Emergency Medical Considerations for FSHD Patients

Attention Clinician: Clinical Considerations for Neuromuscular Disease (NMD) Patients

The information on this page has been reviewed by Joshua Benditt, MD, a board certified physician at the University of Washington Medical Center, where he is medical director of Respiratory Services and a UW professor of Pulmonary, Critical Care and Sleep Medicine. You can also find this information on the FSHD Society website.

You have been referred to this information by a person who has Facioscapulohumeral Muscular Dystrophy (FSHD, **ICD-10 code G71.02**), a type of neuromuscular disease. This individual may have respiratory muscle weakness and may use mechanical/assisted ventilation and other respiratory muscle aids.

- This document is shared in the spirit of positive collaboration and to spread awareness that many treatments that help individuals without NMD may **be harmful** to individuals with NMD such as FSHD.
- This individual and/or their NMD care team are experienced in NMD respiratory involvement and have learned what does and does not work.
- **Check their Medical Alert card for emergency contacts and medical providers.**

Emergency Considerations

- **Beware of:**
    - Supplemental oxygen (O2)
        - Only administer supplemental O2 for sustained periods with monitoring EtC02 capnograph.

        - If given without the individual's mechanical/assisted ventilation, it can cause:
            - Decreased responsiveness;
            - Hypercapnia (an increased level carbon dioxide in the blood and lungs);
            - Suppressed respiratory drive, which can be life-threatening and cause respiratory arrest.
    - General anesthesia medications – e.g. **volatile inhalation anesthetics, and depolarizing muscle relaxants.**
    - Narcotics, sedatives, and opioids can cause - Potentially life-threatening suppression of breathing, especially when mechanical/assisted ventilation is not in use.

General Respiratory Involvement

- Respiratory muscle weakness:
    - Affects muscles between ribs, diaphragm, and sometimes bulbar (mouth and throat) muscles.
        - Can cause orthopnea (discomfort and difficulty breathing while lying down). A semi-reclined position may be required along with their mechanical ventilation during examination and/or therapy.
- Mechanical/assisted Bi-level ventilation:
    - Assists ventilation (movement of air into and **out** of the lungs);
    - Corrects oxygen (O2) **and carbon dioxide (CO2)** gas exchange abnormalities;
    - Can be used continuously via a mask, mouthpiece, or a tracheostomy tube.
- Manual and mechanically assisted cough:
    - Assists a weak and/or ineffective cough;
    - Clears airway secretions and can prevent infection.
- Narrow, restricted airway can cause:
    - Difficult intubation and need for smaller endotracheal tube.

- Reduced lung volume results from:
    - Areas of micro-atelectasis (small areas of collapse) in the lungs;
    - Skeletal abnormalities such as scoliosis (progressive curvature of the spine).
- Discussion with this individual and his or her NMD care team is encouraged.