Lungs matter.

Keith L. Cavanaugh, MD, FAAP, FCCP
April 28, 2018
What we will discuss today

1. The role of a lung doctor
2. The 4 factors
3. Cough & Lung function 101
4. What can we do now?
The 4 factors
1. Ventilation

2. Obstructive Sleep Apnea

3. “Open Lung” Cough ability

4. Swallow Function
When is breathing “Not Normal”?

<table>
<thead>
<tr>
<th>Age</th>
<th>What to expect</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-10 years</td>
<td>Normal</td>
</tr>
<tr>
<td>10-15 years</td>
<td>Inadequate cough</td>
</tr>
<tr>
<td>15-20 years</td>
<td>Inadequate night time breathing</td>
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<tr>
<td>17+ years</td>
<td>Inadequate daytime breathing</td>
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</table>
Diagnosis and management of Duchenne muscular dystrophy, part 2: implementation of multidisciplinary care

Katharine Bushby, Richard Finkel, David J Birnkrant, Laura E Case, Paula R Clemens, Linda Cripe, Ajay Kaul, Kathi Kinnett, Craig McDonald, Shree Pandya, James Poysky, Frederic Shapiro, Jean Tomezko, Carolyn Constantin, for the DMD Care Considerations Working Group*

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British Thoracic Society guideline for respiratory management of children with neuromuscular weakness


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Diagnosis and management of Duchenne muscular dystrophy, part 2: respiratory, cardiac, bone health, and orthopaedic management

David J Birnkrant, Katharine Bushby, Carla M Bann, Benjamin A Alman, Susan D Apkon, Angela Blackwell, Laura E Case, Linda Cripe, Stasia Hadjiannakos, Aaron K Olson, Daniel W Sheehan, Julie Bolen, David R Weber, Learne M Ward, for the DMD Care Considerations Working Group*
How effective is your child’s cough?
Lung Function 101
1. **Spirometry**
   - Forced vital capacity (FVC)
   - FEV1 second (FEV1)
   - Peak expiratory flow (PEF)

2. **Lung Volumes**
   - Total lung capacity (TLC)
   - Functional residual volume (FRC)
   - Residual volume (RV)

3. **Cough peak flow (CPF)**

4. **Maximal static pressures**
   - Inspiratory (MIP)
   - Expiratory (MEP)

5. **Sniff nasal inspiratory pressure (SNIP)**
Lung Volumes and Capacities

- Maximum possible inspiration
- Inspiratory reserve volume
- Vital capacity
- Inspiratory capacity
- Total lung capacity
- Tidal volume
- Maximum voluntary expiration
- Residual volume
- Functional residual capacity
- Expiratory reserve volume
Flow (L/sec)

Inhale

Exhale

Vital Capacity

Peak Expiratory Flow

FEV₁

Vital capacity

Functional residual capacity
Cough Peak Flow

- Less than 160 L/min
- Greater than 160 L/min
- Greater than 270 L/min

Vital capacity

Functional residual capacity
Flow (L/sec)

- Exhale
- Inhale

FVC (L)

FVC (%)

Mayer OH et al
Ped Pulm 2015

N = 60

16-18 years

N = 60

Mayer OH et al Ped Pulm 2015
Forced Vital Capacity less than 50%
“We need to preserve what we’ve got.”
“So for what should I be looking?”

“And what should I do about it?”
The 4 factors
1. Ventilation — Sleep Study
2. Obstructive Sleep Apnea — Sleep Study
4. Swallow Function — Swallow Study
“That is what you can do, but what can I do now?”
Immunize.

Wash your hands.

Don’t smoke.
Mechanically Assisted Breath

Lung Volume Recruitment

“Sip & Puff”

Insufflator Exsufflator
Hill Rom and Respironics Websites
Abdominal thrust or thoracic squeeze

- Do this on an empty stomach
- Scoliosis & contractures of thoracic wall limits effectiveness
Mechanically Assist

Percussive Therapy
Mechanically Assist

Mentions this type of device *once*

“...research is needed to establish efficacy and optimum use...”
What if we started LVR here?
FVC (%) predicted over time

Can we improve FVC?

FVC (%)

Month since LVR

McKim DA et al Arch Phys Med Rehab 2012
FVC (L)

N = 21

Stehling F et al Chronic Resp Disease 2015
PAP Therapy

Restful Sleep
Survival

Eagle F et al, *Chronic Neuromuscular Disorders* 2002

Ventilated

Age at Death

% Survival
Avoid tracheostomy.
Mouthpiece Ventilation
“sip & puff”

Avoid tracheostomy
When to start? How often?

<table>
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<th>Patient’s status</th>
<th>Recommended measurements to be taken during each clinic visit</th>
<th>Frequency</th>
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<td>Ambulatory and age 6 years or older</td>
<td>Sitting FVC</td>
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<td>• Current use of assisted ventilation</td>
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Bushby K et al, Lancet 2010
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*Recommended measurement
Optional measurement
End tidal CO2 (Carbon Dioxide)

“Be cautious with or avoid oxygen use.”

In DMD, hypoxaemia is usually due to hypoventilation, atelectasis, or pneumonia. Therefore, supplemental oxygen therapy should not be used alone.

Type 1 Respiratory Failure: Low oxygen
Type 2 Respiratory Failure: High carbon dioxide
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<th>Assessments</th>
<th>Interventions</th>
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<td><strong>Ambulatory stage</strong></td>
<td>Once yearly: FVC</td>
<td>Immunisation with pneumococcal vaccines and yearly inactivated influenza vaccine</td>
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<td><strong>Early non-ambulatory stage</strong></td>
<td>Twice yearly: FVC, MIP/MEP, PCF, SpO₂, p_a CO₂/pCO₂</td>
<td>Lung volume recruitment when FVC ≤60% predicted</td>
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<td><strong>Late non-ambulatory stage</strong></td>
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<td>Assisted coughing when FVC &lt;50% predicted, PCF &lt;270 L/min, or MEP &lt;60 cm H₂O₁</td>
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<td>Sleep study* with capnography for signs and symptoms of obstructive sleep apnoea or sleep-disordered breathing</td>
<td>Nocturnal assisted ventilation with back-up rate of breathing (non-invasive preferred) when there are signs or symptoms of sleep hypoventilation or other sleep-disordered breathing, abnormal sleep study, FVC &lt;50% predicted, MIP &lt;60 cm H₂O, or awake baseline SpO₂ &lt;95% or pCO₂ &gt;45 mm Hg</td>
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<td>Addition of assisted daytime ventilation when, despite nocturnal ventilation, daytime SpO₂ &lt;95%, pCO₂ &gt;45 mm Hg, or symptoms of awake dyspnoea are present</td>
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Take Home Points

- The 4 factors.
- Wash your hands.
- Take a deep breath.
- Long term goals. This is an investment.

Thank You. Questions??
References