

SPORADIC INCLUSION BODY MYOSITIS

August 2006

What is Sporadic Inclusion Body Myositis?

Sporadic Inclusion Body Myositis (s-IBM) is an inflammatory muscle condition that is characterized by the progressive weakness and wasting of muscles (atrophy). The name was derived from the presence of abnormal bodies containing protein and inflammation in the muscles. s-IBM primarily affects the limbs of an individual, and to a lesser extent, other muscles of the body. It is not unusual for muscle weakness to only affect one side of the body.

s-IBM affects males 2-3 times more frequently than females. Symptoms typically appear over the age of 50 and the individual becomes more progressively disabled throughout the course of the condition. s-IBM does not generally affect life expectancy.

It is a rare, and yet the most commonly acquired, inflammatory muscle condition and affects an estimated 1 in 200,000 individuals.

What are the Features of Sporadic Inclusion Body Myositis?

Initially, s-IBM tends to affect the muscles of the arms and legs, particularly a weakening and atrophy of the quadriceps (front of thigh) and forearms. This results in difficulty in performing every day tasks, such as climbing stairs, getting in and out of chairs, and individuals may be prone to tripping due to foot drop, so have an exaggerated walk due to inability to control the foot at the ankle. Hand and wrist muscles are typically affected, which can make grasping and holding things a challenge. Some individuals will find they fatigue quicker than before on exertion. Muscle loss will gradually be replaced by fatty tissue, increasing the individual's fat content.

Mobility will become progressively restricted and those with s-IBM may have difficulties bending down, walking quickly, and balancing. Individuals with s-IBM often need to resort to a cane or a walker. The severity of the progression of s-IBM differs for every individual, though wheelchairs eventually become a necessity for the majority, usually in 10-15 years from diagnosis.

Speech difficulties are relatively common due to the weakening of speech muscles, and speech therapists can provide assistance. This can also lead to swallowing difficulties (dysphagia). Surgery (cricopharyngeal myotomy) can be beneficial for dysphagia in some older individuals with s-IBM.

As muscle weakness in the speech muscles progresses there is risk of aspiration pneumonia as food and liquid may travel down the airways. This is a life-threatening complication, and therefore monitoring of speaking and swallowing ability is essential. Weakness in the neck muscles can make supporting the head difficult, and facial muscle weakness is sometimes apparent.

1 in every 5 cases experience nerve disease. This can affect multiple nerves and may compromise sensation or feeling in certain parts of the body, with a loss or decrease in tendon reflexes. This may be related to diabetes.

What Causes Sporadic Inclusion Body Myositis?

The cause of s-IBM is currently unknown, though it is likely to be a combination of several interacting genetic and environmental factors. As indicated in the name, s-IBM occurs randomly and is not an inheritable condition. Researchers have developed several hypotheses, as to what causes s-IBM.

s-IBM is sometimes associated with auto-immune disorders. Foreign bodies are normally identified by the immune system to be targeted for destruction. In auto-immune disorders, the immune system identifies the body's own molecules, such as the muscle cells in s-IBM, and attacks them. This is apparent in s-IBM in the inflammation of the muscle cells. Unlike auto-immune disorders, however, immune-suppressant drugs are generally ineffective in treatment.

There are also abnormal protein changes in the cells and some researchers think that these proteins may trigger the abnormal immune response.

Diagnosis of Sporadic Inclusion Body Myositis

Diagnosis usually commences after the identification of symptoms common of s-IBM.

Several tests are available to confirm diagnosis:

- > Muscle Biopsy – most conclusive diagnosis which shows the characteristic presence of round, empty spaces (vacuoles) in the muscle, enclosing clusters of protein (inclusion bodies), and the presence of inflammatory cells.
- > Blood Testing – elevated levels of creatine phosphokinase (CPK) are indicative of muscle problems

- > Electromyography (EMG) – the measurement of electrical activity in muscles should display smaller and shorter duration of activity than normal

Note that s-IBM is often misdiagnosed as polymyositis, another inflammatory muscle disorder.

Management of Sporadic Inclusion Body Myositis

As there is currently no cure for s-IBM, treatment focuses on the management of symptoms.

From an early stage, it is important to undergo regular exercise and stretching programmes, with the help of a physiotherapist, to maintain muscle strength and flexibility.

A good balanced diet with plenty of fresh fruit and vegetables is very important in ensuring excessive weight does not impede mobility. Contact with a physician and/or a nutritionist is valuable for this.

Several drug therapies have also been tested for management of symptoms. Some immunosuppressants have seen reduced inflammation in muscle biopsy and in the testing of CPK levels, however there has been no evidence of clinical improvement. In some individuals, intravenous immunoglobulin therapy (IVIg), another way of suppressing abnormal immune responses, may be effective. Steroids have anti-inflammatory and immune-suppressant functions, and can have positive effects on an individual's condition, though cannot be administered for long periods of time.

Research into Sporadic Inclusion Body Myositis

There are consistencies in the proteins found in the muscle inclusion bodies, in s-IBM, are also present in the brain of those with Alzheimer's disease, suggesting the possibility of a similar mechanism. Further research into this may aid in the discovery of potential treatment strategies for s-IBM.

Because s-IBM is primarily an adult-onset disorder, there is a chance that free radicals, which are molecules that cause accelerated aging, are likely to be implicated in the cause. The use of antioxidants to counter the effects of free radicals is being researched to see if they can aid in protecting the muscle fibres.

As the exact mechanisms of s-IBM become better understood, it will be possible to direct treatment strategies to the cause, rather than simply towards the symptoms.

Support for People with Inclusion Body Myositis

Support is available from the NMA who can offer specialist assessment, information, support, advocacy and referrals to other providers. There is also an active nationwide support network for those interested in meeting with others.

Due to the later onset of s-IBM, most individuals are likely to already have an established career when diagnosed. It may be necessary to have an evaluation if s-IBM is likely to affect an individual's ability to perform his or her job. This is usually performed by a physiotherapist or occupational therapist.

Disability should not hinder alternative employment possibilities, though it is wise to choose a career that does not require heavy physical activity. Any individual has the right to equal pay and equal rights for employment. For more information contact the Employment Relations infoline on 0800 800 863 or visit www.ers.dol.govt.nz.

The government promotes equal employment opportunities in private sector and can be contacted on (09) 525 3023 or visit www.eeotrust.org.nz

Workbridge provides a professional employment service for people with all types of disabilities and administers support funding on behalf of Work and Income. Contact on 0508 858 858 or visit www.workbridge.co.nz

More Information

Muscular Dystrophy Association can be contacted for further information, assistance, advice, support and referrals, on 0800 800 337 or by e-mail at info@mda.org.nz.

The Muscular Dystrophy Association Website also contains information on services available within NZ, our quarterly magazine, contacts, membership details, news and links to other sites - www.mda.org.nz

Further Resources

www.nzord.org.nz – the New Zealand Organisation for Rare Disorders website provides information on a number of rare disorders, a directory of support groups, practical advice, health and disability resources, research information, news and issues.

www.mdausa.org – the MDA USA website has an extensive site with plenty of further information on any muscular dystrophy conditions as well as research news.

www.myositis.org – the Myositis Association website which provides information on several types of inflammatory muscle disease

Information in this fact sheet was primarily sourced from:

National Institute of Neurological Disorders and Stroke (2006) Inclusion Body Myositis Fact Sheet. <www.ninds.nih.gov>

eMedicine (2006) Inclusion Body Myositis. <www.emedicine.com>