Scottish Muscle Network

Limb Girdle Muscular Dystrophy (LGMD) “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

There are more than 30 types of LGMD. The diagnosis is based on the clinical phenotype and is **sometimes** supported by genetic confirmation. Two patterns of inheritance are currently known, autosomal dominant and autosomal recessive.

Clinical features and outlook

Progressive skeletal muscle weakness which affects proximal +/- distal limb muscles. In some Limb Girdle types, the face may also be affected. Joint contractures and scapular winging may be a feature. Some people may eventually require a wheelchair. Life span depends on the LGMD type and may be reduced.

Associated features and anaesthetic risks

LGMDs can be associated with a respiratory muscle weakness and so anaesthesia may carry a higher risk. Malignant hyperthermia (MH) like reactions may occur in some subtypes.

Cardiac manifestations

In some LGMDs, people can develop heart arrhythmias and/or heart block and/or cardiomyopathy. Knowledge of the genetic subtype will inform cardiac risk. Cardiology review is recommended.

Respiratory manifestations

Respiratory weakness can be associated with some types of LGMD. When the genetic diagnosis is not confirmed, symptoms should be monitored with referral to respiratory physician for assessment.

Patients should carry an alert bracelet or similar.

Sources of additional information:

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