Polymyositis and Dermatomyositis are related illnesses affecting muscle and connective tissues of the body. Joints may also occasionally be involved. A rarer form of Polymyositis is known as Localised Nodular Myositis. Inclusion Body Myositis is often unresponsive to treatment but is not as aggressive in the early stages as other forms of Myositis.

In Polymyositis, the main problems are weakness and inflammation of the muscles. (‘Poly’ means many, ‘myo’ means muscle, ‘itis, means inflammation.)

In Dermatomyositis, the problems are similar to Polymyositis but also include skin rashes. (‘Derma’ means skin.)
Localised Nodular Myositis is a condition appearing in single muscle or muscle groups where painful tender lumps can be felt.

Inclusion Body Myositis is usually progressive and very difficult to treat.

In most of these conditions, the voluntary muscles undergo degenerative changes due to inflammation. The main symptom of these illnesses is muscular weakness, which may be progressive and can be severely disabling. Initially it is not usually painful in children and noticeable weakness may develop gradually over several months.

What Causes Myositis

Because Myositis affect individuals differently, it also probably has many causes. Some doctors think it may be an autoimmune disease. This means that it is a result of a defect in the immune system, which is the body’s natural defence against disease. In healthy people, the immune system produces substances to attack disease agents such as bacteria and viruses. In people with an autoimmune disease, there is a defect in the immune system that causes it to turn against the body’s own tissues. Other doctors feel it may be started by a virus or the combination of a viral infection and defective immune system.

What are the Early Symptoms of these Illnesses?

These illnesses can vary greatly from patient to patient, and few cases are identical and follow the same pattern. Some people may have had the disease for months
or even years before it is noticed. However, the majority find within weeks they have developed muscular weakness. This is sometimes accompanied by pain and tenderness especially in adults. The large muscles about the hips and shoulders are usually the first to be affected. The weakness results in difficulty in walking, lifting arms and getting up from the sitting and lying down positions. There may even be some trouble in swallowing and the voice may become nasal in quality. Other muscles sometimes affected are those in the neck making it difficult to raise the head when lying down. Depression and a general feeling of misery particularly in children, is very noticeable and can be an indication of the disease before any sign of muscle weakness.

Who gets Myositis and at what age does it appear?

These illnesses can affect people of any age and sex. PM and DM affect about twice as many women as men in adult life. In children the ratio is equal. IBM usually occurs in middle to later life and is more common in men than women. Although the condition affects adults and children, the childhood form possibly has different underlying causes and behaves very differently from the adult form. Children can be expected to make a complete recovery. However, it is a rare disease in any of its forms.

Can I catch Myositis from other people?

No! There is no evidence to suggest it can be transmitted to other people as an infection. The disease is also not inherited, and as yet there is no way one can predict who in the general population will be affected.

How do these illnesses differ?

The muscular weakness in Dermatomyositis is accompanied by a patchy, dusky red rash, medically termed, an ‘erythematous’ rash. It usually appears over the cheeks, about the eyes, where it can be violet in colour particularly over the eyelids, on the neck, shoulder, and upper chest. It can be quite prominent over the knuckles and elbows and sometimes on the knees and ankles.

Tiny blood vessels may be seen in the skin in the reddened areas, and the skin may become shiny and tight. In severe cases the entire skin may take on a reddish hue. There may also be a degeneration of blood vessels (vasculitis) underlying the muscle damage and the development of calcinosis especially in children. This is where there are deposits of calcium salt in the skin and in the muscle. In the skin these can feel hard and sometimes can erupt and drain a white, chalky fluid. This may lead to a secondary infection which will need to be treated with antibiotics. Calcinosis may require surgical removal of troublesome deposits. This feature is more common in children.
Polymyositis while defined as an inflammation of the muscle can also be a feature of a more general autoimmune illness such as Systemic Lupus Erythematosus, Scleroderma or even Rheumatoid Arthritis. Other possible symptoms of both illnesses include fever and weight loss. A few people have an extreme sensitivity and reaction to cold that is most often felt in the fingers and toes. The problem is called Raynaud’s phenomenon and is caused by a narrowing of the blood vessels in the fingers and toes. This reduces blood flow and turns the fingers and toes white, then gradually to blue. They may also feel numb and may develop shiny red areas around and under the nails. In this situation it is very important to keep hands and feet warm.

There is no association with malignancy in children and only a weak association with an underlying tumour in adults with Dermatomyositis.

### How is the Diagnosis made?

Experienced doctors at a centre familiar to the disease will recognise the disease after a simple examination. It will then need to be confirmed by the measurement of certain blood enzymes and other tests. Blood tests alone will not confirm the disease particularly in the childhood form. Examination of the electrical activity of the muscle, known as electromyography, and in addition a muscle biopsy maybe done which will help make the diagnosis. This involves a small piece of muscle tissue being taken and then examined under a microscope.

### What is the Treatment for Polymyositis and Dermatomyositis?

Once the disease has been diagnosed most patient respond well to steroids. The childhood form is very sensitive to steroids and practically all of them will respond, whereas the adults are much more variable. A second line of treatment is immunosuppressive medication. Like steroids these drugs suppress the body’s immune system and limit the inflammation. These drugs are monitored under a strict regime, with regular blood tests to monitor their effect and progress. In children the steroid treatment has to be carefully tailored as to the child’s needs, as too little treatment might not adequately suppress the condition. If the patient did not respond to these treatments then intermittent (pulsed) treatment can be given intravenously or plasma exchanges would be considered.

### Why this Medication?

Steroids are prescribed because they are thought to be able to ‘modify’ the immune response against tissue. Immunosuppressants are prescribed because they slow down the immune system, reducing its ability to attack disease agents or healthy tissue. The two may be used together with good effect, but the reasons for this are not fully understood.
**Are there Side Effects from these Drugs?**

Steroid drugs and Immunosuppressants are very powerful agents and can have a number of side effects even when correctly administered. These can be shown by weight gain, rounding of the face, increased hairiness, and easy bruising. More serious side effects such as thinning bones, depression, high blood pressure, cataracts, bringing on or worsening of diabetes, and in very rare cases, bleeding from the stomach. These symptoms are rarely encountered if the disease has been managed correctly, and there has been no overlap or influence from another illness. The sufferer, because the immune system has been suppressed, will be open to increased risk of infection from common ailments they would usually fight off. Also the way these ailments express themselves may not follow the normal pattern when these drugs are taken. In children growth may be restricted.

**Can I stop the Drugs if I appear to feel better?**

No! During the medication by steroids, the body slows production of its own steroids. For this reason, any time the dose is to be lowered, the doctor will gradually reduce the level of steroids a patient is taking, tapering off over a period of weeks or months. During this time the body will gradually increase its own steroid production.

**How long do I take Drugs for?**

Opinions vary among doctors as to how long to continue treatment but the majority prefer to give drugs for a least two to three years. During this period regular muscle function tests are given as a guide to the disease activity and to make sure the necessary dose of drugs are prescribed to control the disease. The period of time may need to be longer, although in children this period can be less if the disease has ‘burnt out’, or is under control until it goes away. While the disease remains active the drugs only suppress its activity. As yet there is no totally satisfactory treatment of myositis which is going to effectively cure all of the patients all of the time.

**What would happen if I refused this treatment by Drug Therapy?**

It is possible that the untreated condition may stop deteriorating and may even improve. However, usually this is not the case and muscles may have been permanently damaged and weakened by inflammation as a result of the lack of medication. The muscles may even shrink and cause deformity. The untreated illness may even be severe enough to put the sufferer’s life at risk. Before the days of steroids and particularly in children the disease carried an appreciable mortality, mainly as a result of involvement also of the breathing and swallowing muscles. Approximately a third got better and about a third might have had some general
improvement, but be left with quite marked disability. The prognosis using modern treatment is better now. Therefore, treatment at a centre familiar with these conditions is essential.

**How do I live with the illness?**

During the active phase of the illness rest is probably advisable, but once the inflammation has died down active exercise becomes important, for without exercises the muscles become weak and wasted. Your doctor, in conjunction with your physiotherapist will decide at what point to change from resting to active exercise. The physiotherapist is also very important, as there is a need to stretch muscles to prevent or limit contractures. Contractures are a condition where the muscle fibres have become fibrous and lose their elasticity. This in turn causes a tightness of the muscle to a degree where joint ranges can become restricted.

Rest is also another aspect that is very important. During times of increased muscle weakness you need to take frequent rest periods during the day, and to limit activity to a tolerable level. As the condition improves and inflammation has died down, fewer rest periods are needed. Complete inactivity is harmful and can actually increase muscle weakness. The physiotherapist is invaluable in advising on an exercise programme to balance rest and activity.

**I have Dermatomyositis What can I put on the rash to clear it up?**

The density of the rash is Dermatomyositis can vary from the very faint to the severe involvement of the skin. These can be seen in some case as a very faint hue over the eyelids. In others it can appear as a type of nettle rash, blisters, or bumpiness. Sometimes there can be redness and scaling of the scalp. These conditions are not always permanent and can be treated with topically applied steroids. The more severe the rash the stronger the strength of the steroid cream. Regular baths using oily creams or emollients are advisable, for these will cleanse the skin without removing the natural oil layer. Detergents must be avoided, even ‘simple soap’.

It is also advisable to remember that in suffering from Dermatomyositis you keep out of harsh, bright sunlight for long periods. Firstly it can effect the medication and secondly, the skin, because of the nature of the disease, becomes very sensitive to the effects of direct sunshine. When going on holiday it is advisable to use a total sun-blocking agent.

**If the Disease does go, will it come back?**

There is always a possibility that this can happen. As it is not fully understood how the disease developed initially, it can be triggered off again. In women there
is always a slight chance that there could be a flare up because of hormonal changes particularly in pregnancy and a medical opinion should be sought before this is envisaged. This can also apply when deciding to take oral contraception.

Can a Diet help?

Some sufferers have found by following a diet and eating certain foods it has been beneficial to their general condition. As yet there is not medical proof to support any claims that diet plays a big role in controlling the disease. However, a balanced, healthy diet containing fresh food etc., low in saturated fat and supplemented in certain kinds of fish oil can to a degree be beneficial.

What is my future?

Children may expect their symptoms to go away. In adults a small percentage do get well within two years. For most it is a case of living with the disease and understanding as much about your illness as possible, so that even during the periods of increased pain and weakness a nearly normal life can be led. It cannot be denied that it is a chronic illness but the disease is rarely fatal, and as long as the prescribed medication is taken the future is always hopeful.

Will I end up in a wheel chair?

Some patients may well need a wheel chair from time to time. Other equipment such as callipers or splints may be needed while the illness is at its height. An unfortunate few may even need more permanent help if there is serious damage of the muscles.

Is there any Research into these Illnesses?

Research directed with an emphasis on all forms of myositis is very limited. However, the Support Group is currently funding research projects in London, Manchester and Bath.

The Support Group funds research in various ways and has spent over £320,000 on Myositis projects.
The Myositis Support Group is a registered charity which covers the following illnesses; Dermatomyositis; Polymyositis; Juvenile Dermatomyositis; Inclusion Body Myositis.