

Polymyositis / Dermatomyositis

What are polymyositis and dermatomyositis?

Polymyositis and dermatomyositis are disorders involving inflammation of the voluntary muscles. As a result of this inflammation (myositis), muscles undergo degenerative changes, that may lead to severe disability. In polymyositis, the inflammation is found in many muscles, thus the term "poly". It is present over a prolonged period of time. In dermatomyositis the muscle inflammation is usually accompanied by a skin rash, therefore the prefix "dermato".

What causes polymyositis and dermatomyositis?

Polymyositis and dermatomyositis are believed to be autoimmune disorders. In autoimmune disease, the immunological mechanisms that normally provide protection against infection and foreign substances are disrupted and instead, produce antibodies that attack and injure the body's own tissues. Researchers are looking into the reasons why this immune response occurs in some people.

These are not considered hereditary disorders in the strict sense. Cases are usually sporadic and a history of the disorder in the same family is unusual. However, a hereditary factor, possibly an abnormality of the immune system, may be implicated to some extent. Evidence pointing in this direction is the frequent occurrence of polymyositis or dermatomyositis in one person and another autoimmune connective tissue disease in another member of the same family.

What are the symptoms of each of these disorders?

The cardinal symptom in both of these disorders is muscle weakness, which is usually progressive and may be severely disabling. The weakness develops over a period of days or months, rather than years. Weakness, typically symmetrical, is usually first evident in the large muscles about the hips and shoulder girdle. An individual may notice difficulty in walking, rising from a chair or a bed, climbing stairs and lifting his/her arms. In some cases, as the disease process advances, the pattern of walking becomes clumsy and waddling, there is the tendency to fall and it becomes difficult to turn over in bed. Excessive fatigue may occur after prolonged standing or walking. In polymyositis, there may be an early and severe involvement of the muscles used in swallowing and breathing.

The symptoms of polymyositis and dermatomyositis vary widely from person to person, with respect to severity, rate of progression and complications. Spontaneous remissions may occur, although as a rule the natural course of the disease is progressive unless effective treatment can be found. It has been suggested that the more acute the onset, the better the prognosis. In dermatomyositis a reddish rash appears, typically on the face (cheeks and eyelids), elbows, hands, knees and upper chest. The skin

changes in appearance, becoming shiny, tight and thin. A person's fingers may become very blue in cold weather. In polymyositis, there is no skin rash.

Who can get polymyositis and dermatomyositis?

Anyone. Both disorders can occur at any age and to people of either sex. Both disorders are rare. Polymyositis occurs more commonly in adults, while dermatomyositis is more common in children.

Females are more likely to be affected than males in dermatomyositis while females and males are affected equally in polymyositis.

How do these inflammatory myopathies differ?

Most obviously, the muscular weakness in dermatomyositis is accompanied by a patchy, dusky rash (an "erythematous" rash) that usually appears over the cheeks, about the eyes and on the neck, elbows, knees, knuckles, shoulders and upper chest. The entire skin may take on a reddish hue in severe cases.

Dermatomyositis is seen more frequently in children than polymyositis, and its course is often severe and rapidly progressive. It is accompanied by a degeneration of blood vessels (vasculitis) and with deposition of calcium salts in the skin (calcinosis).

In cases of acute dermatomyositis, the muscles may be tender and swollen so that movement is painful. Polymyositis is less likely to involve pain.

While an association with malignancy has been found in both adult dermatomyositis and polymyositis, it occurs much more frequently in dermatomyositis. This is not the case in juvenile dermatomyositis, here no association between the disorder and malignancy has been identified.

Polymyositis is much more diverse clinically than dermatomyositis and more difficult to define. Some authorities say it is "impossible" to define if that definition is to include characteristics that apply to every case. One distinctive aspect of polymyositis is its frequent association with one or another of the autoimmune collagen (connective tissue) diseases such as systemic lupus erythematosus, rheumatoid arthritis, Sjogren's syndrome, mixed connective tissue disease or scleroderma. According to many authorities, polymyositis is more likely than dermatomyositis to be slowly progressive.

How is the diagnosis made?

Dermatomyositis is a recognizable condition. The primary criteria for diagnosis are clinical and recognizable during a physical exam and detailed history. These criteria include a distinctive erythematous skin rash

What research is being done?

Current research focuses on the origin, causes and course of polymyositis and dermatomyositis and on improved methods of treatment. Reliable tests are being sought for the detection of antibodies that attack muscle tissue. The measurement of such antibodies may make it possible to more precisely monitor a person's response to treatment. Investigators are continuing to evaluate the use of plasmapheresis in treating polymyositis and dermatomyositis, and are also conducting comparative studies of various corticosteroids and immunosuppressive drugs. Studies related to IVIG therapy are examining how the treatment works and the exact way that it impacts on the disease process.

with the typical onset and distribution of muscle weakness. However, because of the rarity of the disorder, especially in children, it may be difficult to diagnose for a physician who has had no prior experience with the disorder. Polymyositis, especially in its slowly progressive form, may be difficult to distinguish from muscular dystrophy of late onset. The most useful test for this purpose is the muscle biopsy. A biopsy showing widespread degeneration and regeneration of muscle fibres, extensive inflammation, and no hypertrophic muscle fibres 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.

Is there any treatment for these disorders?

Yes. Several approaches are followed. Many people respond satisfactorily to prednisone, a cortisone derivative, that is the primary treatment agent in the inflammatory myopathies. A second line of treatment, especially useful in situations where prednisone alone is not sufficient, is the use of immunosuppressive medication, such as cyclosporine, Methotrexate™ and Azothiaprine™. In some cases, prednisone and immunosuppressive medication are used together.

In situations where treatment with prednisone and immunosuppressive medication has not been successful, doctors may use two other approaches in the treatment of polymyositis and dermatomyositis. One is known as plasmapheresis (plasma exchange). The other treatment is intravenous immunoglobulin therapy (IVIG) where immunoglobulins (antibodies) pooled from a variety of donors are injected into a vein.

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.

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