

Myositis Disorders: Polymyositis, Inclusion Body Myositis, and 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 misfolded protein. In dermatomyositis, the muscle inflammation is accompanied by a skin rash, therefore the prefix “dermato,” referring to skin. All three of these conditions are considered rare. They are also known as inflammatory myopathies.

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. 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, 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.

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.

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)

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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. Other types of autoimmune disorders may be common in families with a history of myositis.

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 electromyography, and 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.

Polymyositis may be difficult to distinguish from late onset muscular dystrophy. The most useful test for this purpose is the muscle biopsy. A biopsy showing widespread degeneration and regeneration of muscle fibers, extensive inflammation, and no hypertrophic muscle fibers provides strong support for a diagnosis of an inflammatory myopathy. The presence of one of the autoimmune connective tissue diseases also helps differentiate polymyositis from muscular dystrophy. In the diagnosis of all acutely progressive cases, viral, bacterial and parasitic infections of muscle -- all of which can resemble polymyositis -- must be excluded.

The differentiation between polymyositis and inclusion body myositis can be challenging. In some cases, a patient is diagnosed and treated for polymyositis. If the patient does not respond to treatment, further investigation may lead to a re-diagnosis of inclusion body myositis.

More about Polymyositis

Polymyositis is more common in females than males and usually begins after age 20. It is characterized by a rapid onset of widespread muscle inflammation and weakness. The symptoms of polymyositis are quite diverse and somewhat difficult to define.

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, effective treatments are available. Treatments include medication, physical therapy, exercise, heat therapy (including microwave and ultrasound), orthotics and other assistive devices. The usual treatment for polymyositis involves the administration of a corticosteroid drug, such as prednisone. Immunosuppressant therapy, such as azathioprine and methotrexate, may be used in people who do not respond well to prednisone. Another type of effective therapy involves periodic intravenous infusions of antibodies (immunoglobulin) from donors. Response to treatment is variable.

More about Dermatomyositis

In dermatomyositis, in addition to muscle weakness, a characteristic reddish skin rash develops, 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.

The onset of dermatomyositis occurs rapidly, usually measured in weeks or months. It can occur at any age and in people of either sex. 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. These deposits can cause pain and infections, and may have to be surgically removed. Some cases of dermatomyositis may be accompanied by other autoimmune disorders such as lupus, scleroderma, or vasculitis.

A range of treatments may be used including medication, physical therapy, exercise, heat therapy, orthotics and other assistive devices. The usual

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treatment for dermatomyositis involves the administration of a corticosteroid drug, such as prednisone. Immunosuppressant therapy, such as azathioprine and methotrexate, may be used in people who do not respond well to prednisone. Another type of effective therapy involves periodic intravenous infusions of antibodies (immunoglobulin) from donors.

Most cases respond to some degree, although the disorder tends to be more pervasive than polymyositis and usually requires ongoing treatment.

More about Inclusion Body Myositis

Inclusion body myositis usually affects men more than women and usually strikes after 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. It is a progressive disease, as seen by the progressive weakening of the large skeletal muscles of the arms and legs. Approximately half of cases develop weakness in swallowing. However, the heart and lungs are not usually affected. The degree of disability varies; some remain able to walk, although require a cane for long distances. Others are more

severely affected and require a wheelchair. Unfortunately, immunosuppressant drugs have not proved to be an effective treatment for inclusion body myositis; currently, there is no proven treatment.

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.

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