Breathing Health in FSHD

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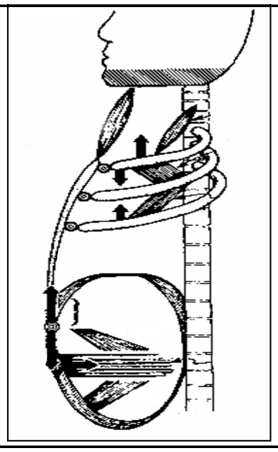
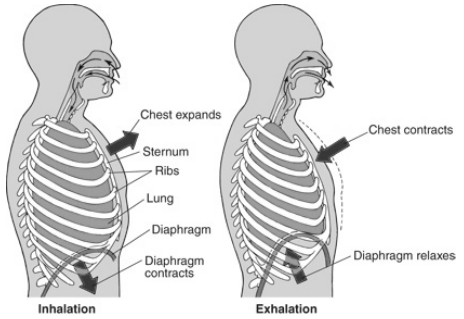
Topics

- Pertinent anatomy
- The problem with sleep and neuromuscular diseases
- Measurement of respiratory function in clinic
- Noninvasive techniques for support
- What you can do to maintain a healthy respiratory system

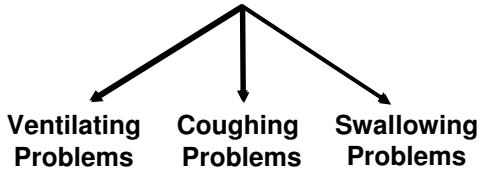
Breathing Muscles



Breathing Movement



Neuromuscular Respiratory Problems

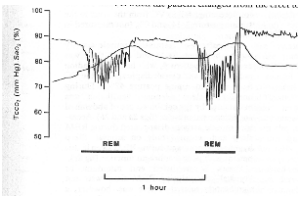


FSHD and Breathing Issues

- Followed 53 patients with FSHD over 10 years.
- 50% of the patients had abnormalities on breathing measurements
  - 13 % had severe involvement
  - 22% had some pulmonary complications
    - *Kilmer Det al. Profiles of neuromuscular diseases: facioscapulohumeral muscular dystrophy. Am J Phys Med Rehabil 1995;74(Suppl); 131-139.*
- More recent study in adults
  - Smaller percentage had breathing problems
  - Patients were adults who may have had a less severe form of disease.
    - *Wolgemuth M, van der Kooi EL, Van Kesteren RG, et al. Ventilatory support in facioscapulohumeral muscular dystrophy. Neurology 2004;63:176-178.*

Ventilating Problems and Sleep

- During sleep, particularly REM, central output to the respiratory muscles decreases
- Weak muscles + decreased drive results in significant hypoventilation with CO_2 retention
- Can result in daytime hypercarbia



- Bye et al. Thorax 1990;45:241-247.

Symptoms of Sleep Problems

- Unexplained nocturnal awakenings
- Vivid nightmares
- Unexplained nightsweats
- Morning headaches
- Daytime hypersomonlence
- Associated:
  - changes in concentration
  - hypertension
  - depression

Ventilatory Assessment

- Forced Vital Capacity
  - Volume of the biggest breath you can take.
  - supine (if diaphragm weakness suspected)
- Maximal Inspiratory Pressure (MIP)
  - Breath in as hard as you can against a pressure measuring device
- Carbon dioxide
  - Arterial blood gas
  - End-tidal (exhaled breath)
  - Transcutaneous

Cough Problems

- Abdominal and rib muscle weakness
- Inability to close glottis
- Reduced peak cough airway flow
- Inability to clear secretions
- Mucous build-up and infection

Expiratory (Cough) Failure Assessment

- Peak Cough Flow
  - Normal > 500 lpm
  - < 160 lpm ineffective clearance
  - < 270 lpm worrisome
    - drops to < 160 seen during infection
- Maximal expiratory pressure
  - ? Minimal value predictive of problems

Cough Peak Flow Meter

Glottic and Swallowing Muscles

- Muscles of mastication
- Tongue
- Pharyngeal and hypopharyngeal muscles
- Laryngeal muscles

Glottic Failure

Airway Protection, Cough and Swallowing

- Intimately related to cough function
- Bulbar muscle involvement
- Portends poorer prognosis
- Difficulty in managing secretions
- Choking episodes and aspiration
- Pneumonia and respiratory failure

Measurement of Swallowing Function

- At bedside or clinic
  - history
  - swallowing challenge
- Barium swallowing study
- Direct endoscopic visualization

Goals of Management

- Ameliorate symptoms
- Improve sleep quality
- Improve quality of life
- Improve and stabilize gas exchange
- Extend survival

Treatment for Ventilatory Problems

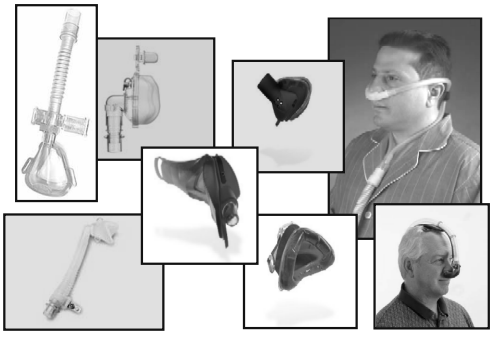
Invasive Ventilation

- tracheostomy

Non-Invasive Ventilation

- NPPV noninvasive positive pressure ventilation (BiPAP)

Interfaces for NPPV-Nocturnal



Nocturnal Ventilators

- Bilevel (pressure support)
- Back-up rate
  - Hypopneas and apneas invariably present
- Modern devices allow for adjustment of inspiratory flow rates
  - "rise time"
- Humidification is important



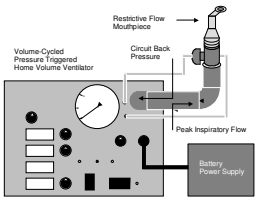
Initiating Nocturnal NPPV

- Screen for symptoms
- Sleep study diagnostic
- or FVC < 50% predicted
- or MIP > -60 cm water pressure
- or $PaCO_2$ > 45 mm Hg
- Noninvasive ventilation
  - BiPAP or VPAP
  - Nasal mask
  - Mouth leak

24 Hour Per Day NPPV Combination Therapies

- Nocturnal ventilation
  - nasal or oronasal ventilation with pressure or volume
  - mouthpiece ventilation (Europe)
- Daytime support
  - If needed
    - dyspnea during day
    - $PaCO_2$ remains elevated despite nocturnal therapy
  - mouthpiece ventilation
  - occasionally nasal ventilation (ALS)
- Oxygen is almost never needed and can be detrimental

Mouthpiece Ventilator Set-up ("Sip Ventilation")



When Must Tracheostomy Be Considered ?

- Significant glottic dysfunction
- Elevated $PaCO_2$ despite optimal noninvasive therapy
- Recurrent pneumonia
- Patient preference
- Lack of experienced healthcare providers for NPPV

Expiratory or Cough Assistance

- Cough augmentation
  - Assisted cough
    - Abdominal or lateral ribcage thrust
    - Postural maneuvers
  - Resuscitation bag
    - breath stacking
    - elastic recoil augments cough function
  - Cough-Assist (Inexsufflator)
    - requires relatively intact bulbar function
  - Mouthpiece ventilator
    - breath stacking also possible

Cough Augmentation
Mechanical Inexsufflation (Cough Assist™)

Manual & Mechanical Cough Augmentation

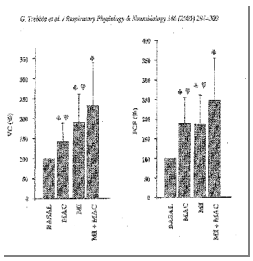
Manual / Mechanical hyperinflation:

- **Increased expiratory flow volume and velocity**
- **Intrathoracic pressure**
- **Portable therapy**
- **Oral strength limitation**
- **Skilled caregiver**

Mechanical + Manual Cough Augmentation

Mechanical Ventilation Supported Insufflation + Manual Assisted Cough

Trebbia G, et al, Resp Phys & Neurobiol, 146 (2005) 291-300

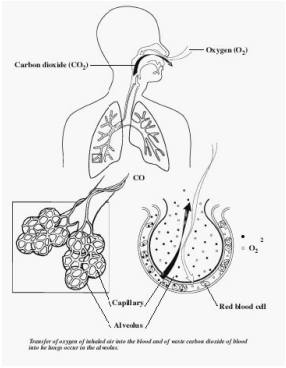


Treatment of Swallowing Problems

- Head down
- Thick liquids
- Avoidance of dry, bulky foods
- Secretion management
- Tracheostomy
- Gastrostomy tube

Oxygen and Carbon Dioxide

- As carbon dioxide goes up, oxygen levels go down
- Weak muscles can cause carbon dioxide to go up
- Giving oxygen when carbon dioxide is up can blunt respiratory drive making CO2 go even higher
- Pulse oximeter finger probes do not measure carbon dioxide but do measure oxygen levels
- If you are sick with respiratory problems in the ER blood gas should be checked not just pulse oximeter



Transfer of oxygen of inhaled air into the blood and of waste carbon dioxide of blood into be lungs occur in the alveolus.

Maintaining Your Respiratory Health

- Avoiding infection
  - Flu shot, pneumonia shot
  - Avoid sick individuals during peak infection season
  - Use hand sanitizer
- Look for symptoms:
  - Of sleep problems
  - Recurrent infections or weak cough
- Assessment
  - Annual vital capacity
  - Other tests as indicated
- ER visit for respiratory infection
  - Oxygen may be given but a check of carbon dioxide must be undertaken
  - Oxygen blunts drive to breathe and may worsen elevated carbon dioxide level