What is Juvenile Myositis?

Juvenile Myositis is one of the conditions in a group of conditions called the Dermatomyositis/Polymyositis Complex.

The conditions in this complex are characterized by muscle damage due to an inflammatory process of the blood vessels that lie under the skin and muscles. Skin changes around the eyelids and over the knuckles and finger joints are also seen.

Juvenile Dermatomyositis (JDM) is the condition most often seen in children around the age of 6 years old. JDM is thought to account for 85% of Juvenile Myositis cases.
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Sometimes children have symptoms of other autoimmune conditions as well as JDM such as rheumatoid arthritis, scleroderma or lupus. This is called "overlap syndrome".

Unlike Dermatomyositis in adults, JDM is not associated with cancer. Heart and lung problems are also less likely in children with JDM.

A special interest group of our committee has been set up for Juvenile Myositis families and patients.

Signs and Symptoms of Juvenile Dermatomyositis

The symptoms of Juvenile Dermatomyositis may resemble other medical conditions or problems. Always consult your child's health professional for a diagnosis.

Symptoms can often appear gradually. At times, there may be a more acute, or intense, onset of symptoms.

The following are the most common symptoms of Juvenile Dermatomyositis; however, each child may experience symptoms differently. Symptoms may include:

- Rash around the eyelids and/or knuckles and finger joints. A rash may also occur on the elbows, knees, and ankles
- Muscle weakness mainly affecting the large muscles around the hips and shoulders
- Fatigue

- Complaints of tummy aches
- Trouble climbing stairs, standing from a seated position, or getting dressed
- Malaise or general tiredness
- Swelling or redness in the skin around the fingernails
- Muscle pain and tenderness
- Weight loss
- Joint pain and inflammation
- Swallowing difficulty
- Calcium deposits under the skin (calcinosis) mouth ulcers
- Fever
- Vasculitic ulcers: Occasionally, in severe cases of JDM, holes in the skin or gastrointestinal tract caused by inflammation of the blood vessels can develop
- Contractures: Shortened muscles that cause a joint to stay in a bent position or have limited movement can sometimes occur
- Loss of body fat: Less than a quarter of children with JDM develop a complication called "lipodystrophy" which is the loss of body fat. If this is widespread, diabetes and high cholesterol can result.

What causes Juvenile Dermatomyositis?

The cause of Juvenile Dermatomyositis has not yet been determined. However, factors which are thought to be associated with Dermatomyositis include dysfunction of the immune system, resulting in infections.

Although JDM is not considered a genetic disease, there may be some genetic factors that make it more or less likely that an autoimmune condition will develop.

Children who develop this disease often have a family history of other autoimmune diseases, such as thyroid disorder, type I diabetes, rheumatoid arthritis, lupus, or Crohn's disease. In these children with a predisposition to develop JDM, it is thought that exposure to certain environment triggers causes the condition to develop. This could include infection with a virus or bacteria or a heavy dose of sun exposure.

How is Juvenile Dermatomyositis diagnosed?

Some of the tests for diagnosing Juvenile Dermatomyositis include:

- **Medical history and physical examination**: The diagnostic process is started by a careful look at your child's medical history and a thorough physical exam. Your doctor may perform muscle strength testing and examine your child's nailfold capillaries for any abnormalities
- **Blood tests**: Blood tests can be used to identify the presence of antibodies, muscle enzymes, and indicators of inflammation in the blood
- **Muscle biopsy**: A muscle biopsy involves removing a small piece of muscle through an incision in the skin which is then examined under a microscope
- **Scans:** Magnetic Resonance Imaging (MRI) may be used to identify muscle damage or inflammation
- **X-rays**: a diagnostic test which uses invisible electromagnetic energy beams to produce images of internal tissues, bones, and organs onto film

- **Electromyography:** A test where wires are attached to the skin to measure the electrical activity of muscles
- **Other tests:** Electrocardiograms (ECGs) and echocardiograms (shortened to Echo) to look at the function of the heart.

Abdominal ultrasound to assess any digestive problems.

Lung function tests (which may include a CT scan) and swallowing tests.

How is Juvenile Dermatomyositis treated?

Juvenile Dermatomyositis cannot be cured. However, with supportive therapy and a multidisciplinary team approach to treatment, remission of the disease may be achievable in time.

Treatment may include:

- Medications such as glucocorticosteroids and methotrexate to treat the inflammatory process and hydroxychloroquine to help treat the skin disease of Dermatomyositis
- Physical and occupational therapy to improve muscle function and strength
- Liberal use of sunscreens to prevent further irritation or damage to the skin.

Please be advised that the information on this page is not a substitute for medical advice.

What research is being done?

Research is ongoing to learn more about Juvenile Myositis and test potential treatments. The details for a number of studies can be found on the clinicaltrials.gov website.

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