About Myositis

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 one of the chronic inflammatory muscle disorders.

Dermatomyositis (DM), polymyositis (PM), inclusion body myositis (IBM) and juvenile forms of myositis (JM) are all inflammatory myopathies or disease where there is inflammation and loss of muscle. Depending on the type of myositis, the first signs of myositis may include general fatigue, trouble standing from a seated position, difficulty climbing stairs and weak grasping of objects.

The underlying causes of DM, PM, IBM and JM are not known, but some doctors believe there is an environmental exposure (perhaps to infection or sunlight) that triggers the disease in someone who has certain specific, but not yet fully defined genes or gene sequences that predispose him or her. Inflammatory myopathies are classified as autoimmune diseases, meaning the body's immune system, which normally fights infections and virus, 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. Inflammatory myopathies are rare diseases and combined, affect an estimated 50,000 people in Canada.

As research continues into the causes, symptoms and effects of this disease, new forms of myositis are brought forward. There are currently treatments for all forms of myositis except IBM. The other forms of myositis can be treated but only about 20% of people affected make a full recovery. Flare ups are often common over the span of a lifetime. What's more, many doctors across Canada have seen so few cases of myositis that often patients go undiagnosed or misdiagnosed. This can lead to complications and at times a longer recovery period. It is estimated there are 2,000 undiagnosed cases in Canada. The average time for diagnosis is 5 years from first reporting of symptoms to diagnosis. Dermatomyositis (DM)

DM affects people of any age or sex but is more common in women than men. DM is the easiest type of myositis to diagnose because it typically has a visible skin rash caused by

inflammation of blood vessels under the skin. The DM rash looks patchy and reddish or purple and is found on the eyelids, elbows, knees and knuckles. Additional rashes may occur on the cheeks, nose, back and upper chest. Some people also have hardened bumps under the skin called calcinosis. Patients usually experience gradual muscle weakness and sometimes pain and they often report the rash well before the muscle weakness. Three sub types of DM are amyopathic DM where the skin is affected but muscles are not involved; cancer associated DM, where cancer and dermatomyositis are diagnosed within two to three years of one another; and overlap, where DM is associated with other autoimmune diseases.

Polymyositis (PM)

PM is found more frequently in adults than children. More women than men have PM. Patients experience muscle weakness gradually over weeks or months, typically beginning with muscles closest to and within the trunk of the body, such as neck, hip, back and shoulder muscles, and affecting both sides of the body equally. Some patients also have weakness in their hands and fingers. PM patients generally do not have skin rashes. Some PM patients have trouble swallowing (dysphagia), difficulty breathing (often associated with interstitial lung disease) and muscle pain. PM may be associated with a malignancy or with other autoimmune diseases.

Inclusion Body Myositis (IBM)

IBM is found in more men than women and is rarely seen in people less than 50 years old. IBM progresses more slowly than other types of myositis, as weakness happens gradually over months or years. Some of the first signs of IBM are falling, difficulty standing from a seated position and weakening grip. Muscles most often affected are those at the front of the thighs, hips, foot-elevators, fingers, wrists, upper arms, shoulders, neck and back. Many IBM patients notice shrinking or atrophy in the arms and thighs as muscles become weaker. Trouble swallowing (dysphagia) is a common problem for IBM patients. Weakness of facial muscles is sometimes seen.

Juvenile Forms of Myositis (JM)

JM occurs in children 18 years old and younger. Juvenile dermatomyositis (JDM) is the most common form affecting children in Canada. Polymyositis in children (JPM) occurs in approximately 10% of children with JM. Signs of JM include the characteristic skin rashes (i.e. a visible, reddish-purple rash over the eyelids or over joints), trouble climbing or lifting the head, weak voice (dysphonia) or problems swallowing (dysphagia). The muscles most affected are those closest to the centre of the body – neck, stomach, upper arms and legs. About half of the children with JM report pain in their muscles. Some children have calcinosis (hardened lumps under the skin) or contractures (when muscle shortens and causes joints to stay bent). Children may have one or more autoimmune disease(s) along with myositis.

Necrotizing Autoimmune Myopathy (NAM)

NAM or Immune-Mediated Necrotizing Myopathy (IMNM) is a newly defined form of idiopathic inflammatory myopathy. In the past, all patients who presented with muscle weakness, elevated creatine kinase levels and other symptoms of myopathy, but who didn't have skin involvement, were classified as having polymyositis. Now, it is recognized that some of these patients have unique findings on their muscle biopsies that distinguish them form those with other forms of myositis.

Patients with necrotizing myopathy have muscle biopsies that show much less inflammation in muscle tissues than polymyositis patients, but they have increased evidence of muscle cell death (necrosis).

Researchers are also beginning to distinguish different categories of necrotizing myopathy that have different risk factors and different treatments. These distinctions are based on the presence of different autoantibodies in the patient's blood and probably mean that these are different diseases. These different forms include:

1. Patients with SRP autoantibodies
2. Patients with HMGCR autoantibodies

3. All other patients with necrotizing myopathy

Like other forms of myositis, patients with necrotizing myopathy may experience:

- Weakness in the muscles closest to the centre of the body, such as the forearms, thighs, hips, shoulders neck and back
- Difficulty climbing stairs and standing up from a chair
- Difficulty lifting arms over head
- Falling and difficulty getting up from a fall
- A general feeling of tiredness

For further information on all types of Myositis such as Detailed Symptoms, Diagnosis, Treatment and Disease Management and Complications, please refer to our friends in the United States at The Myositis Association (TMA) website www.myositis.org , Myositis Support and Understanding (MSU) website www.understandingmyositis.org and the Mayo Clinic website www.mayoclinic.org. All these sites have excellent, detailed reference materials.