Muscular Dystrophy UK

Fighting muscle-wasting conditions

Becker muscular dystrophy

Overview

Becker muscular dystrophy is a muscle-wasting condition, first described in 1956, which usually affects only males. It causes muscles to weaken and waste over time, leading to increasing and often severe disability.

In some cases, Becker muscular dystrophy can lead to life-threatening health problems, as heart and breathing muscles weaken. It is closely related to Duchenne muscular dystrophy, although is generally not as severe.

Symptoms

The average age of diagnosis with Becker muscular dystrophy is 11 years. However, the range is very wide – the diagnosis could be made in early childhood, or well into adult life. The progression and severity of muscle problems can vary significantly from one person to another and some people with Becker muscular dystrophy might have only very mild symptoms throughout their lives.

Symptoms usually begin very mildly in childhood. Cramps during exercise are often the only problem at first, but in some cases, boys may be late in learning to walk. Most people with Becker muscular dystrophy are not very athletic in childhood, and many struggle with school sport. Later, in their teens or 20s, some may find muscle weakness becoming more evident and causing difficulty in running, climbing stairs and getting up from the floor.

As muscle weakness progresses over time, it may involve the shoulders and arms, leading initially to difficulties in lifting heavy objects above waist level and then lifting the arms above shoulder level.

Over a period of many years, some muscles may become weak and wasted, especially certain muscles of the shoulders, upper arms and thighs. Other muscles that are less weak are often enlarged, and this is particularly noticeable in the calf muscles.

Typically, men with Becker muscular dystrophy may – in their 40s, 50s, or even later – become unable to walk, and require a wheelchair. In less frequent and more rapidly progressing cases of Becker muscular dystrophy, this may happen to men in their 20s or 30s.

It is important to be aware that some people with Becker muscular dystrophy may also have heart problems (cardiomyopathy), which can occur at any age. People might still be very mobile but have heart problems requiring treatment, or, as mentioned earlier, the heart problems might be the first sign of Becker muscular dystrophy.

Heart problems in Becker muscular dystrophy often do not cause any symptoms, but they can often be treated. It is therefore important for people with Becker muscular dystrophy to have regular heart monitoring by a specialist. Early treatment of heart problems (with drugs called ACE inhibitors and/or beta-blockers) can protect the heart muscle.

People with Becker muscular dystrophy can also have problems with breathing. This is more likely to be the case, however, in people who are wheelchair users and in later stages of the condition.

The muscles of facial expression, speech and swallowing, and the involuntary muscles (for example those of the bowel and the bladder) are not affected in Becker muscular dystrophy.

People with Becker muscular dystrophy have a higher risk of complications during general anaesthesia. This can be because of their heart and breathing problems, or because of a reaction to some general anaesthetics. It is important, therefore, that people with Becker muscular dystrophy inform doctors of their condition if a general anaesthetic is required.

Some people with Becker muscular dystrophy can show learning and/or behavioural difficulties. As learning difficulties are not progressive, it is important they are identified and addressed promptly (e.g. at school), to offer the child the support he will need to reach his full potential to develop his skills. Family support is essential, and specialists may need to be consulted to address specific issues of learning and behaviour.

Causes

Becker muscular dystrophy is a genetic condition – it is caused by a mutation or mistake in the genetic code (DNA). In Becker muscular dystrophy, the mutation occurs in a gene called dystrophin, which is located on the X-chromosome (girls have two X-chromosomes and boys have only one).

In Becker muscular dystrophy, the genetic mutation causes a faulty (smaller or less abundant than normal) protein called dystrophin to be produced in the muscles. This protein is important to maintain the integrity of the muscles, so when it is smaller or less abundant than normal, the muscle fibres gradually break down and the muscles slowly become weaker.

Some mutations result in the complete absence of the dystrophin protein in muscles and this causes the more severe form of muscular dystrophy, called Duchenne muscular dystrophy.

In some cases, Becker muscular dystrophy is inherited from the mother who is a carrier, but it can also be caused by a new mutation in the child's genes. Each son of a carrier mother has a 50:50 chance of being affected and each daughter has a 50:50 chance of being a carrier.

If a woman carries the gene mutation, then she is known as a 'carrier'. Usually female carriers are not affected because they have a second X-chromosome, which produces the dystrophin protein. A small number of female carriers have a degree of muscle weakness themselves and they are known as 'manifesting carriers' (please see our factsheet called Manifesting carriers).

Genetic advice (counselling) and testing for other family members at risk of being carriers should be provided as soon as possible following a diagnosis of Becker muscular dystrophy. Your clinician

or GP can arrange this for you.

Diagnosis

Unless there is a family history of the condition, Becker muscular dystrophy is usually diagnosed when a person shows signs of the condition. This can occur at any age.

Signs of Becker muscular dystrophy often include muscle problems, such as difficulty running, climbing stairs or getting up from the floor. Sometimes, Becker muscular dystrophy can be diagnosed after finding heart problems, with no other muscle symptoms. If a blood test is done, high levels of a protein called creatine kinase (CK) are seen. CK is normally found in muscle, but when muscles are damaged, such as in the case of Becker muscular dystrophy, it leaks into the bloodstream. The liver enzymes (aminotransferases, ALT and AST) are also often found to be high, but this is a consequence of the muscle damage and not of a liver problem.

Becker muscular dystrophy has to be confirmed by genetic testing, usually through a blood sample. Different types of genetic tests can provide specific and more detailed information about the genetic mutation. Genetic diagnosis is important as it offers a more accurate way of diagnosing Becker muscular dystrophy, and can therefore ensure adequate management. It can also help to identify other family members at risk of carrying the dystrophin mutation.

Your doctor may also recommend a muscle biopsy, where a small sample of muscle is taken for analysis (see our factsheet: Muscle biopsies). Tests on the muscle biopsy can provide information on the amount of dystrophin protein present in the muscle cells, and can help distinguish Becker muscular dystrophy from Duchenne muscular dystrophy and other muscular dystrophies. However, the clinical signs and the genetic test are usually sufficient to confirm the diagnosis, without the need to perform a muscle biopsy.

Treatment

Even though there is currently no cure available for Becker muscular dystrophy, there is ongoing research into increasing or maintaining muscle mass and strength in muscular dystrophies.

Most of the current research is focused on the more severe form of the condition (Duchenne muscular dystrophy). However, some treatments that could be effective in Duchenne muscular dystrophy may also be effective in Becker muscular dystrophy.

Becker muscular dystrophy is a variable condition; symptoms, age of onset and severity can differ significantly from one person to another. It is important, therefore, to manage symptoms and adapt clinical interventions accordingly.

A multi-disciplinary approach, with the input of specialists including physiotherapists and occupational therapists, is important, but the frequency and intensity of the interventions depends on symptoms and can vary over time. Children might require closer monitoring of their development.

It is important to try to keep as fit and active as possible. Active exercise strengthens muscles and this also applies to people with Becker muscular dystrophy. Regular daily exercise is better than occasional sudden bouts of exertion. Swimming is particularly recommended as it ensures gentle exercise to all body muscles, without over-exertion.

People with Becker muscular dystrophy may experience cramps during exercise – often during teenage years. If the cramps are troublesome, it may be worth trying ‘night splints’ (plastic splints to maintain a gentle stretch of the calf muscles overnight), calf muscle massage or compression with air-filled boots. There is however no tried-and-tested treatment for cramps.

Steroids are not routinely prescribed in Becker muscular dystrophy, although they might be considered in some specific, usually more severe, cases. However they have several side-effects, which will need to be taken into account.

In the more advanced stages of Becker muscular dystrophy, a wheelchair is likely to be needed for getting about independently, and for longer distances. There are other pieces of equipment that may be useful and a lot can be done to help both at home and at work to make certain tasks easier. Muscular Dystrophy UK will be able to advise you and put you in touch with the best sources of help.

Disclaimer

If you have feedback about this factsheet or want to request references, please email info@musculardystrophyuk.org.

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@musculardystrophyuk.org.

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