# Current guidelines

## Pulmonary Management

### Ambulatory
- Once yearly: FVC
- Sleep study with capnography* for signs and symptoms of obstructive sleep apnea or sleep-disordered breathing

### Early Non-Ambulatory
- Twice yearly: FVC, MIP/MEP, PCF, SpO2, PetCO2/PtcCO2
- Pneumococcal vaccines and yearly inactivated influenza vaccine
- Lung volume recruitment when FVC ≤ 60% predicted

### Late Non-Ambulatory
- Assisted coughing when FVC ≤ 50% predicted, PCF < 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, hyperomnia, difficulty concentrating, 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, three 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.
Pulmonary function tests

- Spirometry: FVC
- Respiratory muscle strength
- Peak cough flow
- End tidal CO₂ and respiratory rate
Respiratory muscle weakness/impaired cough

- Prolonged cough after respiratory infection
- Weak cough
- Inability to effectively clear secretions
- PCF < 270 L/min is an indication of assisted cough.
RL: 12 years with DMD
RMS: 100%

JR: 10 years with DMD
RMS: 75%
Manual assisted cough

- Deep breath first (Ambu bag)
- Abdominal Thrust or Thoracic Squeeze
Cough assist machine
Prophylactic use and during illness

- Prophylactic use prevents atelectasis, supports chest wall compliance
- Use during respiratory illness - effective airway clearance.
Sleep Disordered Breathing (SDB)

- Polysomnography (sleep study)
  - 1 night in the sleep lab for a diagnostic study
    - Sometimes a 'split night' study will be done
      - First part without support
      - Second part with support
    - If indicated, a second night is done to find the optimal airway pressure to treat SDB
Types of SDB

• OSA: Breathing becomes shallower or stops due to upper airway obstruction
  – Large tongue, obesity, decreased airway tone
• Hypoventilation: Inability to adequately exhale carbon dioxide
  – Muscle weakness
  – May manifest as rapid breathing while asleep
Hypoventilation
Symptoms of Untreated SDB

- Snoring and pauses in breathing*
- Nighttime awakenings +/- dyspnea and palpitations
- Restless sleep
- Unrefreshing sleep
  - Excessive sleep time for age
- Morning headaches
- Excessive daytime sleepiness
- Daytime behavior problems
SDB in DMD

• Inevitable in patients with DMD
  – As early as 12 years old in patients on steroids

• We can treat it!
  – Bilevel Positive Airway Pressure (BiPAP) with a backup rate
  – Higher pressure delivered on inspiration
  – Lower baseline pressure on exhalation and before breathing in again
  – If a patient’s respiratory rate is less than the prescribed rate, the machine kicks in

¹Sawnani H et al. J Pediatr. 2015
BiPAP Masks

NASAL MASKS + XX =
Daytime Assisted Ventilation

• Determined by daytime end tidal CO2 or blood gas

• Nocturnal BiPAP use will have already occurred
  – Ventilator support becomes 24 hours/day
Mouthpiece "Sip" Ventilation

- Essentially a straw connected to the ventilator allowing for breaths on demand.
- Bracket for a wheelchair allows for mobility.
New DMD guidelines

- Sleep study indicated with symptoms of untreated SDB, or FVC < 50%
- Daytime ventilation indicated if end tidal CO2 > 45 or oxygen saturation < 95% on room air
Surgeries and Illnesses

• **Surgeries**
  – If on BiPAP, recommend extubating to BiPAP
    • Airway clearance with cough assist prior to extubation
    • Sip ventilation can also be used if patient is awake and alert enough to initiate breaths

• **Illnesses**
  – Beware of hypoxemia!
    • May be due to high CO2
      – Treat with BiPAP, not just supplemental oxygen
Thank you!

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