

Muscular dystrophy - Duchenne and Becker

Duchenne muscular dystrophy (DMD) and Becker muscular dystrophy (BMD) are often discussed together because they cause similar patterns of weakness and are inherited in the same way.

A gene fault is the cause

The same gene is involved in both Duchenne and Becker muscular dystrophies. A fault in the gene causes a deficiency of a muscle protein called dystrophin. Duchenne muscular dystrophy usually only affects boys.

Early symptoms of Duchenne muscular dystrophy

Duchenne muscular dystrophy is not usually noticeable before the age of two or three. Symptoms and signs include:

- Frequent falls. Difficulty is also experienced rising up from the ground or going up hills or stairs.
- Well developed or excessively large calf muscles. Other muscles are poorly developed.
- A waddling gait.
- A sway-back ('lordosis').
- A tendency to stand and walk on the forefoot, with the heel off the ground. This is often called 'toe-walking'.

Effects on intellectual ability

Intellectual disabilities only affect a minority of boys with Duchenne type muscular dystrophy, but are more common than in other children. The intellect is not affected in Becker type muscular dystrophy.

Duchenne is the most severe form of muscular dystrophy

Duchenne muscular dystrophy can lead to:

- **Wheelchair use** - generally begins from around nine years of age. Total dependence on a wheelchair occurs around the early teens.
- **Restriction of joint motion** - for example, joint contractures. The ankles are usually affected first, and the hips and knees last.
- **Scoliosis** - for example, a sideways curvature of the spine. Corrective surgery is usually required.
- **Difficulty breathing** - caused by weakness of the muscles associated with breathing. In some cases, mechanically assisted breathing helps in the latter stages of the disorder's progress.
- **Heart problems** - in a small number of cases, the dystrophic process can affect the heart muscle.
- **Early death** - most affected people survive into their 20s. Small numbers survive only into their teens or reach more than 30 years.

Becker muscular dystrophy (BMD)

Becker muscular dystrophy is less severe than Duchenne muscular dystrophy:

- People with Becker muscular dystrophy can still walk at 16 years. Some can continue to walk until early adulthood or an advanced age.
- Survival may occur anywhere up to middle age. Some affected people have lived to over 80.
- Scoliosis seldom occurs.
- The effect on lung function is less severe.
- Heart trouble is less frequent, although it is occasionally serious.

Diagnosis of muscular dystrophy

Diagnosis before the age of two or three (before symptoms are very obvious) is possible through:

- **A blood test** - this will show very high levels of a protein called creatine kinase (CPK).
- **A muscle biopsy** - removal of a small piece of muscle tissue for examination. This will show if there is a severe dystrophin deficiency.
- **Electromyography (EMG)** - an examination of muscle activity. It involves inserting a needle into the muscle.

Genetic counselling

Genetic counselling is available to help people understand the hereditary nature of the disorder and the probable risk of them having a dystrophic child. Counsellors can help couples make an informed decision about having children.

Treatment for muscular dystrophy

There is no cure for Duchenne or Becker muscular dystrophies. To help reduce joint contractures, physiotherapists offer advice on stretches and exercises, and the prescription of orthosis. Occupational therapists also provide advice on sitting positions and activities.

Where to get help

- Your doctor
- Muscular Dystrophy Association Tel. (03) 9320 9555

Things to remember

- Muscular dystrophies are inherited muscle diseases, which lead to progressive weakness.
- Duchenne and Becker muscular dystrophies lead to similar patterns of muscle weakness.
- Duchenne muscular dystrophy is more severe and leads to an earlier death than Becker muscular dystrophy.

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Muscular Dystrophy Association

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