

Juvenile Dermatomyositis

Some questions answered for parents who have just learned about the diagnosis.

What is Juvenile dermatomyositis?

Juvenile (childhood onset) dermatomyositis is a condition, which affects a number of systems of the body, although its main effects are seen in the muscles and skin. The underlying process is an inflammation of small blood vessels (vasculitis) which is thought to result from the presence in the blood of circulating immune complexes - combinations of antigen and antibody which are potent stimulators of inflammation.

Juvenile dermatomyositis is very rare. We think that it may arise in children who have a particular genetic predisposition, which may cause problems in clearing certain viruses from the system. However, it is not contagious or catching, and it does not affect other family members. Fortunately, in most children treatment results in rapid improvement although it will need to be continued for some time and monitored carefully to ensure the optimum response.

It is important to stress that the childhood form of dermatomyositis differs from adult polymyositis in which inflammation of muscle, usually without any other organ involvement, is caused by infiltration of muscle by cells called lymphocytes and from the adult form of dermatomyositis which occurs in older people and may be associated with an underlying tumour. Remember to make this distinction if you read other literature.

What are the symptoms?

Children vary in how quickly they develop their symptoms, but in most weakness and discomfort develops in the muscles over a period of days, weeks or even months. Often children also become uncharacteristically miserable and fractious and they may complain of tummy pain. They develop skin changes which vary from an obvious reddening over the cheeks, eyelids, upper chest, knuckles, elbows and knees to a very faint 'violet' discoloration over the eyelids which may not be obvious even to the most discerning parent. There may be some swelling around the eyes and sometimes the skin looks thin and shiny. The weakness mainly affects the large muscles around the hips and shoulders resulting in increasing difficulty with walking, climbing stairs, getting up from the floor and lifting the arms. Many children complain of difficulty in swallowing and their voice may become 'nasal' in quality. Often the initial symptoms are attributed by parents and doctors alike to a minor viral infection, or if misery is a very prominent feature, to the child being upset or just 'difficult'.

Although you may worry that this has led to a delay in diagnosis, it does not prejudice your child's chance of responding to treatment. The diagnosis is a difficult one even for experts to make and the condition is so uncommon that most general practitioners will never see a case.

How is the diagnosis made?

Your child may have seen a number of doctors before the diagnosis is made at a specialist centre. It may be confirmed by taking blood tests, looking particularly at the level of the muscle enzyme creatine kinase (CK), by carrying out an EMG (electromyography) which detects abnormalities of electrical activity in muscle, by muscle ultrasound which shows an abnormal pattern of echoes in the muscle and by muscle biopsy which may demonstrate characteristic changes in the muscle under the microscope. However, it must be stressed that none of these tests is entirely specific; even the muscle biopsy may be normal, as the changes in the muscles can be patchy. The diagnosis will be based on a combination of the clinical picture and results of some, but not necessarily all, of these tests.

What is the treatment?

Left untreated, the condition leads to increasing weakness of muscle, inflammation of skin and the gut and worsening misery. Fortunately, treatment with steroids in relatively low dosage usually results in dramatic improvement, beginning in days with improvement in mood, followed by a gradual return of muscle strength. Once the response to treatment has been established the aim is slowly to decrease the steroids, but treatment may need to be quite prolonged - often for two years or more. Relapses may occur as treatment is being reduced or in association with chance infections.

Many children make a full recovery on steroid treatment alone. When relapses do occur it is often possible to bring the symptoms under control again by increasing or reintroducing the steroids. However, some children prove more resistant to, or are unable to tolerate steroids and in this situation the use of other drugs which suppress the body's immune system is considered. These include azathioprine, methotrex, cyclophosphamide and cyclosporin and intravenous infusions of immunoglobulins. Very rarely, children can become acutely and seriously ill and in this situation plasma exchange has been shown to produce a temporary improvement while the other drugs are beginning to work. Drugs such as azathioprine are much slower in their action than steroids and may take many weeks to produce the desired response.

Are there any side effects of treatment?

Unfortunately most drugs have some side effects, but left untreated juvenile dermatomyositis is a serious disease. Doctors try to minimise side effects of treatment as far as possible by using the lowest effective drug dosage and monitoring each child carefully as an outpatient. This will usually involve regular blood pressure measurements, urine and blood tests. Your doctor will explain the side effects of each drug and what to look out for. Most importantly, if you are unsure or unhappy about any part of your child's treatment - do ask.

Can exercises help?

Once your child is beginning to respond to treatment active physiotherapy becomes an important part of therapy to help strengthen weak muscles and treat or prevent contractures, which lead to stiffness of joints. Contractures can occur early and compound the problem of muscle weakness. Regular physiotherapy will be started in hospital and arrangements will be made for the local services to take over when your child is well enough to go home. Your child

will have a programme of exercises, which will be made as much fun as possible and you will be taught how to help with these at home. What about the future?

Most children make a full recovery from their disease. However the time taken may vary from a few months to several years in some cases. In a minority there may be some permanent weakness and contractures, although with the advent of cyclosporin therapy many such children have made significant progress. A rare complication of juvenile dermatomyositis is calcinosis - the formation of chalky material under the skin. This tends to occur when the muscle inflammation has settled and may improve only very slowly. In some instances surgical removal of the deposits may be necessary. All children should be encouraged to be as active as possible except during the acute disease and, above all, parents should be optimistic about the future.

Families affected by Juvenile Dermatomyositis wanting to meet others may wish to contact the support group:

Dermatomyositis & Polymyositis Support Group,

146 Newtown Road,

Woolston,

Southampton, SO2 9HR

Tel: **023 80449708**

Email: msg@myositis.org.uk

Web: www.myositis.org.uk

MC14

Published: 11/02

Updated: 04/08

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