Preventing Respiratory Complications of Muscular Dystrophy

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When should you see a lung doctor?

- Generally after age 6 (know your resources)
- Annually until full time in a wheelchair
- More frequently in mid- and late-teens
- Measuring lung function
What is a lung function test?

- **Spirometry**: literally, measuring breath
  - FVC: forced vital capacity: how much air can you push OUT
  - FEV1: how much air can you push out within 1 sec
- **Pressures**: Maximal inspiratory and expiratory
- **SNIP**: nasal sniff inspiratory pressure
- **Exhaled carbon dioxide** (ETCO2)
Stages of respiratory involvement in Duchenne MD

1. Normal (age 0-10)
   - Vaccinate, educate
2. Inadequate cough (age 10-15)
3. Inadequate night time breathing (age 15-20)
4. Inadequate daytime breathing (>age 17)
   - These ages vary greatly!
Stage 2: Inadequate cough

- Often asymptomatic until a respiratory tract infection
- Easily predicted with PFT’s and/or measurement of “peak cough flow”
- Peak cough flow <160 L/min associated with failure to extubate
- PEFR/PCF <270 L/m is indication for assisting cough


Assisting cough:
Manual Assisted Cough

• Deep breath first (Ambu bag, G-P breath, or even vent breath)
• Abdominal thrust or thoracic squeeze
  – Do this on an empty stomach
  – Scoliosis and contractures of thoracic wall limits effectiveness of this technique
Manually assisting cough

Photo courtesy of Dr. John Bach
Mechanically assisted cough

• Preferred to direct tracheal suctioning in pts w/ tracheostomy (more effective, too)
• Can be used via mask, mouthpiece, or tracheostomy
• Achieves effective cough flows even in severely weak patients
• Prophylactic use prevents atelectasis, supports chest wall compliance
Special warning: O2 can be hazardous to your health…

- Supplemental O2 can be dangerous
- Suppresses drive, can cause respiratory failure
- Low saturation means increased airway clearance, need for increased ventilation
- Pulse oximeter very helpful – but check CO2!
Obstructive Sleep Apnea

- OSA: when breathing stops in sleep from upper airway obstruction/collapse
- Diagnosed by SLEEP STUDY
- Especially common in DMD:
  - Large tongue
  - Obesity from steroids
  - Decreased muscle tone of upper airway
Treating OSA

• Treated with MASK therapy
  – CPAP: constant positive airway pressure
• Level of support is adjusted in sleep lab
• Make sure device provided can later deliver BiPAP
Stage 3: Nocturnal Hypoventilation

- Often predicted by lung function test
  - FVC < 30%
- Signs include: A.M. headaches, Increasing awakenings, sleepiness, poor school performance, etc
- Low sats on overnight oximetry
- Retaining CO2 on sleep study
What is “BiPAP?”

• It’s a ventilator used via mask
• Bilevel pressure support
  – An inspiratory pressure to help support the size of the breath IN
  – An expiratory pressure to help keep the airway open
• It’s also a brand name (Respironics): VPAP is the same therapy (Resmed)
Note that FVC<30% in DMD correlates to ventilatory failure (but not in SMA)

Selected cohort FVC% and funct score pred of need for MV in DMD not SMA II (youngest pt 9 yrs)
Management of nocturnal hypoventilation

• *Avoid* tracheostomy, avoid oxygen
• BiPAP/VPAP or other positive pressure ventilator
• Avoid CPAP
  – Increases WOB w/o increasing ventilation
• Mask fit is essential!
• Sleep study to titrate (high-span)
Stage 4: 24 hour ventilation dependence

- No longer is tracheostomy required
- Most patients managed non-invasively
- Mouthpiece ventilator
- Newer, lightweight ventilators
  - Mounts to chair
  - Remaining in school or at work
Non-invasive breathing support

Portable ventilator with a mouthpiece attached (like a microphone) to wheelchair

– Trilogy or LTV1150
– Tremendous improvement in energy level and quality of life
Patrick, age 26, graduating from Pitt Law, 2004
Pulmonary emergencies

- Pneumonia/atelectasis
- Low oxygen level from CO2 retention
  - Respiratory failure
- Fat embolism syndrome after broken bone
THANKS FOR WATCHING!