

- The Idiopathic Inflammatory Myopathies (IIM), generally referred to as myositis, is a group of rare incurable autoimmune muscle, skin, lung, and often multi-organ diseases with an unknown cause and characterized by inflammation and weakness of the muscles. Myositis means “muscle inflammation.”
- There is no cure for myositis, but for some forms there are off-label treatments for symptoms of the disease.
- IIMs are thought to be autoimmune diseases, in which the body’s immune system, which normally defends against infection and disease, attacks its own muscle fibers, blood vessels, connective tissue, joints, and organs.
- In some cases, there is evidence that certain drugs, such as cholesterol-lowering drugs (statins), viruses, cancer, and environmental factors may precipitate the onset of myositis.
- An estimated 5-10 people per million per year are diagnosed with an idiopathic inflammatory myopathy.
- Myositis is muscle inflammation and leads to weakness of the skeletal muscles, which are used for voluntary movement.
- Myositis most commonly occurs in adults between ages 40-60 years and in children between ages 5-15 years, although it can affect anyone at any age.
- Diagnosing myositis takes an average of 3.5 years, in part due to the variability in manifestation of each type and each person. Also complicating the diagnosis is the fact that IIMs are rare diseases.
- IIMs include Polymyositis (PM), Dermatomyositis (DM), Amyopathic Dermatomyositis (ADM), Hypomyopathic Dermatomyositis (HDM), Immune-Mediated Necrotizing Myopathy (IMNM), Inclusion Body Myositis (IBM), Antisynthetase syndrome (AS), and the juvenile forms of Polymyositis and Dermatomyositis (JPM/JDM). Each type of myositis has its own symptom criteria, however, each patient may exhibit disease conditions differently.
- Symptoms associated with PM, DM, JDM, and IMNM include symmetric proximal muscle weakness, affecting muscles closest to the body’s core such as the neck, shoulders, upper arms, hips, and thighs, on both sides of the body equally. This causes difficulty getting up from chairs, climbing stairs, lifting objects, and brushing hair. Other symptoms include rashes and other skin-related conditions (DM, CADM, AS, and JDM), difficulty swallowing (dysphagia), difficulty or changes in speaking (dysphonia), Raynaud’s phenomenon (decreased circulation in the fingers and toes), weight loss, fatigue, and muscle pain (myalgia).
- Patients with ADM and HDM have the skin disease of “classic” DM with little-to-no muscle weakness.
- Antisynthetase syndrome is a collection of illnesses characterized by interstitial lung disease (scarring of the lung tissue) and arthritis in addition to the muscle weakness and skin conditions of the other Inflammatory Myopathies, and is associated with the antisynthetase autoantibodies, with anti-Jo-1 the most common.
- People with Inclusion Body Myositis, which typically occurs in those over age 50, usually experience muscle weakness and wasting in the distal muscles such as the fingers, wrists, hands, and thighs, but also affects the proximal muscles. The slowly progressive muscle weakness and wasting leads to significant disability and the need for assistive and mobility devices, while dysphagia may necessitate the need for a feeding tube.
- Patients with some forms of myositis have an elevated risk of cancer, lung and heart disease, and an increased risk of developing other connective tissue diseases such as lupus, rheumatoid arthritis, and scleroderma.

- Diagnosing myositis can be difficult and may take months or years. Blood tests for muscle enzymes that indicate damage can be an early indicator. Electromyogram (EMG) tests for abnormalities in electrical conduction in nerves and muscles. Magnetic Resonance Imaging (MRI) can show evidence of muscle disease. For those with skin symptoms, a skin biopsy may be performed.
- Muscle biopsies, in which small pieces of muscle are removed from the body and examined under a microscope, are considered the gold standard for diagnosing IIM's. Muscle and skin biopsies are used to differentiate the subtypes of myopathies, which have unique characteristics that help determine a specific diagnosis.
- Necrotizing Autoimmune Myopathy is unique in that the muscle biopsy shows little or no inflammation.
- Inclusion Body Myositis is unique in that the muscle biopsy shows "rimmed vacuoles."
- Blood testing for Myositis-Specific Antibodies (MSA) and Myositis-Associated Antibodies (MAA) are now commonly used and may help confirm a diagnosis of myositis. More research is underway but these antibodies may help predict disease severity, potential complications, and best therapies to use. Antibodies are produced by our immune systems to help fight infections and other foreign invaders. Autoantibodies are antibodies that attack our own bodies. Some of these autoantibodies are found only in patients with myositis (MSAs) and others are also found in patients with other conditions (MAAs).
- Treatment for the IIMs usually begins with corticosteroids. Other medications may be used successfully, separately or in combination with steroids, including immunosuppressive agents, chemotherapy agents, monoclonal antibodies (rituximab), and/or Immunoglobulin (Ig) therapy. These medications present patients with risks of harsh side effects and a weakened immune system.
- For patients with skin disease, topical steroids, non-steroidal topical creams, sunscreen, UV blockers and physical sun blockers, avoidance of the sun, and UV protective clothing may help.
- There are no medications used to treat IBM. Physical, occupational, and speech therapy, along with an exercise program may be helpful in maintaining mobility for a longer period of time.
- Physical therapy and exercise are recommended for patients with myositis.
- Myositis patients may require the use of assistive devices for mobility, such as a cane, walker, rollator, or wheelchair, either for short periods such as during flares or for long-term muscle weakness and damage.

The subtypes of the IIMs each have unique and varying symptoms, treatments, and outcomes. This complex group of rare diseases is difficult to diagnose and treat, which is why information and education for patients, medical professionals, and caregivers is so important.

There is a need for additional research to better understand and classify these diseases, leading to more effective treatments.

Those living with the Idiopathic Inflammatory Myopathies may not *look* sick on the outside so look closer and learn more about these rare diseases at www.UnderstandingMyositis.org/myositis