Pulmonary Management of Patients With Duchenne Muscular Dystrophy

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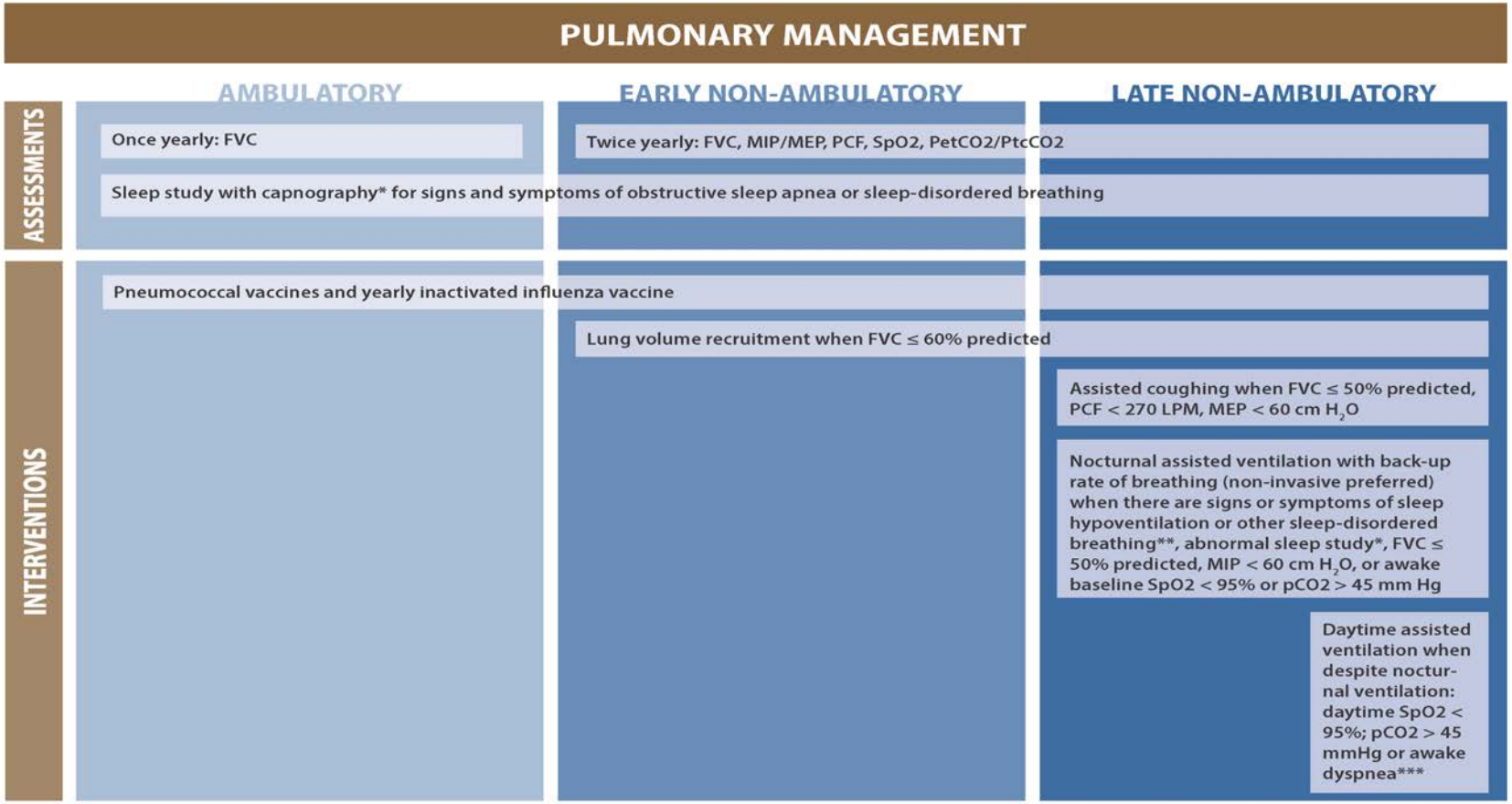
Current guidelines



* See text for definitions.

** Fatigue, dyspnea, morning or continuous headaches, frequent nocturnal awakenings or difficult arousal, hypersomnolence, 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.

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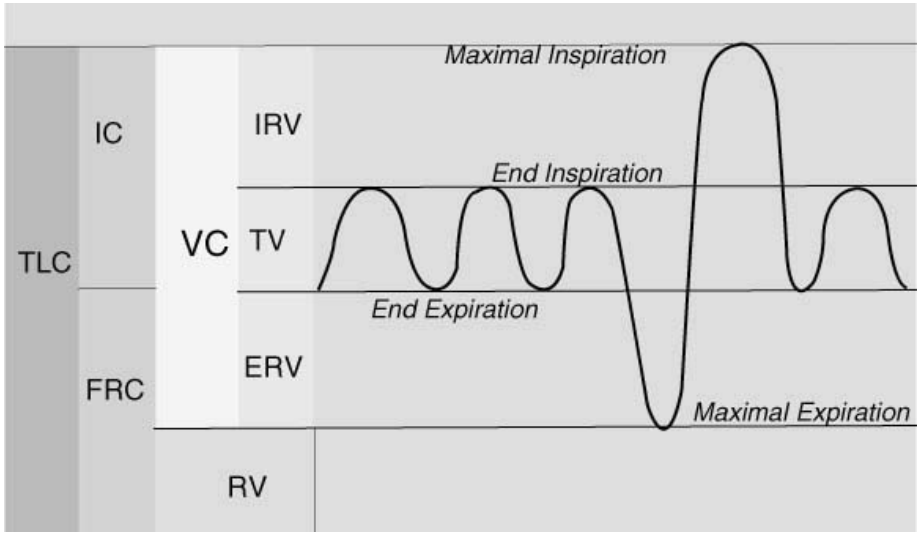
Pulmonary function tests

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



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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.

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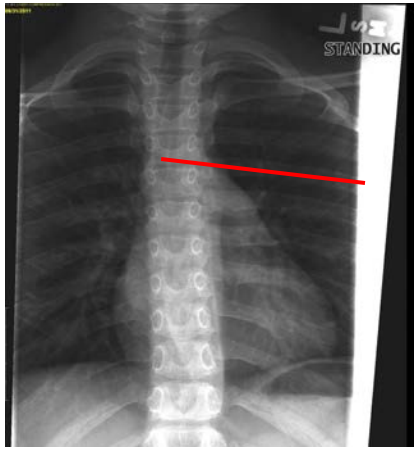
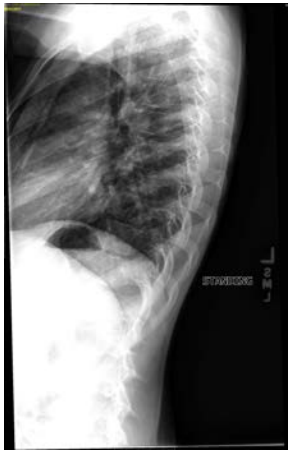
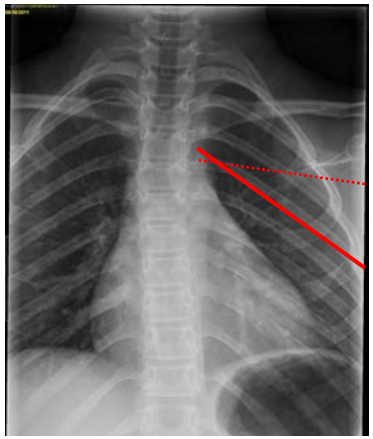
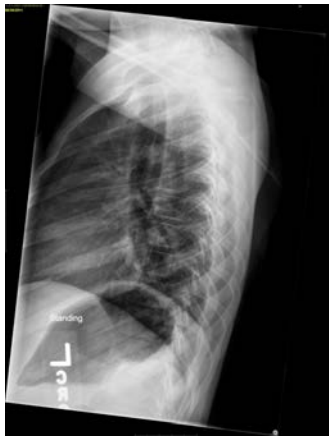




RL: 12 years with DMD

RMS: 100%





JR: 10 years with DMD

RMS: 75%

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Manual assisted cough

- Deep breath first (Ambu bag)
- Abdominal Thrust or Thoracic Squeeze

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Cough assist machine

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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 … gnostic study
    - Sometim … be done
      - First p … part *with* support
  - If indicate … one to find the optimal ai … SDB

Source: Google Images, November 12, 2018

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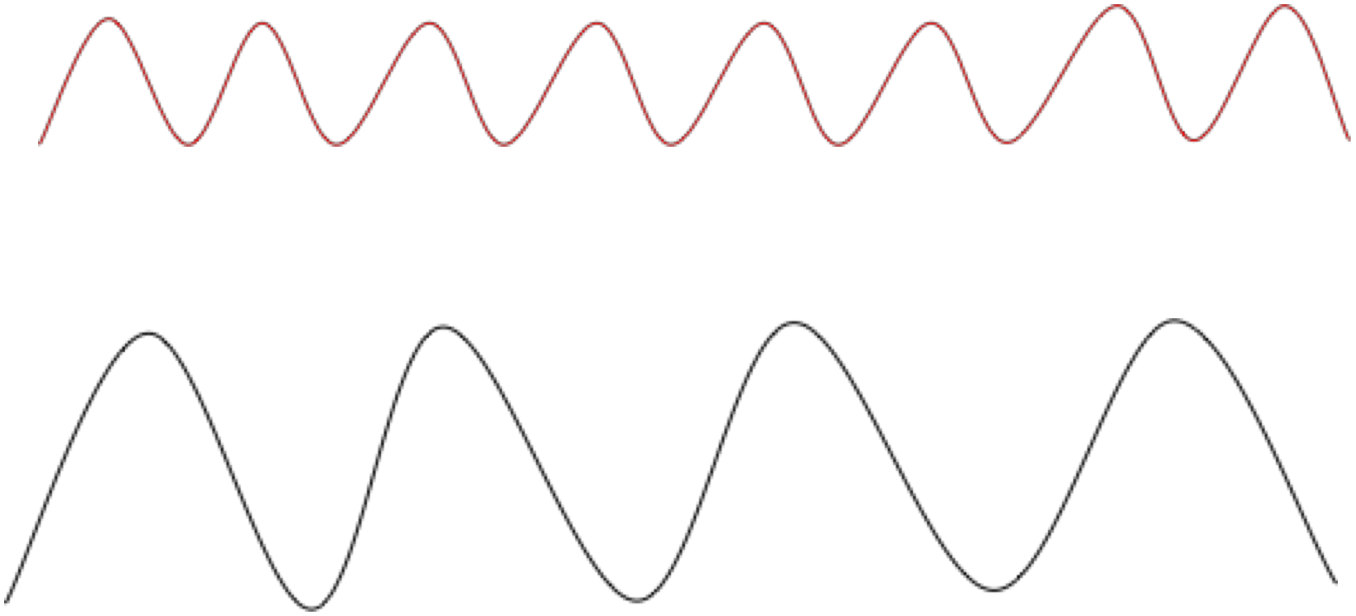
Types of SDB

- 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



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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

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SDB in DMD

- Inevitable in patients with DMD
  - As early as 12 years old in patients on steroids[1]
- 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

[1]Sawnani H et al. *J Pediatr.* 2015

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BiPAP Masks

NASAL MASKS

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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

Source: Google Images, November 12, 2018

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Mouth on

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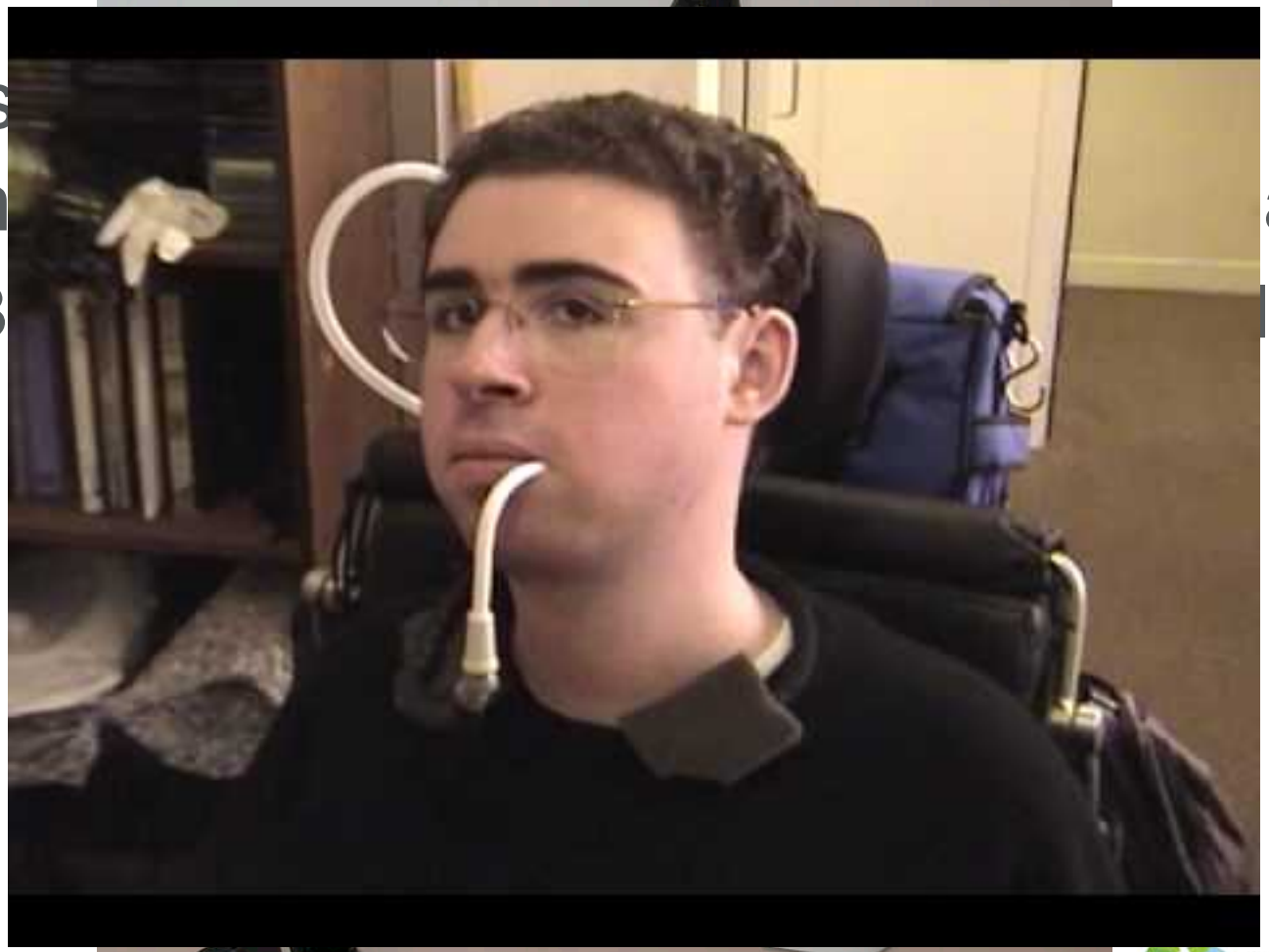

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New DMD guidelines

- 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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