Inclusion body myositis

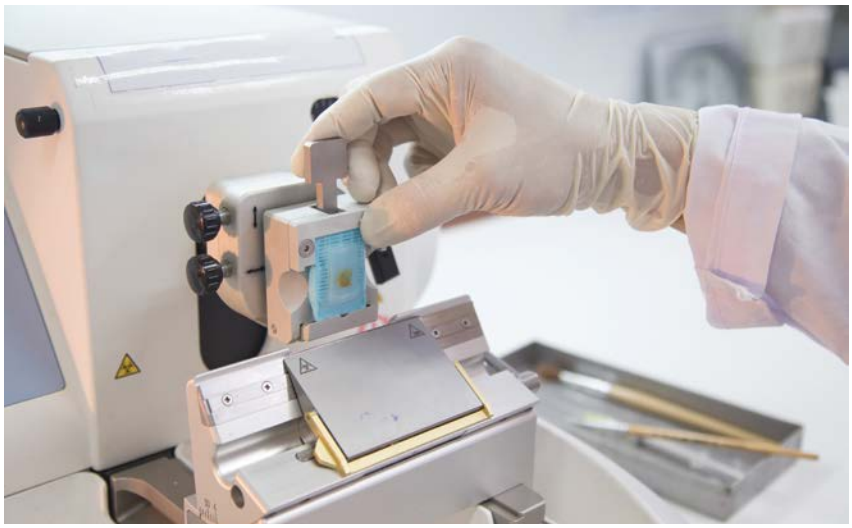
Understanding this inflammatory condition of the muscles that causes weakness.

Inclusion body myositis (IBM) is a progressive muscle disorder characterised by muscle inflammation, weakness, and atrophy (wasting). It is a type of inflammatory myopathy.

The cause of IBM is unknown. The underlying cause of IBM is poorly understood and likely involves the interaction of genetic, immune-related, and environmental factors. It is thought that some people may have a genetic predisposition to developing IBM, but the condition itself is not inherited. More males than females are affected.

Because of the inflammation associated with IBM, some doctors think the disease is a form of autoimmune disorder. In this kind of disorder, the body's immune system goes awry and attacks its own tissues – in this case, the muscles. However people with IBM don't respond at all to immunotherapy, which would normally be helpful in an autoimmune disorder. Some experts have linked IBM to infection by a virus that has yet to be identified. Other researchers believe that the primary problem in IBM is an age-related inability of the muscle to deal with destructive chemicals.

IBM develops in adulthood, usually after age 50. The symptoms and rate of progression vary from person to person. In IBM, the onset of muscle weakness usually is gradual, occurring over months or years and is asymmetrical. Falling and tripping usually are the first noticeable symptoms. For other people, IBM begins with weakness in the hands. People with IBM may have:



IBM develops in adulthood, usually after age 50. The symptoms and rate of progression vary from person to person.

- Difficulty with gripping, pinching, and buttoning
- Weakness of the wrist and finger muscles
- Atrophy (shrinking or wasting) of the muscles of the forearms
- Weakness and visible wasting of the quadriceps muscles (the large muscles on the front part of the thighs)
- Weakness of the lower leg muscles, below the knees

Muscle cramping and pain are uncommon, but some people with IBM do experience this. Most people with IBM progress to disability over a period of years. In general, the older a person is when IBM begins, the more rapid the progression of the condition. Most people need assistance with basic daily activities within 15 years, and some people will need to use a wheelchair. Lifespan is normal, but severe complications (e.g. aspiration pneumonia, bad falls) can lead to loss of life.

In general, the older a person is when IBM begins, the more rapid the progression of the condition.

Usually a muscle biopsy is required to diagnose IBM. After giving an anaesthetic, a doctor takes a sample of tissue from one of the affected muscles to be looked at in a laboratory.

When viewed under the microscope, the muscle cells of persons with IBM contain vacuoles (rounded empty spaces). Within the vacuoles, there are usually abnormal clumps of several proteins including one called amyloid. The protein clumps, or inclusion bodies, give IBM its name. This is the hallmark of IBM.

There is currently no cure for IBM. The primary goal of management is to optimise muscle strength and function. Management may include exercise, fall prevention, physiotherapy, occupational therapy, and speech therapy (for dysphagia).

The NZ NMD Registry currently has 40 people enrolled who have IBM. There have been two enquiries from researchers interested in IBM over the past six months and it is likely more will follow. Contact the registry curator registry@mda.org.nz or speak to the MDANZ fieldworker about enrolling on the NZ NMD Registry.

The power of planning

Receiving an IBM diagnosis was a huge shock for Julie Smith, but she's discovered that with planning and help, she can do many things.

I was a primary school teacher, keen sportswoman and have always led a busy live with family and friends. When diagnosed with IBM in the year 2000 at the age of 59, I was shattered to hear there was no cure or effective treatment for this condition, and in 15 years I would be in a wheelchair!

The first sign of a problem was that I needed to use my hands to help me get up from a chair due to weak quad muscles. To begin with, IBM did not affect my life at all. But gradually, I found difficulty in doing certain things. I stopped playing tennis and took up golf, as I could still walk easily. I played until 2009 when it became unsafe to walk on uneven ground.

I continued to do normal things with the help of disability aids such as a high firm cushion, car fitted with hand controls, raised toilet seat, higher chair at home, and installed a lift and made adjustments to the bathroom. Many good ideas came from my IBM support group.

My muscle strength slowly deteriorated over the years and I have had many falls. I have broken a total of 14 bones - mostly in my legs and ribs. My worst accident was in Paris at the French Tennis Open in 2008. When exiting the Roland Garros Stadium in dim light, I failed to see a step while walking with my stick. I fell and the result was a broken femur

Julie Smith

and three weeks in a Paris hospital where a rod and five screws were inserted in my leg. During this time I used my best school French which created many hilarious situations. The French nurses could only speak a little English and I was asked, "Why do you speak English if you come from New Zealand?" Luckily my doctor spoke good English.

About five years ago my swallowing deteriorated and I could only eat soft foods and liquid. So I ate plenty of soup, yoghurt, custard, mashed vegetables, fish and chicken. The only biscuit I could eat easily was a toffee pop which I loved as a treat.

Early last year, I kept getting chest infections and losing weight. This was because food was getting into my lungs. It was now time for a PEG tube to be inserted into my stomach. I no longer eat any normal food. Overnight I have 1000mls of liquid food pumped

Julie loves visiting her grandchildren in Australia.

I can't drive now so my husband is my chauffeur - but he doesn't appreciate it when I want to go clothes shopping!

into my stomach while I sleep. This works really well and during the day I have water and medication through the same tube. My husband carries out my feeding programme and he is now a real expert.

For about eight years I used crutches to walk everywhere, but last year this changed because I felt unsafe. Now I use a walker inside our home and a wheelchair when I go out. I also have a power chair to use independently. I can't drive now so my husband is my chauffeur - but he doesn't appreciate it when I want to go clothes shopping!

Every year we go to Australia to visit our two daughters and their families. In May we took my wheelchair and hired a walker and a cough assist machine. MDANZ were very helpful in organising this machine and the liquid food for our visit. Our daughter already stores a raised toilet seat and a special chair for our visits.

We went on many outings and my daughters took me clothes shopping - my husband was delighted! Our 9-year-old grandson loved pushing his 5-year-old sisters around in my wheelchair. Great screams of hilarity! And at the pools and playgrounds it was, "Look Gran," and "Watch me Gran."

Our 20-year-old granddaughter was getting a new puppy so we visited animal shops to buy puppy essentials and toys. When it arrived the darling puppy shredded the book Grandad was reading. Three weeks at 25 degrees in Queensland was wonderful. It was certainly worth all the effort and organisation to holiday with our family.

I continued part-time teaching until four years ago. Now I have a carer who helps me shower daily. My arms and hands have become much weaker, but I can still write with two hands holding a pen. Each week I go to an exercise class at a hydrotherapy pool and also play Bridge at the local Bridge club. I have many visits from friends and extended family and enjoy outings with them. I try to keep active as much as I can and keep enjoying social activities. With planning and help I can do many things.

Support us!

Any donation, big or small makes a difference. Donations of $5 or more are tax deductible.

Call: 0900 426 93 to make an automatic $15 donation.

Online: Donate any amount securely online. **www.mda.org.nz**

Post: Make a donation by post. Our postal address is: PO Box 12063, Penrose, Auckland, 1642

Bequests: You can create a lasting difference through making a bequest. Contact us or visit our website for information on how to include MDA as part of your will.

Thank you. We greatly appreciate your support.