Medication and anaesthetic precautions

- It is essential that the anaesthetist is aware of the diagnosis of LGMD1 to allow appropriate pre-operative assessment and post-operative monitoring.
- LGMD1 patients may experience increased sensitivity to sedatives, inhaled anaesthetics and neuromuscular blockade.
- Local anaesthetics and nitrous oxide are safe (e.g. for minor dental procedures).

Fractures and falls

- Owing to weakness, contractures and poor balance, patients with LGMD1 are at high risk of frequent falls.
- If the patient is ambulant before fracture, internal fixation is preferable to casting as it helps to preserve muscle and speeds a return to walking.
- Orthotics input is often important, especially for ankle weakness.
- It is advised to check vitamin D levels and bone mineral density on a regular basis, especially following a fall or fracture.

Subtypes of LGMD1

- **LGMD1B** is caused by mutations in the lamin A/C gene. The phenotype can vary and can selectively involve only the muscle or skin or be multi-systemic. LGMD1B is characterised by predominant proximal weakness in the lower limbs, contractures and **cardiac arrhythmias and dilated cardiomyopathy** with risk of sudden death. Respiratory insufficiency occurs as muscle weakness progresses; NIV may be required in more severely affected patients. CK is moderately elevated.
- **LGMD1C** is caused by mutations in the caveolin 3 gene and is characterised by an onset usually in the first decade, a mild-to-moderate proximal muscle weakness; however distal weakness can also occur. Muscle hypertrophy, especially enlarged calves, is a common feature as are cramps and rippling muscle disease. Cardiac and respiratory function are generally not affected; however, dilated cardiomyopathy has been rarely reported.

TREAT-NMD Neuromuscular Network

Muscular Dystrophy UK
0800 652 6352 / info@musculardystrophyuk.org
www.musculardystrophyuk.org

Registered Charity No. 205395 and Registered Scottish Charity No. SC039445

Muscular Dystrophy UK
Fighting muscle-wasting conditions

Alert card

Limb girdle muscular dystrophy Types 1

Name _____

Date of birth _____ **NHS number** _____

If presenting at an emergency department, contact the neurology/neuromuscular team and respiratory team at:

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Limb girdle muscular dystrophy Types 1

Limb girdle muscular dystrophy Types 1 (LGMD1) are a group of muscular dystrophies that predominantly cause weakness in the shoulder and pelvic girdle and are inherited in an autosomal dominant pattern. The group is further divided into subtypes based on the underlying genetic cause, with a progressive alphabetical letter indicating the chronological order of gene identification.

LGMD1 patients may need to use walking aids, can have difficulties climbing stairs and lose the ability to walk. Creatine kinase (CK) levels can be normal to moderately elevated.

The various subtypes of LGMD1 can differ in terms of condition onset, progression, condition severity and involvement of other systems. Prognosis and management, therefore, are not uniform across the subtypes of LGMD1. Nonetheless, early identification of complications and risk factors is crucial.

Cardiac

- **Cardiomyopathy and/or dysrhythmias** are very common in some subtypes of LGMD1, whereas some other forms don't have cardiac complications.

Respiratory

- Symptoms of nocturnal hypoventilation may signal the development of significant respiratory muscle weakness and need for intervention. Non-invasive ventilation (NIV) may be required. If supplemental oxygen is required during a respiratory crisis, this must be carefully controlled and carbon dioxide levels monitored, especially in the context of chronic respiratory failure.
- Assisted coughing with chest physiotherapy and breath-stacking techniques with an AMBU bag help to clear lower airways secretions. This can also be facilitated by a cough assist device.
- Immunisations should be kept up to date, including the flu and pneumococcal vaccines.

Recommendations and precautions

- **Swallowing difficulties** are rarely reported in LGMD1 patients, however if present, these should be assessed by a SALT.
- **Bowel function** is generally normal in LGMD1 patients; however some patients can experience constipation. If this is severe, it may require specialist input to exclude other causes.
- **Liver** enzymes (AST/ALT/alkaline phosphatase) may be mildly raised on blood tests in up to 50 percent of patients. The clinical setting dictates whether further investigation is indicated.
- Some subtypes of LGMD1 can have **central nervous system** involvement with intellectual disability and/or epilepsy and rarely movement disorders.