

FACT SHEET

IDIOPATHIC INFLAMMATORY MYOPATHIES

Idiopathic Inflammatory Myopathies (IIM) are a group of non-hereditary muscle disorders caused by inflammation in the muscles and can have other organs involved including the skin and lungs.

INCIDENCE: 1 : 40,000 to 1 : 100,000 per head of population across all types

AGE OF DIAGNOSIS: Polymyositis (PM) - often over 20 years of age

Dermatomyositis (DM) - childhood to adulthood

Necrotising Autoimmune Myopathy (NAM) - childhood to adulthood

OCCURRENCE: PM and DM ratio of 3:1 females to males

LIFE EXPECTANCY: average life expectancy. They can be associated with specific antibodies which dictate the risk of associated malignancies

Commonly included in this group are:

- **Polymyositis (PM)** where the cells of the immune system directly attack the muscle fibres.
- **Dermatomyositis (DM)** where the immune cells attack the small blood vessels that supply the skin and muscle.
- **Necrotising Autoimmune Myopathy (NAM)** where immune system cause muscles cell-death, without obvious inflammatory cell involvement on muscle biopsy.

Idiopathic Inflammatory Myopathies (IIM) are caused by the body's own immune system attacking normal healthy cells for unknown reasons. This is called an autoimmune response. It has been suggested that certain triggers such as a virus or drugs can cause myositis. Inflammation occurs while damaged cells try to repair. Inflammation can occur within the muscle cell structure itself or in the surrounding tissues such as the organs or blood vessels.

The inflammation process leads to muscle tissue destruction, progressive and usually symmetrical muscle weakness and sometimes pain especially if surrounding the joints. Over time you may see loss of muscle bulk where the muscle cells have died or shrunk. Major organs of the body such as the heart, kidneys, lungs and even the oesophagus can be affected. IIMs are suspected when there is a history of acute or subacute onset of proximal muscle weakness and symptoms. Symptoms can include difficulty with

stairs, pain in joints and muscles, fatigue, swallowing difficulties, a rash and breathing problems.

Treatment with medication that suppresses the immune system can be effective in these conditions. New research is trying to further understand this group of conditions and develop more successful treatments. People with inflammatory myopathies can experience great pain and discomfort for long periods of time. Prognosis is also dependent on the extent of involvement of other organs. Treatment may continue over several years and a good proportion of patients may completely recover, however some people may continue on low dose autoimmune treatments for life to manage symptoms. The risk of permanent loss of strength, effects from muscle wasting and weakness can be seen in a small proportion of people. Children particularly with dermatomyositis can get permanent stiff joints.

Planning ahead for services & equipment can be difficult as the course of IIMs is not as predictable as with other neuromuscular conditions

Adults with IIMs should be monitored for the possible association with an underlying malignancy. Medical complications can arise therefore the following areas should be monitored: tightening of the joints; respiratory care; sleep studies; heart concerns; stomach and bowel

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problems; and psychological issues as they grapple with the loss of independence and increased reliance on others such as family, friends or paid carers.

It is important that service provision is centered around quality of life outcomes & meets the needs of the individual

Families and individuals affected by chronic conditions, including those that may involve numerous visits to hospitals for treatment, benefit most when empowered to have control over their life choices. Through seeking information about what is available, people will be empowered to make the right decisions to meet their goals. Despite the challenges a person with IIM faces, there are many opportunities available to support people to live fulfilled and productive lives. Goal setting and positive attitudes contribute to achieving success in many facets of life.

CONSIDERATIONS FOR PLANNING SHOULD INCLUDE BUT ARE NOT LIMITED TO THE FOLLOWING:

- A consistent General Practitioner (GP)
- Specialists for monitoring and medical care planning eg. neurologist, rheumatologist, respiratory, heart physicians
- Support coordination for complex care needs
- Physiotherapy eg. hydrotherapy, stretching, tailored exercise program
- Occupational therapy eg. to increase independence, equipment assessment
- Assistive technology eg. scooter, manual wheelchair
- Orthotics eg. leg splints for night time and ankle-foot orthoses (AFO's)
- Nutrition support eg. dietician, supplements
- Opportunities to socialise and be away from home eg. support groups
- Vehicle modifications/transport eg. taxis to and from school or work and the community
- Comfort/sleeping eg. air pressure relieving mattress
- Professional psychological support eg. counsellor, clinical psychologist
- Future goal planning eg. independent living, study, work choices
- Financial entitlements eg. Centrelink, Medicare, Companion Card

USEFUL RESOURCES

MDNSW www.mdnsw.org.au

MDNSW NDIS Toolkit www.mdnsw.org.au/our-services/support-coordination-your-choice/ndis-faqs

The Loop - Your Neuromuscular Resource Hub www.theloopcommunity.org