Speech and swallowing

- Swallowing difficulties are common but often unnoticed. Frequent chest infections and symptoms of food sticking in the throat, coughing and choking while eating, and unplanned weight loss should be investigated. Refer to a speech and language therapy team.
- Strategies to prevent complications include sitting upright, careful chewing, and avoiding talking during meals. Diet modifications and nutritional supplements may be required. In severe cases, a feeding tube may be considered.
- Weakness and contractions in head and neck muscles can affect speech clarity. Encouraging slower speech and extra breaths in conversation can help improve communication.

Cognition

- Cognitive and behaviour changes may occur and can potentially result in neglecting physical health needs and impact ability to make decisions, solve problems, and multitask.
- Apathy is common and individuals may miss appointments. Phone reminders and a lenient approach to discharge following non-attendance may help.
- Excessive daytime sleepiness is common and may require medication. Sleep apnoea and chronic respiratory failure should be considered as potential causes.

Publication date: Jan 2025
Review date: Jan 2028

Mobility and falls

- Muscle weakness and reduced balance impacts ability to walk and perform functional tasks e.g. standing from sitting or using stairs. There is increased risk of falls, injuries, and fractures.
- When managing fractures, consider the individual's functional ability. Internal fixation is preferred to casting to reduce risk of further muscle wasting in those who can mobilise. Early physiotherapy and ongoing rehabilitation are advised. Local HCPs should liaise with specialist neuromuscular teams for advice on long term rehabilitation.

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MUSCULAR DYSTROPHY UK

Alert card

Myotonic dystrophy type 1 (DM1)

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

Myotonic dystrophy type 1 (DM1)

DM1 is a genetic condition that causes progressive muscle weakness and wasting. DM1 can affect multiple parts of the body, and the symptoms can vary in type and severity.

Anaesthetics and sedation

- There is increased sensitivity to sedatives, including opioids and benzodiazepines, as well as inhaled anaesthetics, and neuromuscular blocking drugs. It is essential that the anaesthetist is aware of the DM1 diagnosis to allow appropriate pre-operative assessment and post-operative monitoring. Close liaison between surgical, anaesthetic, and respiratory teams is necessary.
- Local anaesthetics and nitrous oxide are safe for minor dental procedures.

Respiratory

- Chronic respiratory failure can occur. Signs include morning headaches, fatigue, and excessive daytime sleepiness. Respiratory failure may first be noticed after a pneumonia episode or may affect recovery from general anaesthetic.
- In a crisis, supplemental oxygen must be controlled and prompt blood gas tests to assess for respiratory failure. Non-invasive ventilation (NIV) and long-term nocturnal NIV may be required. Contact the respiratory team/ventilation unit for advice.
- Assess secretion management and consider cough augmentation techniques such as assisted coughing, breath stacking with a LVR bag, or cough assist device to clear lower airway secretions.
- Pneumonia is common and requires prompt management and antibiotics. Aspiration of food, drink, or saliva can be a common cause. Pneumococcal, annual flu, and COVID vaccination (if eligible) are recommended.

Cardiac

- Arrhythmias including sinus bradycardia, heart block, atrial fibrillation and flutter, and ventricular tachycardia are common. Symptoms include chest pain, palpitations, dizziness, and difficulty breathing. Some may be asymptomatic. ECG test is required and often shows prolonged PR and QRS interval. Some individuals may have a pacemaker, implantable cardioverter defibrillator, or be on anticoagulants.
- Cardiomyopathy can occur in advanced DM1 but is uncommon in early or mild DM1; other causes may need to be considered.
- Venous thromboembolism is a potentially life-threatening complication. Consider this as a differential diagnosis when cardiac symptoms occur.

Gastrointestinal

- Constipation, diarrhoea, and bloating are common and typically respond to optimising fluid and fibre and establishing a toilet routine. Severe constipation not responding to laxatives or suppositories may need assessment to exclude other causes or gut dysmotility. Chronic diarrhoea requires investigation.
- Liver enzymes (AST/ALT) may be raised on blood tests. Further investigation will depend on the clinical context.