



## THE MYOSITIS ASSOCIATION

The Myositis Association (TMA) is a non-profit, voluntary health agency dedicated to improving the lives of people affected by myositis. Formed in 1993, TMA has grown from 16 patients, who helped form the organization, to more than 6,000 members who have been served by TMA.

TMA is governed by a volunteer Board of Directors that includes patients, family members and interested professionals, bringing diverse strengths and perspectives to the organization.

Through member newsletters, publications, support groups, research and advocacy, TMA helps those who have myositis today and works to prevent any others from having to experience myositis in the future.



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Inclusion-Body Myositis

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“Myositis” describes inflammation or swelling of the muscle tissue. General muscle inflammation can occur after exercising or taking certain medication, or it can be from a chronic inflammatory muscle disorder like inclusion-body myositis (IBM). IBM is a slowly progressive muscle disease causing weakness in certain limb muscles. Scientists think that the IBM weakness is caused by a specific kind of aging-related degeneration within muscle fibers and perhaps also by autoimmune factors. Some doctors believe there may be an environmental exposure (perhaps to an infection or medicine) that triggers the disease in someone who has certain specific but not yet fully defined genes or gene sequences that predispose him or her.

Most cases of IBM do not run in families, although there are rare hereditary types. IBM is sometimes misdiagnosed as amyotrophic lateral sclerosis (ALS, or Lou Gehrig's disease) or passed off as “just old age.” IBM is a rare disease, and all forms of myositis (polymyositis, dermatomyositis, and inclusion-body myositis) combined affect about 50,000 people in the U.S. A considerable number of additional IBM patients are probably misdiagnosed or remain undiagnosed.

IBM is usually found in people older than 50, more commonly in men than women and, infrequently, younger people. IBM is the most common progressive muscle disease in people older than 50. Some of the first signs of IBM are:

- Sudden falling without an apparent reason;
- Difficulty climbing stairs or rising from a seated position;
- “Foot drop” (weakness of muscles holding the front of the foot up) when walking;
- Weak grasping of objects;
- Difficulty swallowing.

The individual muscle fibers of people with IBM contain “vacuoles” (empty spaces) and abnormal clumps of several proteins. These characteristic protein clumps, called “inclusion bodies,” give inclusion-body myositis its name.

The vacuoles and inclusion bodies within the fibers, and the muscle fiber atrophy (shrinkage), reflect the degenerative aspects of IBM and can be identified through a muscle biopsy. Auto-immune inflammatory cells are present in the IBM muscle, like the ones in polymyositis (PM) muscle, but the extent to which they contribute to the muscle weakness in IBM is still uncertain.

Inflammatory myopathies are classified as autoimmune diseases, meaning the body's immune system, which normally fights infections and viruses, does not stop fighting once the infection or virus is gone. The immune system is misdirected and attacks the body's own normal, healthy tissue through inflammation.

### WHAT ARE THE SYMPTOMS OF IBM?

IBM progresses more slowly than other types of myositis, with muscle weakness progressing over years rather than weeks or months. The muscles most often affected are those at the front of the thighs, hips, foot-elevators, fingers, wrists, upper arms, shoulders, neck, and back. Patients typically experience:

- Weakness and noticeable shrinking of the quadriceps muscles (the large muscle at the front of the thigh), sometimes leading to sudden falls;
- Weakness of the muscles below the knees, causing foot drop and tripping (by the toes catching on things);
- Weakness of “flexor” muscles of the fingers, used for gripping and pinching;
- Weakness of the esophageal muscles, which can cause difficulty swallowing (dysphagia) and may lead to choking;
- Possible pain or discomfort as muscles weaken, though often patients report no pain.

### HOW DO DOCTORS TEST FOR IBM?

Your doctor may first ask you questions about your health in general, including your health history and when you first saw signs of muscle

weakness. He or she will then do a physical exam to assess muscle strength and most likely order a muscle biopsy. Other tests that complete the evaluation and rule out another type of disease include:

- Blood tests for muscle enzymes (including CPK and aldolase) and other blood factors
- Electromyography (EMG) and nerve conduction velocities (NCVs)

If you have questions about any test, be sure to talk with your doctor or visit TMA's web site at [www.myositis.org](http://www.myositis.org).

### HOW IS IBM TREATED?

There is presently no significantly effective treatment for IBM. You and your physician should carefully consider the potential benefits and risks of any medicines.

Current research seeks a better understanding of the degenerative processes within the IBM muscle fibers, with the goal of finding drugs to prevent the degeneration, and repair and regenerate muscle fibers, thereby restoring the patient's strength. New immunosuppressant medicines are also being considered.

Since IBM progresses slowly with few fluctuations, your doctor will probably discuss complementary, non-medical treatments, like physical and occupational therapy, assessment for assistive devices, home-environment modification, and fall-prevention techniques. Physical therapy, including efforts to prevent and stretch muscle contractures and a consistent non-fatiguing exercise program appropriate to the individual patient, has been effective in reducing disability and maintaining flexibility. Physicians sometimes treat swallowing difficulty (dysphagia) with dilation or minor surgery.