Endocrine/GI/Liver/Cognitive (EDS) continued

Gastrointestinal

- Constipation, diarrhoea and abdominal pain are common in DM2 but may need assessment to exclude other causes.
- Aspiration pneumonia, secondary to dysphagia, is not common in DM2.
- Patients should be assessed by a SALT if they have swallowing problems.

Cognition

- Excessive daytime sleepiness (EDS) is common. Sleep apnoea and chronic respiratory failure need to be considered as contributing to the cause of EDS.
- Dysexecutive problems and apathy are not common in DM2, in comparison with DM1. Intellectual disability has not been found in DM2, although changes in cognition, behaviour and personality have been described.

We are here to support people living with myotonic dystrophy: Muscular Dystrophy UK – Fighting muscle-wasting conditions; Myotonic Dystrophy Support Group

Fractures and falls

- Owing to weakness and poor balance, patients with myotonic dystrophy are at high risk of frequent falls when their muscles are significantly involved.
- Weakness affects the proximal muscles often more than the distal muscles in DM2 which may cause problems with climbing stairs, getting out of a chair, and off the floor.
- If ambulant before fracture, internal fixation is preferable to casting as it helps to preserve muscle and speeds a return to walking.
- Cataracts are common in DM2 and should be considered in all patients with falls.
- Orthotics input is often important, especially for ankle weakness.
- Consider checking vitamin D levels and bone mineral density, especially following a fall or fracture.

Muscular Dystrophy UK
Fighting muscle-wasting conditions

Alert card

Myotonic dystrophy type 2 (DM2)

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:

_____

Activate your alert card today to receive your vital care plan:

Email info@musculardystrophyuk.org or
call our Freephone helpline 0800 652 6352

Respiratory

- Chronic respiratory failure is relatively uncommon in myotonic dystrophy type 2 (DM2) in comparison to 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.
- 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 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.

Cardiac

- Bradyarrhythmias and tachyarrhythmias are less common in DM2 in than in DM1. They should still 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 DM2, and if present, other causes should be considered.

Anaesthetics / sedation

- Unlike in DM1, studies have suggested that there does not appear to be an increased sensitivity to sedatives, inhaled anaesthetics and neuromuscular blockade in DM2. However, it is essential that the anaesthetist is aware of the diagnosis of DM2 and any background respiratory problems, so that appropriate plans can be made for potential prolonged post-operative monitoring.

Anaesthetics / sedation continued

- Local anaesthetics and nitrous oxide are safe, e.g. for minor dental procedures.
- Detailed anaesthetic guidelines are available for DM1 and may also be applicable in patients with severe DM2. You can find them at: **www.smn.scot.nhs.uk/myotonicdystrophy.html**.

Endocrine/GI/Liver/Cognitive (EDS)

Endocrine

- Increased incidence of type 2 diabetes mellitus in DM2.

Liver

- Liver enzymes (AST/ALT) may be mildly raised on blood tests in up to 50 percent of patients. The clinical setting dictates whether further investigation is indicated.