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

Ventilating Problems
Coughing Problems
Swallowing 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
- More recent study in adults
  - Smaller percentage had breathing problems
  - Patients were adults who may have had a less severe form of disease.
Ventilating Problems and Sleep

- During sleep, particularly REM, central output to the respiratory muscles decreases
- Weak muscles + decreased drive results in significant hypoventilation with CO₂ retention
- Can result in daytime hypercarbia


Symptoms of Sleep Problems

- Unexplained nocturnal awakenings
- Vivid nightmares
- Unexplained night sweats
- Morning headaches
- Daytime hypersomnolence
- 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 PaCO₂ > 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
    - PaCO₂ 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 PaCO₂ 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

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

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