GI/Liver/Cognitive (EDS) continued

Cognitive

- Excessive daytime sleepiness (EDS) is common and is most often owing to CNS involvement. Sleep apnoea and chronic respiratory failure also need to be considered.
- Dysexecutive problems and apathy are common. Patients may miss appointments. Telephone reminders, longer appointment times and a more lenient approach to discharge following DNAs may help.

Precautions / recommendations

- Patients often have facial myopathy, slurred speech and EDS, which may make it difficult for patients to express their feelings. It may also make patients look worse than they feel.

Fractures and falls

- Owing to weakness and poor balance, patients with DM1 are at high risk of frequent falls.
- If 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.
- Consider checking vitamin D levels and bone mineral density, especially following a fall or fracture.

We are here to support people living with myotonic dystrophy:

Muscular Dystrophy UK
Fighting muscle-wasting conditions

Myotonic Dystrophy SUPPORT GROUP

0800 652 6352 / info@musculardystrophyuk.org
www.musculardystrophyuk.org

Health & care information you can trust

Muscular Dystrophy UK
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Alert card

Myotonic dystrophy type 1 (DM1)

Name _____

Date of birth _____

NHS number _____

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

_____

as soon as possible on:

_____

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

Respiratory

- Chronic respiratory failure is common in myotonic dystrophy type 1 (DM1). It may present with early morning headaches, fatigue and excessive daytime sleepiness, but is often first identified following an episode of pneumonia or a difficult or prolonged extubation following general anaesthetic.
- Pneumonia is very common and requires prompt management.
- If supplemental oxygen is required during a respiratory crisis it must be carefully controlled and carbon dioxide levels monitored, especially in the context of chronic respiratory failure. Non-invasive ventilation (NIV) may be required, but is often poorly tolerated.
- Assisted coughing with chest physiotherapy and breath-stacking techniques with an AMBU bag helps 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.

Cardiac

- Bradyarrhythmias and tachyarrhythmias are very common in DM1 and must be considered in patients with palpitations, fainting, dizziness and shortness of breath but may be symptomless. ECG is mandatory and will often demonstrate prolonged PR and QRS interval.
- Clinically significant cardiomyopathy is uncommon in DM1, and if present other causes should be considered.

Anaesthetics / sedation

- There is an increased sensitivity to sedatives, inhaled anaesthetics and neuromuscular blockade, especially in more severe forms of DM1. It is essential that the anaesthetist is aware of the diagnosis of DM1 so that appropriate plans can be made for post-operative monitoring.
- Local anaesthetics and nitrous oxide are safe, e.g. for minor dental procedures. Ideally the surgery should occur in a specialist centre with staff experienced in managing these individuals.

Anaesthetics / sedation continued

- Detailed anaesthetic guidelines are available at: **www.smn.scot.nhs.uk/myotonicdystrophy.html**.

GI/Liver/Cognitive (EDS)

Gastrointestinal (GI)

- Constipation, diarrhoea and abdominal pain are very common in DM1 but may need assessment to exclude other causes.
- Aspiration pneumonia, secondary to dysphagia, is common.
- Patients should be assessed by a SALT (speech and language therapist) if they have swallowing problems.

Liver

- 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.