Scottish Muscle Network

Becker Muscular Dystrophy (BMD) “at a glance” sheet

NOTE
This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined based on all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient’s case notes at the time the relevant decision is taken.

Diagnosis / Genetics

- X-linked recessive, genetic counselling advised especially for potential female carriers
- diagnosis must have molecular confirmation
- Dystrophin, an essential protein in the muscle cell membrane is altered in quality or quantity
- genetic counselling mandatory - some female relatives may be carriers, and some male relatives (including seemingly asymptomatic ones) may be affected

Clinical features and outlook

A limb girdle syndrome with variable prognosis. Ambulation may be lost between the second and fifth decade and later morbidity is associated with cardiac and respiratory involvement. Pain can be a feature and may need to be addressed.

Associated features and anaesthetic risks

Anaesthesia presents a major risk and should be performed with full cardiac and respiratory review.

Cardiac manifestations

Cardiac follow up is mandatory with yearly ECG and 2 yearly ECHO unless abnormalities develop and then at frequency as determined by cardiology. Cardiac disease can occur even when skeletal muscle weakness is not significant.

Respiratory manifestations

Respiratory muscle weakness and resulting symptoms should be monitored yearly with early referral to a respiratory physician. Late respiratory failure usually follows loss of ambulation.

Cognitive manifestations

Some patients may have learning difficulties, behavioural problems or autistic traits. It would be important to address them accordingly.

Patients should carry an alert bracelet or similar.

Sources of additional information:

www.nn.nhs.scot/smn/
www.musculardystrophyuk.org
www.treat-nmd.org/
https://neuromuscular.wustl.edu/