Introduction

• Respiratory complications in MD can be prevented
• Cost of complications >> cost of prevention
• Newer technologies
• Advocacy has led to improvements in care
• Standards of care now exist
Stages of respiratory function in Duchenne MD

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

• Rule of thumb: ambulatory patient does not require assistance with cough or breathing
• Aside from usual risks of anesthesia no special risks
• Pulmonary function testing in pts over age 6
• Immunization (influenza & s. pneumo.)
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


“Airway Clearance”

2 linked portions of airway clearance:

– **Mucociliary clearance**  
  (impaired in cystic fibrosis, smokers, etc)

– **Cough clearance**  
  (impaired in NMD but also by tracheostomy)
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
Fortunately for Sparky, Zeke knew the famous "Rex maneuver."
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
Respironics CoughAssist

“In-exsufflator”
MI-E -- Indications

- Neuromuscular weakness
- Peak cough flow <270 L/min
- Maximum expiratory pressure <60 cm H2O
- History of difficulty clearing secretions
- NOT indicated for CF or others with normal strength
**Special warning: O2 can be hazardous to your health...**

- Supplemental oxygen can be dangerous
- Suppresses respiratory drive
- Can precipitate respiratory failure
- Low saturation means increased airway clearance, need for increased ventilation
- Pulse oximeter very helpful – check CO2!
Pulse oximetry and O₂

- Development of mucus plugs can be insidious and silent
- Patients with MD should have a pulse oximeters (you can now get them on line)
- Saturation < 95% is indication for aid in coughing
- Oxygen is NOT used to treat hypoventilation!
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
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 studies

• Sleep study to ensure normal CO$_2$
• Patient should awaken feeling refreshed
• Availability is limited in SA (see next slide)
• HIGH SPAN BiPAP means high top number low bottom (18/4 or 22/4)
Sleep labs in South Africa

http://www.sleepmedicine.co.za/find.htm
Effect of NIV on diurnal arterial blood gas tensions: improved!

Table 1. Diagnostic categories, mean age at starting noninvasive positive pressure ventilation (NIPPV) and diurnal arterial oxygen ($P_aO_2$) and carbon dioxide ($P_aCO_2$) tensions before and after NIPPV by diagnostic categories

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Age yrs</th>
<th>Before NIPPV</th>
<th>After NIPPV</th>
</tr>
</thead>
<tbody>
<tr>
<td>CMD</td>
<td>11.6±3.7</td>
<td>8.2±1.2</td>
<td>10.65±2.2</td>
</tr>
<tr>
<td>SMA</td>
<td>5.7±4.2</td>
<td>7.9±1.5</td>
<td>11.9±1.1</td>
</tr>
<tr>
<td>CM</td>
<td>7.3±3.3</td>
<td>8.2±0.6</td>
<td>11.3±1.15</td>
</tr>
<tr>
<td>DMD</td>
<td>13.9±1.5</td>
<td>9.3±2.3</td>
<td>12.17±1.46</td>
</tr>
<tr>
<td>Misc</td>
<td>9.4±4.8</td>
<td>9.4±2.3</td>
<td>9.33±1.87</td>
</tr>
<tr>
<td>Overall</td>
<td>8.5±1.8</td>
<td>7.0±1.6</td>
<td>10.9±1.7</td>
</tr>
</tbody>
</table>

Bear in mind:
Many noses -- many interfaces

Dibujo de George Cruikshank (1792-1878)
Various interfaces used for noninvasive positive pressure ventilation. Standard nasal masks in different sizes (Respironics, Inc.) (upper left), oronasal mask with very soft silicone seal (Resmed, Inc.) (upper right), nasal "pillows" (ADAM Circuit, Puritan Bennett, Inc.) (lower left) and mouthpiece attached to lipseal (lower right).
Why non-invasive ventilation?

- AVOIDANCE of tracheostomy and inherent complications
- Parental or personal choice
- End of life issues…
Stage 4: 24 hour ventilation dependence

- In years past, this was indication for trach
- No longer has to be the case!
- Most patients can be managed non-invasively with mouthpiece ventilation
- Newer, lightweight ventilators facilitate portability and remaining in school or at work
Non-invasive Positive Pressure Ventilation (NIPPV)

• First described by Alexander in 1979

• Non-invasive use of PPV best described by Bach
Respiratory Care of the Patient with Duchenne Muscular Dystrophy
ATS Consensus Statement

Stress is on *anticipation* of respiratory care

NON-INVASIVE management also emphasized

Access to *specialty care* important:
- Pulmonologist
- Nutritionist
- Cardiologist
- Orthopedist
- Physical, speech, and occupational therapists; psychiatry, pastoral care as needed
Kevin, age 17, in ventilatory failure, 24 hour vent-dependent..
Patrick, age 26, graduating from Pitt Law
Attorneys and Professionals
M. Patrick Daniels
Staff Attorney
One Oxford Centre
301 Grant Street
Pittsburgh, PA 15219-1410
T 412-562-1612
F 412-562-1041
danielsmp@bipc.com

M. Patrick Daniels is a staff attorney in the Pittsburgh office of Buchanan Ingersoll PC. He is a member of the firm's Commercial Litigation Section.

Before joining Buchanan Ingersoll, Patrick gained experience through several legal externships while attending law school. In 2001, he worked with the Disabilities Law Project, where he drafted a brochure designed for parents of disabled pre-school children to inform them of their rights regarding daycare services. He also drafted a memorandum discussing the use of "testers" in enforcing fair housing laws.

During 2002, Patrick worked in the U.S. Attorney's Office, Western District of Pennsylvania, in the criminal division of the narcotics/violent crime section. There, he drafted memoranda discussing whether specific facts warranted a particular charge, the application of the "knock-and-announce" rule to federal agents and the use of expert testimony in narcotics cases.

In 2003, Patrick added to his experience by working with the Honorable Chief Justice Ralph Cappy in the Pennsylvania Supreme Court. He drafted allocutus reports and researched material for a speech given by the chief justice titled "Candor to the Court."

Patrick earned his J.D. degree from the University of Pittsburgh School of Law in 2004. He received an Outstanding Achievement Award, as well as the CALI Excellence for the Future Award. He also participated in the Murray S. Love Trial Moot Court. Patrick earned his B.A. degree in political science from Wright State University in 2001.
THANK YOU!

I am always happy to help with questions in the future!

finder@pitt.edu