

JUVENILE MYOSITIS

What is juvenile myositis (JM)?

Myositis is a word used to describe inflammation or swelling of the muscles. The two most common forms are juvenile dermatomyositis (JDM) or juvenile polymyositis (JPM). What causes JM is not known, though many doctors believe there is an environmental exposure (like an infection or virus) that triggers the disease in someone with specific genes or gene sequences. It is a rare, chronic disease, affecting only about 3,000 children in the United States.

JM begins in childhood before the age of 18. The first signs are typically a visible, reddish-purple rash (in JDM only); general tiredness; moodiness or crankiness; and trouble climbing stairs or standing from a seated position. With JDM, the skin and muscles are affected, causing a characteristic skin rash. In JPM, there is no skin involvement, but many muscles are affected. Less common forms of juvenile myositis are amyopathic myositis (also called JDM sine myositis), where the skin is affected but the muscles are not involved; and overlap myositis, in which the child has myositis along with at least one other autoimmune disease (like lupus, diabetes or arthritis).

JM is an autoimmune disease, meaning the body's immune system, which normally fights infections and viruses, does not stop fighting once the infection or virus is gone. The immune system then attacks the body's own normal, healthy tissue through inflammation, or swelling. JM is not contagious.

What are the signs and symptoms of JM?

The more common signs and symptoms of JM are:

- Muscle weakness – children may have trouble standing up, climbing steps or lifting their heads
- Muscle pain
- Visible, reddish-purple skin rash, which can be painful and itchy
- Trouble swallowing
- Difficulty catching breath
- Weaker or hoarse-sounding voice
- Fatigue

The muscles most often affected are those closest to the body – neck, stomach and upper arms, for instance.

The skin rash of JDM is caused by vasculitis, or swelling of the blood vessels under the skin. The rash may be painful and/or itchy. The two rashes most characteristic of JDM are Gottron's papules, or bumps found under the skin around the knuckles, elbows and knees; and a heliotrope rash, or a purplish rash around the eyes, shaped like a flower.

What are other complications of JM?

There are other symptoms and complications that occur in some but not all JM patients.

- Calcinosis, or hard, sometimes painful lumps of calcium under the skin that appear on the fingers, hands, elbows and knees.

- Contractures, shortening and tightening of tendons and joints. Less than one-quarter of the patients note this symptom.
- Gastrointestinal problems, like cramping and diarrhea.
- Staph infections, or infections deep in the body and usually widespread. Since JM children are typically on medicine to lower the body's capacity to fight infections, they are more exposed to viruses and bacterial infections.

What medications are used to treat JM?

Pediatric physicians will normally prescribe a high-dose therapy of prednisone, a steroid aimed at suppressing the body's immune system and reducing the inflammation. This relatively high dose tends to bring on a number of unwanted side effects, including weight gain, puffy "moon" face (also called cushingoid), moodiness, and increased risk of infection. Once a child's symptoms begin to subside, the prednisone therapy is slowly reduced over time.

Other medications are also aimed at lowering your body's ability to fight infection. These medicines, especially methotrexate, lead to an increased sensitivity to the sun and other forms of ultraviolet rays, including fluorescent lighting.

Children may be on several medications at once, and some require intravenous administration over a period of time. These treatments often leave children feeling tired and physically drained.

How will JM affect the child at school?

It's important that children with JM, or any rare condition, feel like a "normal" part of the group. However, special accommodations may be necessary at times.

With weakened muscles, it's difficult to walk from classroom to classroom in the allotted time. Allowing a child to leave early gives extra time and prevents unnecessary bumping in crowded hallways. Carrying a heavy load of books is challenging for someone with JM. Depending on the grade level, there are a few options to consider – allowing the child to keep a set of books at school and at home; having other students volunteer to help carry books from room to room; or giving the child an additional place to keep books between classes.

For physical education classes or recess, JM children need to stay out of the sun and whenever they're outside, apply sunscreen liberally. Certain physical activities will not be possible during active disease or during treatments.

You and the child's family may come up with other options that will best meet his or her unique needs. There are numerous possible accommodations, depending on the child's individual situation. Some to consider are:

- Leave class early to get to another class.
- Have a friend help carry books, lunch tray and other items.
- Rest during activities and take time to visit the school nurse, if necessary.
- Sit in a chair during story times when others are on floor.
- Have a friend take notes – or photocopy others' notes, tape class, or use a laptop.
- Choose a locker that is easily accessible to avoid bending down or reaching up.
- Sit away from windows and use sunscreen when outdoors. Place UV filters on fluorescent lighting systems in classrooms.
- Stand and stretch regularly during class time.

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