

Becker muscular dystrophy

What is Becker muscular dystrophy?

The muscular dystrophies are a group of genetic disorders, which cause muscle weakness. The Becker type was first recognised in 1956 and is now known to be a much milder variant of the better known Duchenne type of md. Becker md is generally slowly progressive and affects only males.

What causes it?

A fault in a particular gene (dystrophin) carried on the X chromosome leads to the formation of a faulty protein in muscle fibres. This protein, also called dystrophin, is absent or severely abnormal in Duchenne md. In Becker md a milder fault makes the dystrophin molecule smaller (or occasionally larger) or less abundant than normal. When dystrophin is abnormal the muscle fibres gradually break down and the muscles slowly become weaker. These dystrophin abnormalities in muscle provide a very good test for the diagnosis of Becker MD.

What are the symptoms?

The average age at diagnosis in BMD is 11 years but the range is very wide - sometimes the diagnosis may be made in early childhood or well into adult life. Symptoms usually begin very mildly in childhood; often cramps on exercise are the only problem at first but a few affected boys are late in learning to walk. Most people with BMD are not very athletic in childhood, and many struggle with school sport. Later, in the teens or twenties, muscle weakness becomes more evident causing difficulty in rapid walking, running and climbing stairs. Later still it may be difficult to lift heavy objects above waist level.

Men with typical Becker dystrophy may become unable to walk in their 40s or 50s or even later but there are less frequent and more rapidly progressive variants of Becker dystrophy in which this may happen in the 20s or 30s. Over a period of many years some muscles become weak and wasted, especially certain muscles of the shoulders, upper arms and thighs, while others that are less weak are often enlarged - this is usually particularly noticeable in the calf muscles.

The muscles of facial expression, speech and swallowing and the involuntary muscles (for example those of the bowel and bladder) are not affected in Becker MD It is important to be aware that some people with BMD may have problems with their heart (see the 'heart check' leaflet), and in the long term with the breathing muscles. These often do not cause any symptoms, and watching out for these problems, which can often be treated, is an important reason to keep in touch with a specialist clinic.

Is there a cure?

Unfortunately there is no cure at present. Research is proceeding to try to find a way to induce the muscles to form dystrophin. Any treatment, which may be found to be effective in Duchenne MD, would theoretically be effective also in the Becker type.

So what can be done?

Active exercise strengthens normal muscle fibres (and the great majority are normal in the early years of Becker MD). It is important to try to keep as fit and active as possible. Regular daily exercise is better than occasional sudden bouts of exertion.

Cramps during exercise can bother people with Becker MD at some stage - often especially as a teenager. If they are very troublesome it may be worth experimenting with 'night splints' (plastic splints to maintain a gentle stretch of the calf muscles overnight) or with sessions of calf muscle massage or compression with air-filled boots though there is not yet a properly tried and tested treatment for cramps.

In the later stages a wheelchair is likely to be needed at least for getting about independently over long distances. There is a great deal of other equipment that may be useful to individuals and much can be done to help both at home and at work to make certain tasks easier by careful choice of furniture, bathroom equipment etc. Advice and help with these matters is increasingly available and the Muscular Dystrophy Campaign will be able to put you in touch with the best sources of advice.

What about school?

Most young men with Becker md leave school without having had any major muscle problems except that they are usually slow at running in their teens and not very successful at PE or games. However, sometimes if cramps are a particular problem, keeping the school informed can be a good idea. In many cases the problem is recognised and diagnosed at around the age of 20.

A few boys with Becker md also have learning problems, usually of a mild degree but sufficient sometimes to limit their academic success at school. It is important to realise that this is not true of most affected boys, but when the problem does exist it is sensible to recognise and assess it early and to arrange the best possible plan to provide the right educational help. Unlike the muscle weakness, any learning problem will not get worse as the years go by. Despite the fact that to the outside observer BMD may cause relatively few problems in childhood, there is no doubt that some boys can find it a major psychological problem to be poor at sport in a society where boys are especially encouraged to pursue excellence at sport and where sporting heroes are so much in the news. Promoting self worth in this period is a crucial factor.

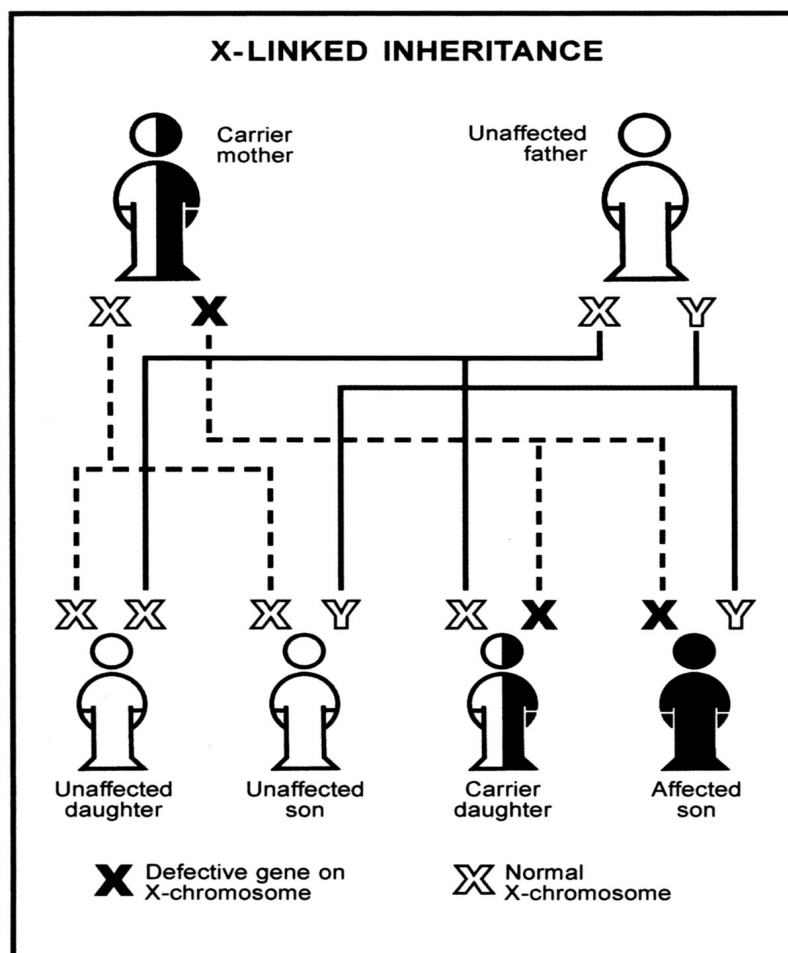
What about work?

People with Becker md have been employed in a range of jobs from steel workers to research scientist though occupations requiring a considerable amount of physical activity are not feasible for most people who have Becker md. It is clearly important for people to plan their careers on the basis that their existing physical capabilities are unlikely to improve and will eventually gradually decline. There is no reason why people with BMD should not work - if any special provisions are necessary the local clinic or family care officer can put you in touch with the right people at the local careers service.

The important principles are to work for the best possible educational qualifications at school, to make good use of any opportunities for further education and then either to plan a career that will depend as little as possible on physical strength and mobility, or to be prepared to re-train and change jobs appropriately as time goes on.

How is Becker md inherited?

The disorder is inherited as an X-linked recessive trait, which means that it affects only males but may be transmitted by unaffected female carriers of the gene to their sons. The sons of carriers each have a 50:50 chance of being affected. The daughters of carriers each have a 50:50 chance of being carriers. The mothers and sisters of affected males may be carriers and may need to be tested. The sons of affected males do not carry the gene and will not be affected or transmit the gene. However, all the daughters of affected males are carriers of the gene and may transmit the disorder to the following generation.



How early can it be diagnosed?

Once Becker dystrophy is known to affect one male in a family it is possible by simple blood tests to identify it or rule it out in any other boys at risk from birth onwards. In most families, but not in all, prenatal diagnosis is also possible, but this is more difficult and if at all possible the situation needs to be fully assessed before a pregnancy is embarked upon.

Can any carriers in the family be identified?

Most carriers can be detected if blood samples from their affected male relatives and certain other key members of the family are available for comparison, using techniques of DNA analysis. Although a simpler blood test for creatine kinase is positive in many carriers, only the DNA studies can rule out the carrier state in a woman at risk (for example in the sister of an affected man). However, in a few families, or if the key samples from relatives are not available, it may be possible only to calculate for each potential carrier her statistical risk of having an affected son.

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