MYOSITIS in Focus

Inflammatory Myopathy
In Focus

- The Idiopathic Inflammatory Myopathies (IIM's), referred to as simply "myositis", is a group of rare autoimmune muscle, skin, lung, and often multi-organ diseases with an unknown cause and is characterized by inflammation and weakness of the muscles. Myositis means "muscle inflammation."
- There is no cure for myositis, but for many forms there are treatments for symptoms of the disease.
- IIM's 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 myositis.
- An estimated 5-10 people per million per year are diagnosed with an Inflammatory myopathy.
- Myositis causes inflammation and muscle weakness of the skeletal muscles, those 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 be diagnosed at any age.
- Diagnosing myositis takes an average of 3.5 years due to the many features and manifestations that can be present, complicating the diagnosis.
- Idiopathic Inflammatory Myopathies include Polymyositis (PM), Dermatomyositis (DM), Amyopathic Dermatomyositis (ADM), Hypomyopathic Dermatomyositis (HDM), Necrotizing Autoimmune Myopathy (NAM), Inclusion Body Myositis (IBM), and the juvenile forms of Polymyositis and Dermatomyositis (JPM/JDM). Each form has its own specific symptoms and each patient may exhibit disease conditions differently.
- Symptoms associated with PM, DM, JDM, and NAM begin with symmetric proximal muscle weakness, affecting muscles closest to the body's core such as 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 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, often anti-Jo-1.
- Patients with Inclusion Body Myositis, which typically occurs in those over 50 years old, usually experience muscle weakness and wasting in the distal muscles such as the fingers, wrists, hands, and thighs and progressing to the proximal muscles. The asymmetric weakness and wasting are severe, leading to significant disability and the need for assistive devices, and may have dysphagia and fatigue.

- Patients with 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 Inflammatory Myopathies. Muscle and skin biopsies are used to differentiate the subtypes of myopathies, which have unique characteristics that lead to specific diagnoses.
- 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 Inflammatory Myopathies 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 current medications used to treat IBM, as patients are generally unresponsive to corticosteroids and immunosuppressive drugs, leading to slow and debilitating progression of the disease. Physical and occupational therapy, and exercise may be helpful in maintaining mobility.
- Physical therapy and exercise are recommended for patients with myositis, but 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 Idiopathic Inflammatory Myopathies each have unique and varying causes, 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 characterize these diseases, leading to more effective treatments and possible cures.

Some patients with Inflammatory Myopathies may not look sick on the outside so look closer and learn more about these diseases at www.UnderstandingMyositis.org/myositis