



# Juvenile Dermatomyositis

*A Parent's Guide*



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The information in this document is intended solely for the person to whom it was given by the health care team.



Developed by the health care professionals of the Pediatric Rheumatology Program with assistance from the Department of Learning & Development.

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## What is Juvenile Dermatomyositis?

Juvenile dermatomyositis is an inflammatory disease of 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 with the average age of onset at 7 years. The disease occurs equally in boys and girls.

## What causes Juvenile Dermatomyositis?

The cause of dermatomyositis is unknown. 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. The end result is inflammation in the blood vessels in the skin and in the muscle. Sometimes other areas of the body are also affected.

## Why my child?



Nothing you did, or did not do, 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. There is no strong genetic factor involved.

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

## Will my child die?

Before the use of steroid therapy (prednisone or methyl prednisone) about 1/3 of children with this disease died, 1/3 were left severely handicapped, and 1/3 recovered completely. With the routine use of prednisone and other medications the majority of children recover. It is now very rare for a child to die, but this remains a risk if vital organs such as the lungs, heart or bowel become involved.

## What are the signs of Juvenile Dermatomyositis?

The signs and symptoms of dermatomyositis vary widely and may be mild or severe. Generally children have mild symptoms which slowly worsen over a number of months. Less frequently, children develop severe weakness very quickly, with or without rash. Specific signs of dermatomyositis include:

- **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 called “Gottron’s papules”. Occasionally the skin may break down. The rash is photosensitive and may worsen with sun exposure.
- **Weakness:** Muscle weakness usually occurs gradually. Often parents notice that their child can’t “keep up” or mistake the weakness for laziness. As the weakness gets worse children often have difficulty going up and down stairs or getting up from the floor. Weakness generally occurs in the muscles of the shoulders, neck, thighs, hips, and trunk. If weakness becomes severe, the muscles of the gastrointestinal tract and chest wall can become involved, causing difficulty swallowing, a nasal sounding voice and, in severe cases, difficulty breathing.



- **Joint pains:** This is a less common complaint but your child may develop stiff and swollen joints. This inflammation does not usually last long or cause severe joint damage.
- **Calcinosis:** Some children can develop small lumps of calcium under the skin or in the muscle. This process usually occurs later in the disease. The lumps may feel like little pebbles under the skin and are generally painless unless they are bumped. Sometimes these lumps break through the skin and drain a creamy white liquid. Usually they disappear over a long period of time.
- **Lipoatrophy:** Lipoatrophy is an uncommon condition associated with juvenile dermatomyositis. It generally occurs late in the course of the disease and seems more likely if the disease is particularly severe at the beginning. It is characterized by a loss of fat from the face and limbs. There may also be problems with sugar metabolism which is why all children have their blood sugars monitored. We don’t know why some children, and not others, develop this unusual complication.



## How is the disease diagnosed?

Dermatomyositis is not always easy to diagnose. In some children the signs develop slowly over a period of months and in others the signs develop much more quickly. The diagnosis is based on both the physical examination and laboratory tests.

The pediatric rheumatologist (a pediatrician who specializes in the diagnosis and treatment of children with inflammatory disease of the joints, muscles, blood vessels and skin) will evaluate the history of your child’s condition, and perform a detailed physical examination.

Blood tests are important in helping to diagnose dermatomyositis and also help to monitor the activity of the disease. Usually a blood test is done at every clinic visit. These tests show if there is inflammation in the muscles or in other areas of the body. Which of these laboratory tests are most indicative of disease activity is a little different in each child. The most common tests include:

- **Muscle Enzymes**  
When muscles are inflamed, certain muscle proteins leak into the blood. The four that are generally measured in dermatomyositis are CK (creatinine kinase), ALT (alanine amino transferase), AST (aspartate amino transferase), and LDH (lactic dehydrogenase).
- **Von Willebrand's Factor**  
Von Willebrand's factor is released from cells lining blood vessels. In some children with dermatomyositis, this protein can be elevated when the disease is active, and can help measure disease activity.
- **Erythrocyte Sedimentation Rate (ESR)**  
The ESR is elevated when there is inflammation in the body. It is not specific to dermatomyositis and can go up in various illnesses including colds and flu.
- **Autoantibody levels**  
There are a number of tests that are done to determine dermatomyositis is not part of a more complex disease such as lupus. These tests are not usually done at every clinic visit, but at the time of diagnosis.
- **Lipid Profile**  
At diagnosis your child will have lipids and blood sugars measured as these can sometimes be abnormal and could mean the development of lipodystrophy.



## What investigations will my child have?

- **MRI (magnetic resonance imaging)**  
This technique can help the doctors identify areas of inflamed muscles and also directs the doctors where to do a muscle biopsy if necessary. MRI may be done later in the disease to assist the doctors in monitoring disease activity. MRI does not use radiation and is a safe procedure. Younger children who need an MRI will need anesthesia (to be put to sleep) for the procedure 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 done under general anesthetic.
- **Electromyography**  
This is a test that measures the electrical activity of the muscles. It is performed by inserting a small needle into the muscle to be tested, and then stimulating the muscle and recording its electrical response. This test can help show whether muscle alone is inflamed or if nerves may be involved as well.
- **Bone Density Scan (Dexa-scan)**  
This is a special x-ray that looks at the architecture of your child's bones (how strong your child's bones are). Inactivity and muscle weakness make bones less strong and one of the medicines, prednisone, can cause your child's bones to be more brittle. We monitor bone density through this special x-ray every 2 years or sooner if abnormal.

## Are there any special shots my child should or shouldn't have?

If your child is on large doses of prednisone your doctor may wish to withhold some childhood vaccinations. If you are unsure always check with your clinical nurse or doctor.

- **Flu Vaccine**

It is wise to have your child, and in fact all your family members, receive the flu shot every year in October or November. This can be given by your family doctor. Flu can sometimes be a very serious illness, and if a child with dermatomyositis on steroids gets the flu they may not be able to fight the infection very well.



## 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 doctor:

- 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 (dermatomyositis can affect the bowel, leading to ulcers or perforation in severe cases)

## 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.

- **Prednisone**



This is the most effective drug to control the disease. It is a steroid but is not the same kind of steroid drug that some athletes take. This drug works by suppressing the immune system and decreasing inflammation.

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

Some side effects you may notice are:

- increase in appetite
- increase in weight
- acne
- mood swings
- stretch marks
- "puffy face"
- hair on face
- slower growth

Your child may also develop side effects that you can't see such as:

- weak or brittle bones (osteoporosis)
- avascular necrosis. This is rare but prednisone can cause the blood supply to the bone to decrease, causing part of the bone to die.
- high blood pressure
- increased susceptibility to infection
- cataracts. This is a cloudy area in the lens of the eye. These cataracts are different from the cataracts older people get, and they usually do not affect vision.



After reading about these side effects, we understand you may feel anxious about prednisone. However prednisone is the best drug for treating dermatomyositis which may otherwise be a fatal or destructive disease. Remember most of the side effects are temporary and will go away as the medication is reduced.

Prednisone is similar to steroids naturally produced by the body. Treatment with prednisone fools the body into shutting off its own steroid production. These chemicals are vital to regulation of normal daily body function. Therefore **never** stop your child's prednisone abruptly as your child could get extremely sick. If your child becomes ill with a vomiting or diarrhea type of illness, his/her body will not produce enough "stress hormones" to deal with the illness and he/she will need more prednisone. If your child is unable to take their prednisone due to a vomiting illness, he/she will need to receive an equivalent measure of prednisone by injection. Please call your doctor if your child becomes sick.

As your child gets better, the rheumatology team doctors will reduce your child's prednisone slowly guided by symptoms and blood tests.

### Intravenous Methyl prednisone

This drug is a steroid similar to prednisone except it is given into a vein in your child's arm. It is often given weekly or monthly in very high doses. It is often given in addition to daily oral prednisone at the beginning of the illness, or if there is a flare.



There are fewer side effects when giving steroids this way but for the long term, it may not be as effective as prednisone by mouth.

### Methotrexate

Methotrexate is an immunosuppressive drug, which means 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

### Cyclophosphamide

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

### Rituximab

Rituximab is an immunosuppressive drug that may be used for children with severe dermatomyositis who have not responded to usual therapies.

### Hydroxychloroquine (Plaquenil)



Plaqueuil is a drug sometimes used to help treat severe rash in dermatomyositis. It is only effective in a small number of patients, but it is a safe

drug with relatively few side effects. Plaquenil very rarely damages the retina of the eye. However at the dosages that we use in our clinic, this has not been a problem. We do ask that you have your child's eyes checked every one to two years by an eye specialist (ophthalmologist).

### Intravenous Immunoglobulin (IVIG)

This infusion is used to help improve the rash in dermatomyositis. It is recommended for a severe rash which has not improved over time. It is still not clear how much benefit, if any, this drug has on the muscle disease.

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

## What special precautions should I take?

### Chicken pox

Children who develop chicken pox while taking prednisone can become very ill. If you child has been in direct contact with a child with chicken pox (and has never had chicken pox, or varicella vaccine) they should receive Varicella Immune Globulin (VZIG) within 72 hours of contact. Your family doctor should be contacted as soon as possible. If your doctor is unavailable, take your child to the local hospital emergency department.

If your child develops chicken pox, contact your family doctor that day. Your child may need an antiviral medication to lessen the severity of the illness.



### Sunscreen

Very often the dermatomyositis rash can become more active with sun exposure. A sunscreen lotion with a sun protection factor (SPF) of 30 or higher is recommended.

### Diet

There is no special diet which will cure your child's illness. A well-balanced diet that includes a variety of foods is key to healthy nutrition. If your child is on big doses of prednisone there are a few key points to remember:

- Avoid foods high in salt, such as fast foods.
- Encourage your child to eat foods high in calcium (e.g. milk, cheese) to help keep bones strong. It can be difficult for some children to receive enough calcium through their diet and doctors may prescribe a calcium supplement such as Tums.
- Remember that prednisone will cause your child to feel hungry. Try and provide foods low in calories and high in nutrients such as fruits and vegetables. Healthy food choices may help slow down prednisone related weight gain.

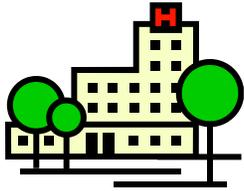


### Complimentary and alternative medicines

There is no evidence that any herbs or natural medicines are helpful in dermatomyositis. Some supplements may actually be harmful. Discuss complementary and alternative medicine use with the doctors or clinic nurse before starting any of these treatments.

### Medi-alert Bracelet

This bracelet (or necklace) will identify your child's diagnosis and the medications they are on. In the event of an accident, it is important for medical staff to know that your child is on prednisone.



## What should I expect when we come to the clinic?

Your child will initially be seen by the clinic nurse. You may then be seen by a pediatric resident (doctor training in pediatrics) or a rheumatology fellow (pediatrician training in rheumatology). You will always see one of the rheumatologists. 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 visits. When your child is first diagnosed, you will be coming to the clinic fairly frequently (usually about every two weeks). As your child improves, the frequency decreases to about every two to three months. You will need to set aside at 1 to 2 hours for each appointment.

The clinic visits are very important as they enable the doctors to monitor your child's disease activity and make adjustments in their medication(s).

- **Physiotherapy and Occupational Therapy**

Your child will usually see our physiotherapist and occupational therapist at each clinic visit. Assessing your child's muscle strength is very important in determining improvement. The therapists will do a formalized muscle test called a CMAS (Childhood Myositis Assessment Scale) at most visits. Early in the disease course passive stretching is recommended to prevent the muscles from tightening up. Once the active inflammation has been well controlled, strengthening exercises and range of motion exercises may be recommended. Restrictions on your child's activities are almost never necessary and in fact we encourage your child to return to their activities as soon as they feel able.



- **Research**

Research is an important part of our clinic and you may be asked to participate in research studies related to your child's dermatomyositis. Remember, you are never obligated to participate in research and your child's care will not differ whether you decide to participate in research studies or not. Participation in research helps us understand dermatomyositis and may benefit your child and other children with dermatomyositis.



## What can I do to help?

Raising a child can be a challenge 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's illness and grow from this experience.

- Take one day at a time. You do not have to learn all medical terms and procedures in one day. Your nurse and physicians are always available to answer any questions.
- Take good care of your own physical and emotional needs. You will be most helpful to your child when you are feeling your best.
- Talk things over regularly with all members of your family and share your feelings. Communicating openly helps prevent tensions and fears that can add stress to family life.
- Do not overly protect your child. On the one hand you will want to protect your child; on the other hand you will want to encourage your child's independence. It is best to strike a balance.

- Discipline must not change for your child with an illness. It is sometimes easy to feel sorry for your child. However, your rules and expectations should continue as they would have before your child got ill. Your child does not want to be treated “different”.
- Assign chores that your child can manage. When you have the expectation that the child contributes to the “family work” you give them the message that they are valuable contributing members of the family.
- Remember to discuss your concerns and questions with the health care team. Write your question down and bring them to the clinic. Sometimes speaking to another parent for support can be helpful.
- **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 half time for a week or two after treatment has been started, but it is rare to need to miss school after this time, except for clinic appointments. The occupational therapist and nurse can help communicate with the school if any adjustments need to be made to your child’s school program, such as missing gym for a while or needing extra time to get around the school.

### What about the future?

Dermatomyositis is variable in its severity and it is difficult to know how long your child will require treatment. Generally children are started on a high dose of prednisone and then the dose is decreased over time depending on your child’s response. The goal is to be able to discontinue the prednisone, and continue on methotrexate for a period of time for disease control;

however it is difficult to predict when this will occur for your child. Most children require medication for at least two years. Some children may have a disease flare as the medications are being withdrawn; there is usually no particular trigger for disease flares. Some children may require treatment longer than two years, or require additional drugs. Most children will recover completely from dermatomyositis with no ill effects.

Unfortunately a small proportion of children continue to have active disease for many years. They may develop muscle wasting and contractures (shortening of the muscle) which may leave them with some physical limitations.

Once your child has completely recovered from the disease it is very rare for the disease to recur.

Hopefully this booklet has answered some of your questions regarding dermatomyositis. A good understanding of the disease will enable you to help your child cope well with their illness with little if any changes to your family’s lifestyle.