

FACT SHEET

BECKER MUSCULAR DYSTROPHY

**MUSCULAR
DYSTROPHY**
NSW BUILDING STRENGTH
REACHING POTENTIAL

Becker Muscular Dystrophy (BMD) is a milder form of Duchenne Muscular Dystrophy.

INCIDENCE: 1 : 18,000 to 1 : 30,000 live male births

AVERAGE AGE OF DIAGNOSIS: 5 - 15 years of age

OCCURRENCE: mostly in boys due to its inheritance pattern. It is not uncommon for there to be more than one sibling affected. Female carriers can experience symptoms of mild muscle weakness and heart problems

LIFE EXPECTANCY: normal life expectancy depending on severity, cardiac and or respiratory complications

Becker Muscular Dystrophy (BMD) is a genetic condition that infants are born with but may not cause symptoms until five to fifteen years of age. BMD is caused by an insufficient amount of the protein called dystrophin in the muscle cell which causes the muscle to breakdown over time. The muscle cell cannot repair itself effectively to work as it should. The damage in the muscle leads to progressive weakness of the muscles that support the spine and the body such as the arms, legs, the breathing muscles and the heart muscle. People with BMD produce small amounts of dystrophin but it is not in sufficient amounts to repair the muscle damage. Having some functional dystrophin slows the progression of muscle cell damage but does not totally prevent it. Progression of weakness is delayed compared to Duchenne MD where there is a total absence of dystrophin.

Children/adolescents with BMD will often present with muscle cramps, leg pain, fatigue with exercise and on examination may walk up on their toes, have large calves, a waddling gait and difficulties getting off the floor. BMD is usually a slowly progressive condition and those affected will most likely lose the ability to walk in their 40s. They may use mobility aids/equipment later in life depending on the severity of fatigue and weakness and usually have a normal life expectancy unless medical complications arise.

Learning difficulties may be seen in some people with BMD and usually affect attention focusing, verbal learning, memory and emotional interaction. Cognitive assessments should be conducted if there are any difficulties suspected and appropriate supports for learning put in place.

There is currently no cure for BMD. Treatments for BMD focus on keeping the child/adult healthy and active for as long as possible. Regular physiotherapy aims to prolong function, walking and maintain the muscles used

for breathing. Occupational therapy improves the skills of everyday living and focuses on equipment to enhance mobility. The person with BMD will need an experienced physiotherapist and occupational therapist to ensure their needs are being met.

As BMD progresses, needs will change, and the amount of support and care the person needs will increase. It is important that they are assessed at least annually to plan their future medical, physical, social and psychological supports.

Planning ahead for services and equipment is essential, particularly later in adult life as BMD progresses

As children become adolescents and adults, they will have goals and desires similar to their peers. This may include relationships, work options, study or planning for holidays and so on. Access and inclusion are paramount to ensure independence for education and community involvement. Issues related to puberty, relationships and sexuality may require professional intervention.

In adulthood, the weakness associated with BMD can progress significantly and may affect the person's ability to function at home and or at work. Adults with BMD who experience increased physical difficulties will require support to help them overcome barriers such as physical access, transport, social exclusion, financial constraints, health risks, low self-esteem and daily support needs. Given their changing needs, men with BMD are at risk of becoming depressed, so it is important that a variety of opportunities are encouraged, for example accessible work options, sport and recreation.

An older person with BMD may need to rely on others for a variable level of care support, which may have an effect on family dynamics. Enough support needs to be provided

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to the person with BMD so that their carer can have a break. Independent living is achieved by the majority of people with BMD and they often go on to lead normal lives, work, marry and have families of their own without too much difficulty. All of this can be achieved with the right supports and planning.

As a person with BMD ages, medical complications can arise, therefore the following areas should be monitored: tightening of the joints and feet deformities; respiratory care; sleep studies; non-invasive mechanical respiratory support; heart concerns and annual reviews; and psychological issues as they grapple with the loss of independence and increased reliance on others such as family, friends or paid carers.

It is important that service provision is centered around quality of life outcomes and meets the needs of the individual

Genetic testing and counselling should be offered to people with BMD, parents and family members for future pregnancy planning. Families and individuals affected by chronic life limiting conditions benefit most when empowered to have control over their life choices. Through seeking information about what is available, people will be empowered to make the right decisions to meet their goals. Despite the challenges a person with BMD faces, there are many opportunities available to support people to live fulfilled and productive lives. Goal setting and positive attitudes contribute to achieving success in many facets of life.

CONSIDERATIONS FOR PLANNING SHOULD INCLUDE BUT ARE NOT LIMITED TO THE FOLLOWING:

- A consistent General Practitioner (GP)
- Specialists for monitoring and medical care planning eg. neurologist, respiratory, heart, sleep physicians
- Support coordination due to progressive nature of BMD and complex care needs
- Physiotherapy eg. hydrotherapy, stretching
- Occupational therapy eg. to increase independence, equipment assessment
- Assistive technology eg. scooter, manual wheelchair, shower chair, hoist, slings
- Orthotics eg. leg splints for night-time and ankle-foot orthoses (AFO's)
- Home modifications for access to all areas eg. ramps, lifts, bathrooms, bedrooms, kitchen, outdoors areas, pool
- Vehicle modifications/transport eg. taxis to school, work and community
- Nutrition support eg. dietician, supplements
- Opportunities to socialise and be away from home eg. MDNSW Camps, support groups, accessible travel
- Comfort/sleeping eg. air pressure relieving mattress, electronic bed with high/low options
- Assistive ventilation devices eg. CPAP or BiPAP machines, cough assist machines
- Adapted sporting and recreation equipment eg. boccia equipment, sports wheelchair
- Support workers eg. meal preparation, personal care
- Professional psychological support eg. counsellor, clinical psychologist
- Future goal planning eg. independent living, study, work choices
- Financial entitlements eg. Centrelink, Medicare, Companion Card

USEFUL RESOURCES

MDNSW www.mdnsw.org.au

MDNSW NDIS Toolkit www.mdnsw.org.au/ndis