



Inclusion body myositis (IBM)

“I felt the start of the condition many years before I did anything about it. I put it down to ‘just getting old’, and only once I got the correct information did I realise the slow decline was a result of having IBM.

“Having the correct information about my condition is really important. Not only for me and other people with IBM, but also for healthcare professionals who may not have heard of the condition because it’s so rare.”

David Morgan, who has IBM.

What is inclusion body myositis?

Inclusion body myositis (IBM) is a muscle-wasting condition, which causes muscles to become thin and weak. It was recognised as a condition in its own right in the 1960s. It usually occurs in mid to later life and is more common in men than women.

For muscle specialists, it is the most common muscle-wasting condition diagnosed in those who are over the age of 50 but it is sufficiently rare that most general practitioners (GPs) will not have looked after patients with IBM before, and many hospital doctors will not have heard of the condition.

How will it affect me?

IBM is a slowly progressing condition causing a gradual deterioration in muscle strength over years. The muscles are often affected in an asymmetrical way with the muscles on one side of the body being weaker than those on the other side. The pattern of weakness in IBM is characteristic. The most frequently affected muscles are the quadriceps (the thigh muscles, which straighten the knee joint) and forearm muscles (that flex the wrists and fingers). Accordingly, people affected by IBM may fall and can have difficulty climbing stairs, getting out of a chair and poor hand-grip. Although many people with IBM do present with this characteristic pattern of weakness, it is not evident in everyone, particularly early in the condition. In most people the quadriceps muscles are affected first but in others the forearm or other muscles are affected first.

Swallowing muscles are affected in some people, but this is rarely a significant problem early on and most do not encounter severe swallowing problems. The condition does not affect the heart, eyes, gut or bladder. It does not affect the function of the brain or sensation, and speech is rarely affected.

The condition itself does not cause pain. However, weakened muscles can predispose to problems such as falls resulting in injuries affecting bones, joints and soft tissues.



What causes IBM?

The short answer is that no-one knows. The presence of inflammatory cells in some muscle samples has led to the hypothesis that muscle is damaged by inflammation, caused by a virus or a misdirected immune system.

It has also been suggested that IBM is primarily a degenerative muscle condition with the deposition of abnormal proteins in the muscle that may trigger an inflammation of the muscles as a secondary part of the condition's progression. Researchers from around the world continue to investigate the cause of IBM.

What is the prognosis?

Life-expectancy is not reduced in those with IBM, and the condition usually progresses slowly, over many years. People with IBM experience varying degrees of disability as the condition progresses, and usually require adaptations to the home or work environment, as well as the use of mobility aids.

How is IBM diagnosed?

A number of tests can be carried out together to make the diagnosis of IBM.

1. Blood test: when muscles are damaged, they release a protein into the bloodstream, called creatine kinase (CK). This can be detected in a routine blood test and may help indicate that there is a muscle condition, but not which one. In most, but not all, people with IBM, the level of this protein in the blood is raised. The raised level of CK may not be high enough to immediately suggest a muscle condition. This means that the CK level cannot be used to make a specific diagnosis of IBM. Nor can a normal CK level be used to rule out a diagnosis of IBM.

Researchers have detected certain antibodies in the blood of those with IBM. Further research is needed to know whether these antibodies are detected in all cases of IBM especially in the early stages of the condition. We also need to know how specific these antibodies are for IBM. If they are not specific for IBM, these will lead to false positive results. Until we know this information, these antibody tests are not yet suitable for routine diagnosis of IBM.

2. Electromyography (EMG): when healthy muscles contract, they fire off a co-ordinated pattern of electrical impulses that can be detected by a tiny needle positioned in the muscle. When muscles that are not healthy contract, abnormal electrical impulses can be detected. However, although EMG may be helpful in highlighting the presence of a muscle condition, it cannot diagnose the specific condition.

3. Muscle imaging: imaging muscle, for example by MRI (magnetic resonance imagery), may be helpful in suggesting a possible diagnosis of IBM. However, this cannot be used to make a definitive diagnosis.



4. Muscle biopsy: this is a definitive test for IBM. It involves taking a small sample of muscle under local anaesthetic, for analysis in a laboratory. This process involves the use of a series of dyes and reactions, which will highlight different aspects of muscle structure and function. In IBM, muscle cells appear damaged and there is evidence of inflammation. In addition, the hallmark of IBM is the *inclusion body*, which is an abnormal clump of proteins, which can be seen in damaged cells with the use of specific dyes. This appearance will allow the pathologist and clinician to confirm the diagnosis of IBM. In some people, an initial biopsy may not be sufficient to make the diagnosis, and a second biopsy may be necessary.

Despite all these tests, early signs and symptoms of IBM may not be recognised readily, which can delay the diagnosis for some people. Equally, in some cases, the pattern of muscle weakness may be so typical for IBM that further tests or biopsies may not be essential.

Are other members of the family at risk of IBM?

Although there are examples of two close relatives getting IBM, e.g., siblings or parent-offspring, this is extremely rare. Generally, IBM is considered an acquired rather than a genetic condition so it is very unlikely that more than one person in a family will have IBM.

Are there any treatments for IBM?

Usually doctors rely on the evidence of clinical trials to know whether any treatment will help those with any condition. As the rate of deterioration in muscle strength seen in IBM is slow, this means that helpful clinical trials for IBM need to recruit large numbers of patients with IBM, use sensitive measures for the condition, and last long enough to detect treatment benefit or lack of benefit.

A recent systematic review of all the clinical trials that have tested various potential treatments for IBM concluded that these trials were too small, too short and, most did not use sensitive enough measures for monitoring the condition. These trials therefore cannot tell us whether any of the treatments they tested do or do not work. This means that experts have no evidence upon which they can say any treatment works for IBM. Thus, decisions on treatment are left to the expert's discretion and in fact many do not treat IBM with any drugs.

The presence of inflammatory cells in some biopsies led to suggestions that steroids and other drugs that suppress the immune system might be beneficial in this condition. However, given the lack of evidence, using these immunosuppressant treatments is controversial. Some doctors are of the opinion that these drugs can give short-term improvement and possibly long-term benefit in slowing the rate of progression, although all agree that these drugs will not prevent muscles from continuing to weaken in the long term. Other experts argue that any benefits are transient and are outweighed by long-term side-effects of the drugs.



Researchers have also studied intravenous infusions of human immunoglobulin (IVIG) in IBM. Again, the trials on its benefit are inconclusive and, given its costs and side-effects, it is not a routine treatment for IBM. An exception to this is that NHS England Commissioning policy and *National Immunoglobulin Guidelines* allow the use of IVIG specifically for severe swallowing problems seen in some cases of IBM.

In summary, there is currently no proven treatment for IBM. Further research into the cause of the condition will help the development of effective therapies. Clinical trials re-testing previously trialled or new treatment ideas may be ongoing, so it is worthwhile keeping in touch with Muscular Dystrophy UK, other support groups and your specialist to hear of any new developments.

What other support is available for people with IBM?

Physiotherapy: appropriate exercises can help maximise the efficiency of the relatively unaffected muscles. When walking is affected, physiotherapists can advise on walking aids (sticks, etc.). Physiotherapists can also teach people how to transfer between chairs, beds and wheelchairs if and when this becomes necessary.

Occupational therapy: occupational therapists (OTs) can provide advice and equipment to assist in tasks that may become increasingly difficult with weakened muscles. Typically, OTs will observe people in their own homes so they can advise on assistive strategies, aids and equipment. Hospital doctors and GPs can make referrals to OTs, or people can refer themselves, through Social Services.

Speech therapy: speech and language therapists (SALTs) also have expertise in supporting people with swallowing difficulties (dysphagia), which may affect some people with IBM. Dysphagia may cause fragments of food or drink to enter the windpipe, resulting in coughing after meals, or chest infections. Some people with IBM reduce their food intake, which results in significant weight loss. SALTs can advise on strategies to help swallowing, which may include other techniques, such as IVIG (see above) or permanent tube-feeding to ensure adequate nutrition.

What can I do to maintain health and wellbeing?

Even if you have IBM, it is generally beneficial to remain as physically active as possible. However, this needs to be undertaken safely as falls can cause injuries, which worsen disability. A physiotherapist familiar with muscle-wasting conditions is best-placed to advise on how best to optimise physical activity within sensible and safe limits.

The disability caused by weak muscles will be aggravated if one is overweight, so achieving and maintaining an ideal weight is helpful. As weight control can be more difficult when physical activity is limited, preventing weight gain in the first place is best.



Clinical trials

Research into IBM is ongoing in the UK and internationally, and you may wish to get involved in the testing of new potential treatments and therapies in clinical trials. If you are interested, mention this to your physician. You can also find out more about ongoing research and clinical trials on the Muscular Dystrophy UK website at www.musculardystrophyuk.org/research or contact our research team at research@musculardystrophyuk.org

Other related publications and resources

- ▶ Inheritance
- ▶ An introductory guide for families with a child newly diagnosed with a neuromuscular condition
- ▶ Inclusive Education Guide
- ▶ Wheelchair Provision for Children and Adults with Muscular Dystrophy and other Neuromuscular Conditions
- ▶ Muscle biopsies factsheet
- ▶ Heart check factsheet
- ▶ Anaesthetics factsheet
- ▶ Manifesting carriers factsheet
- ▶ Benveniste O, Guiguet M, Freebody J, Dubourg O, Squier W, Maisonobe T, et al. Long-term observational study of sporadic inclusion body myositis. *Brain*. 2011 Nov;134(Pt 11):3176–84.
- ▶ Mann HF, Vencovský J. Clinical trials roundup in idiopathic inflammatory myopathies. *Curr Opin Rheumatol*. 2011 Nov;23(6):605–11.

We're here for you at the point of diagnosis and at every stage thereafter, and can:

- ▶ give you accurate and up-to-date information about your or your child's muscle-wasting condition, and let you know of progress in research
- ▶ give you tips and advice about day-to-day life, written by people who know exactly what it's like to live with a muscle-wasting condition
- ▶ put you in touch with other families living with the same muscle-wasting condition, who can tell you about their experiences
- ▶ tell you about – and help you get – the services, equipment and support you're entitled to.

If you would like your GP or other health professional to have more information about IBM, we have some relevant materials. We've developed an online training module for GPs, as well as one for physiotherapists working with adults with muscle-wasting conditions. Contact our helpline or email us to find out more.



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Here for you

The friendly staff in the care and support team at the Muscular Dystrophy UK's London office are available on **0800 652 6352** or **info@muscular dystrophyuk.org** from 8.30am to 6pm Monday to Friday to offer free information and emotional support.

If they can't help you, they are more than happy to signpost you to specialist services close to you, or to other people who can help.

www.muscular dystrophyuk.org