Pulmonary Testing for the DMD Population

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Conflicts of Interest

• None to report
Outline of Talk

• Review respiratory physiology
• Pulmonary decline in DMD
• Pulmonary testing used in DMD
• Sleep disordered breathing in DMD
• Current pulmonary guidelines
The Muscles of Respiration

**Inspiration**
- Muscles that expand the thoracic cavity
  - Diaphragm
  - External intercostals
  - Accessory muscles

**Expiration**
- Muscles that compress the thoracic cavity
  - Mainly elastic recoil of chest wall – no muscle involvement
  - Internal intercostals
  - Abdominal muscles
https://en.wikipedia.org/wiki/Muscles_of_respiration
Normal Airway Clearance

- **Conducting airways**
  - Mucociliary elevator
  - Airflow
    - Cephalad airflow bias
    - Cough

- **Respiratory zone**
  - Chemical absorption
  - Alveolar macrophages
Mucociliary Transport

- Gel Layer (high viscosity and elasticity)
- Sol Layer (low viscosity and elasticity)
- Rapid movement in extension to propel material in gel phase
- Slow return in flexion to starting position, traveling through sol phase
- Most effective in small airways due to large cross-sectional area

60 μm s⁻¹
Cephalad Airflow Bias

- Decreased airway diameter during exhalation results in increased flow velocity
- Increased airflow velocity shears secretions and drives material in direction of flow
- Present in large and small airways but is the primary mechanism of transport in smaller airways
Cough

• 3-Phase process
  - Inspiratory
  - Compressive
  - Expiratory

• Debris clearance
  - Dynamic airway compression
  - Shearing forces at airway wall
Rate of Pulmonary Decline

• Age at loss of ambulation is key

  – Associated with age at peak forced vital capacity (FVC)
    • Loss ambulation < 8 years, peak FVC between 8-11 years at 1.5 L
    • Loss of ambulation > 11 years, peak FVC around 16 years at 2.5 L

  – Associated with rate of decline in FVC \(^{(1,2)}\)
    • Peak FVC < 1.7 L  \(\downarrow\) 8-9% per year
    • Peak FVC > 2.5 L  \(\downarrow\) 4-5% per year

  – Rate of decline in FVC similar in steroid naïve and steroid treated patients once non-ambulatory \(^{(3,4)}\)
Phases of Respiratory Failure

- **Sleep disordered breathing**
  - No hypercapnia / Brief Hypoxemia with OSA
  - Hypercapnea / Hypoxemia during REM sleep only
  - Hypercapnea / Hypoxemia during all stages of sleep

- **Diurnal chronic respiratory insufficiency**
  - Hypercapnea / Hypoxemia when awake/asleep (5,6)
To Measure Lung Function…..

• Age of the Patient
• Cognitive ability
• What is being tested
  – Volume of air (FVC)
  – Airway pressure (MIP/MEP)
  – Flow of air (peak cough flow)
Volume Measurements

Pulmonary Function Testing
Volume Measurements
Pulmonary Function Testing

• Pulmonary Function Testing
  – Forced vital capacity: FVC
  – Forced expiratory volume in 1 second: FEV1
  – FEV1/FVC ratio

• Information
  – Inspiratory muscle strength
  – Expiratory muscle strength
Volume Measurements

Lung Volumes

- Body Plethysmograph needed to measure total lung capacity and residual volume
- Not practical for patient’s in wheel chairs
- Doesn’t give you better information than the FVC measurement
Airway Pressure Measurements

Force Meter

- **MIP**
  - Lowest pressure developed during a forceful inspiration of an occluded airway
  - Measures inspiratory muscle strength
  - -60 or higher is problematic

- **MEP**
  - Highest pressure developed during forceful expiration
  - Measures expiratory muscle strength
  - +60 or lower is problematic
Airway Pressure Measurements

- Maximal Inspiratory Pressure (MIP)
- Maximal Expiratory Pressures (MEP)
- Significantly more variable
- Testing reliability is poor
  - Requires a very experienced, consistent RT
  - Already affected in most young children with DMD
  - Slow rate of decline over time
Airflow Measurements
Peak Expiratory Flow Rate / Peak Cough Flow

- Directly measures expiratory muscle strength
- Indirectly measures inspiratory muscle strength
- Can be measured when performing FVC maneuver
- Lower than 270 L/min associated with respiratory muscle weakness
- Lower than 160 L/min associated with failure to extubate after surgery
Things to Consider

• Pulmonary function tests depend on
  – Patient effort / cooperation
    • At least 2 second exhalation
    • Reproducibility
  – Patient ability to understand / follow commands
    • Cognitive impairment
  – Accurate height / weight
    • Wheelchair dependent
    • Scoliosis
Sleep
Signs of Sleep Disordered Breathing

- Snoring
- Pauses in breathing
- Restless sleep / frequent awakening
- Headache upon awakening that passes quickly
- Daytime drowsiness or hyperactivity
- Increased napping
- Mood lability
At Risk Populations

• **Early Childhood (OSA)**
  - Decreased pharyngeal and upper airway tone
  - Tonsil/Adenoid enlargement in small airways

• **Childhood (OSA)**
  - Obesity from steroid use

• **Teenager (OSA / Alveolar Hypoventilation)**
  - Pharyngeal muscle weakness/upper airway tone
  - Macroglossia
  - Obesity
  - Scoliosis → affecting chest wall mechanics
  - Diaphragmatic weakness
When To Screen?

• Each visit ask about signs of sleep disordered breathing
  • Screen + → obtain a sleep study
• When FVC is around 50%
  • obtain a baseline sleep study
Identify Problems Early

• Undiagnosed Sleep Problems Cause
  • Blunting of the peripheral /central chemoreceptors
    – depresses the respiratory drive
  • Aspiration of secretions
  • Lower lung volumes with sleep
    – Worsening atelectasis
    – gas exchange impairment
  • Cardiac impact
    – Worsening heart strain
    – Risk for arrhythmia
    – Pulmonary hypertension
Recommendations?
# PPMD Pulmonary Management Recommendations

<table>
<thead>
<tr>
<th>AMBULATORY</th>
<th>EARLY NON-AMBULATORY</th>
<th>LATE NON-AMBULATORY</th>
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<tr>
<td><strong>ASSESSMENTS</strong></td>
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<tr>
<td>Once yearly: FVC</td>
<td>Twice yearly: FVC, MIP/MEP, PEF, SpO₂, PetCO₂/PtcCO₂</td>
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<td>Sleep study with capnography* for signs and symptoms of obstructive sleep apnea or sleep-disordered breathing</td>
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| INTERVENTIONS | | |
|---------------|||
| Pneumococcal vaccines and yearly inactivated influenza vaccine | Lung volume recruitment when FVC ≤ 60% predicted | Assisted coughing when FVC ≤ 50% predicted, PEF < 270 LPM, MEP < 60 cm H₂O, 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 ≤ 50% predicted, MIP < 60 cm H₂O or awake baseline SpO₂ < 95% or pCO₂ > 45 mm Hg, Daytime assisted ventilation when despite nocturnal ventilation: daytime SpO₂ < 95%, pCO₂ > 45 mmHg or awake dyspnea*** |

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* See text for definitions.

** Fatigue, dyspnea, morning or continuous headaches, frequent nocturnal awakenings or difficult arousal, hypersomnolence, difficulty ventilating, awakenings with dyspnea and tachycardia, frequent nightmares.

*** We strongly endorse the use of non-invasive methods of assisted ventilation instead of tracheostomy to optimize patient quality of life. Indications for tracheostomy include patient preference, patient cannot successfully use non-invasive ventilation, there failed extubation attempts during a critical illness despite optimal use of non-invasive ventilation and mechanically assisted cough, or failure of non-invasive methods of cough assistance to prevent aspiration of secretions into the lungs due to weak bulbar muscles.
The Pulmonary Action Plan

Well Plan
- Cough at baseline
- Mucus at baseline

Medications:
- Singular – 1 tablet, daily
- Flonase – 2 squirt each nare, daily
- Zyrtec – 1 tablet, daily

Airway Clearance:
cough assist/suction up to 2 X a day

Comments:
cough assist settings are 40/40, 5 cycles of 5, ending on an inspiratory breath.

Step up to sick plan until symptoms resolve or as directed, then return to the well plan

Sick Plan
- Increase cough
- Increase mucus
- Change in mucus color
- Change in mucus consistency

Medications:
- Green Zone Medications
  - Afrin – 1 squirt each nare, before bedtime for up to 3 – 5 days
  - Albuterol – 2 puff or 1 neb, before airway clearance

Airway Clearance:
cough assist/suction up to 4 X a day
May use NIV after airway clearance for 15–20 min

When to Call
- Decreased oxygen saturations. Respiratory symptoms accompanied by fever lasting more than 7 days
- Call Pulmonary Clinic: 816–983–6490

*** Call 911 if lips or fingernails are blue or if you feel your child’s condition worsens ***
References


Questions?