



Managing Respiratory Symptoms in Neuromuscular Disease

Kimberley W. Cawley, RRT
Registered Respiratory Therapist
University of Rochester

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Incidence of Respiratory Complications in FSHD

- Documented incidence is rare but due to the nature of the disease, we feel that being aware of and monitoring for respiratory insufficiency is important.
- Increased risk with:
 - Kyphoscoliosis
 - lumbar hyperlordosis
 - Pectus Excavatum
 - Wheelchair dependency

Identifying patients at Risk for Developing Respiratory Insufficiency

- Monitor FVC and MIP annually and as needed in between.
- Pulmonary function studies before any surgery requiring general anesthesia.
- Routine questioning during examination to include: incidence of shortness of breath, orthopnea, morning headaches, snoring or evidence of sleep apnea
- Special attention to patients with severe kyphoscoliosis and those that have become wheelchair bound.

Managing Symptoms

- Shortness of breath
- Orthopnea
- Morning Headaches, snoring, or evidence of sleep apnea.
- Decreased Maximum Inspiratory Pressure or Negative Inspiratory Force (MIP or NIF)

Non-Invasive Ventilation

- Initial equipment is a Bi-level positive pressure machine, or BIPAP (Respironics TM)
- Bi-level therapy consists of a positive pressure mask connected to a machine that delivers two different set pressures.

Inspiratory Pressure

- The Inspiratory Pressure is delivered at the start of inhalation and will decrease the work of breathing on the muscles responsible for inhaling. This pressure also dictates the size of the breath, or tidal volume that is delivered to the patient.
- In managing respiratory insufficiency or failure, this pressure is set to decrease Carbon Dioxide levels in the blood.

Expiratory Pressure

- Expiratory Pressure acts as a “splint” in the airway to keep it open. When treating sleep apnea without the presence of neuromuscular disease, this pressure is known as “CPAP”
- Expiratory Pressure also improves oxygenation by maintaining a patent airway and recruiting alveoli to make them more available for gas exchange.

Determining Settings

- Patient comfort. If therapy cannot be easily tolerated, patients will be non-compliant and if it's not worn, it doesn't help!
- Titrating
- Formal Sleep studies with titration.
- Patient's often "know" that they need and increase in pressure and it's usually more comfortable to increase both pressures at the same time.
- Supplemental Oxygen can be "bled" into system if needed.

Non-Invasive Ventilation with a conventional ventilator

Used when unable to adequately achieve ventilation goals with Bi-level.

Utilizes similar mask to Bi-level but exhalation ports on mask must be blocked.

Can adjust tidal volume, pressure, flow, and oxygen levels.

More sophisticated alarm system than Bi-level.

Internal battery and FDA approved to connect to external battery if needed.

Invasive Ventilation via Tracheostomy

- Final Option when Non-Invasive Ventilation fails or cannot be tolerated.
- Advantages include patent airway, more settings to “tweak” to achieve ventilation goals, and more options for alarm settings.
- Disadvantages are that trained caregivers become a necessity, and independent living can be a challenge.

Cough Assist

- Sometimes called an “in-exsuffalator”
- Manually produces an adequate cough
- Used daily for prophylaxis and frequency of use is increased with any respiratory symptoms.
- Recommended “treatment” is 10 cough cycles.

Emerson Cough-Assist Machine





Questions?

kwcawley@rochester.rr.com

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