

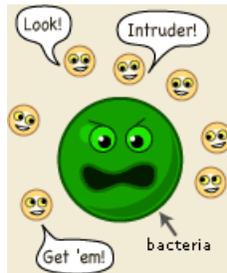
Juvenile Dermatomyositis - What does it mean to me?

(Information for children and young people with JDM)

Juvenile Dermatomyositis is a long name so we often call it **JDM**. Being told you have JDM can leave you wanting to ask many questions. The team looking after you will always be happy to answer your questions. This leaflet is to help you learn more about JDM and the experiences you may have.

What is Juvenile Dermatomyositis (JDM)?

JDM is an autoimmune disease (illness). The immune system protects the body against infections. In autoimmune diseases, the immune system attacks the body itself. This means it starts to damage the body rather than protect it. JDM affects the very small blood vessels. This can cause problems in the skin (**derm**), and muscles (**myo**), causing them to become inflamed (red and sore). This is **Dermatomyositis**. Dermatomyositis can occur in adults (grown-ups) but this is often different from the diagnosis in young people. Therefore, if you are under 16 years old we call this disease Juvenile Dermatomyositis or JDM.



Picture courtesy of www.nobelprize.org

What symptoms (complaints) will I get with JDM?

You might have noticed you have some symptoms (complaints) of JDM, so some things in this list will be familiar. Most people with JDM will not have all the symptoms below.

Muscle pain and weakness

This occurs more often in the muscles near the centre of the body. These include the upper arms, thighs and neck. It can affect any muscle in the body. Some people have difficulty swallowing or a change in the sound of their voice. You may have noticed you have had problems getting up from bed, climbing up stairs, brushing your hair and getting up from the floor or a chair.

Skin rashes

Rashes often appear over the face, knuckles, elbows, knees and ankles. They may be photosensitive which means they become worse in sunlight. They may appear before, after the muscle weakness, or at the same time. Some of these rashes have special names. All of the rashes (and other complaints) get better over time with treatment.

A purple rash can occur over the eyes - called a **heliotrope rash**, named after a heliotrope flower. A rash can occur on the face called a **malar rash** - also called a 'butterfly rash.'



Heliotrope rash



Heliotrope flower



Malar rash



Scaly patches can occur on the knuckles, elbows, knees or ankles called **Gottron's papules**. The tiny blood vessels around the nails can also become red and irregular called **nailfold changes**. Lumps of calcium can appear under the skin (**calcinosis**) and sometimes leak through the skin. These can be tiny or bigger lumps or sheets.



Gottron's papules



Nailfold changes



Calcinosis



Other symptoms (complaints)

- 👉 Tiredness
- 👉 Irritability
- 👉 Joint pain (**arthralgia**) and / or swelling or stiffness in the joints (**arthritis**)
- 👉 Temperatures
- 👉 Mouth ulcers
- 👉 Headaches
- 👉 Hair thinning or hair loss (**alopecia**)
- 👉 Change of colour of the hands in the cold (from white to blue to red - called **Raynaud's phenomenon**)
- 👉 Chest pain
- 👉 Tummy pain / bowel problems (such as diarrhoea or blood in the poo)

Some of these symptoms occur in people who do not have JDM.

What causes JDM?

We do not know exactly what causes JDM. Lots of doctors and scientists are working to try to find out why some people get JDM. We know that everyone has thousands of genes in their body that contain messages that play a part in what they look like. Your parents have passed genes to you. It is likely that some of the genes you inherit from your parents mean you are more likely to have an autoimmune disease like JDM but this is not the whole story. We think that there may be certain things that 'trigger' the start of JDM. These could include some infections (germs) or sunlight. This means that when you come into contact with a trigger it can lead to your immune system getting confused and attacking the body causing JDM. JDM is not contagious – this means that you do not 'catch it' from somebody else and you cannot pass it onto another person.

Why me? Is it my fault?

JDM occurs by chance. It is not your fault or your parents' fault. It is not something that you have done (or not done) to make you have JDM.

Can JDM be prevented?

We do not know how to stop people getting JDM. There is nothing that you can do to stop you getting JDM, but you can help to make JDM go away by taking your medicine and following the advice that your doctor gives you. Your parents will help you do this.

Is JDM the same for everyone?

Everyone's experience of JDM is different. Some people will have a very mild disease (illness) with few symptoms (complaints). Others will have a more serious disease that makes living a normal life much harder. Some people need strong treatment to stop them having problems in the future. You can ask the team looking after you to explain to you how serious your disease is.

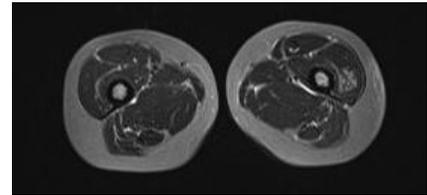
How does my doctor know I have JDM?

(Investigations for JDM)

The symptoms you have will have led your doctor to think you may have JDM. Blood tests are very important to help find out what is wrong. They also help to keep an eye on the disease and treatments. Several blood tests are needed but they can normally be taken from one sample when you attend clinic.

Other tests – you may or may not need some or all of the following tests:

MRI can look closely at the muscles in your thigh. It consists of a large tube with powerful magnets. You will need to lie still in this tube for 20-30 minutes. The scan does not hurt but it is quite noisy.



MRI of thighs with JDM

X rays may look at your chest or joints. They can also show calcinosis (lumps of calcium under the skin that show up on X-ray) in the arms or legs. Only some people with JDM have calcinosis and so you may not need to have this done.

ECG and ECHO look at the function of your heart. Neither test hurts. An ECG involves sticking some leads to your chest using sticky plasters and monitoring how the heart beats. An echocardiogram involves an ultrasound of the heart. (Women have ultrasounds of their tummy when they are pregnant).



ECG: Picture courtesy of www.nobelprize.org

Abdominal Ultrasound looks inside your tummy. It does not hurt but the jelly may feel a bit cold.

Lung function tests look at how well your lungs work. If you have this done, you will need to blow into a special machine.

CT scans look inside your chest. This involves lying still in a tube (a bit like an MRI) for a short period. It does not hurt.



CT picture of the Lung

A Speech and Language Therapist may see you if you are having difficulty swallowing. In some cases, an X-ray assesses swallowing - called a video **fluoroscopy**. It involves swallowing some special liquid whilst taking x-rays.

Muscle biopsy takes a very small bit of muscle from the top of your leg to look at it under a microscope - usually while you are asleep under a general anaesthetic.

Skin biopsy takes a small sample of skin to look at under a microscope - at the same time as a muscle biopsy or at a different time.

EMG is a test to see if there is a problem with your muscles or not. Very small needles called electrodes are put into a muscle to measure its electricity. This can be a bit uncomfortable but you may not need to have this test.

What happens in clinic?

Every time you come to clinic, you will see your doctor and may see a physiotherapist and a nurse. They will ask you questions about how you have been feeling since your last clinic

appointment. The doctor will then examine you and will want to listen to your heart and lungs, feel your tummy and examine your joints. The doctor / physiotherapist / nurse will also want to see how strong you are and they may test muscle strength in your arms, legs, neck and tummy. Blood tests are usually taken when you come to clinic to monitor your illness and the medicine that you are taking. You may need blood tests regularly at the start of your illness but in time, you will not need blood tests as often. You will be offered a gel to numb your skin before the blood test so that the needle does not hurt. If you have a flare (worsening) of your disease, you may need to undergo more investigations such as an MRI, X-rays, ECG, ECHO or CT.

How will I get better?

(Treatments for JDM)

A team of people will be there to look after you and will make you feel better. This team is likely to include doctors that specialise in JDM, specialist nurses and specialist physiotherapists. You may also see other professionals as part of a team including occupational therapists, psychologists and pharmacists. There are medicines to help you get better. These medicines – called immunosuppressants - reduce the activity of the immune system. They aim to reduce inflammation (pain, redness, swelling) caused by the immune system attacking the body. Your treatment may change over time depending on how JDM is affecting you. It is very important that you take your medicine regularly. Medicines for JDM work in the background over time - so you may not notice a difference if you miss one dose but the medicines will not work as well if you miss several doses.

The immune system is there to help your body fight infections and stress. In JDM, the immune system is over-active and so the medicines that you take reduce the action of the immune system. This may mean that you get infections because your immune system may not be able to fight infections as well as it otherwise would. If you become unwell you should see your doctor. The medicines for JDM may have some side effects (give you new symptoms). Your medical team can talk to you about these before starting any treatments. The medicines used are the best for treating JDM. Your doctors will use the lowest doses possible to try to limit side effects. It is important you follow advice from your doctor about how to reduce your treatment as some of them must not be stopped suddenly.



Picture courtesy of www.nobelprize.org

Is there anything I can do to help make my JDM better?

As well as taking your medicines as you should do there are other things you can do to help improve the symptoms of JDM.



Sun Protection

Sunlight can make JDM rash worse or can lead to a flare of the illness. To prevent this, use a sunscreen of factor (SPF) of 50 or more. Apply it 30 minutes before going out even on cloudy days and do not forget your ears. Protect exposed areas with a hat and long sleeved shirts. Sometimes the rash may get worse with indoor lighting. So check whether lights in the home or school could be affecting you.

Pace Yourself

Tiredness is very common in JDM. You may have a limited pot of energy and so it is important that you do not exhaust yourself. You should ask for help when needed. It is all about taking things at a steady pace and gradually increasing activity over time.



Exercise

It is always important to exercise, but during a flare of JDM, it may become very difficult. The physiotherapist will help by teaching you the right kind of exercises that will help build up muscle strength and energy and increase the movement in your joints. Once you are feeling better, it is important to get back to doing regular physical activity but take things at a steady pace and increase gradually.



Diet

There is no special diet to cure or prevent a flare of JDM. We suggest a well balanced diet that includes different kinds of foods. Remember that some medications can cause you to feel hungry and this increase in appetite is not your fault. Trying to replace crisps and sweets with fruit, vegetables, nuts and seeds may help to slow down any weight gain.



Sleep

You may feel more tired than usual due to JDM. This will get better over time with treatment. Having a regular sleeping pattern and routine to your day will help you to have more energy.



Vaccinations

We have vaccinations to help stop us getting certain infections. While you are taking medicine for JDM, there are some vaccinations you cannot have. However, it is very important you have the vaccinations that you can have safely to help avoid nasty infections. You may also need extra vaccinations like the flu vaccination because of your disease and the medicines that you take.

Will I be on treatment forever?



Everyone's experience of JDM is different. Some people will only have symptoms for a short time while others may have the disease for many years. It is very unlikely that you will need to take medicine forever. Your doctor will try to control your disease so you can lead a normal life and do everything you wish. It is only possible to reduce or stop your medication when you have been very well with no symptoms (complaints) for some time (usually at least a year). Stopping your medication too quickly can cause a flare (worsening) of your disease. It is important you talk to your medical team about when and how to reduce your treatment in the safest way to stay well.

How do I know if I am having a flare (worsening) of JDM?

You may feel weak, have a rash or feel generally unwell. If you are worried about the possibility of a flare (worsening), you should see your specialist.

Will JDM affect me in school?

When the disease is active, you may become more tired than usual at school and may find it difficult to concentrate. Muscle weakness can make it difficult to walk around school. Some children may need to have shorter days at school but this will increase to full days in time. You should take part in exercise but when the disease is active, you may need to miss PE. Your physiotherapist will be able to guide you on this.

Can I get extra help from school if needed?

Schools can help to make it easier for you to join in. You may need to focus on core subjects, reduce your workload, and have time to catch up or rest or have extra time to get to classes. Your medical team can help by talking to the teachers at school if you want them to. If you ask them to, your medical team can write to teachers and provide more information about JDM. Many young people with JDM have gone on to university and many have gone on to have good jobs.

Is there anything that I will not be able to do because of my JDM?

Initially you may struggle with everyday activities. It may take a while for you to get 'back to normal'. With treatment, over time, you should be able to lead a normal life carrying out all of the activities that you wish.

Will my puberty or growth be affected by JDM or by the treatments?

JDM can affect growth and puberty. The best way of improving growth is to control the illness. Some medications can also affect growth, but doctors will ensure that you have as low dose as possible or use alternative medications where possible. When there is

inflammation (redness, pain, swelling), puberty (bodily changes when you become an adult) often occurs a little bit later than usual. If this happens, it actually means that you will have more time to grow! Some children or young people may see a specialist growth doctor called an endocrinologist.

What is in the future for me?

You should be able to lead a normal life, firstly taking medicines and in time, without medicine. We would expect you to gain normal muscle strength and energy over time and be able to carry out all activities, including sporting activities. Many children with JDM will have one episode of JDM which may last for 2-3 years and then goes away (enters remission). Other children will have a several episodes where the disease can come back after a period of remission. Some children have a more prolonged disease than can last many years (a chronic course). Occasionally people may have long-term problems due to damage in their muscles or skin. It is important for a specialist to treat JDM early and properly (or fully) to help avoid problems later in life.

Will I be able to have a family in the future?

You should be able to have a healthy child in the future, despite having JDM. Some people cannot have children for other reasons. Some medicines can affect fertility (your ability to have a child). However, this only happens with very few medicines when high doses are given and doctors will ensure that they give you as low a dose possible to treat the disease. There are some medicines used to treat JDM that should not be taken whilst pregnant as they may damage an unborn child. It is therefore important that you talk to your doctor if you want to become pregnant. Whilst you are taking these medicines, you should be using contraception to ensure that you do not become pregnant if having sexual intercourse.

Will I meet other children with JDM?

JDM is very rare. It only affects approximately four children in every million each year. However, your specialist will be treating other children with JDM. If you would like to meet them or talk to them, ask your specialist.

