www.printo.it/pediatric-rheumatology/IE/intro

Juvenile Dermatomyositis

Version of 2016

1. WHAT IS JUVENILE DERMATOMYOSITIS

1.1 What kind of disease is it?

Juvenile dermatomyositis (JDM) is a rare disease that affects muscles and skin. A disease is defined as "juvenile" when it starts before the age of 16.

Juvenile dermatomyositis belongs to a group of conditions that are thought to be autoimmune diseases. Usually the immune system helps us to fight infections. In autoimmune diseases, the immune system reacts in a different way and becomes overactive in normal tissue. This immune system reaction leads to inflammation, which causes tissues to swell and can lead to possible tissue damage.

In JDM, the small blood vessels in the skin (dermato-) and the muscles (myositis) are affected. This leads to problems such as muscle weakness or pain, especially in the muscles of the trunk and those around the hips, shoulders and neck. Most patients have typical skin rashes as well. These rashes can affect several areas of the body: face, eyelids, knuckles, knees and elbows. The skin rash does not always occur at the same time as the muscle weakness: it can develop before or after it. In rare cases, small blood vessels in other organs can also be involved.

Children, adolescents and adults can all develop dermatomyositis. There are some differences between adult and juvenile dermatomyositis. In ~30% of adults with dermatomyositis, there is a relation to cancer (=malignancy), whereas in JDM there is no association with cancer.

1.2 How common is it?

JDM is a rare disease in children. Approximately 4 in 1 million children will develop JDM each year. It is more common in girls than in boys. It most often starts between the ages of 4 and 10 years, but children of any age can develop JDM. Children from all around the world and from all ethnic backgrounds can develop JDM.

1.3 What are the causes of the disease and is it inherited? Why does my child have this disease and can it be prevented?

The exact cause of dermatomyositis is not known. Internationally, there is a lot of research going on trying to find the cause of JDM.
JDM is currently regarded as an autoimmune disease and is probably caused by several factors. These could include a person's genetic predisposition in combination with exposure to environmental triggers such as UV-radiation or infections. Studies have shown that some germs (viruses and bacteria) can trigger the immune system to react abnormally. Some families with children affected by JDM suffer from other autoimmune diseases (diabetes or arthritis, for example). However, the risk of a second family member developing JDM is not increased.
Presently, there is nothing we can do to prevent JDM. Most importantly, there is nothing you could have done as a parent to prevent your child from getting JDM.

1.4 Is it infectious?

JDM is not infectious, nor is it contagious.

1.5 What are the main symptoms?

Each person with JDM will have different symptoms. Most children have:

Fatigue (tiredness)

Children are often tired. This may lead to a limited capacity to do exercise and eventually to potential difficulties in day-to-day activities.

Muscle pains and weakness

Muscles close to the trunk are often involved, as well as muscles in the

abdomen, back and neck. In practical terms, a child might start to refuse walking longer distances and doing sports, small children may "become fussy", asking to be carried around more. As the JDM gets worse, climbing stairs and getting out of bed might become a problem. In some children, the inflamed muscles become tight and shorten (called contractures). This leads to difficulties in fully straightening the affected arm or leg: the elbows and knees tend to be in a fixed bent position. This can affect the movements of the arms or legs.

Joint pain and sometimes joint swelling and stiffness

Both large and small joints can be inflamed in JDM. This inflammation can cause swollen joints as well as pain and difficulty in moving the joint. This inflammation responds well to treatment and it is uncommon for it to result in damage to the joints.

Skin rashes

The rashes seen in JDM can affect the face with swelling around the eyes (periorbital oedema) and a purple-pink discolouration of the eyelids (heliotrope rash); there can also be redness over the cheeks (malar rash) as well as on other parts of the body (top of the knuckles, knees and elbows) where the skin can become thickened (Gottron’s papules). Skin rashes can develop long before muscle pain or weakness. Children with JDM may develop many other rashes. Sometimes doctors can see swollen blood vessels (appearing as red dots) in the child’s nail beds or on their eyelids. Some JDM rashes are sensitive to sunlight (photosensitive), whereas others can result in ulcers (sores).

Calcinosis

Hard lumps under the skin containing calcium may develop during the course of the disease. This is called calcinosis. Sometimes it is already present at the onset of disease. Sores may develop on top of the lumps and a milky liquid made of calcium can drain out. Once they have developed they are difficult to treat.

Abdominal pain or tummy ache

Some children have problems with their bowels. These can include tummy aches or constipation, and occasionally severe abdominal problems if the blood vessels to the gut become affected.

Lung involvement

Breathing problems may occur because of muscle weakness. Muscle weakness can also cause changes in a child’s voice, as well as swallowing difficulties. Sometimes there is inflammation of the lungs, which can result in shortness of breath.

In the most severe forms, virtually all muscles attached to the skeleton (skeletal muscles) may be affected, which results in problems with breathing, swallowing and speaking. Therefore, voice changes, difficulties with feeding or swallowing, coughing and shortness of breath are important signs.

1.6 Is the disease the same in every child?

The severity of the disease varies with each child. Some children may have just their skin affected with no muscle weakness (dermatomyositis sine myositis), or with very mild muscle weakness that may only be apparent on testing. Other children may have problems with many parts of their body affected: skin, muscles, joints, lungs and intestines.