Scottish Muscle Network

Limb Girdle Muscular Dystrophy type 1B (LGMD1B) "at a glance" sheet

Also known as Laminopathy

Basis / molecular abnormality

- Autosomal Dominant
- Mutation in LMNA/C gene, encoding lamin A/C.
- Genetic counselling mandatory

Clinical features and outlook

Progressive skeletal muscle weakness which may affect proximal or distal limb muscles. Contractures may be prominent. Onset and severity of weakness varies widely.

Associated features and anaesthetic risks

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

Cardiac manifestations

There is a **very high risk** of cardiac conduction block, atrial and ventricular arrhythmias and sudden cardiac death. This can occur with no or minimal skeletal muscle weakness. Pacemakers do not reduce this high risk. Implantable cardioverter defibrillators (ICDs) should be considered. These individuals must have annual cardiac follow up with at least 24 hour ECG and echo and should be under the care of a cardiologist.

Respiratory manifestations

Respiratory muscle weakness and respiratory failure may occur. Be aware of symptoms and refer early to respiratory physician for assessment.

Patients should carry an alert bracelet or similar

Sources of additional information

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

NOTE

This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined on the basis of 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.