Polymyositis and dermatomyositis

This booklet provides information and answers to your questions about these conditions.
Polymyositis and dermatomyositis are conditions that cause inflammation in the muscles. They both cause pain and weakness in many muscles, but dermatomyositis occurs with a skin rash. In this booklet we’ll explain about the symptoms, causes, tests and treatments.

At the back of this booklet you’ll find a brief glossary of medical words – we’ve underlined these when they’re first used.
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What are polymyositis and dermatomyositis?

Myositis means inflammation of the muscles. Polymyositis affects many muscles and especially the larger ones such as those around the shoulders, hips and thighs. Dermatomyositis causes similar muscle symptoms along with a particular skin rash. Both conditions are autoimmune diseases. This means the immune system attacks your body’s own tissues.

What are the symptoms?

The severity of the symptoms of polymyositis and dermatomyositis differ from person to person. Most people will only have mild symptoms. These typically include:

- weak and tired muscles
- muscles that feel tender to the touch and painful
- feeling generally unwell, weight loss and night sweats.

Who gets them?

Polymyositis and dermatomyositis are rare diseases, affecting only 6–8 people out of 100,000. It mostly affects adults, although children can be affected by a type of dermatomyositis called juvenile dermatomyositis.
How are they diagnosed?

The symptoms for polymyositis and dermatomyositis can be similar to many other conditions, so your doctor may base the diagnosis on:

- an examination
- blood tests
- an electromyography (EMG) test
- a muscle biopsy
- a magnetic resonance imaging (MRI) scan of the muscles with the most symptoms.

What treatments are there?

In many cases disease-modifying anti-rheumatic drugs (DMARDs) such as methotrexate, azathioprine, ciclosporin or cyclophosphamide can control the condition. A small number of people have been treated with biological therapies such as infliximab and rituximab when other treatments haven’t been effective.

Infusions of immunoglobulin

Antibodies collected from blood donations given by healthy people can help stop your immune system from attacking your own tissues. However, most people won’t need this treatment.

Exercise

Once the disease is controlled you’ll be given exercises which will help to restore muscle strength.

Drugs

Steroids are normally the first choice of treatment for reducing the inflammation.
What are polymyositis and dermatomyositis?
Myositis means inflammation of the muscles (myo = muscle, itis = inflammation). Polymyositis affects many areas (poly = many), mainly the larger muscles like those around the shoulders, hips and thighs. When polymyositis develops alongside a skin rash, the condition is called dermatomyositis (derm = skin).

What are the other types of myositis?
Post-infectious reactive myositis can occur after some viral infections. After killing the virus, the immune system may cause inflammation in some parts of the body for a few months. If the inflammation occurs in muscles, it causes myositis.

The symptoms of post-infectious reactive myositis are similar to those of polymyositis (muscle pain/weakness), and you may get a positive result for some of the tests that help to diagnose polymyositis, but post-infectious reactive myositis is usually mild and will settle without treatment.

Inclusion body myositis (IBM) causes muscle weakness like polymyositis and dermatomyositis, and tests to diagnose IBM show similar results to the tests for the other two conditions. However, in IBM the weakness usually affects muscles near the ends of the limbs (hands, forearms and calves).

The key difference in IBM is the presence of abnormal lumps of protein (inclusion bodies) in muscle cells, which aren’t found in polymyositis and dermatomyositis. These lumps are so tiny that they can only be seen with special microscopes.

IBM doesn’t respond well to steroids, so if you’re diagnosed with polymyositis and steroids aren’t working you may need further tests for IBM. IBM only rarely causes muscle pain and it generally affects people over 50.
What are the symptoms of polymyositis and dermatomyositis?

The symptoms of polymyositis and dermatomyositis vary between individuals. Most people will only have mild and short-lived symptoms. These can include:

- **weak and tired muscles** – making normally easy tasks very tiring
- **inflamed muscles** – causing pain (known as myalgia) and feeling tender to the touch
- **generally feeling unwell (malaise)**
- **weight loss**
- **night sweats.**

If you have dermatomyositis (myositis with a rash), you may get some of the above symptoms as well as the ones below:

- **a red/pink rash** on the upper eyelids, face and neck, and on the backs of the hands and fingers (see Figure 1)
- **swelling** of the affected skin, causing a characteristic puffiness and colouring around the eyes.

Myositis has similar symptoms to fibromyalgia, another condition that causes muscle pain and fatigue. The conditions aren’t related and, unlike myositis, fibromyalgia isn’t an autoimmune disease.

See Arthritis Research UK booklet *Fibromyalgia.*

![Figure 1](image)

A red/pink rash occurs in people with dermatomyositis.
Other conditions can mimic polymyositis. These include:

- the side-effects of some medications (for example steroids and cholesterol-lowering drugs such as statins)
- toxic effects of long-term alcohol excess
- hormonal conditions (for example under- or overactive thyroid)
- low vitamin D levels, or abnormal calcium or magnesium levels
- infections
- other, rarer nerve-muscle diseases (for example muscular dystrophies).

What causes polymyositis and dermatomyositis?

Polymyositis and dermatomyositis are autoimmune diseases. This means that the immune system, which normally protects the body against infections, attacks the body’s own tissues. This causes inflammation in the muscles and skin. We don’t yet know why this happens.

What is the outlook?

The outlook for people with polymyositis and dermatomyositis is good. Symptoms aren’t often severe and in most cases the pain and feelings of being unwell can be eased fairly quickly with drug treatment. Even in severe cases myositis responds to treatment. The condition frequently becomes inactive. Muscle strength will take longer to recover, but once the disease is controlled with treatment you’ll be given exercises to counter any muscle wasting and to improve the strength of the recovering muscle.

What are the possible complications?

In a few cases of polymyositis and dermatomyositis, drug treatment doesn’t work fully and the muscles can remain weak. We’re still doing research into why this happens, but moderate exercise (so you start breathing faster but aren’t straining yourself) still helps the recovery of your muscle strength.

Occasionally polymyositis can also affect breathing and swallowing. This may occur at the start of severe cases when the muscles used become very weak. It may also cause weakening of the heart, and inflammation of the lungs may cause scarring. This affects how the lungs work. Lung and heart conditions can cause long-term breathlessness.

Children with dermatomyositis may develop painful calcium deposits in damaged muscles. These deposits, combined with immobility, can occasionally result in permanently bent joints (flexion contracture).
Polymyositis occasionally occurs in patients who also have another autoimmune rheumatic disease such as rheumatoid arthritis, lupus, and scleroderma. The myositis is then said to be part of an overlap syndrome.

See Arthritis Research UK booklets Lupus; Rheumatoid arthritis; Scleroderma.

On rare occasions, myositis can be associated with cancer. Most people with polymyositis and dermatomyositis don’t develop cancer, but your doctor might arrange tests such as a chest x-ray or an ultrasound scan of your abdomen and pelvis to be on the safe side.

How are polymyositis and dermatomyositis diagnosed?

Your doctor will talk to you about your symptoms and examine you, but because the symptoms of polymyositis and dermatomyositis are similar to many other conditions you’ll probably have blood tests and other examinations.

Blood tests

Creatine phosphokinase
Creatine phosphokinase is an enzyme that leaks out of damaged muscle cells. It’s likely that you’ll have a high level of this enzyme in your blood if you have polymyositis or dermatomyositis, but the level should fall as the disease is treated and brought under control. You’ll need to have repeat measurements taken throughout the course of the condition to tell the doctor how well it’s responding to treatment.

Erythrocyte sedimentation rate
Erythrocyte sedimentation (ESR) detects and measures inflammation by assessing how quickly blood cells settle at the bottom of a test tube. Inflammation occurs in other conditions so a high ESR alone won’t confirm polymyositis or dermatomyositis.

Autoantibodies
Antibodies are part of the immune system and usually help us to fight off infections by recognising and attacking viruses or bacteria. Some people’s immune systems develop autoantibodies, antibodies that attack the body’s own tissues. This can contribute to causing autoimmune diseases like polymyositis and dermatomyositis.

No autoantibody test proves you definitely have polymyositis or dermatomyositis, but there are tests that can be helpful in making the diagnosis. One of these is the anti-nuclear antibody test (ANA). This tests whether a patient’s blood contains antibodies to the nucleus (the central part) of cells. About 80–90% of people with polymyositis and dermatomyositis test positive for ANA, but healthy people can also test positive so it’s only helpful alongside all the other information about your symptoms.
Other autoantibody tests include ENA (extractable nuclear antigens) and the anti-Jo-1 test. These tests are sometimes positive in people with polymyositis and dermatomyositis. There are almost 20 autoantibodies which occur specifically in myositis. Tests for these can be useful in predicting people’s responses to treatment.

**Other tests**
These tests may not be able to diagnose polymyositis or dermatomyositis, but they can help to rule out other conditions. You may need other tests to confirm a diagnosis.

**Electromyography (EMG)**
A thin electrode is inserted into the muscle to record electrical discharges from nerve endings that cause your muscles to move. An unusual pattern of electrical activity in a number of different muscles suggests you have polymyositis or dermatomyositis. This test isn’t painful but may be uncomfortable.

**Muscle biopsy**
A small sample is taken from one of your larger muscles (for example at the front of your thigh) and examined under a microscope. The site of the biopsy might be chosen using an MRI scan to assess the area. The part of the muscle that looks most badly affected on the scan will be the site of the biopsy. You’ll be given a local anaesthetic to numb the area while the sample is taken, but there may be some discomfort for a few days afterwards.

The biopsy may show white blood cells sticking to the covering of the muscle and damaging the proteins that make your muscles contract, which causes the weakness and fatigue.
You may need to have a repeat biopsy if your condition doesn’t improve with standard treatments. This is to check for the proteins found in inclusion body myositis (IBM).

You may see different specialists depending on which symptoms are giving you the most trouble. You could be referred to one or more of the following people:

- a nerve specialist (neurologist) for muscle weakness
- a muscle specialist (rheumatologist) for muscle pain
- a skin specialist (dermatologist) for rashes.

See Arthritis Research UK booklet Meet the rheumatology team.

**What treatments are there for polymyositis and dermatomyositis?**

Treatment for polymyositis and dermatomyositis includes a combination of drug treatment and exercise.

**Drugs**

The first choice for treatment is steroids, which are usually given in higher doses to begin with. They can be given as tablets or injections. They should reduce the inflammation very quickly and settle muscle pain and the feeling of being unwell.

High doses of steroids can have side-effects so your doctor will reduce the dosage as quickly as possible. Other drugs may be prescribed alongside the steroids to reduce the risk, for example to help prevent developing osteoporosis.

See Arthritis Research UK booklet and drug leaflets Osteoporosis; Local steroid injections; Steroid tablets.

Sometimes the inflammation can flare up when the steroid dose is lowered. Your doctor may prescribe other drugs to help reduce the inflammation.

The first choice for treatment is steroids. When taking steroid tablets you must carry a steroid card.
The most common of the drugs used are methotrexate, azathioprine and ciclosporin. Cyclophosphamide may also be prescribed. A newer drug now being used is called mycophenolate mofetil. These are all types of disease-modifying anti-rheumatic drugs (DMARDs), and you will need regular blood tests to check for any possible side-effects.

See Arthritis Research UK drug leaflets Azathioprine; Ciclosporin; Cyclophosphamide; Methotrexate; Mycophenolate.

When taking steroid tablets you must carry a steroid card, which records your dose and how long you’ve been taking it. If you become ill, or are involved in an accident in which you’re injured or become unconscious, it’s important for the steroids to be continued, and the dose might need to be increased. This is because the treatment may prevent your body from being able to produce enough natural steroids in response to stress. Your doctor, rheumatology nurse specialist or pharmacist can give you a steroid card.

Occasionally these drugs aren’t able to control the disease. A small number of people with severe muscle disease and complications have therefore been treated with biological therapies, which are a newer type of DMARD. They work by blocking the process of inflammation. Examples include infliximab and rituximab. As yet there is little scientific evidence on which biological therapy is the best for myositis.

See Arthritis Research UK drug leaflets Infliximab; Rituximab.

Infusions of intravenous immunoglobulin might also be given, though most people won’t need these. Immunoglobulins are antibodies that stop your immune system attacking your body’s own tissues. Infusions are given in hospital at monthly or 3-monthly intervals. Sometimes you might feel a little unwell (as if you have flu) during the infusion.

See Arthritis Research UK drug leaflet Intravenous immunoglobulin.
Even in severe cases myositis usually responds well to treatment, although many people need life-long drug treatment to keep their condition under control.

**Exercises and physiotherapy**

It’s probably best to rest when your myositis is very active, but once it’s calmed down you’ll need to start exercises. Aerobic exercise (any exercise that makes you breath more heavily and your heart beat faster) is especially important to help restore muscle strength and improve stamina. At first this should be done under the supervision of a physiotherapist, who’ll give you a tailored programme to suit your specific needs. Very strenuous exercise should be avoided.

Children with juvenile dermatomyositis will need more vigorous physiotherapy to prevent flexion contracture. This is a condition where the joints, especially the knees, become permanently bent. Some people with severe inflammation never fully recover but most people do, though it may take several months for muscle strength to improve. It may take weeks or months for your body to repair your muscles and you may get tired more quickly than normal, so don’t worry if you’re not back to full strength straight away.

**Research and new developments**

A lot of research is being done on polymyositis and dermatomyositis. Because the conditions are so rare, research centres are joining forces for their studies. One of these studies is looking at the effectiveness of steroids in various combinations with other drugs. It aims to find the best combination of drugs that are currently available for treating polymyositis and dermatomyositis. The UK Myonet collaboration is starting to find out what genetic factors might make some people more likely to get myositis. This may help us understand the processes that cause muscle damage, which will hopefully lead to the development of new and more effective treatments. New tests are also being developed to help tell myositis apart from other muscle diseases.
Glossary

Autoimmune disease – a disorder of the body’s defence mechanism (immune system), in which antibodies and other components of the immune system attack the body’s own tissue rather than germs, viruses and other foreign substances.

Antibodies – blood proteins that form in response to germs, viruses or any other substances that the body sees as foreign or dangerous. The role of antibodies is to attack these foreign substances and make them harmless.

Biopsy – the removal of a small amount of living tissue from the body. The sample can help diagnose illness when examined under a microscope.

Disease-modifying anti-rheumatic drugs (DMARDs) – drugs used in rheumatoid arthritis and some other rheumatic diseases to suppress the disease and reduce inflammation. Unlike painkillers and non-steroidal anti-inflammatory drugs (NSAIDs), DMARDs treat the disease itself rather than just reducing the pain and stiffness caused by the disease. Examples of DMARDs are methotrexate, sulfasalazine, gold, infliximab, etanercept and adalimumab.

Erythrocyte sedimentation rate (ESR) – a test that shows the level of inflammation in the body. Blood is separated in a machine with a rapidly rotating container (a centrifuge), then left to stand in a test tube. The ESR test measures the speed at which the red blood cells (erythrocytes) settle.

Fibromyalgia – a long-term (chronic) form of widespread pain in the muscles and soft tissues surrounding the joints throughout the body.

Immune system – the tissues that enable the body to resist infection. They include the thymus (a gland that lies behind the breastbone), the bone marrow and the lymph nodes.

Immunoglobulins – a class of blood proteins that are responsible for immunity to specific infections.

Inflammation – a normal reaction to injury or infection of living tissues. The flow of blood increases, resulting in heat and redness in the affected tissues, and fluid and cells leak into the tissue, causing swelling.

Lupus (systemic lupus erythematosus or SLE) – an autoimmune disease in which the immune system attacks the body’s own tissues. It can affect the skin, the hair and joints and may also affect internal organs. It’s often linked to a condition called antiphospholipid syndrome (APS).

Magnetic resonance imaging (MRI) – a type of scan that uses high-frequency radio waves in a strong magnetic field to build up pictures of the inside of the body. It works by detecting water molecules in the body’s tissue that give out a characteristic signal in the magnetic field. An MRI scan can show up soft-tissue structures as well as bones.
Osteoporosis – a condition where bones become less dense and more fragile, which means they break or fracture more easily.

Physiotherapist – a therapist who helps to keep your joints and muscles moving, helps ease pain and keeps you mobile.

Rheumatoid arthritis – an inflammatory disease affecting the joints, particularly the lining of the joint. It most commonly starts in the smaller joints in a symmetrical pattern – that is, for example, in both hands or both wrists at once.

Systemic sclerosis (scleroderma) – a medical condition characterised by hardening and tightening of the skin. It often affects other parts of the body as well – including the connective tissues that surround the joints, blood vessels and internal organs.

Ultrasound scan – a type of scan that uses high-frequency sound waves to examine and build up pictures of the inside of the body.

Where can I find out more?
If you’ve found this information useful you might be interested in these other titles from our range:

Conditions
- Fibromyalgia
- Lupus
- Osteoporosis
- Rheumatoid arthritis
- Scleroderma

Therapies
- Meet the rheumatology team
- Occupational therapy and arthritis
- Physiotherapy and arthritis

Self-help and daily living
- Fatigue and arthritis
- Keep moving
- Looking after your joints when you have arthritis
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• Intravenous immunoglobulin
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You can download all of our booklets and leaflets from our website or order them by contacting:

Arthritis Research UK
Copeman House
St Mary’s Court
St Mary’s Gate
Chesterfield
Derbyshire S41 7TD
Phone: 0300 790 0400
Email: enquiries@arthritisresearchuk.org
www.arthritisresearchuk.org

Related organisations
The following organisations may be able to provide additional advice and information:

Arthritis Care
Floor 4, Linen Court
10 East Road
London N1 6AD
Phone: 020 7380 6500
Helpline: 0808 800 4050
Email: info@arthritiscare.org.uk
www.arthritis.org.uk

Myositis Support Group
146 Newtown Road
Woolston
Southampton
Hampshire SO19 9HR
Phone: 023 8044 9708
Email: msg@myositis.org.uk
www.myositis.org.uk

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Please note: We’ve made every effort to make sure that this content is correct at time of publication. If you would like further information, or if you have any concerns about your treatment, you should discuss this with your doctor, rheumatology nurse or pharmacist.
We’re here to help

Arthritis Research UK is the charity leading the fight against arthritis.

We’re the UK’s fourth largest medical research charity and fund scientific and medical research into all types of arthritis and musculoskeletal conditions.

We’re working to take the pain away for sufferers with all forms of arthritis and helping people to remain active. We’ll do this by funding high-quality research, providing information and campaigning.

Everything we do is underpinned by research.

We publish over 60 information booklets which help people affected by arthritis to understand more about the condition, its treatment, therapies and how to help themselves.

We also produce a range of separate leaflets on many of the drugs used for arthritis and related conditions. We recommend that you read the relevant leaflet for more detailed information about your medication.

Please also let us know if you’d like to receive our quarterly magazine, Arthritis Today, which keeps you up to date with current research and education news, highlighting key projects that we’re funding and giving insight into the latest treatment and self-help available.

We often feature case studies and have regular columns for questions and answers, as well as readers’ hints and tips for managing arthritis.

Tell us what you think

Please send your views to: feedback@arthritisresearchuk.org or write to us at: Arthritis Research UK, Copeman House, St Mary’s Court, St Mary’s Gate, Chesterfield, Derbyshire S41 7TD

A team of people contributed to this booklet. The original text was written by Dr Robert Cooper, who has expertise in the subject. It was assessed at draft stage by consultant rheumatologist Dr Lorraine Croot and clinical nurse specialist Tracy French. An Arthritis Research UK editor revised the text to make it easy to read, and a non-medical panel, including interested societies, checked it for understanding. An Arthritis Research UK medical advisor, Dr Ben Thompson, is responsible for the overall content.
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- volunteering
- supporting our campaigns
- taking part in a fundraising event
- making a donation
- asking your company to support us
- buying products from our online and high-street shops.

To get more actively involved, please call us on 0300 790 0400, email us at enquiries@arthritisresearchuk.org or go to www.arthritisresearchuk.org