



# Myositis 101

Your Guide to Understanding Myositis

THE MYOSITIS ASSOCIATION

## **Myositis 101**

Patients who are informed, who seek out other patients, who develop helpful ways of communicating with their doctors, and who are able to advocate for themselves or loved ones have better outcomes. Because myositis is such a rare disease, The Myositis Association 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.

The opinions expressed in this publication are not necessarily those of The Myositis Association. We do not endorse any product or treatment that we report. We ask that you always check any treatment with your physician.

# Table of Contents

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<b>Myositis basics</b> .....	<b>1</b>
Dermatomyositis.....	2
Polymyositis.....	3
Necrotizing myopathy .....	3
Sporadic inclusion body myositis .....	4
Juvenile myositis .....	5
<b>Diagnosis</b> .....	<b>7</b>
Blood tests.....	11
<b>Treatment and disease management</b> .....	<b>15</b>
Medications .....	15
Exercise and physical therapy .....	21
Complementary and self-care therapies.....	23
<b>Complications</b> .....	<b>29</b>
Antisynthetase syndrome .....	29
Calcinosis .....	30
Cardiovascular disease.....	32
Dysphagia .....	33
Infection .....	34
Interstitial lung disease .....	37
Overlapping autoimmune diseases.....	38
<b>Prognosis</b> .....	<b>41</b>
<b>Become your own best advocate</b> .....	<b>43</b>
<b>Glossary</b> .....	<b>47</b>





## Myositis basics



The term “myositis” refers to a general inflammation or swelling of the muscle. Many people have experienced sore muscles after vigorous exercise, a condition that is temporary and improves with rest. Other conditions that can cause muscle weakness and pain include infection, muscle injury from medications, inherited diseases, electrolyte imbalances, and thyroid disease.

More often, however, the term myositis is used to refer to a disease involving chronic inflammation of the muscles, often occurring together with other symptoms. These conditions are also known as idiopathic inflammatory myopathies (IIM). Myositis is highly variable and has been classified into a number of forms, including dermatomyositis (DM), polymyositis (PM), necrotizing myopathy (NM), sporadic inclusion body myositis (sIBM), and juvenile forms of myositis (JM).

Inflammatory myopathies are rare autoimmune diseases. This means that the body’s immune system, which normally fights off foreign invaders such as infections and viruses, is misdirected and begins to attack the body’s own normal, healthy tissue. All forms combined affect an estimated 50,000 to 75,000 people in the United States. While it is still unclear what causes myositis, some scientists believe certain individuals have a genetic predisposition to develop an autoimmune disease, which, when triggered by an environmental exposure, such as infection, virus, toxin, or sunlight, can lead to myositis. Actual triggers, however, are currently unknown.

Symptoms of weakness and sometimes muscle pain often appear gradually. Long before patients are diagnosed, they may have trouble getting up from a low chair, climbing stairs, combing their hair, dressing, or grasping objects with their hands. Patients may fall, find it difficult to raise their arms up, or other symptoms. Myositis may, however, appear within weeks, especially in children.

As the disease progresses, some patients also have trouble swallowing (dysphagia) or difficulty breathing, which can be a sign of an inflammation in the lung tissue called interstitial lung disease (ILD). Many patients feel pain as well as weakness in their muscles. Patients may have more than one autoimmune disease overlapping with myositis.

Myositis is often difficult to diagnose, because many physicians are unfamiliar with the disease and its symptoms. It is a rare disease, so it is also difficult to enroll enough patients to conduct adequate research of new treatments, and there are no clear guidelines in the medical community on how to effectively manage patients with myositis. Nevertheless, myositis is a serious and often treatable illness that, in most cases, needs to be managed aggressively. With inadequate or no treatment, myositis can cause significant disability and even death. While symptoms can be managed and remission can occur, there is no true “cure” for any of the forms of myositis, and it is a lifelong illness.

## **Dermatomyositis**

Dermatomyositis (DM) affects people of any age and sex but is more common in women. Muscle weakness, if it occurs, develops over days, weeks, or months, and usually occurs in the limbs closest to the core (shoulders and hips). Creatine kinase (CK) levels in the blood are usually markedly elevated, which reflect breakdown of muscle. CK levels, however, can be normal in some cases.

DM is the easiest type of myositis to recognize, because patients usually have a visible skin rash caused by inflammation of blood vessels under the skin. Patients often notice the rash well before the muscle weakness.

The rash is usually patchy and reddish or purple, and it can be seen characteristically on the eyelids, or overlying the knuckles, elbows, and knees. It may also be present around the nailbeds and on the cheeks, nose, back, or upper chest. Patients also often experience intensely itchy skin, tender areas under the skin, joint pain, itchy scalp, and hair loss. Some patients, especially children, also develop hard lumps—calcium deposits—under the skin called calcinosis.

Some people with DM, however, never exhibit muscle weakness, and laboratory tests do not indicate muscle damage as they do in other forms of myositis. This form of DM is known as amyopathic dermatomyositis (ADM).

DM in adults can also be associated with an increased risk of malignancy. Cancer-associated DM occurs when cancer and dermatomyositis are diagnosed within two or three years of one another.

## **Polymyositis**

Polymyositis (PM) most often affects adults and is more common in women than men. Muscle weakness usually develops over days, weeks, or months. It begins with muscles closest to and within the trunk of the body, such as the muscles of the neck, hips, back, and shoulders. Some PM patients may also experience muscle pain, breathing problems, and trouble swallowing. Creatine kinase (CK) is usually markedly elevated and correlates with the degree of muscle inflammation.

Like DM, PM in adults may be associated with other autoimmune diseases. Polymyositis is often confused with NM or sIBM, especially if muscle biopsy is not done or if biopsy results are not definitive.

## **Necrotizing myopathy**

Necrotizing myopathy (NM), also known as necrotizing autoimmune myopathy (NAM) or immune-mediated necrotizing myopathy (IMNM), has long been thought to be extremely rare. Recently, however, it is being recognized as a common form of inflammatory muscle disease.

Like other forms of myositis, NM usually develops over days, weeks, or months, causing muscle weakness in the limbs closest to the core and often extremely elevated creatine kinase (CK) levels. Patients also report weakness in other muscles, including breathing and swallowing muscles. Some patients have difficulty holding their head in an upright position. Heart muscle involvement is rare. NM can also be associated with cancer in adults.

The exact mechanism underlying this disorder is not known, but as many as 60% of patients with NM have autoantibodies directed against HMGCR (HMG-CoA reductase) or SRP (signal recognition particle). HMGCR is the enzyme targeted by statin medications (prescribed for many adults to reduce cholesterol levels), and it has been suggested that statin medications or dietary statins may trigger NM. Many patients with NM, however, have never taken statin medications.

Necrotizing myopathy is often confused with PM. Unlike other forms of myositis, however, muscle biopsies in NM show noticeable death (necrosis) in muscle cells with little or no accompanying inflammation. Nevertheless, the presence of autoantibodies and the fact that symptoms improve when treated with anti-inflammatory drugs or intravenous immune globulin support the idea that NM is an autoimmune disease.

### **Sporadic inclusion body myositis**

Sporadic inclusion body myositis (sIBM) is the most common acquired myopathy in patients over the age of 50. It is unlike all other forms of myositis in terms of symptoms, treatment, and who it affects.

More men have sIBM than women, and the disease is rarely seen in people younger than 50 years of age. Symptoms of sIBM progress more slowly than the other types of myositis, with weakness increasing gradually, sometimes over years. It is not uncommon for patients to realize that they have been experiencing symptoms for many years before they were diagnosed. The slow nature of the disease and the age of typical patients often means some physicians dismiss the symptoms as “old age.”

Unlike other forms of myositis, muscle weakness with sIBM often occurs asymmetrically (on one side of the body more than the other). In addition to weakness and muscle wasting (atrophy) in the upper legs, sIBM also frequently involves the flexor muscles of the wrist and fingers. Forearms and feet are also affected, and dysphagia (difficulty swallowing) occurs in about two-thirds of patients. Creatine kinase levels can be normal or elevated but never more than ten times the norm.



There is currently some debate among myositis experts about whether or not sIBM is actually an inflammatory disease. On one hand, inflammatory cells are present in muscle tissue from patients with sIBM, especially earlier in the disease process, but their role in causing muscle weakness is unclear. In addition, autoantibodies have been found in about one-third of the patients with sIBM, suggesting an autoimmune process.

On the other hand, the weakness also appears to be the result of a degenerative process within the muscles, and the disease does not respond fully to treatment with anti-inflammatory medications. Most experts agree, however, that optimal treatment and future cure will likely require attention to both inflammation and muscle degeneration.

Sporadic inclusion body myositis should not be mistaken for hereditary inclusion body myopathy (hIBM). Although muscle biopsy findings in the hereditary myopathies share some of the same features seen in sporadic IBM—rimmed vacuoles and inclusions in muscle cells—these two conditions are otherwise quite different. The hereditary form of the disease is caused by a gene defect, not inflammation. The average age of onset in hIBM is between the teenage years and mid-twenties, not in older age. Muscle weakness in hereditary inclusion body myopathy is usually distal (in the extremities) and may include other areas of weakness. CK levels range from normal to slightly elevated.

## Juvenile myositis

Juvenile Myositis (JM) is found in children under the age of 18 and affects 3,000 to 5,000 children in the United States. The most frequent form of JM is juvenile dermatomyositis (JDM), in which children experience marked muscle weakness and characteristic DM skin rashes.

Unlike the adult forms, JM is not associated with cancer. Polymyositis and necrotizing myopathy in children are very rare, and IBM is not seen. Like adults, children may have more than one autoimmune disease or overlapping diseases.

The first sign of JDM 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 JDM have many of the same rashes as adult DM patients, including red rashes on the cheeks and face, anterior neck, forearms and thighs, and dilation of capillaries around the fingernails. Children with juvenile polymyositis do not experience these skin symptoms.

JDM patients can have weak muscles at the same time they see the skin rash, or the weakening muscles may develop after the rash over days, weeks, or months. The weaker muscles are usually those closer to the body, in the neck, shoulders, back, and hips. The child may have 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. Children with developing DM can feel miserable.

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

Some children may develop calcinosis, hardened lumps or sheets of calcium under the skin. Contractures can also occur in which the muscle becomes shortened, causing the joint to stay flexed. Exercising the muscles and joint range of motion can prevent contractures.

# 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's 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's 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 physician is trained to recognize the signs and symptoms of muscle malfunction.

## Medical and family history

Physicians usually begin the process of deductive reasoning through a medical history, and you are a partner in this process. Your input provides some of the most important clues to what may be causing your symptoms. It's important to be as complete as possible, therefore, as you respond to questions about your health in general, and the details of your personal and family health histories.

The details of your symptoms are also important clues: when you first saw signs of the skin rash, what made you first notice muscle weakness, whether you did anything to treat these symptoms yourself, whether there are certain things (foods, activities, weather) that make the symptoms better or worse, whether you had an infection or other illness around the time the symptoms started, or whether you used particular medications such as statins. All of these clues can point toward a direction for further investigation.

## **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 sIBM. Observation of weakness in finger flexor muscles is thus an important clue to the possibility that a patient may have sIBM.

Similarly, polymyositis (PM) and dermatomyositis (DM) involve muscles closer to the trunk, for example shoulder, hip, back, and abdominal muscles. Patients with DM also have mild to severe characteristic rashes that help your doctor to distinguish DM from PM. The physician assembles such observations (“findings” in medical speak, “clues” in Holmes’s 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. So 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. Finally, a blood test can be taken to identify “myositis specific antibodies.” These antibodies are becoming increasingly important in diagnosing myositis.

## **Electromyogram (EMG)**

Although few who have experienced this test 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. These are the sounds of electrical signals in the nerves and muscles, and patterns of electrical abnormalities can indicate the presence of an inflammatory disease.

Keep in mind that the EMG is unlike other tests, such as an MRI, in that it is “operator dependent.” In that way, it is like the physical exam in which the physician performing the test interprets the results as the test is being done. This often means that an EMG will need to be repeated if you go to a myositis specialist or other physician for a second opinion. The EMG is also helpful in guiding the physician to an appropriate site for muscle biopsy.

## **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, physicians will perform MRI scans on patients who are suspected to have myositis. This study, too, can provide evidence about selective involvement of muscles. MRI can often indicate where muscle damage is most significant, which can guide the physician when deciding which muscle to target for best diagnostic results when performing a muscle biopsy.

## **Muscle or skin biopsy**

Ultimately, a biopsy of one of the muscles and/or skin 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 that might produce similar abnormalities. Since decisions about treatment hinge on having an accurate diagnosis, the 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 tissue, it is really a rather minimal procedure. For a muscle biopsy, 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.

Similarly for a skin biopsy, the doctor will remove a small piece of affected skin by first numbing the area with an injection of local anesthetic and then removing a small piece of the tissue.

Once obtained, the muscle or skin sample is examined by a physician called a pathologist who has special training in interpretation of biopsies. The tissue sample must be processed and stained with various chemical compounds before it can be interpreted under the microscope. This processing can take a week or more. Occasionally, the information obtained on the biopsy is inconclusive, and a second biopsy is sometimes necessary.

Other diagnostic tests may be done to further clarify the diagnosis or to rule out another disease or condition that has symptoms similar to myositis or that might be present in addition to myositis. It is also recommended that doctors routinely screen newly diagnosed patients for cancer or lung disease (see Complications).

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

## Blood tests

In the process of diagnosing a disease, the physician will need to narrow down the possibilities that arise during the history and physical. There are a number of blood tests the doctor may choose to help this process. The following is a list of some of the blood tests that tell the doctor something about muscle disease or inflammation.

Unless otherwise noted, all of these tests require taking a sample of blood by using a needle. Some blood tests require that you do not eat or drink, except for water, for about eight hours before the test. Because exercise can affect some test results, you might need to limit exercise for several days before the test to avoid falsely high results. Certain medications may also affect the results of these tests, so be sure to check with your doctor if you are taking any type of medication at all, including anti-inflammatory drugs or over-the-counter medications and supplements.

**Aldolase** is an enzyme found especially in the liver and skeletal muscles. When the liver or muscles are damaged, levels of aldolase in the blood will be elevated. Since muscle weakness can be caused by problems with either the nerves or the muscles, this test identifies weakness caused by muscular problems. Aldolase will not change when weakness is caused by neurological problems.

**Antinuclear Antibodies** (also known as ANA) is a screening blood test to determine if you have an autoimmune disease. Normally there should be no ANA detectable in the blood in a healthy individual, so the presence of ANA may indicate some type of autoimmune disease, such as myositis. A positive ANA will not, however, identify the specific disease.

**Creatine Kinase** (also known as CK, or Creatine Phosphokinase [CPK]) is an important diagnostic blood test for myopathies. CK is an enzyme that is especially active in skeletal muscle, heart tissue, and the brain. When muscle tissue is damaged, CK levels in the blood are elevated. Higher levels of serum CK can indicate muscle damage from chronic disease or acute muscle injury. In myositis, it is not uncommon for CK levels to far exceed the upper limit of normal. If the CK test indicates muscle damage, more tests will be needed to find exactly where the muscle damage occurred and why.

CK levels are often used to evaluate the progress of disease after treatment. This is not a reliable measure of disease activity in all cases, however. Myositis patients sometimes wonder why they feel better or worse than their CK levels indicate. Levels may lag behind the improvement or worsening of the disease, and they may be affected by activity or other factors.

**Liver enzymes**, including alanine aminotransferase (ALT or SGPT) and aspartate aminotransferase (AST or SGOT), are enzymes found in many tissues, including muscle and the liver. When muscle tissue is damaged, muscle cells leak these enzymes into the blood so that a blood sample will show increased levels of muscle enzymes.

### **Myositis-specific autoantibodies**

In addition to the conventional blood tests above, it is helpful for a patient to be tested for myositis-specific autoantibodies (MSAs). These antibodies, and others called myositis-associated autoantibodies (MAAs) have been found to be present in about 50-70 percent of myositis patients. Most have excellent sensitivity and almost never occur unless a patient has one of these diseases. They assist in confirming the diagnosis and may also be associated with complications (see below). Because patients who have the same autoantibodies follow certain clinical patterns, the presence of a particular MSA or MAA offers some insight into the possible course of the disease and how to treat it.

Table 1 shows the myositis-specific autoantibodies that are currently known and the clinical features they are associated with. Be sure the panel your doctor orders includes all of these MSAs.



**Table 1.**

## MSAs and clinical features associated with them

Autoantibodies	Clinical features
Anti-ARS	AS syndrome with moderate to severe muscle weakness, high muscle enzyme levels, RP, mechanic's hands, fevers, arthritis, and ILD
Anti-Jo-1	Chronic continuous disease course, with clinical symptoms for > two years after diagnosis; mean five-year survival rate = 65%, usually due to ILD; AS syndrome features
Anti-PL-7	AS syndrome with higher frequency of ILD
Anti-PL-12	AS syndrome with higher frequency of ILD
Anti-EJ	Dermatomyositis and ILD
Anti-OJ	Myositis and ILD
Anti-KS	ILD with less myositis
Anti-Ha	NA
Anti-Zo	Myositis and ILD
Anti-SRP	Acute onset necrotizing myopathy with severe weakness, high CK, cardiac involvement; treatment resistant
Anti-Mi-2	Adult DM and JDM with hallmark cutaneous disease, milder myositis with good response to treatment
TIF1-γ (Anti-p155/140)	CAM in adult DM; severe cutaneous disease in adult DM and JDM
Anti-SAE	Adult DM; may present with CADM first
Anti-MDA5 (anti- CADM-140)	CADM; rapidly progressive ILD
Anti-NXP-2	Predominantly JDM with subcutaneous edema, calcinosis, and a severe muscle phenotype with contractures; increased risk of cancer in some adult DM studies
Anti-HMGCR	NM; associated with statin use in adults; severe proximal muscle weakness; partially responsive to immunosuppressive medications; better response to IVIg
cN-1A (Mup44, NT5c1A)	IBM and other forms of myositis; higher mortality risk in IBM, more severe disease in JDM; occurs in 40-60% of IBM patients

ARS = aminoacyl-tRNA synthetase; AS syndrome = antisynthetase syndrome; RP = Raynaud's phenomenon; ILD = interstitial lung disease; SRP = signal recognition particle; TIF1-γ = transcriptional intermediary factor 1-gamma; NXP-2 = nuclear matrix protein NXP-2; SAE = small-ubiquitin-like modifier activating enzyme; MDA5 = melanoma-differentiation associated gene 5; CAM = cancer-associated myositis; CADM = clinically amyopathic DM; NA = not applicable/no data.



## Treatment and disease management



Myositis diseases vary widely from one patient to another, so no one treatment will work for everyone. There are a number of medications available that may be used individually or in combination to treat most forms of the disease. Sometimes, some trial and error may be needed to settle on the best treatment plan, and the risks of serious side effects must often be balanced against the therapeutic benefit of these powerful drugs. Regrettably, for some forms of myositis, particularly IBM, there are no fully effective treatments available yet.

Because myositis and its treatments are so variable, it is also extremely important that you communicate well with your physician about your treatment, its effectiveness, and any side effects you may experience.

Keep in mind that medications are only one step in treating myositis diseases. There are many other therapies that patients find helpful and are recommended to assist you to live life to the fullest, despite the challenges of chronic disease.

### Medications

For most patients, treatments can effectively control and improve symptoms, and a small minority of patients can become symptom free even without medication over time. Ongoing research continues to add new medications for treating myositis diseases, which gives patients and physicians a choice in deciding what medication is best based on the balance of risks and benefits for that individual.

While most of these medications are not approved for myositis by the FDA, they are all approved for other uses and are used “off label” in myositis. Nevertheless, their use in myositis is based on research, including small clinical trials, expert experience, and individual reports in the literature of effectiveness in myositis.

## **Corticosteroids**

**Corticosteroids**, also called adrenal corticosteroids, glucocorticoids, or simply steroids, are commonly used as first-line treatment with fairly rapid results in DM, PM, NM, and JM. Corticosteroids, such as prednisone, slow the body's immune system and stop the inflammatory attack on muscle, skin, and other body systems. These medicines control the inflammation, ease pain, and increase muscle strength.

The specific dose varies from patient to patient, but doctors tend to prescribe relatively high doses (40-80mg/day) for an extended period of time (sometimes months) and decrease the dose very slowly as the symptoms improve. Doses depend on the patient's weight and disease severity. Prednisone can be given orally or intravenously. Often, doctors begin patients, especially children and those with severe symptoms, on high-dose intravenous corticosteroids for a few days followed by regular oral doses.

While corticosteroids are very effective, they come with frequent severe side effects, some of which can become permanent conditions, especially with high doses and long-term treatment. They include brittle bones (osteoporosis), cataracts and glaucoma, stomach upset, weight gain, mood swings, diabetes, adrenal insufficiency, a condition known as Cushing's syndrome (round red face, a fatty lump between the shoulders, abdominal weight gain, acne), and other symptoms. Long-term use of corticosteroids in children may cause problems with growth and development.

To reduce these complications, doctors try to taper the dose as quickly as possible while still effectively combating the symptoms. Nevertheless, this tapering usually takes months and should not be rushed, because tapering too quickly can cause symptoms to flare. Many physicians will start patients on another anti-inflammatory medication while they taper off prednisone to avoid causing symptoms to flare.

In some cases, patients may not be able to stop taking corticosteroids completely. Many myositis patients must continue to take low doses

(5-10 mg) of prednisone a day to control their symptoms. This low dose can be maintained indefinitely with few severe side effects.

You should never discontinue or reduce your dose of corticosteroids without checking with your physician. Tapering or slowly lowering your dose is essential to allow your body to begin to produce cortisol again on its own.

**Acthar gel** is a synthetic, long-acting form of corticotropin hormone, also known as adrenocorticotrophic hormone (ACTH). It is injected either subcutaneously (under the skin) or intramuscularly (into the muscle) to treat DM and PM. It is the only drug that is approved by the FDA for treating myositis diseases.

Corticotropin is a naturally occurring hormone produced in the pituitary gland and causes the release of cortisol from a patient's adrenal glands, thus mimicking the effects of taking corticosteroids by mouth. ACTH may also interact at a number of receptors throughout the immune system and may reduce immune overactivity responsible for causing DM and PM.

Patients taking Acthar gel should be monitored closely for side effects, which are similar to those found in long-term corticosteroid use.

## **Immunosuppressants**

Immunosuppressants are drugs that inhibit or prevent the activity of the immune system. In addition to treating many autoimmune diseases, they are also used to prevent organ transplant rejection, as chemotherapy for cancer treatment, and as treatment for other conditions.

In treating myositis diseases, immunosuppressants are often introduced as second-line medications or used in combination with corticosteroids. This allows patients to have better improvement, taper off the steroids more quickly, and avoid some of the unwanted side effects. The choice of which immunosuppressant is used depends on the patient's symptoms and what medications target those symptoms. Different drugs may be added or changed, depending on how well symptoms are being controlled.

Keep in mind that all immunosuppressants carry an increased risk of infection, because they dampen the body's response to attacks from microorganisms such as bacteria and viruses. *You will need to try to avoid getting an infection and report signs of infection to your doctor immediately.* (See Complications.)

The following immunosuppressants are currently used to treat myositis diseases.

**Methotrexate** was developed as a cancer treatment in the 1940s, but it has since become one of the most common medications used, in much smaller doses, to treat rheumatoid arthritis, another autoimmune disease. It can be taken orally (in pill form or liquid) or by injection (either under the skin or into the muscle). Symptoms may take two to three months to resolve. Based on a large randomized trial in JDM and other studies, methotrexate is now given as part of first-line treatment to patients with moderate to severe JDM. Methotrexate is not intended to be taken every day; most people take it only once a week at a dose of 15-25mg/week. Taking this medication every day may cause severe side effects, and overdose can be fatal. Some physicians avoid methotrexate for myositis patients who also have ILD, because some studies have indicated there is a very small risk of pulmonary fibrosis.

**Azathioprine** (Imuran) is also used to treat rheumatoid arthritis and as an antirejection medication for people who have had an organ transplant. In myositis, it is usually taken orally, and is best taken with food to avoid stomach upset. Doses are usually started at 50mg twice daily, then increased by 50mg every two to four weeks. It may take up to three or four months for symptoms to improve. This medication also has an effect on the blood clotting mechanism in the body, so you may notice that you bruise more easily, and it may take longer for a wound to stop bleeding. *Contact your doctor immediately if you have bleeding that doesn't stop.* Azathioprine may be a better choice for some patients who have ILD. It is also suggested that patients be tested for the enzyme thiopurine methyltransferase (TPMT) before starting this medication as some people have a genetic deficiency that places them at great risk of developing severe, potentially life-threatening bone marrow toxicity.

**Mycophenolate mofetil** (CellCept) is also used to prevent rejection in organ transplant patients and to treat other autoimmune diseases. In myositis, it is usually taken orally at a dose of 250-500mg twice per day, increasing gradually to a dose of 2,000-3,000mg per day. Patients with kidney problems should use a lower dose. Mycophenolate mofetil has been used with good results in patients who have interstitial lung disease and in difficult-to-treat DM skin disease, especially in combination with prednisone or other immunosuppressants.

**Cyclosporine** is used to prevent rejection in organ transplant patients and to treat other autoimmune diseases. It is used as a second- or third-line treatment in myositis, especially for patients who have ILD. It is taken orally.

**Tacrolimus** (Prograf) is a drug that was developed as an antirejection medication for organ transplants. It is used as a second- or third-line treatment in myositis, especially for patients who have ILD. It is taken orally.

**Cyclophosphamide** (Cytoxan) was developed as a chemotherapeutic agent to treat cancer. It is also used as an antirejection medication for organ transplants. Because it can be fairly toxic, its use in myositis is limited to very difficult cases with ILD, gastrointestinal or skin ulceration, or other severe patients resistant to other medications. It is given orally or intravenously.

**Hydroxychloroquine** (Plaquenil) is an antimalarial drug that also has anti-inflammatory qualities and is used to treat certain autoimmune diseases, including rheumatoid arthritis and Sjögren's disease. It can be a helpful adjunctive medication in myositis, especially as part of the treatment for DM skin rashes. It is taken orally.

## **Immunoglobulin**

Immunoglobulin is a blood product derived from large pools of donated human plasma that contains the part of the blood that has antibodies. It is usually given intravenously (IVIg), but it can be given subcutaneously (SCIg) (under the skin). Doses are based on the patient's weight, initially with a dose of 2g/kg given over 2-5 days,

then 1g/kg/month for several months, then tapered depending on the response.

Because immunoglobulin is very expensive, it is usually reserved for cases that are resistant to other treatments. However, it is often especially effective for patients with DM skin symptoms that do not respond to other treatments. Some physicians also use immunoglobulin alone as first-line therapy in patients who have NM with the anti-HMGCR autoantibody or as part of first-line therapy for moderate to severe juvenile myositis, including patients with severe swallowing troubles.

Some physicians use IVIg to treat sIBM, although the effectiveness of this is questionable. Nevertheless, some patients who have dysphagia (swallowing problems) do seem to benefit.

## **Biologic agents**

Biologic agents are different than drugs. While drugs are created chemically, biologics are created biologically. Hormones, insulin, and red blood cell-stimulating products are examples of biologic agents that have been used in medicine for decades. Recent technology has provided the means to create other very large, complex molecules that target specific parts of the immune system.

Keep in mind that all biologic agents used to treat myositis diseases suppress the immune system's response to germs (bacteria, viruses, and other organisms) and therefore place the patient at an increased risk for serious infection. Biologic agents may also increase the risk of developing certain types of cancer, especially in older adults.

A number of biologic agents are currently being studied for use in treating myositis diseases.

**Rituximab** (Rituxan) was developed as a treatment for certain types of cancer, but it has been approved for use in rheumatoid arthritis and other autoimmune diseases. It has shown promise in treating myositis diseases, especially for those whose disease does not respond well to other treatments. Patients with anti-SRP necrotizing myopathy,



antisynthetase syndrome, or interstitial lung disease, which can be resistant to other medications, are more likely to respond to rituximab.

Rituximab is not used as first- or second-line treatment. It is given intravenously, with a second dose in two weeks. It can take up to three months for symptoms to begin to respond to rituximab, and it continues to have an effect in the body for as long as 12 months or more. If rituximab does seem to be effective, these doses can be repeated every 6 to 18 months.

## **Exercise and physical therapy**

Exercise and physical therapy are important parts of standard myositis treatment plans. Physical exercise has been shown to reduce inflammation, reduce fatigue, increase stamina, and build muscle, even in patients with myositis.

There is a strong association between aerobic capacity and general health, both in healthy individuals and those with myositis. Regular physical activity and exercise can improve one's quality of life and reduce the risk of serious chronic diseases, such as type II diabetes, osteoporosis, hypertension, and cardiovascular disease. These are all complications of myositis diseases or their treatment, so exercise is doubly important.

## **Myositis and the muscles**

One of the reasons myositis patients feel fatigued is because their muscles are weakened by the disease. Movement relies on many muscles working together, so when one muscle or muscle group becomes weak, other muscles must work harder to compensate. This causes fatigue.

It is natural to want to avoid movement when muscles are painful and fatigued. Sometimes people are even afraid that exercise will cause damage or increase the progression of the disease process. But avoiding movement only makes the pain and weakness worse.

Muscles that are not exercised regularly become weaker and can even atrophy (waste away). Muscles will begin to lose strength within

24-48 hours of inactivity. Maximum muscle strength is lost within the first six weeks. The only way to recover muscle strength is by using your muscles. If muscles atrophy, they may not be built back up again.

That's why exercise is so important. Exercise keeps weak muscles from atrophying and helps the body compensate for weakness by strengthening the surrounding musculature.

Stretching is also important, especially for patients with fixed weakness. Active stretching by the patient and passive stretching, in which a care partner moves the limbs for the patient, can maintain mobility.

### **Starting an exercise program**

1. Have your physician clear you for exercise. Do not start exercising if you are currently in a severe or worsening inflammatory flare.
2. Seek expert guidance from a physical therapist who understands myositis diseases. Have the therapist work with you to develop an individualized treatment plan.
3. With the guidance of the therapist, learn how to do the exercises correctly. After that, you can probably do them on your own.
4. Each person must find the proper exercise program that is right for them. Exercise should be designed to stretch joints, build aerobic endurance, and strengthen muscles and muscle groups that are affected by your disease process, while protecting the muscles that are not affected.
5. The proper program means you will notice improved efficiency with essential daily activities and be able to perform those activities without overuse of involved muscles. If you find you are working much harder to perform normal, everyday functional activities, the exercise is probably causing undue stress to involved muscle groups, and you should consult with your physical therapist to see how best to change your program.

6. If your physical or occupational therapist is unfamiliar with myositis and the exercise needs of myositis patients, there is information for these professionals available on The Myositis Association's website ([www.myositis.org](http://www.myositis.org)).

## Complementary and self-care therapies

According to the NIH's National Center for Complementary and Integrative Health (NCCIH), more than 30% of Americans—including many with myositis—use health care approaches that have been developed outside of conventional Western medicine. Most people use these alternative or traditional therapies in combination with mainstream methods for a complementary approach.

While many of these methods and products have not been rigorously tested for safety and effectiveness in the same way that drugs are tested, and while others have been found to be ineffective or shown mixed results, many have few risks and significant traditional support to recommend them.

If you are considering incorporating complementary or non-conventional approaches into your treatment regimen, it is important to discuss this with your medical doctor (rheumatologist, neurologist, dermatologist, primary care provider), so they can see the whole picture of what might be influencing your symptoms and recovery. And you should always take your conventional medications as prescribed; never stop or change these medications without first consulting your doctor.

While TMA cannot vouch for the effectiveness of any complementary treatment, TMA members offer the following suggestions about complementary practices for your consideration. Please keep in mind that many of these ideas are not prescriptive, and some people may find them more valuable than others.

### Diet and nutrition

**A healthy diet** can go a long way toward improving overall health. Some TMA members have found that their symptoms are greatly

improved when they adopt special diets such as paleo or vegan. While these eating plans may take some special commitment, the anti-inflammatory plan (sometimes referred to as a Mediterranean diet) is easily adaptable and is one that all people, but especially those with an autoimmune disease, can benefit from. It includes the following:

- Avoid processed and fast foods, including those with high fructose corn syrup, artificial ingredients, preservatives, and pesticides. Instead, opt for a wide variety of brightly colored fresh fruits and vegetables and unrefined foods.
- Reduce the number of foods made with wheat flour and sugar, especially bread, pasta, and most packaged snack foods. Choose instead foods containing whole grains, such as brown rice and bulgur wheat.
- Decrease intake of saturated fat by eating less animal fats and products made with palm kernel oil.
- Use extra-virgin olive and expeller-pressed canola, sunflower, and safflower oil. Do not use partially hydrogenated oils.
- Include avocados and nuts, especially walnuts, cashews, almonds, and nut butters made from these nuts.
- Increase omega-3 fatty acids in your diet by eating salmon, sardines, herring, black cod, omega-3 fortified eggs, hemp and flax seeds, or take a fish oil supplement.
- Eat more vegetable protein, especially from beans and soy, and choose fish, cheese, and yogurt more often than you choose animal proteins.
- Avoid drinking soda—including diet sodas—and choose tea instead of coffee.
- If you drink alcohol, red wine is preferable.
- Eat chocolate (in moderation) with a minimum of 70% cocoa.

### **Special dietary considerations for those taking corticosteroids.**

People who must take corticosteroids (prednisone), especially in high doses and for longer periods of time, face a number of complications that carry considerations for what you eat.

- Prednisone increases appetite. To avoid weight gain, avoid high-calorie foods and eat frequent small meals to help maintain steady blood sugar levels. Getting plenty of exercise will also help.
- To reduce the risks of high blood pressure and fluid retention, limit salt intake to less than 1,500mg per day. You can do this by eating fresh rather than processed or canned foods and avoid adding salt to food.
- If you take medication for high blood pressure, you may need to increase intake of foods high in potassium, such as bananas, apricots, cantaloupe, baked potatoes, and tomatoes.
- Prednisone can also irritate the stomach, so it is important to take it with food, not on an empty stomach.
- Diabetes is also a risk when taking corticosteroids. To keep blood sugar levels within the normal range, avoid foods high in simple carbohydrates like sugar, and keep carbohydrate intake to between 45 and 60 mg per day.

**Dietary supplements** have a wide range of products and recommended uses. The following specific recommendations are offered for those who have myositis diseases:

- **Calcium** is a concern for those who take prednisone. You should eat foods rich in calcium, such as milk, yogurt, cheese, leafy green vegetables (spinach, kale, bok choy), almonds, and broccoli. Calcium supplements may also be needed to minimize bone loss and osteoporosis.
- **Vitamin D** is a hormone produced in the skin in response to sunlight. It is important in calcium absorption and many other processes. Because most people do not spend enough time in the sun (and DM patients should not spend time in the sun), most people need to take dietary supplements of Vitamin D.
- **Folic acid** (also called folate) is a B vitamin that is abundant in leafy green vegetables, such as spinach, kale, broccoli, and other sources. Because methotrexate interferes with the way folic acid is used in the body, those who take this anti-

inflammatory medication need more folic acid than can be consumed from dietary sources, so they should take folic acid supplements.

- **Omega-3** fatty acids, which are anti-inflammatory, and omega-6 fatty acids, which are pro-inflammatory, should be in balance in the body. Most Americans, however, eat far more omega-6 foods (vegetable oils, safflower oils, meat, poultry, and eggs), causing a more pro-inflammatory state. To bring this back into balance, you should eat more foods containing omega-3 fatty acids, including salmon, mackerel, sardines, leafy green vegetables, flaxseed, canola oil, walnuts, and enriched eggs. Omega-3 supplements are also available.

## Sun protection

Protecting the skin from too much sun exposure is recommended for all people, especially those with light skin, in order to prevent skin cancer. Those who live with DM, however, must be especially vigilant about avoiding sun exposure to the skin. For about 20% of DM patients, sun exposure, even for brief periods, can cause symptoms to flare. TMA members recommend the following extreme sun protection measures:

- Always wear sunscreen with SPF 50 minimum, even indoors and year round. Apply it to every inch of exposed skin.
- Repeat application of sunscreen 30 minutes before going out of doors, and reapply often.
- Apply sunscreen even to your hands, and be sure to reapply every time you wash your hands.
- Wear hat, sunglasses, gloves, long sleeves, and long pants/skirt when outside, even when driving.
- Wear double layers or clothing that is specifically designed to be sun protective.
- Be aware of unexpected places where reflected UV rays from the sun may also affect your skin: moonlight, metallic rocks (such as mica), water, passing cars, and windows and other glass.

- Be aware that glass does let UV rays pass through, so shades and heavy curtains or special window coatings are recommended for home and office.
- Light bulbs emit UV rays, so LED lightbulbs are recommended for use everywhere.
- Avoid going outside between 11 am and 4pm.
- Use a chemical with your laundry detergent that has UV protection included, called RIT sunguard.

## **Mind and body practices**

Mind and body practices include a variety of procedures or techniques administered or taught by a trained practitioner or teacher. The amount of research on these approaches varies widely depending on the practice. For example, many studies have been done using acupuncture, yoga, spinal manipulation, and meditation, but there have been fewer studies on some other practices.

TMA is unaware of scientific studies related to the effects of various mind and body practices on myositis specifically, and TMA does not specifically endorse any of these practices. Some TMA members, however, have found one or more of these approaches to be helpful.

Keep in mind that, when trying to decide which complementary approaches to use, most patients find the process to be an ongoing learning experience with lots of trial and error. There is also a great deal of variation in technique and effect from different practitioners of these approaches.

- Acupuncture
- Chiropractic and osteopathic manipulation
- The practices of traditional healers
- Ayurvedic medicine
- Traditional Chinese medicine
- Homeopathy
- Naturopathy

**Stress reduction and adequate sleep** affect more than your mood. Both have been shown to have a significant impact on your physical health, including immune system function. For people who live with myositis, high levels of stress and lack of sleep can cause flares in muscle weakness and pain and in skin symptoms. Limiting stress and getting enough sleep and rest are, therefore, therapeutic.



## Complications



Most autoimmune diseases are complex, multifaceted conditions that have no simple solutions. In addition to the symptoms of DM, PM, NM, sIBM, or JM, each myositis patient may experience other symptoms that don't appear to be part of the disease itself.

The following are some of the complications that may occur together with or because of myositis.

### Antisynthetase syndrome

Antisynthetase syndrome is a set of symptoms that typically occur for patients who have myositis together with one of several specific autoantibodies known as antisynthetase antibodies (see Table 1). These autoantibodies are immune system proteins that target and attack tRNA synthetase enzymes. You can find out if you have one of these autoantibodies through a blood test that screens for a number of myositis-specific autoantibodies.

**Symptoms** associated with antisynthetase syndrome include the following:

**Interstitial lung disease (ILD)** – As many as 90% of those with antisynthetase syndrome have interstitial lung disease. (See page 37 for more about ILD.)

**Muscle inflammation** – Antisynthetase syndrome typically affects those with adult or juvenile dermatomyositis or polymyositis.

**Inflammatory arthritis** – Patients may experience pain, stiffness, swelling, redness, or warmth in and around joints, often the small joints of the hands. It is unusual, however, to experience the sort of joint deformity seen in rheumatoid arthritis.

**Fever** – About 30% of patients with antisynthetase syndrome will experience fever that is unrelated to infection or other causes.

**Raynaud’s phenomenon (or syndrome)** – This is a condition in which spasms of the arteries cause episodes of reduced blood flow, typically involving the fingers and toes. Nose and ears can also be affected. These episodes are usually triggered by cold or stress. They can last several minutes to several hours and can cause the affected part to feel numb and cold and to turn white or blue.

**Mechanic’s hands** – This is a condition that involves thickened, dry, cracked skin on the sides of the fingers and palms, which can be painful. The term “mechanic’s hands” is used, because the condition makes the hands look rough and dirty, resembling those of a manual laborer.

**Treatment** of antisynthetase syndrome is similar to treatments recommended for other inflammatory myositis diseases. Early and aggressive treatment with corticosteroids (prednisone) is especially important with this condition. Physicians also often start second-line therapy together with steroids right from the beginning.

Patients with ILD may need a multidisciplinary approach to their care that includes regular visits with a pulmonologist and monitoring of the lungs with chest CT and pulmonary function tests.

Antisynthetase syndrome can go into remission, especially for those with milder lung disease, but patients are more likely to remain symptom free while continuing treatment. Flares tend to occur when medications are tapered too rapidly.

## **Calcinosis**

Dystrophic calcinosis is the abnormal collection of calcium salts in or under the skin and in the fat tissue, muscles, or tendons, even when levels of calcium in the blood are normal. It occurs in some patients with DM. These lesions may also appear in patients with overlapping autoimmune diseases, including systemic sclerosis, systemic lupus erythematosus, and mixed connective tissue disease.

Calcinosis appears more often in juvenile dermatomyositis, where as many as 40% of JDM patients may be affected. In adults with myositis, about 20% of patients report this complication. Calcinosis usually appears within the first three years after diagnosis, however, it is sometimes the first symptom to appear. Other times, it doesn't occur until many years after diagnosis.

Calcinosis appears as hard, irregular nodules (lumps) or linear deposits along tendons and muscles in or under the skin in any area of the body. These lumps can be especially uncomfortable when they appear on the face, around joints, or on pressure points, such as the buttocks, feet, or wrists. The nodules can cause functional disability, contractures, skin ulcers, and pain. Needless to say, calcinosis can have a significant negative impact on the patient's quality of life.

Why these nodules develop is still not well-understood. It is hypothesized that tissue damage from inflammation and the blood vessel changes of DM may lead to these abnormal mineral formations in the skin.

**Treatment** of dystrophic calcinosis can be challenging. There is no treatment that is effective for everyone. Inadequate initial treatment of DM may play a role in the development of calcinosis lesions. Therefore, early and aggressive treatment of the underlying muscle and skin disease with immunosuppression is strongly recommended.

In addition, increasing blood flow to the extremities, through smoking cessation, decreasing stress, and limiting exposure to cold, may be helpful. Appropriate sun-protection is always important in DM, but it may also prevent calcinosis, since sun exposure can stimulate the immune system, which may contribute to calcinosis. Calcinosis occasionally clears on its own with no intervention.

Smaller lesions may respond to medications such as ceftriaxone and intravenous immunoglobulin (IVIg). Larger lesions have been treated with varying degrees of success with medications such as diltiazem, bisphosphonates, probenecid, and aluminum hydroxide.

Surgical excision may be used to remove smaller lesions that are confined to a single area, especially those in painful or troubling areas

of the body. Unfortunately, the nodules often grow back after surgical removal. Indeed, sometimes surgery seems to stimulate calcinosis to grow back. Having the dermatomyositis under good control may help minimize calcinosis return.

## Cardiovascular disease

The heart is a muscle, so just as myositis causes inflammation in other muscles in the body, inflammation can, in some cases, occur in the heart muscle. Recent research suggests that, in a myositis patient, the process of inflammation in the heart occurs in the same way it does in skeletal muscles. Other recent research findings have identified several autoantibody biomarkers, including anti-signal recognition particle (SRP) and anti-mitochondrial antibody (AMA), that are associated with severe heart disease in myositis patients. And recently developed immune checkpoint inhibitors used in cancer treatment have been shown to cause inflammatory diseases, including myositis and myocarditis.

Inflammation in the heart muscle (called myocarditis) can lead to fibrosis (scarring). Myositis patients can develop a number of cardiovascular problems as a result of inflammation and fibrosis, including cardiomyopathy (weakened heart muscle), arrhythmias (irregular heart beat), congestive heart failure (fluid build up around the heart and lungs), and atherosclerosis (hardening of the arteries).

While not all myositis patients develop cardiac complication, all myositis patients should be evaluated regularly for heart disease, especially if they have risk factors.

**Treatment** of cardiovascular disease in myositis is best when the plan of care is coordinated between the rheumatologist or neurologist (who attends to the myositis side of the disease) and the cardiologist (who attends to the heart disease).

Treatment of the underlying myositis is of primary importance. Immunosuppressant medications are used to treat both myositis and cardiovascular inflammation. There are, however, some special considerations that must be addressed in the treatment plan for myositis with cardiovascular involvement.

Management should also include addressing those cardiovascular risk factors that can be modified, such as quitting smoking, controlling diabetes and high blood pressure, and addressing high cholesterol levels.

## **Cholesterol, heart disease, and myositis**

Treating elevated cholesterol levels with statin medications (atorvastatin [Lipitor], fluvastatin [Lescol, Canef, Vastin], lovastatin [Mevacor], pitavastatin [Livalo], pravastatin [Pravachol or Selektine], rosuvastatin [Crestor], and simvastatin [Zocor]) is a topic of debate within the myositis community. Patients should, therefore, consult with their myositis physician before they start this treatment.

Lowering blood cholesterol levels is an important factor in reducing the risk of cardiovascular disease, and statins are widely used and effective in lowering cholesterol levels and therefore the risk of heart attack. One of the common side effects of these drugs, however, is muscle weakness and pain, similar to that experienced by myositis patients. Most times, these symptoms go away once the patient stops taking the medication. In a small number of patients, however, these symptoms do not resolve, even after the medication is stopped.

It is now clear that there is a causal relationship between statin use and inflammatory myopathy. An autoantibody to HMG-CoA reductase (HMGCR), the target of statin medications, has been found to be a myositis-specific biomarker for NM, suggesting statins trigger this immune-mediated myopathy.

Not all cases of NM are related to statin use, and not all myositis patients who have the HMGCR autoantibody have taken statin medications. However, myositis patients who have the HMGCR autoantibody or who have had worsening symptoms while taking statins should not take these medications.

## **Dysphagia**

Dysphagia is not specific to myositis; there are many reasons why someone might have trouble swallowing food or fluids. For the myositis patient, dysphagia is usually caused by weakness in the

muscles of the esophagus. It occurs in about one-third of myositis patients.

Dysphagia can occur in all forms of myositis, however it is most common in patients with sIBM and JDM. Especially in sIBM patients in whom symptoms progress very slowly over several years, dysphagia may be the first sign of the disease.

Dysphagia is a very serious complication because it can lead to aspiration pneumonia, caused by bits of food or fluid “going down the wrong pipe” and entering the lungs. Because eating and drinking are so difficult for those who have dysphagia, they may also suffer from inadequate nutrition or dehydration and may start to lose weight unintentionally.

There are many different causes of dysphagia, and treatment is guided by the cause. It is important, therefore, for dysphagia to be fully evaluated by a speech pathologist to be sure that myositis is the actual cause.

## **Infection**

The immune system is a complex system comprising many biological structures and processes that protect the body against disease. When a foreign protein, such as a virus or bacteria, is detected in the human body, a whole cascade of processes is activated that are intended to deactivate and remove that invader.

For reasons that are still not well understood, some people develop autoimmune diseases in which this process is activated against the person’s own cells. In order to treat autoimmune diseases, like myositis, doctors attempt to suppress some of these immune system processes with medications. Suppressing the immune system, however, places the patient at an increased risk for developing a serious infection.

For the vast majority of myositis patients, long-term immunosuppression is the only treatment for their muscle and skin symptoms. As a result, myositis patients must always be aware of prevention, early detection, and aggressive treatment of infection.

## Signs of infection

Immunosuppression reduces the body's normal response to infection. Therefore, typical signs of infection, such as fever, will be weakened and delayed. Consult your primary care physician immediately if any of the following symptoms develop:

- Frequency or burning with urination
- Signs of respiratory infection such as coughing, sneezing, nasal discharge, sputum production, fever, aching muscles or joints, malaise
- Diarrhea, vomiting, fever, malaise
- Signs of skin infection, especially at the site of a cut or injury, such as redness, swelling, warm to touch, discharge, fever

## Preventing infection

Prevention is by far the best treatment for infections. Patients and their care partners should always practice these infection prevention measures:

**Wash your hands.** Nothing prevents the spread of infection better than vigorous, frequent hand washing.

**Pay attention to skin wounds.** When you get a cut or scrape, wash the wound immediately with soap and water, and apply a bandage with antibiotic ointment. See your doctor if the area develops redness, swelling, foul-smelling discharge, or doesn't show signs of healing.

**Avoid contact with others who are sick.** This may include avoiding crowds and close public spaces. It also includes using safe sex practices.

**Avoid tick and mosquito bites,** which may transmit infections such as Lyme disease and West Nile virus. Use insect repellent and cover your skin when out of doors during the summer, especially in the evening.

**Avoid foods that may carry bacteria**, including raw eggs, unpasteurized milk, and insufficiently cooked or raw meat. And always wash raw vegetables before eating them.

**Be aware of unsafe drinking water**, especially if you travel outside the US. Drink only bottled water while away, use it for rinsing your toothbrush, and don't eat or drink anything that has not been cooked or peeled.

**Discuss travel-related infections** with a specialist if you plan a trip to developing countries where certain infectious diseases, such as yellow fever and malaria, are prevalent.

**Vaccines** are another way to prevent certain infections. A vaccine is a biological preparation that provides active acquired immunity to a particular disease. A vaccine often contains weakened or killed forms of the bacteria or virus that causes the disease. The vaccine stimulates the immune system to recognize that microorganism as a threat, to destroy it, and to recognize and destroy that microorganism if it encounters it in the future.

Patients with a suppressed immune system should never be given vaccines made with live or attenuated organisms, because it could cause the patient to become sick with the disease. The shingles vaccine and the nasal flu vaccine are live attenuated vaccines.

Immune suppressed patients can get inactive vaccines, which include the flu shot (but not the nasal flu vaccine) and pneumococcal, meningococcal, and pertussis vaccines. Care partners and others living with the patient should also be immunized.

## **Other considerations**

Prior to starting immunosuppressive treatment, it is recommended that patients be tested to see if they have tuberculosis (TB). If immunosuppression is started before the TB test is done, this treatment will also suppress the response to the TB test. If TB is present and untreated, this infectious disease may flare and spread to other areas of the body when immunosuppressive medications are started.



It is also recommended that testing for myositis specific antibodies (MSAs) be done prior to starting immunosuppressive treatment.

Patients with lung disease or those receiving high doses of prednisone, often in combination with other immunosuppressants, are at greater risk for developing an opportunistic infection called pneumocystis jiroveci pneumonia because of immunosuppression. These patients may be prescribed the antibiotic Bactrim to prevent this opportunistic infection of the lung.

## Interstitial lung disease

Interstitial lung disease (ILD) is a group of diseases that affect the tissue and spaces (interstitial) around the air sacs (alveoli) in the lung. This is the place where oxygen that we breathe in is passed into the blood stream and carbon dioxide passes from the blood to the lungs to be breathed out. When these spaces are obstructed by inflammation, this exchange is impaired. When inflammation is untreated for too long, it can result in pulmonary fibrosis, in which the lungs are scarred causing serious breathing problems.

Except in IBM, interstitial lung disease is the most common and serious complication of the inflammatory muscle diseases in adults. Researchers estimate that 30-40% of myositis patients have some form of lung disease.

There is also a strong association between interstitial lung disease and antisynthetase antibodies. Of those patients who have interstitial lung disease, about 75% have anti-Jo-1 antibodies. Autoantibodies to MDA5 and PM-Scl (an antibody prevalent in polymyositis/systemic scleroderma overlap) are also associated with interstitial lung disease.

**Symptoms** can be variable, including shortness of breath, cough (usually a dry cough with no sputum), or no symptoms at all. Symptoms usually progress slowly, but respiratory distress can also occur quickly. Interstitial lung disease can appear before muscle symptoms become apparent. In addition, the severity of muscle or skin disease is not necessarily an indication of the severity of lung disease. Muscle and skin symptoms may be mild or even nonexistent, but interstitial lung disease may be severe.

**Treatment** of interstitial lung disease is best when the plan of care is coordinated between the rheumatologist (who attends to the myositis side of the disease) and the pulmonologist (who attends to the lung disease). Ongoing care typically includes the following:

**Immunosuppressive medications** to treat both myositis and lung inflammation. Some medications also have special considerations for lung disease associated with antisynthetase syndrome.

**Antibiotics**, including Bactrim DS, Dapsone, or pentamidine, may be prescribed on a long-term basis to prevent infections in the lung.

**Frequent pulmonary function tests (PFTs)** to monitor disease progression or progress.

**High resolution CT (HRCT) scans** may be done to monitor disease progression or progress.

**Echocardiograms or an ultrasound** of the heart may be done to be sure there is no pulmonary hypertension.

Many patients need **supplemental oxygen**.

## **Overlapping autoimmune diseases**

Autoimmune diseases often express similar symptoms in different conditions. This is one of the reasons it is so difficult to diagnose these diseases. Dermatomyositis and lupus, for example, both have a characteristic rash over the bridge of the nose. Patients with antisynthetase syndrome experience the same Raynaud's phenomenon as those with scleroderma.

Sometimes, however, patients can experience the whole range of clinical symptoms and laboratory findings of two well-defined autoimmune diseases at the same time—even those that are not part of one or the other disease. When this happens, they are said to have overlap syndrome. Myositis overlap syndromes tend to appear more in patients with DM, PM, and NM.

Common conditions that tend to occur together with myositis include the following:

**Systemic sclerosis (scleroderma)** is the most common overlapping disease with myositis. It is a progressive disease of skin and connective tissue (cartilage, bone, fat, and the tissues that support the nerves and blood vessels). It is caused by the accumulation of collagen (a structural protein) in the inner walls of the small arteries that causes skin and the tissues of internal organs to become sclerotic (thickened, hardened).

**Rheumatoid arthritis (RA)** is a systemic autoimmune disease that causes inflammation and deformity in many joints throughout the body. It can also involve other organs, especially in patients who are not treated.

**Systemic lupus erythematosus (SLE or lupus)** is a chronic autoimmune disease that damages tissue in many parts of the body, including skin, joints, lungs, and kidneys. Some of the symptoms of lupus include hair loss, mouth ulcers, swollen lymph nodes, fatigue, and a characteristic red rash that spreads over the cheeks and nose.

**Sjögren's syndrome** is an immune-mediated destruction of the endocrine glands, most prominently the tear and salivary glands, causing dry eyes and dry mouth. Other glands and organs can also be affected.

**Mixed connective tissue disease (MCTD)** is a syndrome that involves a variety of manifestations of several different autoimmune diseases without definitive diagnostic features of any one disease.



# Prognosis



Prognosis for the different forms of myositis varies greatly and often depends on the presence of other conditions, such as interstitial lung disease or certain autoantibodies.

While sIBM is a progressive disease, life expectancy for those with sIBM is usually the same as for those without the disease. In fact, IBM patients don't die from the disease, but from complications (often preventable) that are associated with it. Patients who develop impaired swallowing, for example, are at greater risk for choking and aspiration (inhaling food and fluids into the lungs), which causes pneumonia and may lead to death. Similarly, injuries that occur as a result of falling, such as hip fractures and head injuries, also increase the likelihood of dying.

For DM, PM, and NM, the progression of the disease is more complicated and harder to predict. More than 90 percent of these patients are still alive more than five years after diagnosis. About one-third experience only one period of acute illness in their lifetime; others struggle with symptoms for years or experience relapses or "flares" (waxing and waning of symptoms over time).

One of the biggest problems in treating myositis is obtaining an accurate diagnosis. The average myositis patient visits five doctors over three-and-a-half years before receiving an accurate diagnosis.

A too-frequent cause of early death is not the myositis itself, but infection due to suppressed immunity as a result of treatment. Cancer, most frequently in patients with DM, can also lead to early death.

Your physician may be able to determine more likely complications by knowing your autoantibody status. Complications contributing to mortality include myositis-associated cancer, dysphagia (trouble swallowing), interstitial lung disease, and heart disease.



# Become your own best advocate



Rare disease patients are sometimes in a difficult position with regard to those who are caring for them. Because myositis is a rare disease, many health care professionals, including physicians and physical therapists, do not have a complete understanding of the needs and challenges involved in treating these patients. That's why it's important for myositis patients to be an active partner in their own care and to advocate for themselves to get the care they need.

Here are some pointers from TMA patients and the myositis experts on our Medical Advisory Board:

- **Know your own disease.** Learn everything you can about your own disease and what helps you feel better. Be assertive in asking for what you need. TMA offers resources on its website for helping you become an expert on your disease.
- **Learn more about myositis.** Read as much as you can about your illness, medications, treatments, and the research related to it. Be careful, however, where the information comes from; not everything that is found on the internet is reliable. The TMA website is a valuable source of scientifically based information.
- **Talk to other myositis patients** at TMA's Annual Patient Conference, at TMA support group meetings, and on the TMA Community Forum. These are people who have experienced the same things you are going through and can offer tips and suggestions for dealing with the challenges of this disease. The knowledge you gain will also help you understand what the doctor tells you and help you ask better questions.
- **Help your doctor help you.** Physicians rely on patient input when they make treatment decisions. So it's helpful, for example, if you can notice how your function has changed since the medication started. What can you do today that you

couldn't do two months ago? What other symptoms or side effects have you noticed? Even things that seem unrelated to your disease, such as sleep disturbance or irritability, could be side effects of the treatment.

- **Ask questions when you see the doctor.** It's a good idea to come to office visits with a list of questions or concerns that you want to talk about. Writing them down helps you remember everything you wanted to ask. And if the doctor uses terminology that you don't understand, ask for clarification.
- **Ask for copies of your medical records.** Sometimes the different doctors you see don't communicate with each other about your care. If you keep a copy of all your own medical records, you can share them with all of your physicians so they are aware of all of the findings from all physician exams, lab tests, and procedures.
- **Know your medical history.** Tell your doctor if you have other medical conditions you have been diagnosed with and surgeries you have had in the past. The doctor will also want to know about medical conditions of other members of your family.
- **Know your medications.** Provide a written list of all the medications you are currently taking at each doctor's visit. This should include non-prescription medications, vitamins, and supplements, as well as herbal and homeopathic preparations.
- **Understand your options.** If treatments are discussed, make sure you understand the length of time they will be needed, benefits, side effects, and probability of success for each one. Take notes about the treatment information.
- **Bring someone along to the doctor's office.** These visits can be stressful, and patients often forget much of what the doctor says. Having a friend or care partner there with you can not only be calming, but it adds another set of ears to hear what is said. This person can also take notes for you, so you can refer to them later when additional questions arise.
- **Document your progress.** As your treatment progresses, keep your own written records of tests performed, where they were performed and when, and what the results were.



Take notes on the effects and side effects of medications and treatments. Pay attention to any concern expressed by your pharmacist, and notify your doctor before taking the medication. Write down any change in your health—good or bad.

- **Be a partner.** As a patient, it's best to work together with the doctor as partners for your health. It's your right to understand any tests, treatments, what to expect with the disease, and what you can do to support your own health. Work to form an open, comfortable relationship with your physician. If you find it impossible to communicate, it may be time to look for another doctor.
- **Keep your doctor informed.** Use your written records and notes when you tell your doctor about any side effects or any changes in your health after starting a medication or treatment. If a serious change or complication arises, notify the doctor immediately. Your doctor needs to know your reactions in order to treat you properly.
- **Find out about drug trials.** TMA notifies its members when information about human drug trials becomes available. 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,” meaning they will receive a placebo. Other trials provide the drug to one set of patients at first, then offer the drug to the control patients later in the trial.
- **Visit TMA's online Community Forum.** TMA members often post remarks on the Forum 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.
- **Ask TMA's medical experts.** TMA offers periodic Live Discussions on our website with medical experts. These online chats offer the opportunity for members to submit questions in advance and join the live discussions.

- **Find support.** One of the most important sources of support for those with chronic diseases are others who live with the same disease and understand what you're going through. TMA has a network of support groups and other resources to help you connect with myositis patients.

For more information on these and other disease management topics, visit [www.myositis.org](http://www.myositis.org), or contact TMA at [tma@myositis.org](mailto:tma@myositis.org) or 1-800-821-7356.

## Glossary



**Alternative therapy:** any therapy used in place of or in addition to conventional or more traditional treatments. [See complementary therapy.]

**Amyopathic dermatomyositis:** dermatomyositis that involves the skin only without muscle inflammation.

**Antibody:** a protein produced by the body that acts against an antigen (substances foreign to the body) in an immune response. In a typical immune response, lymphocytes of the immune system identify the antigens as invaders and stimulate the formation of antibodies to react with the foreign cells and render them harmless.

**Antigen:** a 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:** a myositis specific autoantibody that is associated with antisynthetase syndrome.

**Antisynthetase syndrome:** a rare condition involving the presence of antisynthetase autoantibodies in the blood and associated with interstitial lung disease, DM, PM, and other autoimmune diseases.

**Arthritis:** inflammation or swelling of the joints.

**Aspiration:** inhaling or breathing in food or other substances into the lower respiratory tract and lungs. This can lead to aspiration pneumonia, an often serious infection in the lungs caused by these bits of inhaled substances.

**Autoantibodies:** proteins produced by the immune system that are directed against one or more of a person's own proteins.

**Autoimmune disease:** a disease in which the body's immune system, which normally fights off infections and viruses, is misdirected and mistakenly attacks its own healthy tissues through inflammation. In myositis, the immune system attacks otherwise healthy muscles and blood vessels.

**Biologics (also called biologic response modifiers):** pharmaceutical compounds derived from natural, living sources, in contrast to most drugs that are chemically synthesized. Biologics include vaccines, blood and blood components (such as IVIg), gene therapies, monoclonal antibodies, and other products.

**Biopsy:** a test in which a small piece of muscle, skin, or other tissue is removed to view under the microscope, looking for abnormalities that may help in diagnosing a condition.

**Calcinosis:** hard, often painful lumps of calcium that form under the skin's surface, especially in DM.

**Cataract:** clouding of the lens of the eye that blocks light and distorts vision. Cataracts can be a side effect of prednisone and other medicines used to treat myositis. Regular eye exams are encouraged.

**Complementary therapy:** any therapy used in addition to treatment prescribed by your physician. Complementary approaches to health may include exercise, yoga/tai chi/qi gong, hypnosis, movement therapies, meditation, acupuncture, dietary supplements, other botanical products, homeopathy, special diets, and more.

**Contracture:** a permanent shortening of a muscle or tendon, causing a joint to remain bent. Range-of-motion exercises may be prescribed to help prevent or improve contractures.

**Conventional therapy:** traditional treatments that are most commonly prescribed by physicians. For many people with myositis, for example, conventional therapy includes prednisone and/or other immunosuppressant medications.

**Corticosteroids (also called steroids):** medications aimed at slowing the immune system, reducing inflammation, and relieving swelling, itching, and discomfort. Prednisone and Solu-medrol are corticosteroid medications.

**Disease damage:** long-term effects or changes caused by the disease or medication side effects and not by current disease activity.

**Disease-modifying antirheumatic drugs (DMARDs):** a category of drugs used to treat rheumatic diseases to slow the progression of the disease.

**Distal:** located away from the center of the body, such as muscles in the fingers, hands, and feet. [See also proximal.]

**Dysphagia:** difficulty swallowing.

**Dysphonia:** problems with the voice, often resulting in weaker or hoarse sounds.

**Electromyogram (EMG):** a diagnostic test to evaluate and record the electrical activity produced by skeletal muscles.

**First-line treatment:** medication considered the first or best choice for treating a certain 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 medication too quickly or overexerting oneself through exercise or stress. People with DM may also notice flares when they have been exposed to sunlight.

**Gottron's sign (also Gottron's papules):** red, sometimes scaly, eruptions on the knuckles, elbows, and knees. Gottron's papules are a characteristic skin symptom of DM.

**Heliotrope rash:** blue-purple discoloration on the upper eyelids with swelling. Heliotrope rash is a characteristic skin symptom of DM.

**Idiopathic:** of unknown cause. Idiopathic inflammatory myopathy (IIM) is the broader medical term used to describe all forms of myositis.

**Immune response:** the body's response to the presence of a foreign substance in order to keep it from harming the body. Inflammation or swelling is one type of immune response.

**Immune system:** a network of cells, tissues, and organs that work together to defend the human body against attacks by harmful foreign invaders, such as bacteria, parasites, fungi, and viruses that can cause infections.

**Immunoglobulin (also gamma globulin or immune globulin):** a substance made from human blood plasma obtained from donated human blood that contains antibodies and is used to treat certain diseases, including some forms of myositis. Immunoglobulin can be given intravenously (IVIg) or subcutaneously (SCIg).

**Immunosuppressant:** drugs that suppress or reduce the strength of the body's immune response. Immunosuppressant drugs are often used to treat autoimmune disorders such as myositis.

**Inflammation:** part of the immune system's natural response to heal an injury or fight an infection. Typical signs of inflammation include heat, pain, redness, swelling, and loss of function.

**Integrative therapy:** an approach to healing that uses both conventional medication and complementary methods in a coordinated way.

**Interstitial lung disease (ILD):** a group of lung diseases causing scarring and thickening of the tissues around the air sacs of the lungs. Except in sIBM, ILD is the most common and serious complication of IIMs.

**Intravenous:** a method for giving medication or fluids through a needle directly into the vein.

**Local:** affecting only a part of the body where medication is applied, as opposed to the whole body. Local treatments include creams applied to a DM skin rash.

**Lymphocyte:** a white blood cell that is a part of the immune system.

**Magnetic resonance imaging (MRI):** a medical imaging test used to form pictures of the organs and processes in the body in both health and disease. MRI is used to detect inflammation in muscles in myositis.

**Maintenance dose:** the smallest amount of medication a person needs in order to keep symptoms from returning or worsening.

**Malar rash:** a butterfly-shaped rash on the cheeks and bridge of the nose often seen in DM.

**Manual muscle testing (MMT):** test of a person's muscle strength, or ability of the muscle to move a part of the body against resistance. A doctor or therapist will assess muscle strength in individual muscles, and the results show which muscles are weak and the pattern of the weakness. MMT is often performed throughout the disease course to follow the patient's progress over time.

**Mechanic's hands:** roughened and cracked skin on the sides of the fingers and palms, resulting in irregular, dirty-appearing lines that resemble those of a manual laborer. Mechanic's hands are a characteristic skin symptom of antisynthetase syndrome.

**Mixed connective tissue disease (MCTD):** an autoimmune disorder in which there are signs and symptoms of multiple connective tissue diseases, such as lupus, scleroderma, and PM.

**Monoclonal antibody medications:** compounds that use natural immune system functions to target certain cells in the body to treat disease. These compounds are called biologics (rather than drugs) and have names that end in -mab, such as rituximab and infliximab.

**Muscle enzymes:** proteins found mostly in muscle tissue cells. When the muscle tissue cells are injured (as they are through the inflammation of myositis), the cells break open and leak enzymes into the bloodstream. Blood tests are used to measure the levels of these enzymes in the blood to help doctors determine what may be happening and also how well you are responding to treatment.

**Myositis-associated autoantibodies:** proteins found in the blood of myositis patients as well as in the blood of patients with other autoimmune diseases. These autoantibodies are not specific to myositis, but they can help guide treatment and understand the likely course of the disease. [See myositis-specific autoantibodies.]

**Myositis-specific autoantibodies:** proteins found only in the blood of myositis patients, therefore these autoantibodies are specific to myositis. Anti-Jo-1 and anti-SRP are myositis-specific autoantibodies. Because they are unique to myositis, finding one of these autoantibodies in the blood is diagnostic for myositis. The specific type of MSA also helps guide treatment and provides clues about the likely course of the disease. [See myositis-associated autoantibodies.]

**Occupational therapy:** therapy designed to restore or maintain a patient's ability to perform tasks used in daily living, often through developing ways to modify or adapt activities.

**Osteoporosis:** a disease characterized by decreased bone density, resulting in thinning of bone tissue and decreased mechanical strength.

**Overlap syndrome:** an autoimmune disease in which a person presents with symptoms of two or more diseases. The patient's symptoms must meet the diagnostic criteria of all of the diseases to be considered overlap. Some common diseases that overlap with myositis are rheumatoid arthritis, lupus, scleroderma, and Sjogren's disease.

**Physical therapy (also physiotherapy):** treatment of a disease through physical activity with goals of restoring and/or maintaining functional activities.



**Proximal:** located toward the center of the body, such as muscles of the neck, stomach, upper arms, and upper legs. [See distal.]

**Range-of-motion (ROM):** the full amount of movement at each joint. ROM exercises focus on maintaining flexibility and movement in the joints.

**Raynaud's phenomenon (also Raynaud's syndrome):** a condition in which spasms of arteries cause episodes of reduced blood flow, typically involving the fingers and toes. The episodes last several minutes to several hours and cause the affected part to feel numb and cold and to turn white and then blue. Raynaud's is a characteristic symptom of antisynthetase syndrome.

**Refractory:** resistant or unresponsive to treatment.

**Remission:** a state or period during which a patient shows no symptoms of disease and has been off all medications.

**Rheumatic disease:** an umbrella term used to describe chronic conditions affecting the joints and/or connective tissue. Rheumatology is the medical specialty concerned with the study, diagnosis, and treatment of rheumatic diseases. Myositis is considered a rheumatic disease.

**Rimmed vacuoles:** small areas of destruction in the muscle cells in IBM and other muscle disorders.

**Second-line treatment:** medication chosen after a patient fails to respond to the first medication given or when side effects from the first medication are too great. Methotrexate is often a second-line treatment for myositis after initial treatment with prednisone.

**Shawl sign:** a widespread, flat, red rash that appears on the upper back, shoulders, and back of the neck.

**Side effect:** an unwanted reaction to a medicine. For example, weight gain is a common side effect of corticosteroids.

**Taper:** the process of gradually lowering the dosage of medication to reach a maintenance dose or to stop the medication completely. Corticosteroids in particular require a tapering schedule to allow the body to adjust to the lower dosage.

**Ulceration:** break in the skin or mucous membrane.

**Vasculitis:** inflammation of the blood vessels causing restricted blood flow. Vasculitis is believed to be the underlying cause of DM.

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