The Art of Respiratory Care in Muscular Dystrophy

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Muscles Affected

**Breathing Muscles**
- Accessory muscles
- External Intercostals
- Internal Intercostals
- Diaphragm
- Abdominal muscles

**Muscles of Inhalation**

**Muscles of Exhalation**
Four Respiratory Stages In MD

1. Initially: normal breathing
   - Birth to age 10 or so
2. Normal breathing, but weak cough
   - Early teens
3. Normal breathing during daytime, but poor breathing asleep (Hypoventilation)
   - Mid teens
4. Poor (weak) breathing awake and asleep
   - Mid-late teens
Pulmonary Function Testing
Helps Us To Track Changes

TERMS YOU SHOULD KNOW

1. Forced Vital Capacity
2. Mean Inspiratory Pressure
3. Mean Expiratory Pressure
4. Cough Flow
5. Carbon Dioxide levels
1. **Manually assisted cough**: abdominal thrust following a deep breath (or assisted breath with ambu-bag or other device).
   - Fair at best

2. **Mechanically assisted cough**: a few different brands exist on the market (Respirronics and Hill Rom)
   - HELPS YOU COUGH

Get one/demand one when needed
The Vest

• Approved in other diseases like cystic fibrosis to mobilize secretions
• No great evidence that it works well in DMD
• Probably works well in a hospital situation
• May be beneficial to the patient with recurrent pneumonia
• HIGH cost
There is good evidence that if we correct the problem with surgery that patients do better overall and that if combined with other respiratory support there is even more improvement.
Sleep Studies

Your Sleep Study
What to expect

1 - Patient arrives
2 - Electrode application
3 - Head electrodes
4 - Body electrodes
5 - Lights out
6 - Tech monitors patient
7 - Patient sleeping
8 - Lights on
9 - Patient leaves
Breathing Support

- Non-Invasive Ventilation (BIPAP)
  - helps you to get a good breath in
  - helps to keep your lungs open
  - overall helps make breathing easier at night
- Invasive Ventilation (with a tracheostomy)
  - some people choose this for more help
  - some people don’t
Bottom Line

• Tell us about any breathing problems you are having at each visit and call if they happen in-between visits
• Get pulmonary function testing done at least yearly
• Watch your weight and nutrition
• Have the support at home to help keep you out of the hospital
• Discuss issues with your back at these visits
• Talk about how you are sleeping disordered at each visit and sleep studies can be done at the first symptoms
• Having an emergency plan