

JUVENILE
DERMATOMYOSITIS

JDM



NEW ZEALAND
PAEDIATRIC RHEUMATOLOGY SERVICE



What is Juvenile Dermatomyositis?

Juvenile dermatomyositis is an inflammatory disease of the small blood vessels (“vasculitis”) that primarily affects the skin and muscle.

This inflammation leads to a skin rash and muscle weakness. It is a rare disease which occurs equally in boys and girls.

What causes Juvenile Dermatomyositis?

The cause of dermatomyositis is not yet known. We do know that the symptoms are caused by an abnormal reaction of the body’s immune system. Our immune system protects our bodies against infections caused by foreign invaders such as bacteria and viruses.

In a child with an auto-immune disease like dermatomyositis, their immune system attacks

their body’s own cells. This results in inflammation in the blood vessels in the skin and in the muscle. Sometimes other organs and body systems are involved.

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Why my child?



There is nothing that you did, or did not do, that caused your child to get dermatomyositis. Dermatomyositis is not contagious which means your child did not catch it from someone else, nor can anyone catch it from your child.

Dermatomyositis almost never occurs in other family members, so there is almost no chance of your child’s brothers or sisters getting the disease.



How is JDM diagnosed?

The diagnosis is based on both the physical examination and laboratory tests.

In some children the signs develop slowly over a period of months and in others the signs develop much more quickly. The doctor will ask many questions about your child's symptoms and perform a detailed examination paying special attention to skin and muscles.

What investigations will my child have?

Blood tests

Blood tests help to diagnose JDM and to monitor the activity of the disease. The most important of these are muscle enzymes, such as Creatinine Kinase (CK), transaminases (ALT & AST) and Lactic Dehydrogenase (LDH).

Magnetic Resonance Imaging (MRI)

This technique can help the doctors identify areas of inflamed muscles and also directs where to do a muscle biopsy if necessary. MRI does not use radiation and is a safe procedure. Younger children may need an anaesthetic so they do not move while the test is being performed.

Muscle Biopsy

This is where a small piece of muscle is removed through a small needle or surgical incision and then examined under a microscope. This is usually not required.

Other tests

Usually other tests are done to check for involvement in other organs. For example:

Heart-electrocardiography (ECG) and echocardiogram (ECHO)
Lungs-Chest Xray and High resolution CT scan and lung function tests
Swallowing-Speech Language Therapy assessment and Video fluoroscopy.

What are the signs of Juvenile

Dermatomyositis?

Rash

The typical rash is a purple-reddish discoloration of the upper eyelids known as a heliotrope rash. Sometimes there is swelling around the eyes. The knuckles, elbows and knees are also involved with red, thick scaly patches. Occasionally the skin may break down. The rash often gets worse with sun exposure.

Muscle Weakness

This usually occurs gradually and can be preceded by muscle pain. Weakness generally occurs in muscle groups close to the trunk such as the muscles around the shoulders, neck, thighs and hips. Common movements like climbing stairs and getting up from the floor or chair may become difficult. If weakness becomes severe, all muscles can be affected including those involved in breathing, swallowing and talking.

Joint Pains

This is a less common complaint but your child may complain of stiff and sore joints. The joints or the muscles around them may become inflamed. This does not usually last long or cause joint damage.

Calcinosis

Some children can develop small lumps of calcium under the skin. They can vary in size and feel like little rocks under the skin. Sometimes they break through the skin and drain. Usually they disappear

Other important things to think about.

Physiotherapy/Occupational Therapy

It is important to strengthen weak muscles & stretch stiff joints. The physiotherapist will assess your child and give an individualised exercise programme. The occupational therapist will assess how you are coping with daily activities at home, school and the things you enjoy doing.

The therapists will do a formalised muscle test called a CMAS (childhood myositis assessment scale) at most visits.

Complimentary and alternative medicines

There is no evidence that any herbs or natural medicines are helpful in dermatomyositis. If you would like to take an alternative therapy please tell a member of the Rheumatology team.

Diet

There is no special diet which will cure your child's illness. a well balanced diet that includes a variety of foods is the key to healthy nutrition.

What medications will my child have to take?

There is no cure for dermatomyositis but there is treatment to control the inflammation and prevent damage to the muscle and skin. Your child's treatment regimen may change from time to time depending on how dermatomyositis is affecting his/her body. The common medicines we use are explained below.

Corticosteroids(Prednisone and Methylprednisolone)

These are the most effective drugs to control the disease, wherever it is in the body. They are steroids but not the same kind of steroid drug that some athletes take. They act by suppressing the immune system and decreasing inflammation. If they need to work fast they can do so. Unfortunately they do have side effects, which is why your doctors try to control the inflammation using other medication in the long term.

The side effects are related to the dose and duration your child is on the medication. At the beginning you child will probably have high doses of prednisone, but as they improve the dose will gradually be lowered.

Some common side effects you may notice are increase in appetite, weight gain, acne, mood swings, hair on face, "moon face".

Methotrexate

This is an immunosuppressive drug, which means that it helps to suppress the cells that activate inflammation. Methotrexate is usually used in combination with prednisone and allows doctors to decrease your child's prednisone while keeping the disease in control. It is well tolerated with few side effects. It is given once a week as a pill or subcutaneous injection.

Cyclophosphamide

Cyclophosphamide is a strong immunosuppressive drug that may be used in more severe forms of dermatomyositis.

Intravenous Immunoglobulin

This contains human antibodies concentrated from blood and is given into a vein as an infusion. It is recommended for a severe rash which has not improved over time.

There are other immunosuppressive drugs which may be prescribed to help control our child's illness and these will be discussed in detail by your doctor.

What signs should I watch for when the disease is active?

If your child experiences any of the symptoms listed below you must notify your GP and the Rheumatology Team.

- a change in your child's voice(nasal or softer sounding)
- trouble chewing foods or swallowing liquids
- choking or excessive drooling
- trouble breathing or shortness of breath
- tummy pains (JDM can affect the bowel leading to ulcers or perforation in more severe cases)

Juvenile Dermatomyositis

What should I expect when I come to clinic?

There are a number of people that make up the Rheumatology team. Your child will be seen by the Nurse Specialist, Paediatric Rheumatologist, Physiotherapist, Occupational Therapist and perhaps by a rheumatology fellow (a doctor training in Rheumatology). It is necessary to examine your child at each visit and this will include strength testing. A blood test will also be done, as these tests help the doctor in determining disease activity.

Your child will have to miss school for scheduled clinic visit. When you are first diagnosed you will be coming to the clinic fairly frequently (usually, weekly -two weekly) As your child improves the frequency decreases to about once every two to three months. Each appointment will last 1-2 hours.

What can I do to help?

Raising a child can be challenging at times. Raising a child with a chronic illness

requires even greater patience and understanding. It is only natural to have concerns and worries. Some parents feel guilty and may over indulge their child. Far too often parents expect too much from themselves and feel they are responsible for their child's illness. Families can learn to adapt to their child illness and grow from this experience.

School

It is important that your child get back into a normal school routine as soon as possible. Most children with dermatomyositis can manage school full time. A few children have to miss school or only go part-time for a week or two after treatment has been started. It is rare to need to miss school after this time, except for clinic and therapy appointments.

What special precautions should we take?

Chicken Pox

Children who develop chicken pox when taking prednisone or other immunosuppressants can become very ill. If your child is in direct contact with a child with chicken pox they may need to have Varicella Immune Globulin (VZIG) within 72 hours of contact. If your child develops chicken pox they may need an antiviral medication to lessen the severity of the illness.

Sunscreen

In New Zealand we are all more than aware of the harmful effects of the sun. Kids with JDM need to pay particular attention as the rash can become more active with sun exposure.



Slip, Slop, Slap!!

If you have questions or concerns at any time please feel free to contact us.

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