAs most often checked for include antibodies against Jo-1, KS, KJ, PL-7, PL-12, OJ, Mi-2, and SRP.



COMMON QUESTIONS

Now that you've been diagnosed, you are sure to remember questions that you didn't think of in the doctor's office. The following questions are some of those most commonly asked by newly diagnosed patients. You will find more detailed answers on the other pages of this publication, and you are welcome to call TMA if you need more information on these topics or any others.

HOW LONG WILL IT TAKE TO RESPOND TO TREATMENT?

Patients with polymyositis and dermatomyositis generally respond to treatment in a month or two, showing improvement in blood tests and in physical strength. More difficult cases may have flares that last a year or longer, and some cases respond immediately, with no reoccurrence. IBM patients have a slow-progressing disease, with no effective treatment. Patients diagnosed with polymyositis who don't respond to treatment should ask to be tested for inclusion-body myositis.

WILL I BE CURED?

There is no cure for myositis. A review of many dermatomyositis and polymyositis cases found that 20% of patients recover completely, although they are not considered "cured." These patients will never have another active period, or flare. 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.

The average active period of the disease, after initial onset, is usually 2-3 years in both children and adults, but patients with cardiac or pulmonary complications have a longer active period than others.

WHAT IS REMISSION?

Often doctors will say that people are “in remission” when a patient is able to discontinue all medications and there is no sign of disease activity.

WHAT IS A FLARE?

People with polymyositis, dermatomyositis and juvenile myositis typically have periods when the disease activity is greater, either before treatment, after tapering medication, or after a period of remission. (See Glossary, page 38.) Inclusion-body myositis patients weaken gradually and do not have the same pattern of remission and flares.

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 disease, cardiac involvement or cancer. Talk to your doctor about cancer screenings, and see your doctor immediately if you are having trouble swallowing, if you have symptoms of pneumonia, or if you notice other changes in your health.

WILL MY CHILDREN BE AT RISK FOR MYOSITIS?

While people inherit a tendency that makes it more likely for them to get an autoimmune disease, it is very unlikely that children of a myositis patient will ever get myositis. The exception is the rare hereditary form of IBM, which is passed down in certain families. Scientists know that a complicated relationship of genetics, environment and personal health history determines who will get myositis.

HOW CAN I FIND A PHYSICIAN FAMILIAR WITH MYOSITIS?

TMA's medical advisors are very familiar with myositis and many of them accept patients. To find one near you, visit "About TMA" on www.myositis.org, and click on "Medical Advisory Board."

You can also visit the American Colleges of Rheumatology web site, at www.rheumatology.org; or the American Academy of Neurology web site, at www.thebrainmatters.org, to find specialists listed by city and state.



TREATMENT

CORTICOSTEROIDS

Prednisone and methylprednisolone are commonly used with fairly fast results in polymyositis, juvenile myositis and dermatomyositis patients. Although they were originally hailed as miracle drugs, we now know that corticosteroids have negative side effects, particularly in high doses. Some side effects are brittle bones, cataracts, stomach upset, weight gain, and changes in blood sugar. Because of these troubling side effects, doctors generally prescribe the lowest dose possible, and for as short a time as possible. In many cases, corticosteroids are literally life-saving and work quickly. Some physicians prescribe them every other day, rather than every day, and those working with patients who experience insomnia suggest patients take them in the morning. Some patients are unable to take corticosteroids because of extreme side effects. IBM patients very rarely respond to steroid treatment, but it may be tried for a short period, sometimes along with methotrexate or azathioprine.

Physicians are likely to increase the dose in the event of a flare. Increasingly, doctors are prescribing combinations of drugs in order to reduce the amount of corticosteroids. There is a wide range in the size of the steroid doses that physicians prescribe, depending on the weight of the patient and the severity of the disease. Never discontinue or reduce your dose without checking with your physician.

DISEASE-MODIFYING ANTI-RHEUMATIC DRUGS

None of the drugs used in myositis treatment were originally developed for inflammatory myopathies but were taken from treatment protocols for other diseases. Plaquenil was developed to combat malaria; methotrexate and chlorambucil were borrowed from oncologists who used them for treating cancer; and cyclosporine was developed by transplant specialists as an anti-rejection drug.

Cyclosporine (Neoral, Sandimmune) is an alternative to prednisone for long-term immunosuppression and relatively rapid onset of benefit. Side effects are uncommon.

Azathioprine (Imuran) provides long-term immunosuppression with few side effects. It is used to reduce needed doses of corticosteroids.

Mycophenolate (CellCept) may be used for long-term immunosuppression as the primary agent or to reduce side effects from other medications.

Methotrexate (Rheumatrex or Trexall) is often used in combination with prednisone, to reduce the dose required. It is also used alone and with other drugs. With routine monitoring of liver function, serious side effects are uncommon. If you drink alcohol, discuss this with your doctor.

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Cyclophosphamide (Cytoxan) is used in cases with life-threatening features, especially with lung involvement. It is used carefully because of possible side effects.

Hydroxychloroquine (Plaquenil) is also used to reduce steroids and is especially helpful for dermatomyositis skin rashes.

BIOLOGIC RESPONSE MODIFIERS

These drugs were borrowed from rheumatoid arthritis treatment. Some used by myositis patients are etanercept (Enbrel) and infliximab (Remicade). These drugs are very expensive and many insurance companies do not pay for them. They provide a more targeted treatment and are being studied in several clinical centers.

Rituximab (Rituxan) has been used by physicians both alone and after other drugs have failed. There are multi-center clinical trials underway for both adults and children. More will be known about side effects as the drug becomes more widely used.

ACTHAR

Acthar is a preparation of ACTH in 16% gelatin formulation which is used either subcutaneously or intramuscularly in the treatment of polymyositis and dermatomyositis. Acthar is the only FDA approved drug for PM and DM other than corticosteroids. It is a naturally occurring hormone produced in the pituitary gland and has several potential mechanisms as to how it may benefit patients with PM or DM. ACTH causes the release of cortisol from a patient's adrenal glands, thus mimicking the effects of taking corticosteroids by mouth. ACTH also interacts at a number of receptors throughout the immune system and may reduce immune overactivity responsible for causing PM and DM. Although Acthar has FDA approval for treating PM and DM, there is very limited data on the effectiveness of the therapy. Ongoing studies are being performed to evaluate the role for Acthar in the treatment of PM and DM.

BLOOD PRODUCTS

Plasma exchange (plasmapheresis, PE) and human immune globulin (IVIG) are used for rapid onset, short-term benefit when patients have life-threatening signs such as respiratory insufficiency, dysphagia, or severe weakness. IVIG is increasingly used in DM when other drugs fail or are poorly tolerated. Availability and access to IVIG fluctuates. Side effects are rare and usually related to delivery rather than the drug itself.

treatment



DISEASE MANAGEMENT

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Although your doctor will monitor your medication, there are many things that you can do yourself to maintain or improve your health.

- ▶ **Stay as active as possible.** Physical therapy and exercise are very important in the treatment of myositis. Even if you cannot leave your bed, gentle stretching and movement can help you. If you can exercise on your own, you should follow a program of strengthening that starts out very slowly and increases as you regain some strength. Children as well as adults should go about their normal routines as much as possible.
- ▶ **Treat swallowing difficulties.** If you have trouble swallowing (dysphagia), you must address this seriously and consult with your doctor. Your physicians may advise you to prepare foods a certain way, work with a speech therapist, avoid foods that give you trouble, or use a feeding tube.
- ▶ **Ask your doctor about cancer screening.** While myositis doesn't cause cancer, physicians have found that a higher than normal percentage of dermatomyositis (DM) and polymyositis (PM) patients also have cancer. TMA's medical advisors suggest that newly diagnosed patients, especially those older than 50, be screened for cancer. When cancer is successfully treated, the myositis symptoms usually disappear. If there is more than a five-year lapse between the two diagnoses, they are not considered to be related.

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- ▶ **Report lung involvement immediately.** Myositis patients, especially those showing certain patterns in blood tests, are at a greater risk for interstitial lung disease (ILD). Ask your doctor if your blood test shows this susceptibility, and report any difficulties with breathing, speaking or swallowing immediately.
 - ▶ **Watch for overlapping diseases.** Like other autoimmune disease patients, people with myositis sometimes show symptoms of more than one illness. Some overlapping illnesses found in myositis patients are rheumatoid arthritis, lupus, scleroderma, and Sjogren's Syndrome. Other patients report symptoms of fibromyalgia, which is itself a poorly defined and understood disease. TMA's medical advisors have observed that myositis symptoms are often less severe in patients who have another autoimmune disease. In general, medications for the overlapping diseases are often the same as those for treating myositis, so treatment will be similar. Since none of these autoimmune diseases has a cure, your doctor will treat the symptoms and monitor your progress carefully.
 - ▶ **Know the side effects of prednisone.** If you are being treated with prednisone, you are at risk for osteoporosis, weight gain, diabetes, mood swings, and skin and vision problems. Report any side effects immediately to your doctor. You can manage many of these effectively with diet, lifestyle changes, and supplements; or your doctor may decide to use other medications.

- **Eat nutritious food.** If you limit sodium, sugar, and excess fat, you may be able to avoid some of prednisone's side effects, particularly the weight gain. As researchers find out more about the anti-inflammatory properties of certain foods, it appears that a diet based on fruits, vegetables, whole grains, healthy fats, and lean sources of protein can help people with chronic disease achieve the best health possible.
- **Manage stress.** Learning you have a chronic illness can be upsetting. Many patients with myositis find new ways to increase hope and meaning in their lives through prayer, meditation, volunteering, spending time with nature, adopting a pet, gardening, or pursuing other interests.
- **Find support.** No matter what the disease, you will do better if you find a community that understands and supports you. TMA provides many options for support.
- **Become an advocate.** TMA members raise funds for research; they lead campaigns for better access to care and research dollars; they mentor new patients through TMA's support groups and online message boards; they educate the public and policymakers about the need for quicker diagnosis and better treatment through "Myositis Awareness Day" and building relationships with the local media.

For more information on these and other disease management topics, visit www.myositis.org, or contact TMA at tma@myositis.org or 1-800-821-7356.



BE AN INFORMED PATIENT

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The more you know about your disease, the better your outcome will be. Some of these suggestions from The Myositis Association's physicians and patients may help:

- ▶ **Ask questions.** Be sure to ask all the questions on your mind. Write them down before your appointment so you won't forget anything. Don't wait for your doctor to ask for your questions (he/she may not!). Ask while the questions are still fresh in your mind.
- ▶ **Listen carefully.** Doctors often explain things with scientific language instead of everyday words, so remember any words you don't understand and ask the doctor to explain them. Ask more questions until you do understand. There's a lot of information to remember, so take notes on what the doctor says. Ask for copies of your medical records to keep at home.
- ▶ **Learn more about myositis.** Read as much as you can about your illness. You can begin at TMA's web site, using information from the site or asking questions on the message boards. This will help you understand your doctor's responses and help you ask better questions.

- **Know your medical history.** Tell your doctor if you have other medical problems, and provide a written list of your medications. Include non-prescription medications, vitamins and other supplements, including herbal and homeopathic preparations. Your health history is important.
- **Understand your options.** If treatments are discussed, make sure you understand the length, benefits, side effects, and probability of success for each one. Take notes about the treatment information. Sometimes it is helpful to bring someone along to do this for you. Refer to the notes later and write down additional questions as they come up.
- **Document your progress.** As your treatment progresses, keep your own written records of tests performed...where, when, and by whom. Note any side effects of medications. Pay attention to any concern expressed by your pharmacist, and notify your doctor. Write down any change in your health — good or bad.
- **Be a partner.** As a patient, you're working with the doctor. It's your right to understand your tests, treatment, what to expect with the disease, and what you can do. Work to form an open, comfortable relationship with your doctor — you are partners in your health care. Many patients call TMA with questions about the best treatment for them. While staff are always glad to hear from you and share information, it's important for you to discuss your concerns with someone who actually knows your case history. If you find it impossible to communicate, it may be time to look for another doctor.

- ▶ **Keep your doctor informed.** Use your written records when you tell your doctor about any side effects or any changes in your health after starting a medication. Don't delay if there's a serious change. Your doctor needs to know your reactions in order to treat you properly.
- ▶ **Find out about drug trials.** When information about human drug trials becomes available, TMA provides a link on its site, www.myositis.org. Ask your physician if you qualify for any ongoing trials; or call the recruitment number directly. Be aware that some trials do not provide the drug to everyone who participates: Some patients are used as "controls" and receive a placebo. Other trials furnish the drug to one set of patients at first, then switch, furnishing the drug to the control patients.
- ▶ **Visit TMA's Community Forum.** TMA members often post remarks on the Forum talking about physicians they find especially helpful, day-to-day coping strategies that work for them, new ideas from media health coverage, or side effects they experience from particular medications. There is a Forum for each type of disease as well as a general Forum at TMA's web site.

TMA also offers periodic live discussions with medical experts on the web site, and the opportunity for submitting questions in advance and joining the online discussions. These discussions are a benefit of TMA membership.



GLOSSARY OF TERMS

The following are general definitions for terms used in connection with myositis. They may be explained in more detail within the rest of the publication. These terms may also have other definitions depending on the situation.

**alanine aminotransferase
(ALT, also called serum glutamate
pyruvate transaminase [SGPT]):** see Blood Tests

aldolase: see Blood Tests

alternative therapy: any therapy used in place of conventional, or more traditional, treatments; examples are special diets instead of chemotherapy for cancer, or homeopathic remedies. (See also complementary therapy.)

**amyopathic dermatomyositis
(also called dermatomyositis sine
myositis):** type of dermatomyositis in which there is a noticeable skin rash but no evidence of muscle weakness. Muscle may become involved over time, or the skin rash may be the only indication of dermatomyositis.

antibody:	protein produced by the body that acts against antigens (foreign proteins) in an immune response.
antigen:	foreign protein that stimulates an immune response in the body. An immune response is the body's reaction to a foreign substance in order to keep it from harming the body.
anti-Jo-1 antibody:	protein, found in about 25 percent of myositis patients, that is associated with a higher incidence of interstitial lung disease (ILD).
antinuclear antibody (ANA):	see Blood Tests
anti-rheumatic:	medicine that acts against disease with inflammation or pain in muscles or joints.
aspartate aminotransferase (AST, also called serum glutamic-oxaloacetic transaminase [SGOT]):	see Blood Tests

autoimmune disease:

disease in which the body's immune system, which normally fights off infections and viruses, is misdirected and mistakenly attacks its own healthy tissue through inflammation. In myositis, the immune system attacks otherwise healthy muscle and skin tissue.

biopsy:

test in which a small piece of muscle or skin is removed to view under the microscope. A muscle biopsy is often used to confirm a diagnosis of myositis.

calcinosis:

hard, often painful lumps of calcium that form under the skin's surface, especially in juvenile dermatomyositis.

complementary therapy:

any therapy used in addition to treatment prescribed by your physician. Examples are massage, aromatherapy, and tai chi used along with your prescribed treatments. (See also alternative therapy.)

contracture:

stiffening of the joint, causing it to shorten and stay bent. Range-of-motion exercises may be prescribed to help prevent or improve contractures.

conventional therapy:

traditional treatments that are more commonly prescribed by physicians. For many people with myositis, for instance, conventional therapy is prednisone and/or methotrexate.

**corticosteroids
(also called steroids):**

medicines aimed to slow the immune system, reduce the inflammatory response, and relieve redness, swelling, itching, and discomfort. Prednisone and Solumedrol are corticosteroid medicines.

**creatine phosphokinase
(CPK, also called creatine kinase
[CK]):**

see Blood Tests

dysphagia:

trouble swallowing; difficulty moving food or liquid from your mouth to your stomach.

efficacy:

effectiveness of a particular medicine to treat the disease or condition for which it is being tested.

electromyogram (EMG):

test in which a small needle is inserted into muscle to measure electrical activity as you relax and tighten that muscle. Changes in the electric activity pattern help determine whether you have a muscle disease.

erythrocyte sedimentation rate (ESR, also called sed rate):

see Blood Tests

first-line treatment:

medicine that the doctor chooses to try as the first treatment for your condition.

flare:

return of past symptoms or increase in current symptoms after a period of remission or slower disease activity. This may occur when tapering medicine too quickly or overexerting yourself through exercise or stress.

flow cytometry:

see Blood Tests

generic:

type of drug manufactured to mimic brand name medicine at less cost. Generic medicines are typically less expensive than their brand name counterparts.

**Gottron's sign
(also Gottron's papules):** unusual redness of the knuckles with a raised, scaly eruption; characteristic skin symptom of adult and juvenile dermatomyositis.

heliotrope rash: blue-purple discoloration on the upper eyelids with swelling; characteristic skin symptom of adult and juvenile dermatomyositis.

immune system: your body's system that protects you from foreign substances through immune response (i.e. inflammation). In myositis, the immune system is thought to be overactive, causing your body to attack its own healthy tissue.

immunosuppressant: medicine that lowers the body's ability to fight infection by slowing the body's immune system. For myositis, the goal is to keep the immune system cells from fighting healthy tissues.

inflammation: response to injury that results in redness, swelling, pain, and sometimes loss of function.

integrative therapy:

combination of complementary therapy to treat symptoms and conventional therapy to treat underlying disease.

intravenous:

method of giving medicine through a needle directly into your vein.

intravenous port:

device that remains in your vein to make giving medicines by needle less painful or uncomfortable. Intravenous medicines are then given by inserting the needle into the port rather than having to find a vein in which to stick the needle each time.

lactate dehydrogenase:

see Blood Tests

local:

affecting only a part of the body where medicine is applied or given. Local treatments include topical creams for the DM skin rash.

lymphocyte:

cell originating from bone marrow that plays a role in immunity.

magnetic resonance imaging (MRI): test using powerful magnets to create images of muscle to determine if there is inflammation present.

maintenance dose: very small amount of medicine a person may need in order to keep symptoms from returning or worsening.

malar rash: butterfly-shaped rash on the cheeks and bridge of the nose in dermatomyositis.

mechanic's hands: dilated capillary loops at the base of the fingernails with irregular, thickened, and distorted cuticles, or cracked, "dirty" horizontal lines at the side and palm areas of the fingers.

monoclonal antibodies: antibodies from a single cell, in large numbers, that act against a particular antigen. Enbrel and Remicade, for example, target tumor necrosis factor (TNF), a protein that is believed to increase in myositis patients.

glossary of terms

pulse:	dose of medicine given intravenously (through IV needle) over a short period of time. Pulse doses are often higher doses given to jump-start treatment.
refractory:	resistant or unresponsive to treatments.
relapse:	return of symptoms after a period of remission or no additional disease activity.
remission:	period of time when patient shows no symptoms of disease and has been off all medicines for six months or longer.
retrospective:	type of study that looks at patients' past experiences to determine if a certain treatment has worked.
rheumatic disease:	disease in which there is swelling or pain in the muscles or joints.

second-line treatment:

medicine chosen after a patient fails to respond to the first medicine given or when side effects from the first medicine are too great. Methotrexate is often a second-line treatment after prednisone.

shawl sign:

a flat red rash on the back and shoulders.

**steroid myopathy
(also called type 2 muscle fiber atrophy):**

weakness caused by long-term use or corticosteroid medicines.

systemic:

therapy that affects the body as a whole, as in medicines taken orally or intravenously.

taper:

process of slowly lowering your dosage of medicine to reach a maintenance dose or stop taking the medicine completely. It is important to follow a tapering schedule especially with corticosteroids to allow your body to adjust properly.

glossary of terms

vacuoles:	holes in the muscle fibers evident in muscle biopsies of inclusion-body myositis (IBM) patients.
vasculitis:	swelling of blood vessels under the skin that causes a visible rash.
von Willebrand factor antigen:	see Blood Tests

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This reprinting of Myositis 101 is made possible through a charitable contribution from Mallinckrodt Pharmaceuticals. Mallinckrodt has not provided any input into the content presented.



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