SARAA
South African Rheumatism & Arthritis Association

Inflammatory myopathies

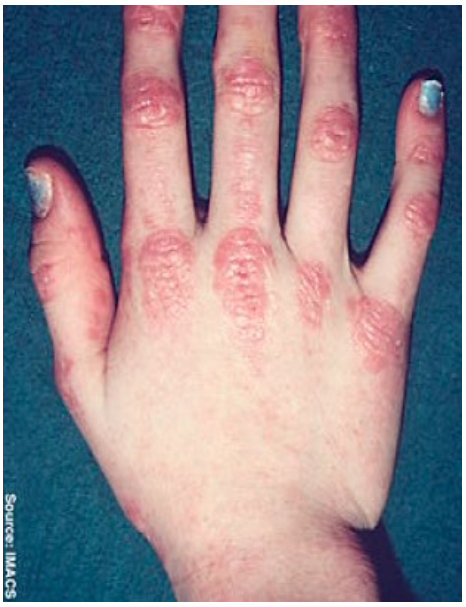
Myopathy is a general medical term used to describe a number of conditions affecting the muscles. All myopathies cause muscle weakness.
The idiopathic inflammatory myopathies are a group of diseases in which inflammation occurs in muscles and often in organs and tissues other than muscle. muscle weakness, and, in some cases, muscle pain.

They include
• Dermatomyositis
• Polymyositis
• Juvenile dermatomyositis
• Juvenile polymyositis
• Myositis that occurs with other systemic (body-wide) rheumatic diseases, such as mixed connective tissue disease, lupus, and scleroderma
• Autoimmune necrotizing myopathy
Some muscle diseases occur when the body's immune system attacks muscles. The result is misdirected inflammation, hence the name inflammatory myopathies.
People with inflammatory myopathies may have these features:
• Proximal weakness in the large muscles around the neck, shoulders and hips
• Trouble climbing stairs, getting up from a seat, or reaching for objects overhead
• Little, if any, pain in the muscles
• Choking while eating or aspiration (intake) of food into the lungs
• Shortness of breath and cough

Sometimes patients can have the rash with no sign of muscle disease. Doctors call this form of the disease amyopathic dermatomyositis.
People with dermatomyositis may also have lung inflammation that causes cough and shortness of breath.
Children with the disease may have an inflammation of the blood vessels (vasculitis) that can result in skin lesions.

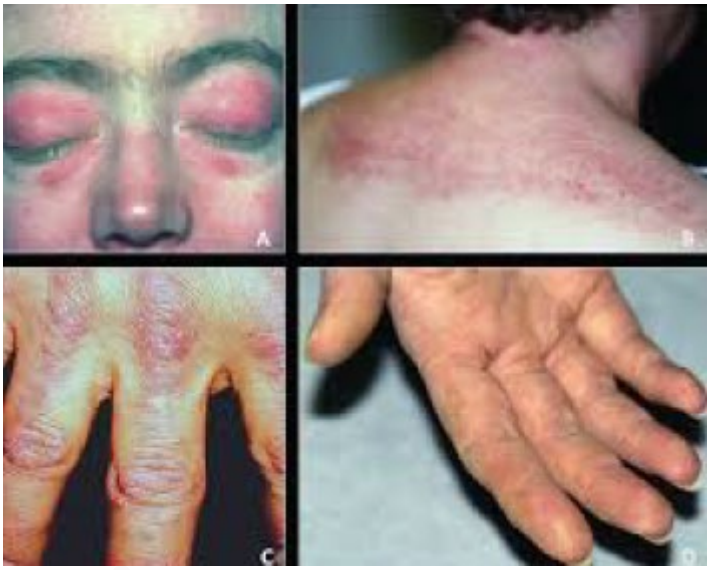
Skin changes — People with dermatomyositis often develop a rash or other change in the skin.

- Gottron's sign or Gottron's papules – Gottron's sign is a flat red rash over the back of the fingers, elbows or knees. Gottron's papules are red, often scaly, bumps overlying the knuckles of the fingers.

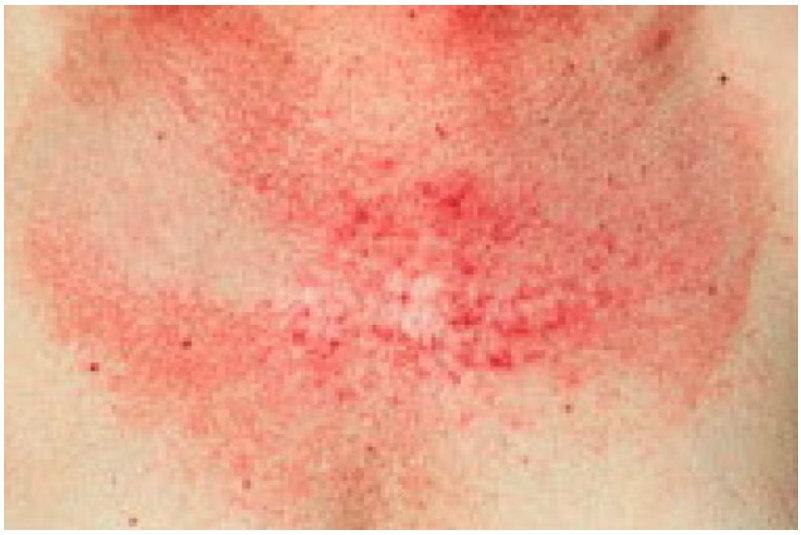


- Shawl sign – The shawl sign is a widespread, flat, reddened area that appears on the upper back, shoulders, and back of the neck. It can worsen with exposure to ultraviolet light.

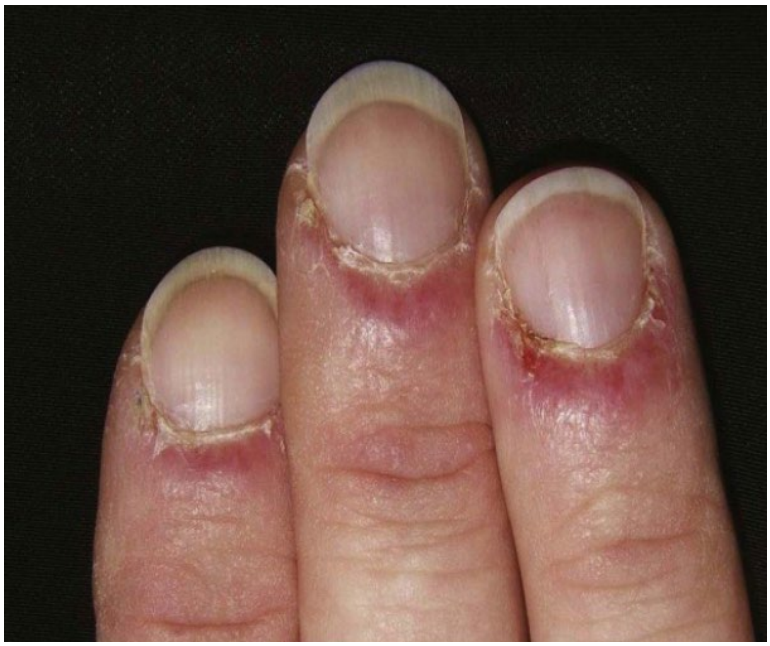
- Heliotrope rash – The heliotrope rash is located on the upper eyelids and is often accompanied by eyelid swelling. The heliotrope sign is named for the heliotrope flower, which is violet-coloured.



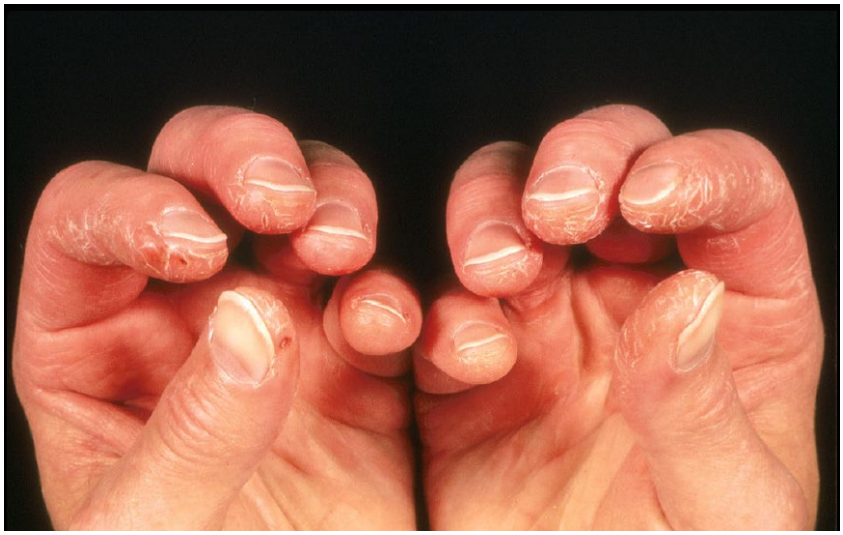
- V sign – The V sign has an appearance similar to that of the shawl sign, but appears on the front of the chest in the area of skin exposed by a V-necked sweater.



- Nail abnormalities – The nailfolds (the skin around the fingernails) may become reddened and may develop changes in the blood vessels.



- Mechanic's hands – People with dermatomyositis or polymyositis may develop "mechanic's hands," a roughening and cracking of the skin of the tips and sides of the fingers, resulting in irregular, dirty-appearing lines that resemble those of a manual labourer.



- Juvenile dermatomyositis — Children with inflammatory myopathy usually but not always have a dermatomyositis rash. They differ from adults most importantly in that painful calcium deposits can form on the skin and on the fibrous tissue that wraps around muscles, called fascia.
- There are many causes of muscle disease other than inflammation. They include infection, muscle injury due to medicine, inherited diseases that affect muscle function, disorders of electrolyte levels, and thyroid disease. It is unknown what causes inflammatory myopathies.
- Inflammatory myopathies are rare.
- Polymyositis and dermatomyositis occur in about one person per 100,000.
- All ages can get these diseases.
- The peak ages 5 to 10 in children, and 40 to 50 in adults.
- Women get inflammatory myopathies about twice as men.

How to diagnose

- A blood test to measure the level of various muscle enzymes (creatine kinase ck, aldolase) and myositis-specific antibodies.
- An electromyogram - often referred to as an EMG - to gauge electrical activity in muscle
- A biopsy of a weak muscle (a small piece of muscle tissue is removed for testing)
- Magnetic resonance imaging - more often called MRI - to try to show abnormal muscle.
- In adults, dermatomyositis and, to a much lesser extent, polymyositis at times may be linked to cancer. Therefore, all adults with these diseases should have tests to rule out cancer.

Treatment

- Corticosteroids: Often, the first treatment is an oral (by mouth) corticosteroid, such as prednisone, at a high dose. (Beware of sight effects).
- Disease Modifying Antirheumatic Drugs (DMARDs) - methotrexate or azathioprine. This gives better long-range control of the disease and helps avoid long-term side effects of corticosteroids.
- • Patients whose disease is severe or who do not respond to standard treatment have other options. They include intravenous immunoglobulin, cyclosporine, tacrolimus (Prograf), mycophenolate mofetil (CellCept), and rituximab (Rituxan).

Prognosis

- The severity of disease is highly variable, ranging from mild weakness that responds well to treatment to a rapid progression of symptoms that are unresponsive to all treatments.
- Less commonly, people with these conditions improve spontaneously without any treatment.
- People with dermatomyositis or polymyositis tend to have a better outcome if they are treated promptly, have mild muscle weakness, have no difficulty swallowing, and have no signs of disease in other organ systems such as the heart and lungs.

References

1. Uptodate
2. Google images