

Inflammatory Myopathy

What is Inflammatory Myopathy?

Inflammatory Myopathy is an autoimmune disease that causes muscle inflammation, leading to weakness.

There are many types of myositis. Common ones are:

- Polymyositis
 - Muscle inflammation without rash
- Dermatomyositis
 - Muscle inflammation with rash, usually around the eyelids

Who are at Risk of Inflammatory Myopathies?

Inflammatory Myopathies are rare diseases that affect every five to ten people in a million worldwide.

It can affect people of any age, but the two main clusters are:

1. Children between 5 to 15 years old
2. Adults between 30 to 65 years old

Women are twice as likely to be affected compared to men.

In adults, dermatomyositis may be related to underlying cancer. Therefore, all adult patients with these disorders should receive appropriate testing to rule out cancer.

Signs and Symptoms of Inflammatory Myopathy

- Muscle weakness/pain is one of the main symptoms, which usually affects muscles located at the centre of the body (e.g., neck, shoulders, hips and thighs).

Patients may experience:

- Difficulty in climbing stairs
 - Difficulty in standing up from a seated position
 - Difficulty in grooming (e.g. Combing hair/dressing)
 - Difficulty in swallowing (may result in choking or accidental inhalation of food into the lungs when eating)
 - Coughing and shortness of breath due to lung inflammation
 - General symptoms include lethargy, fatigue, weight loss and poor appetite.
- Patients with Dermatomyositis develop a rash in addition to muscle symptoms. It can appear as a purple or red discolouration that affects the face, area around the eyes, knuckles, elbows or knees.



Left: Gottron's Papules; Right: Mechanic's Hands

Photos retrieved from Kelley & Firestein's Textbook of Rheumatology.



Left: Heliotrope Rash; Right: Shawl Sign

Photos retrieved from Myositis Support & Understanding.

How is Inflammatory Myopathy Diagnosed?

Diagnosis of Inflammatory Myopathy takes time, and several visits to the doctor may be needed to identify the type of myositis you have.

You should try to identify the location of muscle weakness and pain and the duration of your symptoms. A complete physical examination will be performed on you to determine your muscle strength and the extent of the disease.

The following tests may be performed:

- Routine Tests: To check your full blood count, level of muscle enzymes and your body's inflammatory status.
- Auto-antibody test: To identify different types of antibodies in your blood to determine how your disease will develop overtime and its association with cancer or lung inflammation.
- Electromyogram (EMG): To look out for abnormal findings to identify potential muscle inflammation.
- Muscle Biopsy: To confirm the presence of muscle inflammation if presentation is abnormal. This biopsy is performed only when required.
- Radiological Imaging: To determine the diagnosis (e.g. MRI to identify abnormal muscles) and evaluate extent of disease (e.g. chest X-ray and high resolution CT scan to detect lung involvement).

Further procedures may be required to check for hidden cancers (e.g. endoscopy, mammogram, CT scans).



How is Inflammatory Myopathy Treated?

Medications

Corticosteroids and immunosuppressants are commonly used treatments. In severe cases, intravenous immunoglobulins (antibody drips through the veins) or rituximab (a biologic) may be used.

Adjunct Therapy and Diet Modification

Physical therapy involves participating in specially designed physiotherapy programs to regain muscle strength. Those with swallowing difficulties may require a modified diet or temporary use of a feeding tube and subsequent follow-up(s) with the speech therapist.

General Dermatomyositis

For patients with Dermatomyositis, sun protection is important as ultraviolet (UV) light exposure can worsen the rash.

Cancer-Associated Dermatomyositis

In this group of patients, treatment of cancer should take place concurrently. This may include surgery, chemotherapy, radiotherapy sequentially or in combination.

Can This Disease Be Cured?

The extent of recovery depends on the type of myositis. Most patients with Dermatomyositis and Polymyositis are able to regain most of their muscle strength with early treatment.

The control of myositis in patients with concurrent cancer usually depends on the control of the cancer. This means that the myositis symptoms tend to resolve when the cancer responds to treatment.

What Can I Do Now That I Have This Condition?



- **Stay Active**

- Work with the physiotherapist to regain muscle strength.

- **Pace Yourself**

- You may feel more tired than usual. Overdoing things will only set you back.



- **Know Your Disease**

- Read about your disease, talk to our nurse clinicians and do not be afraid to ask your doctor questions about your disease.



- **Be Part of the Team**

- It is important to follow the treatment plan that both you and your doctor agreed upon.



If you encounter any problem with your treatment, please contact your doctor.

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