limb weakness. If applicable, consider the rising seat option to help the individual get in and out of the chair independently and preserve limited ambulation capabilities for as long as possible.

Respiratory

- Respiratory muscles are typically unaffected by GNE myopathy, but monitoring of lung function is advisable.
- Individuals with an early onset of the condition may be at higher risk of respiratory weakness and respiratory failure. Symptoms may include morning headaches, fatigue, and reduced appetite.
- Annual flu and COVID-19 vaccinations (if eligible) are recommended.

Anaesthetic precautions

- Individuals with neuromuscular conditions could be more sensitive to certain anaesthetics and may take longer to wake up from surgery. The anaesthetist should be aware of the diagnosis of GNE myopathy and any respiratory weakness to allow for a pre-operative assessment.
- There is no evidence at present that GNE myopathy is associated with malignant hyperthermia (a severe reaction to certain anaesthetic drugs and muscle relaxants).
- Local anaesthetics and nitrous oxide are safe for minor dental procedures.

Publication date: Jan 2025
Review date: Jan 2028

www.musculardystrophyuk.org
Registered Charity No. 205395 and Registered Scottish Charity No. SC039445

MUSCULAR DYSTROPHY UK

Alert card

GNE myopathy

Name ..... Date of birth .....

NHS/CHI/H&C number .....

If presenting at A&E, contact the specialist team at:

.....

as soon as possible on: .....

For information and support, contact us on our helpline 0800 652 6352 or email info@musculardystrophyuk.org

GNE myopathy

GNE myopathy is a rare condition that causes progressive weakness in the muscles used for movement. Symptoms usually occur in adulthood.

GNE myopathy is also known as hereditary inclusion body myopathy (HIBM), distal myopathy with rimmed vacuoles (DMRV), or Nonaka myopathy.

The condition usually does not affect the heart, lungs, or muscles used for speech and swallowing.

Muscle weakness

- GNE myopathy often starts with distal muscle weakness in the lower legs. The front calf muscles are affected at the very early stages in the majority of individuals. This makes it difficult to lift the feet, known as footdrop. Weakness is expected to progress and will affect both upper and lower limbs. This can affect the individual's mobility and ability to complete day-to-day activities.
- Quadricep muscles may remain strong despite other muscles in the legs weakening. This is unique to GNE myopathy.

Physiotherapy

- Staying active is important and physiotherapists can provide advice on how to adapt to the progression of the condition and come up with a suitable exercise plan. Regular activity and exercise can help maintain muscle strength and functional abilities.
- It is important to increase the mobility of muscles and prevent tightening by doing stretches regularly. An orthotics assessment and splints may be beneficial. Combining stretches with the use of splints, when applicable, can help manage contractures (joint and muscle tightness).
- Contractures are more prevalent in wheelchair users – physiotherapists can prescribe wheelchair-based exercises and assess posture and postural management.

Mobility and falls

- Individuals with GNE myopathy are prone to falls due to muscle weakness and reduced balance. Getting up from the floor can be very difficult or impossible without help.
- Minimisation of fall risks in all environments is necessary, and a risk assessment can be carried out by a physiotherapist or relevant healthcare professional. Aids, such as a walking stick or orthosis, can improve stability and independence. These can be provided by a physiotherapist or occupational therapist.
- Wheelchair users may be at a higher risk of developing osteoporosis and be more susceptible to fractures.
- Individuals may need to use a powered wheelchair at the later stages of the condition due to the presence of upper