

## **Polymyositis, dermatomyositis and sarcoid myopathy**

### **What are polymyositis & dermatomyositis?**

They are a group of conditions in which there is inflammation of the muscles. Resulting disability is very variable: sometimes weakness is mild but at the other extreme there may be major disability. Generally speaking, polymyositis (PM) and dermatomyositis (DM) respond to treatment. DM occurring in childhood may have important differences and a separate MDC leaflet is available entitled 'Juvenile Dermatomyositis'. Inclusion Body Myositis (IBM) is another important cause of chronic muscle inflammation, mainly in older male subjects and is also dealt with in another MDC leaflet. With very rare exceptions PM and DM are not inherited. Both PM and DM can affect individuals at any age.

The exact causes of these disorders are as yet to be established. In PM and DM there is thought to be an abnormal immune reaction in the body which causes muscle inflammation and weakness. In DM there is evidence that involvement of the small blood vessels in muscle may be important. What triggers off the inflammation in these disorders is unclear. Many cases of PM and DM occur in isolation but some cases are associated with what are called "connective tissue disorders" such as rheumatoid arthritis, systemic lupus erythematosus (SLE), Raynaud's disease, scleroderma or Sjögren's syndrome. Some cases of PM and DM, usually in older patients, may be associated with an underlying malignancy, such as lung and stomach cancer. There are other causes of muscle inflammation such as viral infection; bacterial infection (in tropical countries) and abnormal reactions to prescribed drugs such as those used to treat high blood cholesterol/lipid levels. These conditions are usually self-limiting and, with drug-induced myositis, get better when the drug is stopped.

### **What are the symptoms?**

In PM and DM the weakness mainly affects the proximal limb muscles (the muscles around the shoulders, upper arms, pelvis and thighs). The weakness sometimes comes on rapidly, over a period of days, but is usually more chronic, coming on over months or even years. The weakness is usually painless but sometimes aching, discomfort, or mild tenderness of the affected muscles is noted. Pain is uncommon except in acute cases. The pattern of weakness tends to cause difficulty, for example, in standing up from a sitting position, especially from a low chair, rising from a squatting position and managing stairs.

With the arms there is difficulty with lifting or using the hands with the arms elevated. Both conditions can affect the throat muscles, causing swallowing difficulties. Sometimes, severe and acute cases of PM and DM can affect the respiratory muscles (including the diaphragm). In rare cases severe PM and DM can affect cardiac muscle and the lungs. In Dermatomyositis there is skin involvement typified by a rash, involving the face, exposed parts of the chest, and hands (knuckles and nail beds). The rash is made worse by exposure to sunlight.

### **How is the diagnosis made?**

These conditions are relatively rare and it is important that doctors consider the diagnosis in any patient presenting with significant muscle weakness where no other explanation is apparent. It is important that General Practitioners refer such patients to a specialist, usually a neurologist. A useful, but non-specific blood test is the serum CK. This is an enzyme that is present in high concentration in muscle and is measurable in blood. If there is muscle damage the level of CK in the blood is usually elevated and if very high, strongly points to a muscle disorder. Occasionally, however, the CK can be normal or only slightly raised.

The two important tests to establish a definite diagnosis are an electromyogram (EMG) and a muscle biopsy. The EMG, which is done in a hospital department of neurophysiology, involves a very small needle being inserted into several different limb muscles. The electrical activity recorded will show whether muscle weakness is due to a primary disorder of the muscles (a "myopathy") or of nerves supplying the muscles (therefore not PM or DM). Inflammatory muscle disorders may also show a characteristic EMG pattern of spontaneous electrical discharges.

Muscle biopsy is a minor surgical procedure done with a local anaesthetic. This can be done with either a punch/needle biopsy on the ward, or with a small surgical incision in the operating theatre. The author favours the latter as muscle inflammation can be patchy, and the larger specimen obtained with an open biopsy gives more information. The biopsy is usually frozen in liquid nitrogen and a large number of tests carried out to confirm muscle inflammation and to exclude other causes of myopathy. Usually part of the frozen biopsy is stored and can be re-examined at a later date if necessary. Electron microscopy is important in excluding IBM.

### **What is the treatment?**

Most cases of PM and DM respond to corticosteroid drugs such as prednisolone. Unfortunately these drugs have significant side effects if a large dose is used for more than a few weeks. For this reason, corticosteroids are often given with another immunosuppressive drug (to control the muscle inflammation), and there is evidence that the addition of these drugs lessens the dose and duration of steroid treatment needed. The drugs used include azathioprine, methotrexate, cyclophosphamide and cyclosporin. Unfortunately, partly because PM and DM are uncommon disorders, there is little clear evidence as to which of these drugs is the best. There is also limited evidence to suggest that high dose intravenous immunoglobulin (a naturally occurring human blood protein) may be helpful in acute or severe cases. Due to the risk of osteoporosis with steroids, it is important to consider supplements of calcium and vitamin D, and HRT in post-menopausal women. If long-term steroid treatment is likely, a group of drugs called bisphosphonates may help prevent steroid-induced osteoporosis. In some centres osteoporosis can be identified and monitored by a simple bone scan called bone densitometry.

Physiotherapy is important in maintaining the full range of muscle and joint movement, and encouraging as much normal muscle activity as possible. Many patients with significant limb weakness find activities such as swimming helpful in maintaining function.

The results of treatment are variable. Most patients improve, but not all patients make a full recovery. There is some evidence that patients who have slowly progressive weakness of long duration before treatment do less well. If there is no response to treatment, or continuing progression of weakness, the diagnosis should be reviewed. Particularly in older male subjects, the possibility of IBM should be considered. Paradoxically, long-term high dose corticosteroid treatment can cause proximal muscle weakness.

### **Does exercise help?**

Once treatment has been started, active exercise is important in improving muscle strength and mobility. The advice of physiotherapy services is often helpful. Prolonged sitting or bedrest is undesirable as this may increase the risk of muscles becoming weaker and wasted, and also of secondary complications such as osteoporosis or leg vein thrombosis. On the other hand, extreme or prolonged exercise may be harmful. It is important to avoid weight gain. This is not always easy if an individual is relatively immobile or on steroids.

### **Other help**

More severely affected individuals with restricted mobility will benefit from assessment and help from an Occupational Therapist, who may be either hospital- or Local Authority-based. Sometimes special aids and home adaptations are needed. Occasionally wheelchairs are helpful, particularly for longer trips outdoors.

### **The future**

Active research, particularly in the field of immunology, will undoubtedly tell us more about why individuals develop PM and DM and also explain the mechanism of the muscle damage and weakness. Equally important is improving drug treatment, especially identifying which treatments are most effective and free of significant risks and side effects. There is an important need for scientifically sound controlled trials of the various drug treatments already available, let alone new or experimental treatments.

### **Sarcoid Myopathy**

Sarcoidosis is a disease of unknown cause, which is characterised by small areas of tissue reaction, called "granulomas". The condition often affects several organ systems in the body and can affect almost any organ. The most common presentation is with involvement of the lungs, lymph glands or skin.

Muscle involvement as a clinical problem is relatively rare in patients with sarcoid although sarcoid granulomas may be found in as many as 50% in patients with active sarcoidosis if they have a muscle biopsy. The large proportion of these individuals do not have any muscle symptoms.

Some patients can develop an acute myositis (inflammation of muscles) with stiffness, pain and tenderness of the limb muscles. In some this may lead to a more chronic myopathy which tends to affect mainly the proximal muscles (shoulder, upper arm and thigh). This can also develop without an initial acute stage and may lead to wasting of the muscles and chronic weakness. Sometimes sarcoidosis can cause small localised swellings in muscle "nodular myositis". These are sometimes noted by chance but can be tender.

Sarcoidosis can also affect the central nervous system (the brain or spinal cord) and this can cause weakness as a *secondary* effect. Secondly, sarcoidosis can affect one or more peripheral nerves in the arms or legs and this will result in focal weakness (and also impaired sensation) in the area supplied by the affected nerves.

There is no known “cure” for sarcoidosis but some of the acute episodes get better without any treatment being needed and many patients may have only isolated episodes or long remissions. There is no specific treatment for sarcoid myopathy and treatment is largely dictated by severity and by which other organ systems are involved. Most cases respond to corticosteroid drugs (such as prednisolone). If long-term treatment with corticosteroids is likely to be needed, patients should be advised about calcium and vitamin D supplements (and HRT in post-menopausal women) and specific drugs to help prevent steroid-induced osteoporosis. If the condition cannot be controlled with steroids, or if long-term treatment with higher doses is needed, the addition of an immunosuppressive drug may be considered. There is little controlled data about the best drug but methotrexate is often used in this situation.

### Support Group

The **Polymyositis and Dermatomyositis Support Group** offers support and encouragement to families and individuals.

For further information contact:

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### Sarcoid Myopathy

Further information about sarcoidosis can be obtained from the (US-based) website

[www.sarcoidcenter.com](http://www.sarcoidcenter.com).

MC37

Published: 06/02

Updated: 04/08

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