
Polymyositis

Summary

- Polymyositis is a connective tissue disease that triggers inflammation and muscular weakness.
 - The cause is unknown, but polymyositis is thought to be an autoimmune disorder, possibly triggered by a viral infection.
 - Since symptoms differ between individuals, polymyositis is hard to diagnose and may be mistaken for muscular dystrophy.
 - Treatment options include corticosteroids, immunosuppressive drugs and physical therapy.
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Polymyositis is a connective tissue disease that triggers inflammation and muscular weakness. The cause is unknown, but polymyositis is thought to be an autoimmune disorder, possibly triggered by a viral infection of muscle tissue.

The disease is more common in women than men and tends to develop between the ages of 50 to 70 years, although anyone of any age or either sex can be affected. Usually, the muscle weakness develops gradually over the course of a few weeks or months. The inflammation may spread to other areas of the body including the heart.

Since symptoms differ between individuals, polymyositis is hard to diagnose and may be mistaken for muscular dystrophy. In many cases, polymyositis is associated with other autoimmune disorders of connective tissue such as scleroderma, systemic lupus erythematosus, rheumatoid arthritis and Sjogren's syndrome.

Symptoms of polymyositis

The symptoms include:

- The shoulders and hips usually affected first
- Muscular weakness
- Muscle wastage
- Muscle pain
- Fatigue
- Breathlessness
- Swallowing difficulties
- Tremors, particularly of the hands
- Wide-footed stance and walking style
- Clumsiness
- Tendency to fall over.

Progression of weakness

Polymyositis develops gradually over weeks or months. By the time the person experiences symptoms, they have already lost around half of their muscle fibres to the disease. The head, hands and feet are usually untouched by the disease. Difficulties may include:

- Profound muscular weakness in affected body parts, such as being unable to lift the arms above shoulder height or lift the head off a pillow
 - Voice changes, if the muscles of the larynx are affected
 - Problems with swallowing if the oesophagus is affected, with a tendency to regurgitate food
 - Peristalsis (the muscular contractions of the bowel) may falter, leading to constipation
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- In its later stages, abnormal shortening of muscles (contracture).

A malfunction of the immune system

The cause of polymyositis hasn't been found, but there is good evidence to indicate that it is likely to be an autoimmune disorder, which means the immune cells mistakenly attack the muscle fibres of the body. If a small tag (biopsy) of affected muscle is examined in a laboratory, it looks withered and studded with immune system cells. Muscle tissue could be damaged by the immune system as it tries to rid the tissue of a viral infection.

As people with polymyositis are likely to have other connective tissue disorders, such as rheumatoid arthritis or scleroderma, there could be a genetic susceptibility. People with polymyositis also tend to have family members with connective tissue disorders, which further supports the theory of an inherited tendency.

Possible complications of polymyositis

The person may experience difficulties with breathing or swallowing if the associated muscles weaken too severely. Some of the other possible complications of polymyositis include:

- Heart inflammation
- Heart attack
- Breast cancer
- Lung cancer
- Lung disease
- Pneumonia
- Respiratory failure.

Diagnosis of polymyositis

Polymyositis is sometimes mistaken for muscular dystrophy, so careful diagnosis is important. Some of the tests for polymyositis include:

- **Medical history** – people with other connective tissue diseases, such as scleroderma, are at greater risk of polymyositis.
- **Physical examination** – this includes general tests.
- **Electromyography** – wires attached to the skin measure the electrical activity of muscles.
- **Muscle biopsy** – a small tag of muscle tissue is removed and examined in a laboratory for evidence of degeneration and abnormal immune system activity. This is the definitive test for polymyositis.
- **Scans** – these include magnetic resonance imaging.
- **Blood tests** – an antibody titre is included, since polymyositis is characterised by the overproduction of particular antibodies. There are also higher than normal amounts of a muscle enzyme indicating muscle damage (creatine phosphokinase) circulating in the bloodstream, which can be measured by a specific blood test.
- **Other tests** – these are used to exclude other causes such as bacterial or parasitical infections of muscle tissue.

Treatment for polymyositis

The outlook for polymyositis is hard to predict. Some people will recover, most will respond satisfactorily to treatment, while others will die from complications. Treatment options include:

- **Corticosteroids** – used to dampen the activity of the immune system and reduce inflammation. For some people, corticosteroids contribute to muscle weakness, so other medications have to be used instead.
- **Immunosuppressive drugs** – include drugs such as azathioprine and methotrexate.
- **Plasmapheresis** – the antibodies responsible for attacking muscle tissue are removed from the bloodstream by plasmapheresis. Blood is taken from the patient and the blood cells are separated from the plasma. Only the blood cells are returned to the patient, leaving the antibodies behind in the plasma.
- **Immunoglobulin** – a special protein obtained from the plasma of blood donors, and administered intravenously, has sometimes had good results in difficult cases.

- **Physical therapy** – helps strengthen muscles.
- **Ongoing monitoring** – includes blood tests, so that medications can be adjusted if necessary. Generally, the initial doses are high, then gradually tapered down.

Dermatomyositis

Dermatomyositis is not the same disease, but is often grouped with polymyositis. They share many similarities such as muscle inflammation and weakness, immune system involvement and similar treatment therapies.

Apart from muscular weakness, other symptoms of dermatomyositis include:

- Blotchy, dark red skin rash on the cheeks, throat, shoulders and chest
- Sometimes, the whole skin surface may appear reddened
- Swollen and painful muscles
- More likely to be an associated cancer
- Children aged between five and 15 years most commonly affected.

Where to get help

- Your doctor
- Rheumatologist
- Muscular Dystrophy Association Tel. (03) 9320 9555 or 1800 656 632
- **Myositis Association** Tel. (02) 4464 2043

Things to remember

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Muscular Dystrophy Association

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