

Facial weakness

- Weak facial muscles can make it difficult to close the eyes while sleeping and can lead to dry eyes and, in severe cases, corneal ulcers. Using eye lubricants can protect the eye and prevent irritation.
- Substantial facial muscle weakness can impact the ability to display typical emotional expressions, particularly in those with severe childhood-onset FSHD.

Mobility and falls

- Individuals with FSHD are prone to falls due to muscle weakness and reduced balance. Using aids such as a walking stick can help improve balance and stability, reducing the risk of falls.

- Orthotics input is important for ankle weakness. Orthotics can also support weakened core muscles and muscles around the shoulder blades.
- If a fall leads to a fracture, immediate specialist attention is required. Contact the specialist neuromuscular team.
- For individuals who can undergo surgery, internal fixation with early rehabilitation is preferable to casting. Referral to hospital therapy teams and community teams once at home is vital to preserve muscle strength and function. This will facilitate a return to baseline function, whether that is transferring, walking, or all activities including sports. It is optimal to avoid periods of bed rest and immobility as this can lead to rapid loss of muscle strength and function.

Anaesthetic precautions

- Some individuals may be more sensitive to certain anaesthetics. It is essential that the anaesthetist is aware of the diagnosis of FSHD and any respiratory weakness to allow pre-operative assessment and post-operative monitoring.
- Local anaesthetics and nitrous oxide are safe for minor dental procedures.

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**MUSCULAR
DYSTROPHY
UK**

Alert card

**Facioscapulohumeral
muscular dystrophy (FSHD)**

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

NHS/CHI/H&C number

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

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as soon as possible on:

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

Facioscapulohumeral muscular dystrophy (FSHD)

FSHD is a progressive muscle wasting condition. Muscle weakness is usually in the face, shoulders, and upper arms. Lower legs, forearms, wrists, ankles, abdominal, and back muscles can also be affected at later stages in the condition. The severity of the condition and the muscles affected varies between individuals.

Respiratory

- Respiratory function varies among individuals with FSHD depending on the severity of their condition. In some cases, type 2 respiratory failure may occur because of weak breathing muscles and a restrictive chest wall, causing shortness of breath and shallow breathing at night (nocturnal hypoventilation).

- If breathing function is impaired and supplemental oxygen is required during a respiratory crisis, aim for SpO₂ over 94% and monitor CO₂ levels with a blood gas test. Non-invasive ventilation (NIV) may be required if breathing rate is greater than 20 breaths per minute (if usual vital capacity is less than 1L) or pH is less than 7.35 and pCO₂ is higher than 6.0kPa.
- Chest infections could be common, and those with swallowing difficulties (dysphagia) might also be at risk of aspiration pneumonia.
- Cough augmentation techniques such as assisted coughing with chest physiotherapy, breath stacking techniques with a LVR bag, and/or a cough assist device may help clear lower airway secretions during severe chest infections.

Cardiac

- Heart function is typically unaffected. Further investigation is necessary if individuals experience cardiac symptoms like palpitations, fainting, dizziness, or shortness of breath.

Physiotherapy and pain

- Staying active is important and specialist clinics can provide advice on healthy activities and suitable exercise. Regular activity and exercise can help maintain muscle strength of non-affected or minimally affected muscles and manage pain. This can also, potentially, help individuals to remain independent for longer.
- Individuals with FSHD often have chronic musculoskeletal pain, indirectly caused by the symptoms of FSHD, especially in the neck, shoulders, and lower back.

- Weakened core muscles can lead to an exaggerated inward curve of the spine (lumbar lordosis) due to compensatory postural adjustments.
- Physiotherapists and occupational therapists can advise on simple pain-relieving strategies, postural management, and fatigue management.

Gastroenterology

- Symptoms like abdominal discomfort and constipation seem to occur more frequently. They typically respond to strategies of optimising fluid and fibre intake and establishing a toilet routine.
- The liver is usually not affected. If a liver issue is suspected, appropriate investigations are needed. CK and liver enzymes (AST/ALT) may be elevated due to muscle damage.