Inclusion Body Myositis

What is inclusion body myositis?

Inclusion body myositis (IBM) is a disease of muscle, which causes muscles to become thin and weak. It was recognised as a disease in its own right in the 1960's. It usually occurs in middle to late life and is more common in men than women. We don't know how many people in the UK are affected but it is the most common muscle disease diagnosed after the age of 50. Nevertheless, it is sufficiently rare that most general practitioners will not have looked after patients with IBM before, and many doctors will not have heard of the condition.

How will it affect me?

IBM is a slowly progressive condition causing a gradual deterioration in muscle strength over the years. Most limb muscles can be affected. In particular the quadriceps (the thigh muscles which extend the knee joint), and forearm muscles that flex the wrists and fingers are commonly weak. Accordingly, patients often notice difficulty with stairs, getting out of a chair and a poor grip. Swallowing muscles are affected in some patients, but most do not encounter severe swallowing problems. The disease typically does not affect muscles of the heart, eye, gut or bladder. It does not affect the function of the brain or sensation, and speech is rarely affected. In general patients do not die of the disease, but most meet with some degree of disability as the disease progresses.

The disease itself is painless. However, weakened muscles can predispose to injuries affecting bones, joints and soft tissues.

What causes it?

The short answer is that no one knows! A number of theories have been put forward over the years though none have stood the test of time. In some families the condition is inherited, often with earlier onset than the more common sporadic (non-inherited) form. The presence of inflammatory cells in some muscle samples has led to the theory that muscle is damaged by inflammation, caused by a virus or a misdirected immune system.

No conclusive evidence to support these theories has been provided. Efforts to suppress the immune system have not led to clear benefit. More recently it has been suggested that IBM is primarily a degenerative disease of muscle with inflammation only secondary to this process. Many researchers in several countries continue to accumulate evidence to shed light on the cause of the disease.

How is it diagnosed?

There are a number of pieces of evidence that go together to make the diagnosis of IBM:

Blood test: When muscles are damaged they release a protein into the blood stream called creatine kinase. This can be detected in a routine blood sample. In some people with IBM the level of this protein in the blood is slightly raised. This blood test may therefore alert the physician to the possibility of muscle disease.

Electromyography (EMG): When healthy muscle contract they fire off a co-ordinated pattern of electrical impulses that can be detected by a tiny needle positioned in the muscle. When sick muscles contract abnormal electrical impulses can be detected. However, although EMG may be helpful it cannot make a definite diagnosis.

Muscle biopsy: The definitive test for IBM is a muscle biopsy. The biopsy involves taking a small sample of muscle under local anaesthetic. Laboratory analysis includes a series of stains and

reactions, used to highlight different parts of the muscle. In IBM, muscle cells appear damaged. The hallmark of IBM is the inclusion body, which is an abnormal clump of proteins seen in damaged cells. This appearance will allow the pathologist and clinician to confirm the diagnosis of IBM.

Because of the indolent nature of muscle weakness in IBM, a diagnosis is sometimes delayed for years after the onset of weakness. In some patients, the initial biopsy may not disclose the diagnosis, and a second biopsy may be necessary.

In other forms of myositis steroid treatment may be helpful. If steroids fail to help then this may give rise to consideration of IBM as a possible diagnosis.

Are other members of the family at risk of IBM?

Every patient requires individual assessment. The inherited form appears to be rare in the UK. However if IBM is of typical age of onset without evidence of similar disease previously in the family, the risk to other members of the family is very small.

Is there any treatment?
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. This immunosuppressant treatment is controversial. Some neurologists 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 from the drugs.

Recent trials have studied intravenous infusions of human immunoglobulin (IVIG) in IBM. Results have been contradictory, but provide no firm evidence of enduring benefit. Further trials continue, but currently the costs and side-effects do not justify routine treatment of IBM patients with IVIG.

In summary, there is currently no proven treatment. Further research into the cause of the disease will hopefully allow a rational basis to develop effective therapies.

What other help is there?
Despite the absence of a cure for IBM there are a number of therapists who can help:

Physiotherapy: Physiotherapy is not able to make weak muscles strong again. However, appropriate exercises can help to 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 they become necessary.

Occupational therapy: Occupational therapists (OTs) can provide advice and equipment to assist in tasks that become increasingly difficult with weakened muscles. More than any other professional they are in a position to provide valuable help in overcoming the everyday practical problems that IBM patients face. Typically they will observe a patient in their own home before advising on strategies, aids and equipment. Examples include cutlery with chunky handles to make gripping easier, aids to help stair climbing and advice on bathing or showering difficulties. OTs are employed in hospitals and by Social Services. Referral to OTs can be made by hospital doctors, GPs or patients themselves through Social Services.

Speech therapy: Speech therapists also have expertise in swallowing difficulties (dysphagia). In some IBM patients weakened swallowing muscles may cause dysphagia. This may cause fragments of food or drink to enter the windpipe resulting in coughing after meals or chest infections. Some IBM patients reduce their intake resulting in significant weight loss. Speech therapists can advise on strategies to help swallowing. Occasionally it is necessary to use other techniques to give adequate nutrition.

Can I help myself in any other way?
Exercise is generally helpful, and helps to get the most out of diseased muscles. Falls and injuries however can cause substantial disability. Patients therefore have the difficult task of undertaking

regular exercise within their capability but avoiding injury through accident. Because weakened muscles cannot carry an excess load, keeping to an ideal weight is helpful. This may be obvious, but weight control is more difficult when exercise is limited. It is likely that in the future therapeutic trials of drugs in IBM will be organised. If you are interested you should mention this to your physician.

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