



Polymyositis and dermatomyositis

"When I was 50, I was becoming weak, had a rash that didn't get better and shortness of breath. 'Old age,' my doctor said. No-one knew about dermatomyositis. My symptoms got worse over six months. I am so glad there is more awareness of myositis now, and people are getting the help they need more quickly."

Christine Nixon, who has dermatomyositis

What are polymyositis and dermatomyositis?

Polymyositis (PM) and dermatomyositis (DM) are 'autoimmune' conditions. This means that the immune system – which usually protects the body against infection/germs – becomes overactive and starts attacking the body's own tissues rather than protecting them.

PM and DM are part of a range of conditions known as myositis-spectrum disorders, which also include necrotising myositis and anti-synthetase syndrome. The key feature linking these conditions together is 'myositis' – this means inflammation of the muscles, which can lead to weakness of the arms and legs.

DM occurring in childhood may have important differences – please see our [Juvenile dermatomyositis](#) factsheet for more information on the childhood form of DM.

What causes PM and DM?

The exact cause of PM and DM is unknown. One theory is that autoimmune conditions start because of contact with a 'trigger', which makes the body's immune system overreact. A variety of triggers may spark off events that lead to PM and DM and these include environmental factors such as sunlight, some infections or medications.

In some people, the trigger is an underlying cancer, which may already have been diagnosed, or might be discovered during the investigation of the myositis. Research has found that some people with myositis spectrum disorders have an increased risk of developing cancer. If you have any concerns about this, please get in touch with your specialist.

What are the symptoms?

Muscle weakness is common and mainly involves the 'proximal' limb muscles (the muscles around the shoulders, upper arms, pelvis and thighs). This sometimes comes on rapidly, over a period of days, but usually over months or even years. The pattern of weakness means that some people struggle to climb stairs or get up out of chairs. With the arms, there is often difficulty lifting or using the hands with arms elevated, for example in reaching overhead cupboards or with washing hair.



A few people may experience muscle pain, particularly where the onset of symptoms is quick. Sometimes there can be aching, discomfort, or mild tenderness of the affected muscles.

Myositis can cause swallowing difficulties and can affect the breathing muscles (including the diaphragm), heart or the lungs. If you experience shortness of breath, chest pains, palpitations, or changes in your breathing, discuss this with your GP or specialist.

The skin is affected in DM, usually by a rash, on the face, exposed parts of the chest, and hands (knuckles and nail beds). Exposure to sunlight can make the rash worse. People with anti-synthetase syndrome can also experience skin changes, particularly cracking of skin on the side of the fingers (called 'mechanics' hands').

How is the diagnosis made?

These conditions are relatively rare and diagnosis is usually made by a specialist, such as a neurologist or a rheumatologist. For a more detailed overview of different diagnostic and genetic tests, please see our [Diagnostic tests](#) factsheet.

Two important tests that can help to diagnose myositis are an electromyogram (EMG) and a muscle biopsy. The EMG, which is done in a hospital, involves a small needle being inserted into several different limb muscles. The electrical activity recorded will show whether muscle weakness is caused by a muscle disorder (as would be expected in myositis) or by the nerves supplying the muscles (suggesting an alternative diagnosis).

A muscle biopsy is a procedure in which a small sample of muscle – about the size of a pea – is taken and examined under a microscope. It is considered a 'minor' procedure, and is usually done under a local anaesthetic. Please see our [Muscle biopsies](#) factsheet for more information.

What is the treatment?

It is important to find the optimal treatment regime as there can be risks with long-term high-dose treatment.

Most people with myositis respond to corticosteroid ('steroid') drugs such as prednisolone. Although often effective and necessary at the start of treatment, these drugs can have side-effects, especially if they are taken in large doses for extended periods (several months or even years). For this reason, specialists may also prescribe other immunosuppressive drugs to help control the muscle inflammation and reduce and eventually stop the steroids.

Your specialist may suggest other treatments if the condition is proving hard to treat. For example, high-dose intravenous immunoglobulin (a naturally occurring human blood protein) may be helpful in acute or severe cases.



Steroids can lead to an increased risk of thin bones (osteopenia or osteoporosis). Your GP or specialist will need to assess your bone health before starting you on steroids. They may do this by checking the vitamin D levels in your blood and/or by arranging a bone scan.

For most people, the symptoms improve with treatment, but the responses to treatment vary. Those who have developed weakness over a long period may not respond so well to treatment.

Does exercise help?

When you're having treatment, active exercise is important in improving muscle strength and mobility. Keeping active will lessen the risk of muscles weakening and wasting, and of other complications such as osteoporosis or leg vein thrombosis.

Your specialist will probably suggest physiotherapy, as it will help you maintain the full range of muscle and joint movement. Many people with significant limb weakness also find activities such as swimming helpful in maintaining muscle function. Exercise will also help avoid weight gain.

Other help

Individuals with more severe forms of myositis and restricted mobility will benefit from assessment and help from an occupational therapist (OT) at the hospital or your Local Authority. They may recommend special aids and home adaptations. Some people might find using a wheelchair helpful, particularly for longer trips outdoors.

The future

Active research, particularly in the field of immunology, will tell us more about why individuals develop myositis, and explain how muscle damage and weakness occur. Improving drug treatment is equally important, particularly in identifying which treatments are most effective and free from significant risks and side-effects. There is a need for scientifically sound, controlled trials of the various drug treatments already available, as well as new or experimental treatments.

Find out the latest news on research by visiting our website at www.muscular dystrophyuk.org/news/news/research/

To find out more about the research that Muscular Dystrophy UK is funding please visit: www.muscular dystrophyuk.org/progress-in-research/research-projects/current-grants/

If you have any questions, please email our research team at research@muscular dystrophyuk.org



If you would like your GP or other health professional to have more information about PM and DM, we have some relevant materials. We've developed an online training module for GPs, as well as one for physiotherapists working with adults with muscle-wasting conditions. Contact our helpline or email us to find out more.

We're here for you at the point of diagnosis and at every stage thereafter, and can:

- ▶ give you accurate and up-to-date information about your or your child's muscle-wasting condition, and let you know of progress in research
- ▶ give you tips and advice about day-to-day life, written by people who know exactly what it's like to live with a muscle-wasting condition
- ▶ put you in touch with other families living with the same muscle-wasting condition, who can tell you about their experiences
- ▶ tell you about – and help you get – the services, equipment and support you're entitled to.

If you have feedback about this factsheet or would like to request any references used to produce it, please email info@musculardystrophyuk.org.

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Here for you

The friendly staff in the care and support team at the Muscular Dystrophy UK's London office are available on **0800 652 6352** or info@musculardystrophyuk.org from 8.30am to 6pm Monday to Friday to offer free information and emotional support.

If they can't help you, they are more than happy to signpost you to specialist services close to you, or to other people who can help.

www.musculardystrophyuk.org