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Myositis Disorders:
Polymyositis / Inclusions Body Myositis / Dermatomyositis

What is myositis?

Myositis is an inflammation of the muscle. Polymyositis, inclusion body myositis and dermatomyositis are disorders characterized by inflammation of the voluntary (skeletal) muscles. These conditions impact the muscle leading to weakness and in some cases, severe disability. In polymyositis, the inflammation is found in many muscles, thus the term "poly". As the name implies, in inclusion body myositis, the muscle is characterized by abnormal inclusions – accumulations of mis-folded protein. In dermatomyositis the muscle inflammation is accompanied by a skin rash, therefore the prefix "dermato," referring to skin.

What are the symptoms of these disorders?

The primary symptom of these disorders is muscle weakness, which is usually progressive and may be severely disabling. In polymyositis and dermatomyositis, the onset of weakness is rapid, usually measured in weeks or months. The onset of inclusion body myositis is measured in months or years. Within each of these disorders, the symptoms vary widely from person to person, with respect to severity, rate of progression and complications. In addition, the three disorders differ markedly in terms of response to treatment. Weakness is generally first evident in the large muscles around the hips and shoulder girdle. An individual may notice difficulty in walking, rising from a chair or a bed, have difficulty turning over in bed, climbing stairs and lifting his or her arms. In some cases, as the disorder progresses, the pattern of walking becomes clumsy and there is a tendency to fall. Excessive fatigue may occur, especially after prolonged standing or walking.

Who can get myositis?

Dermatomyositis can occur at any age and in people of either sex. Polymyositis usually occurs after the age of 18. Inclusion body myositis usually affects men more than women and usually strikes after 50. All three of these conditions are considered rare.

A synopsis of each disorder.

Polymyositis: polymyositis is characterized by a rapid onset of widespread muscle inflammation and weakness. In polymyositis, a characteristic immune response occurs: the muscle cells display a “flag” that draws the attention of the immune system that responds by killing the muscle cell. The symptoms of polymyositis are quite diverse and somewhat difficult to define, some people respond well to treatment while others display a more resistant and severe disorder, culminating in significant disability. There may be involvement of the muscles used in swallowing and breathing. Spontaneous remissions may occur, although as a rule, the natural course of the disease is progressive unless effective treatment can be instituted. Polymyositis may be associated with other connective tissue disorders, vascular disorders or autoimmune diseases, such as lupus. Polymyositis may also be associated with infectious disorders, such as HIV-AIDS, or with Lyme disease or toxoplasmosis.

Although polymyositis tends to be progressive, it often displays a positive response to treatment, in many mild cases, leading to a full resolution. A range of treatments may be used for polymyositis, including medication, physical therapy, exercise, heat therapy (including microwave and ultrasound), orthotics and other assistive devices, and rest. The usual treatment for polymyositis involves the administration of a corticosteroid drug. Immunosuppressant therapy, such as azathioprine and methotrexate, may be used in people who do not respond well to prednisone.

Dermatomyositis: In dermatomyositis, in addition to muscle weakness, a characteristic reddish skin rash develops, either preceding or accompanying muscle weakness, typically on the face (cheeks and eyelids), elbows, hands, knees and upper chest. The entire skin may take on a reddish hue in severe cases. Dermatomyositis may affect children and there is a well recognized juvenile form. The course of dermatomyositis is often severe and rapidly progressive, sometimes involving the heart and lungs. It is often accompanied by degeneration of blood vessels (vasculitis) and with the formation of calcium deposits (calcinosis) under the skin or in the muscle that can cause pain and infections and that may have to be surgically removed. Some cases of dermatomyositis may be accompanied by other autoimmune disorders such as lupus, scleroderma, or vasculitis.

In cases of acute dermatomyositis, the muscles may be quite painful. Prior to the advent of modern therapy, dermatomyositis had a substantial mortality rate. For some reason, dermatomyositis is often associated with cancer and may signal a pre-existing cancer, in many cases, if the cancer is resolved, the dermatomyositis also remits.

A range of treatments may be used for dermatomyositis and most cases respond to some degree, although the disorder tends to be more pervasive than polymyositis and usually requires ongoing treatment. Treatment options may include medication, physical therapy, exercise, heat therapy (including microwave and ultrasound), orthotics and other assistive devices, and rest. The usual treatment for dermatomyositis involves the administration of a corticosteroid drug. Immunosuppressant therapy, such as azathioprine and methotrexate, may be used in people who do not respond well to prednisone. Periodic use of intravenous immunoglobulin may also improve recovery.

Inclusion body myositis: Inclusion body myositis is a late onset disorder, usually appearing after the age of 50, although 20% of cases are diagnosed before the age of 50. Inclusion body myositis displays the same general immune system involvement as seen in polymyositis, however, in inclusion body myositis there is also a progressive deterioration of the muscle characterized by the accumulation of abnormal proteins that is not seen in polymyositis. Inclusion body myositis is not generally associated with other conditions but is relentlessly progressive, characterized by progressive weakening of the large skeletal muscles of the arms and legs. Approximately half of cases develop weakness swallowing. Inclusion body myositis invariably progresses to severe disability. Although rare, inclusion body myositis is the most common muscle disorder seen after the age of 50. The common form is sporadic although there is an extremely rare hereditary form called inclusion body myopathy that has been linked to a specific genetic mutation.

Unfortunately, there is no standard treatment, inclusion body myositis has not responded to any immunosuppressant drugs and the search continues for an effective treatment.

[BOX] What causes myositis?

These conditions were initially classified together because they all displayed muscle inflammation. Although the cause of each of these illnesses remains unknown, they are distinct clinical conditions. Polymyositis and inclusion body myositis show some similarities in terms of their pathological characteristics, dermatomyositis appears to a different type of disorder altogether.

One theory is that there is an autoimmune component to these illnesses. In autoimmune disorders, the immunological mechanisms that normally provide protection against infection and foreign substances are disrupted and instead, attack and injure the body's own tissues. Researchers are looking into the reasons why this abnormal response occurs in some people. These are not considered hereditary disorders in the conventional sense. Cases usually just appear (sporadic) and a history of the disorder in the same family is unusual. However, a hereditary factor, the presence of a group of genes associated with a predisposition to developing an autoimmune disorder, is implicated. In families with patients having myositis disorders, other members may frequently display other forms of autoimmune disorders. [END BOX]

[BOX]What research is being done?

Muscle research is ongoing, the exact mechanisms involved in normal muscle function are still poorly understood. Research on the myositis disorders focuses on the origin, causes and course of these disorders and on improved methods of diagnosis and treatment. A great deal of research is centered on the steps in the progression of these disorders to help reveal potential opportunities for treatment. [END BOX]

How is the diagnosis made?

In diagnosing muscle conditions, several standard tests are utilized including a blood test of creatine kinase, an electrical test of muscles called an electromyograph and in some cases, a muscle biopsy. Generally speaking, the most useful test for diagnosis is the muscle biopsy, although it is also the most invasive for the patient. Dermatomyositis tends to be a recognizable condition due to the characteristic rash seen. The clinical presentation of muscle weakness is important in these disorders and often recognizable during a physical exam and detailed history.

The differentiation between polymyositis and inclusion body myositis can be challenging. Many cases of inclusion body myositis are initially diagnosed as polymyositis and treated. When the patient does not respond to treatment, further investigation leads to a re-diagnosis of inclusion body myositis. Polymyositis may also be confused with other types of muscular dystrophy of late onset.

How can I help?

Muscular Dystrophy Canada conducts year-round fund raising campaigns to support our diverse programs. Your gift will help the Association provide the dollars necessary to assist individuals living with neuromuscular disorders, and fund much needed medical research and educational information. Please make a gift through our National office or any Regional or Community Muscular Dystrophy Canada office.

All Muscular Dystrophy Canada Information Sheets are available on our website:
www.muscle.ca
Ce feuillet d’information est aussi disponible en français.

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