Respiratory Complications of Neuromuscular Disease: Evaluation and Management

Nicholas Hill MD
Tufts Medical Center
Boston MA

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Outline

- Pathophysiology
- Evaluation
- Therapy
  - Noninvasive Ventilation
  - Tracheostomy ventilation
  - Cough Assist
- Outcomes

Respiratory Muscle Involvement: Major Groups

- Inspiratory muscles
  - Diaphragm
  - Accessory muscles
- Expiratory muscles
  - Abdominals
  - Intercostals
- Bulbar muscles
  - Speech swallowing

Key Muscles of the Respiratory System

- Inspiratory muscles
  - diaphragm, intercostal and accessory muscles (neck and shoulder muscles)
- Expiratory muscles
  - abdominal and intercostal muscles
- Bulbar muscles (tongue and upper airway)
  - Responsible for speech and swallowing

Question: Which is least useful to detect bilateral diaphragm paralysis

1)Orthopnea

2)Accessory muscle use

3)Abdominal paradox

4)Sniff fluoroscopy

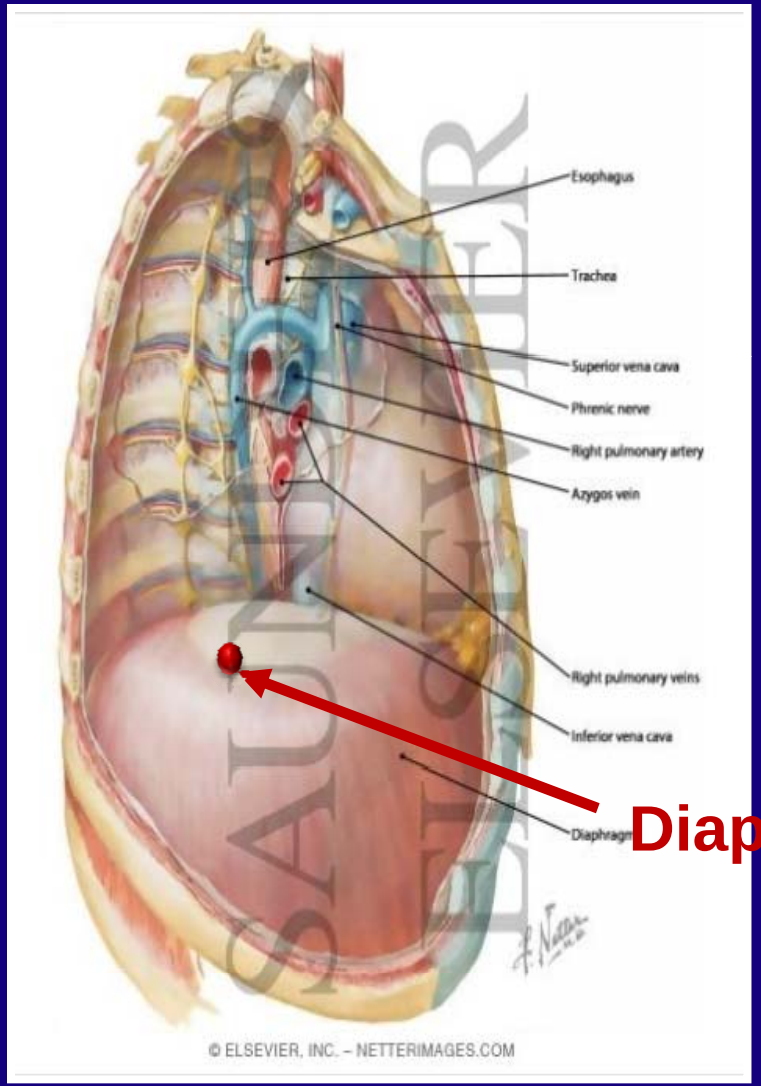
5)Trans diapgragmatic pressure

- Answer: Sniff fluoroscopy

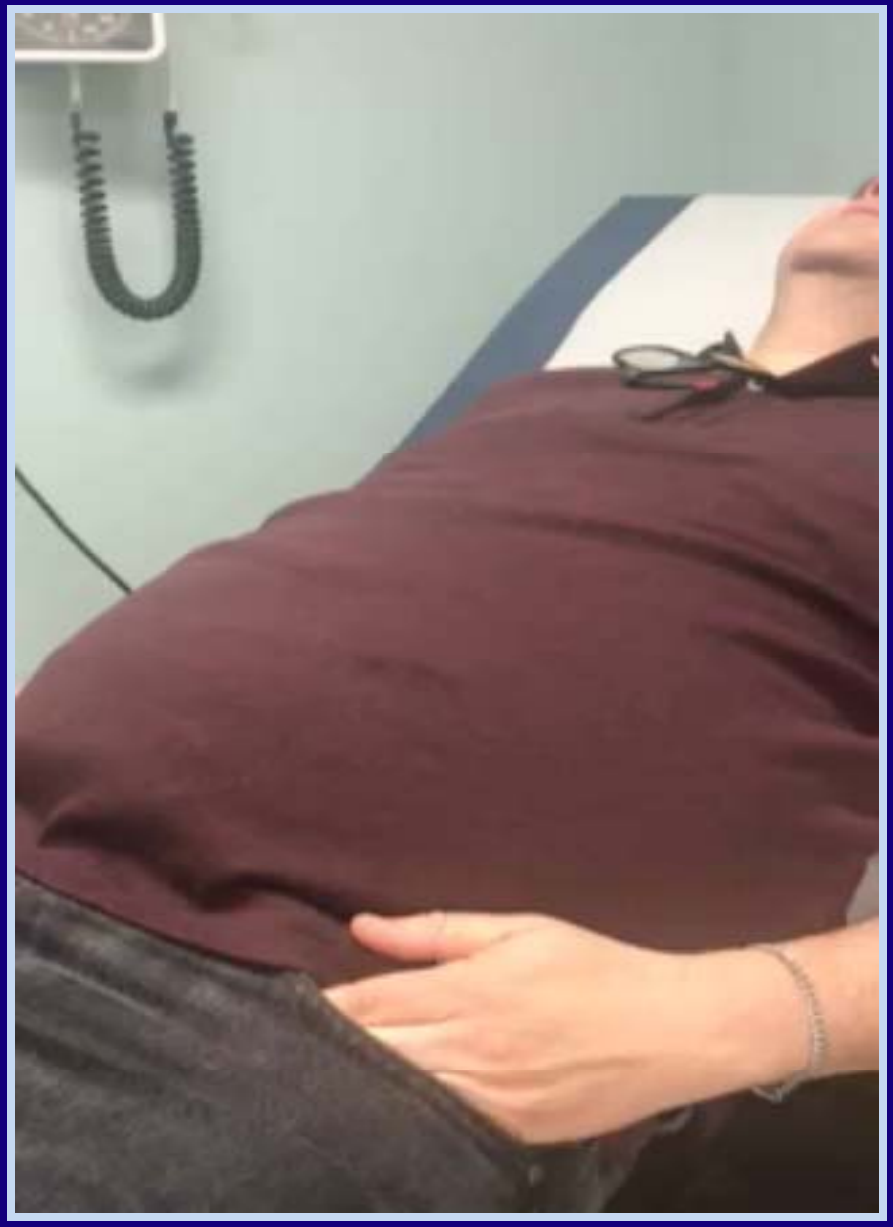
Side view of Diaphragm in Thorax

A weak diaphragm causes:

- **Orthopnea** – Shortness of breath when lying flat
- **Dyspnea on exertion** –labored breathing with increased activity



Abdominal Paradox

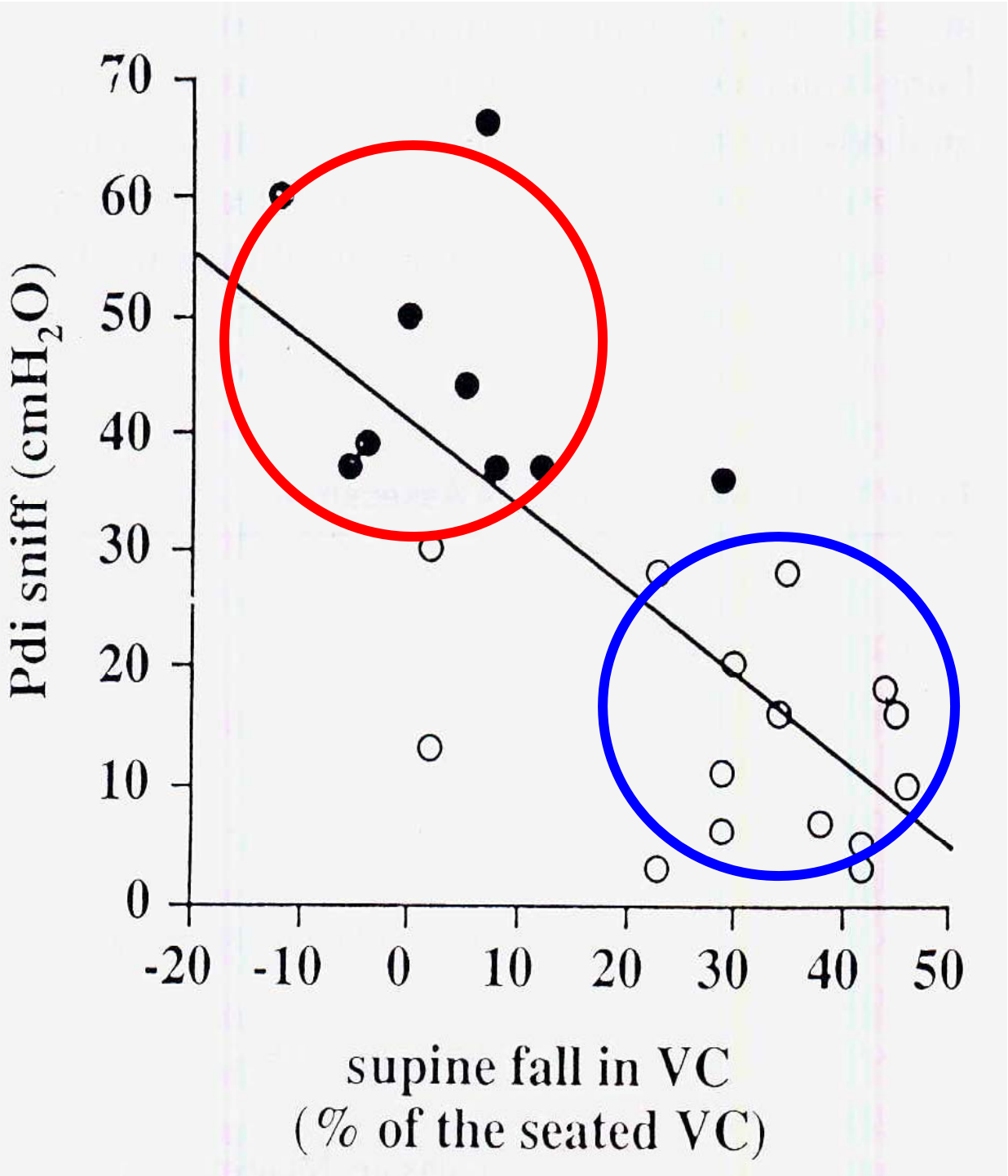


Upright FVC
55% pred

Supine FVC
28% pred

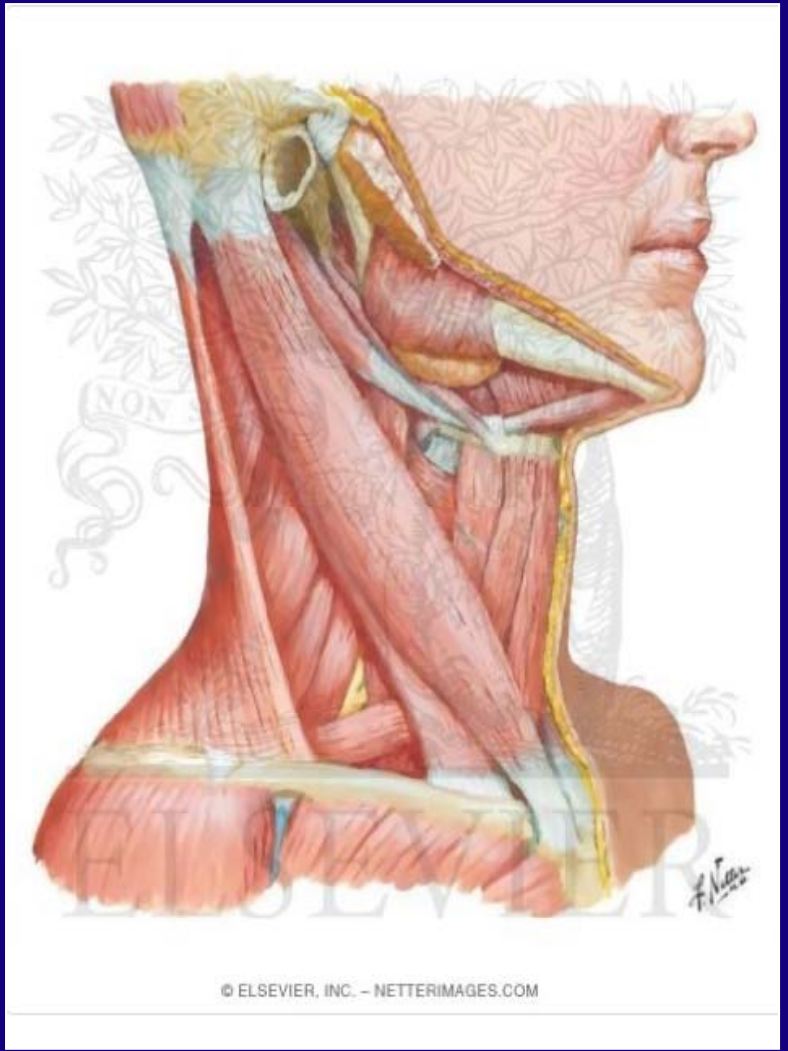
Sniff test more reliable than MIP in NMD

Fromageot et al, Arch Phys Med Rehabil, 2001



Accessory Muscles of Breathing

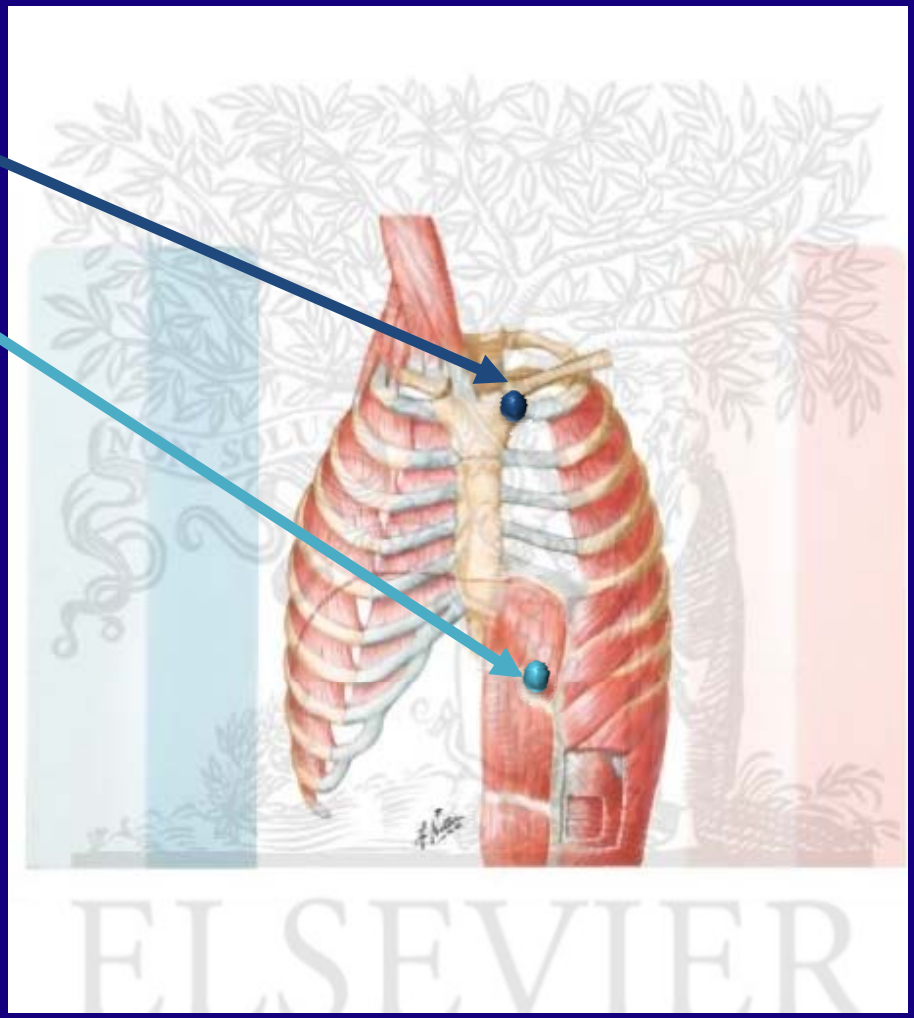
Accessory inspiratory (neck and shoulder) muscles are recruited with a very deep breath. with exercise or tocompensate for a weakened diaphragm



Expiratory Muscles

Intercostals

Abdominals



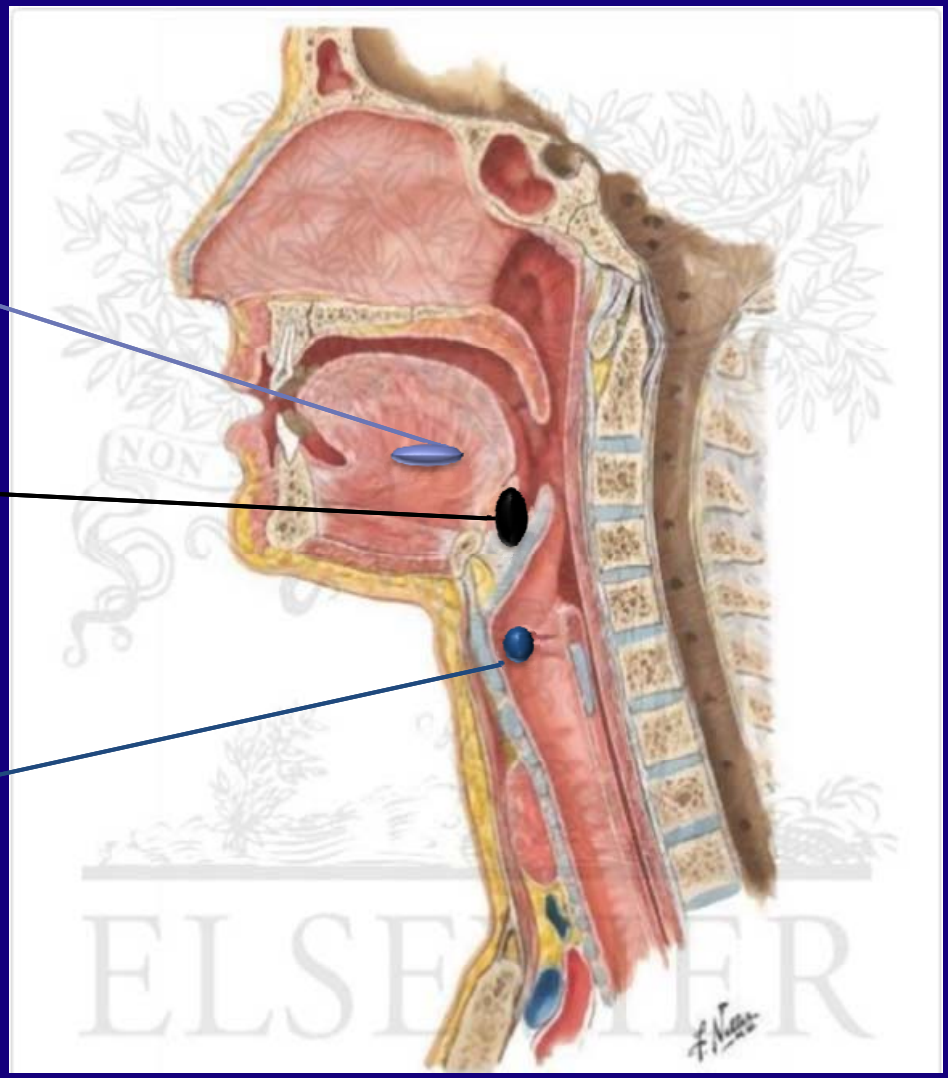
For an effective cough, well functioning inspiratory and bulbar (mouth and throat) muscles are needed in addition to the expiratory muscles

Upper Airway

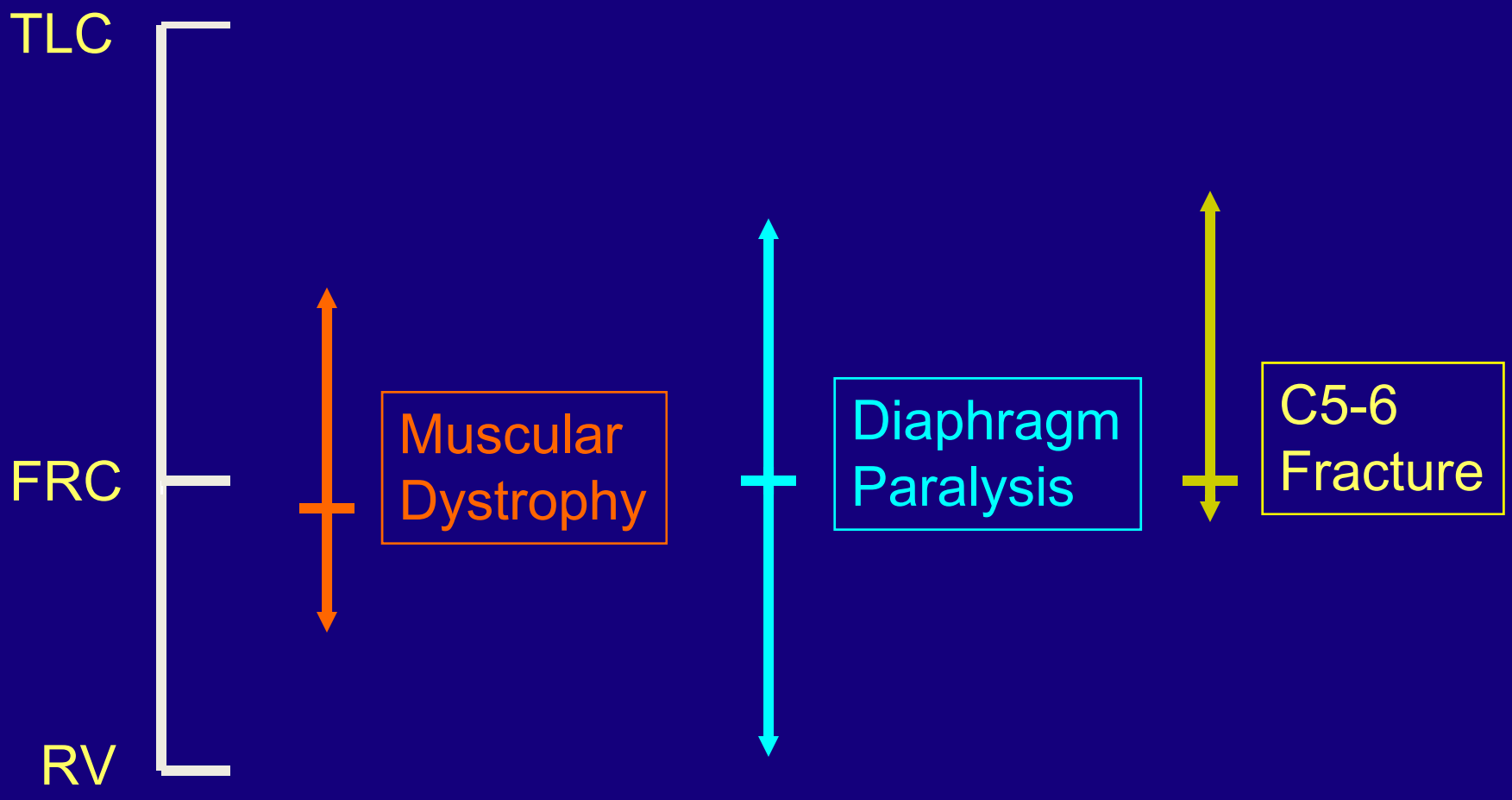
Bulbar Muscles – Used for speech and swallowing

Epiglottis – prevents aspiration

Vocal Cords – vibrate to vocalize; can obstruct airflow



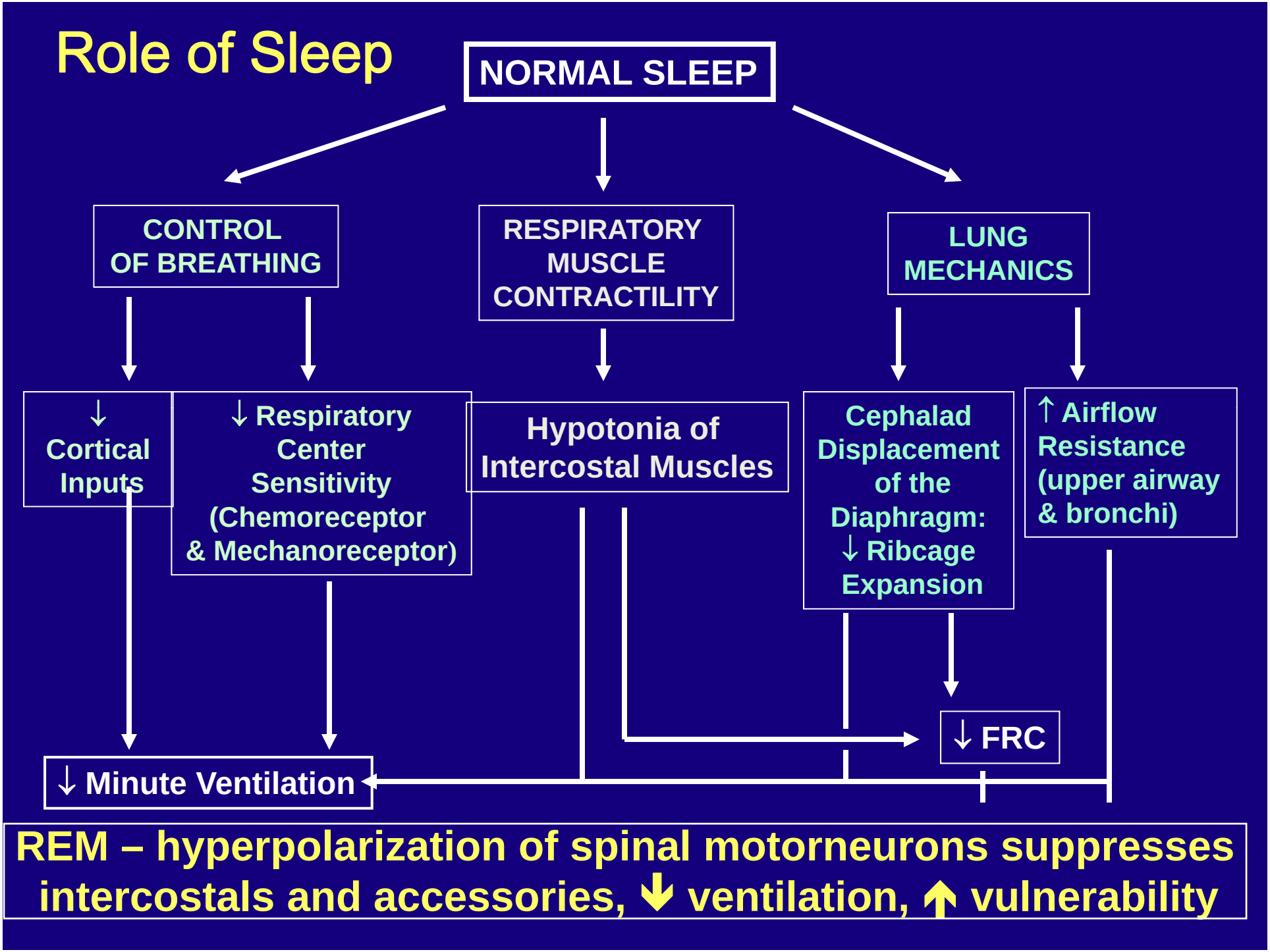
Effects of Resp Muscle Weakness on Pulm Functions



PFTs for NMD

- Quantification and characterizes of functional defect
- Diaphragm involvement: ≥ 25% reduction when supine
- Useful for tracking progression
- BUT:
  - Bulbar involvement – difficulty with lip seal
  - Lack of cooperation in some patients
  - Lack sensitivity – maximal inspiratory and expiratory pressures more sensitive

Role of Sleep



Patterns of SDB in NMD

- Magnification of normal responses
- Upper airway resistance syndrome
- Obstructive sleep hypopnea/apnea
- Central apneas
- Oxygen desaturations
- Global hypoventilation; particularly during REM if diaphragm weak

SDB in NMD

- Occurs in an estimated 42% of pts
- Risk factors:
  - Pulmonary dysfunction
  - Kyphoscoliosis (>120°)
  - Increased body mass index
  - Macroglossia (Duchenne MD)
  - Bulbar muscle involvement (ALS)

Lebanowski et al, Neurology, ’96; 47:1173

Neuromuscular Diseases for NIV: Commoner conditions (Adults)

- Amyotrophic Lateral Sclerosis (Motor Neuron Disease)
- Muscular Dystrophies
  - Duchenne, Limb Girdle, Fasciosculohumeral
  - Spinal Muscular Atrophies (SMA)
  - Myotonic Dystrophy
- Neuropathies
  - Charcot Marie Tooth
  - Nemaline
- Post-polio Syndrome
- Spinal cord injuries
- Pompe's Disease – glycogen storage

NMD with Respiratory Insufficiency: Presenting Features (Subtle)

- Symptoms
  - Morning headaches
  - Daytime Hypersomnolence
  - Fatigue
  - Dyspnea
  - Orthopnea
- Signs
  - Tachypnea
  - Accessory muscle use
  - Diaphragm dysfunction/paradox
  - Chest wall paradox
- Labs
  - Oximetry, bicarb, VBGs, ABGs, $PtcCO_2$

CMS Guidelines: NPPV for NMD

- Symptoms
  - Morning HA, Daytime hypersomnolence
- PFTs
  - FVC < 50% pred, MIP > -60 cm H2O
- Gas exchange
  - Daytime PaCO2 > 45 mm Hg, O2sat 88% or less for > 5 min nocturnally

Case Presentation

48 yo with hx of polio (iron lung use) at age 2. Quadriparetic, kyphoscoliosis. Uses wheel chair. Prior several mos, had increasing snoring, fatigue, am headaches and hypersomnolence

On exam, no resp distress, P 70s, RR 20, BP 130/70, Overwt (BMI 34) Mallampati 4, + access musc use, lungs clear, cor nl, no abd paradox, back severely scoliotic, edema Neuro: quadriparetic

Post-polio Syndrome

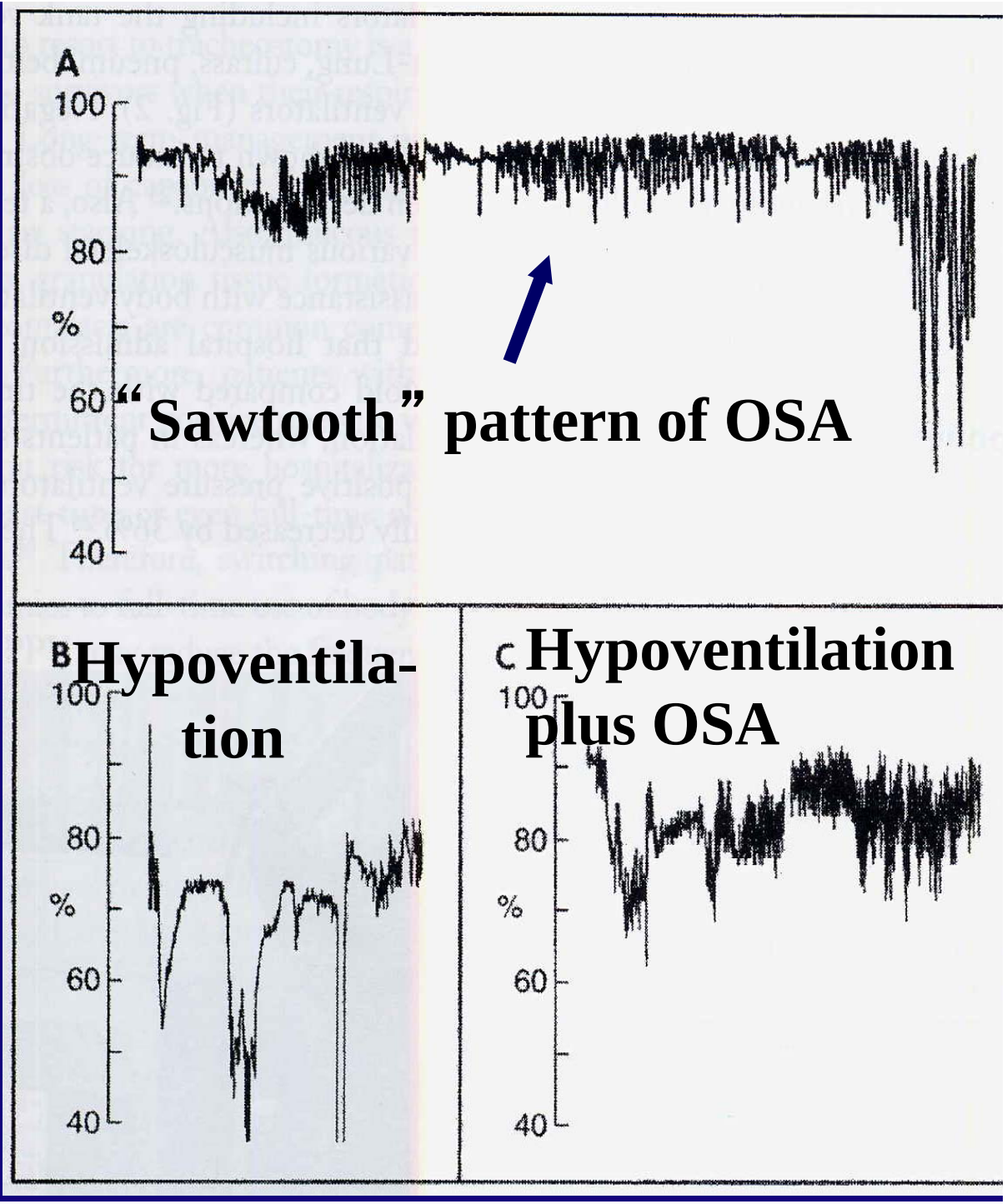
- Progressive weakness of muscles associated with chronic pain and fatigue, often decades after acute illness
- Sleep-disordered breathing very common:
  - Obstructive events associated with obesity
  - Central apneas with bulbar involvement
  - ↓ventilation with respiratory muscle weakness and scoliosis
- Very slow progression

Evaluation

- CBC hct 48
- CXR severe scoliosis
- PFTs: FVC 0.97 (34%), FEV1 0.79 (34%)
- ABG (RA) 7.35/PaCO2 72/PaO2 58 mm Hg
- Cardiac echo: RV dilatation,
  - est PA 45 mm Hg

Polysomnogram: arousal index 14, AHI 34, severe sustained desaturation (70-89%)
Dose Titration: IPAP 11, EPAP 3, rate 15
After 2 mos, PaCO2 67, Sx no better

Oximetry Patterns of Sleep-disordered Breathing in Post-Polio Syndrome



Question: What went wrong with our patient?

1) EPAP too low
2) Backup rate too low
3) IPAP too high
4) IPAP – EPAP too low

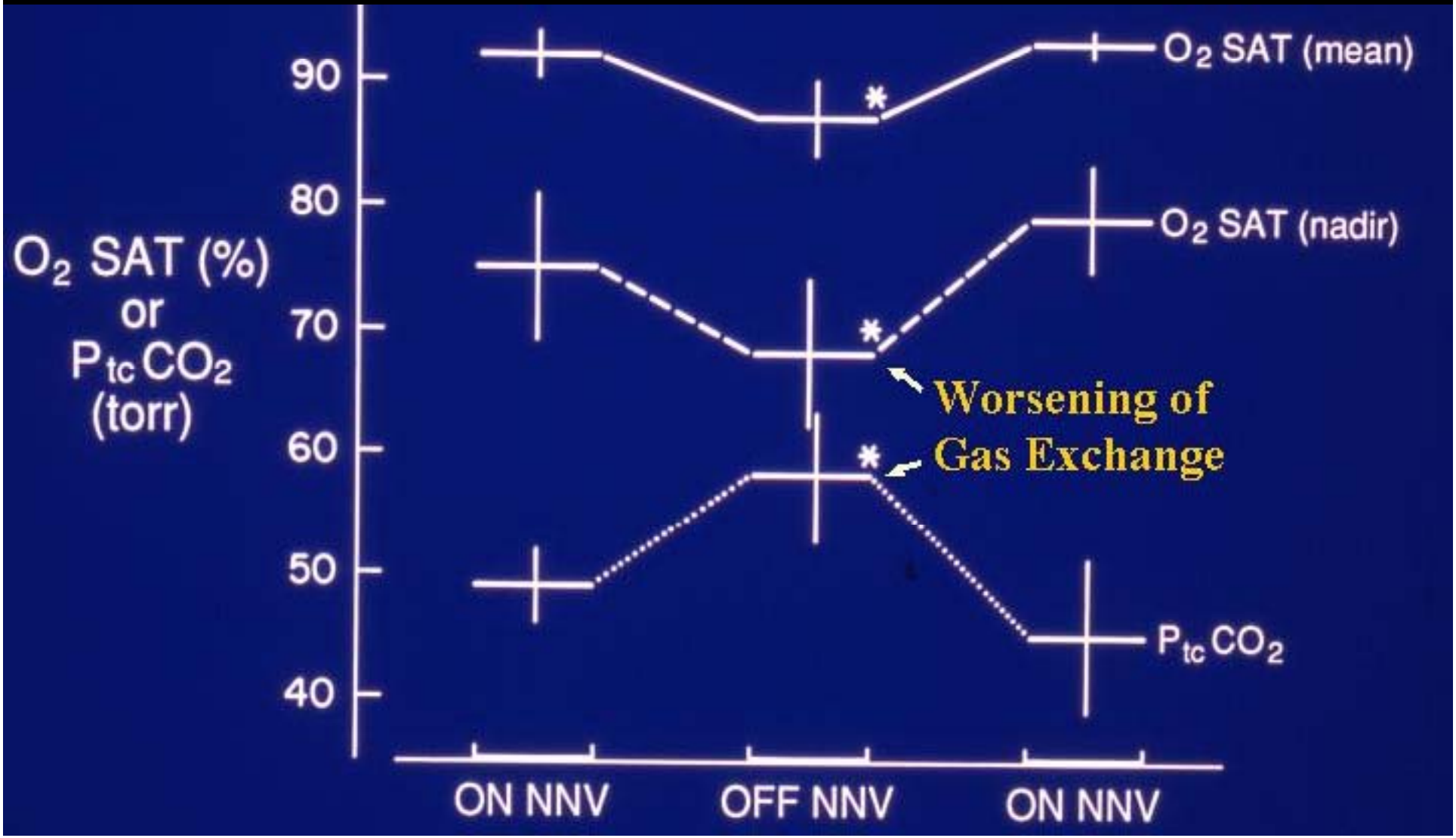
Answer #4

Is a Polysomnogram necessary?

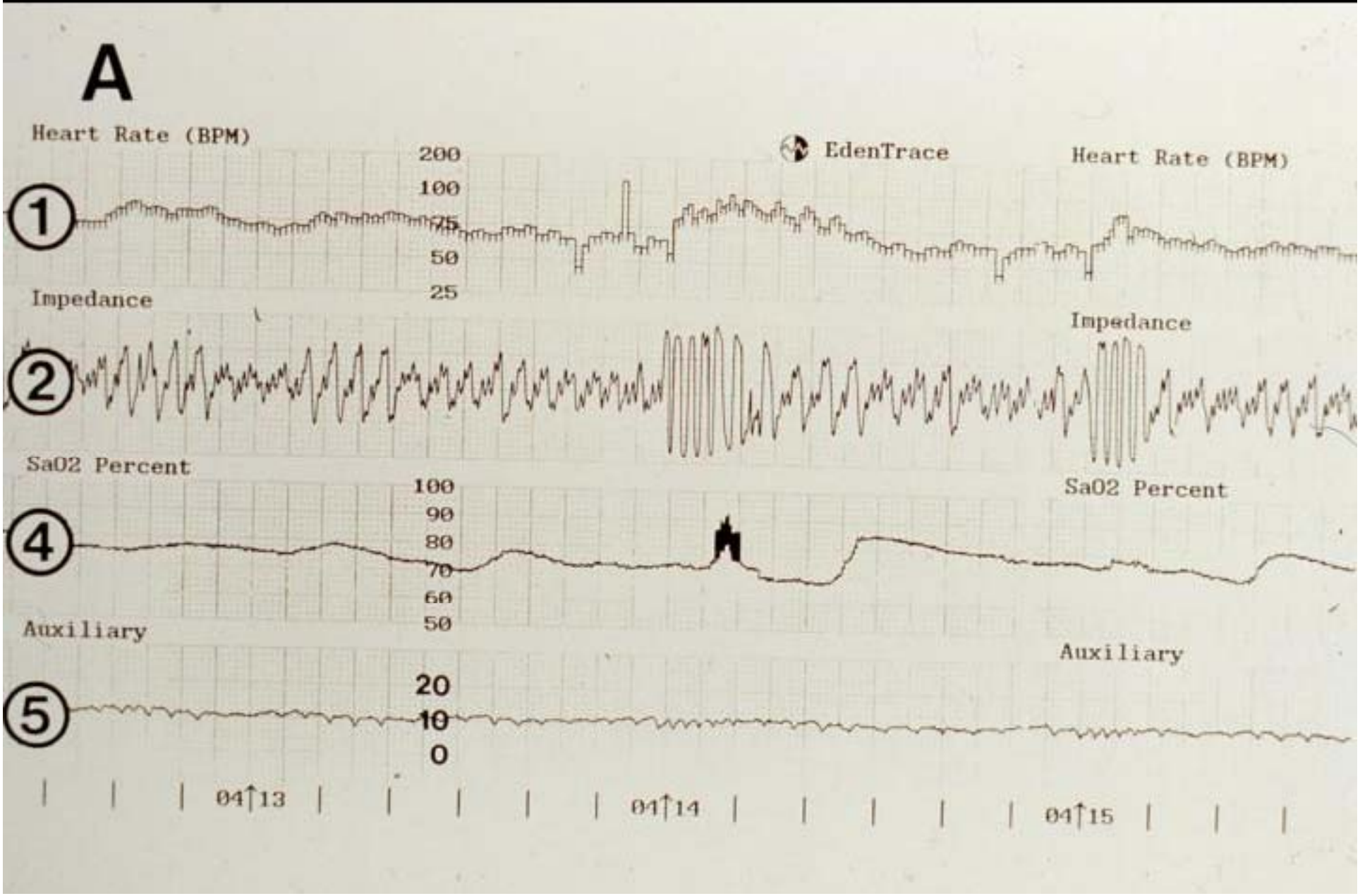
- Yes – if OSA likely (symptoms, but PFTs less than severe (FEV1 > 1-1.5L))
  - unexplained symptoms or CO2 retention
  - Risk factors (obesity, scoliosis)
  - To titrate pressures or assess suboptimal response
- Not necessarily – If daytime hypercapnia or severe pulmonary dysfunction – if many physical limitations, may be challenging

HOW DOES NIV WORK FOR NMD?

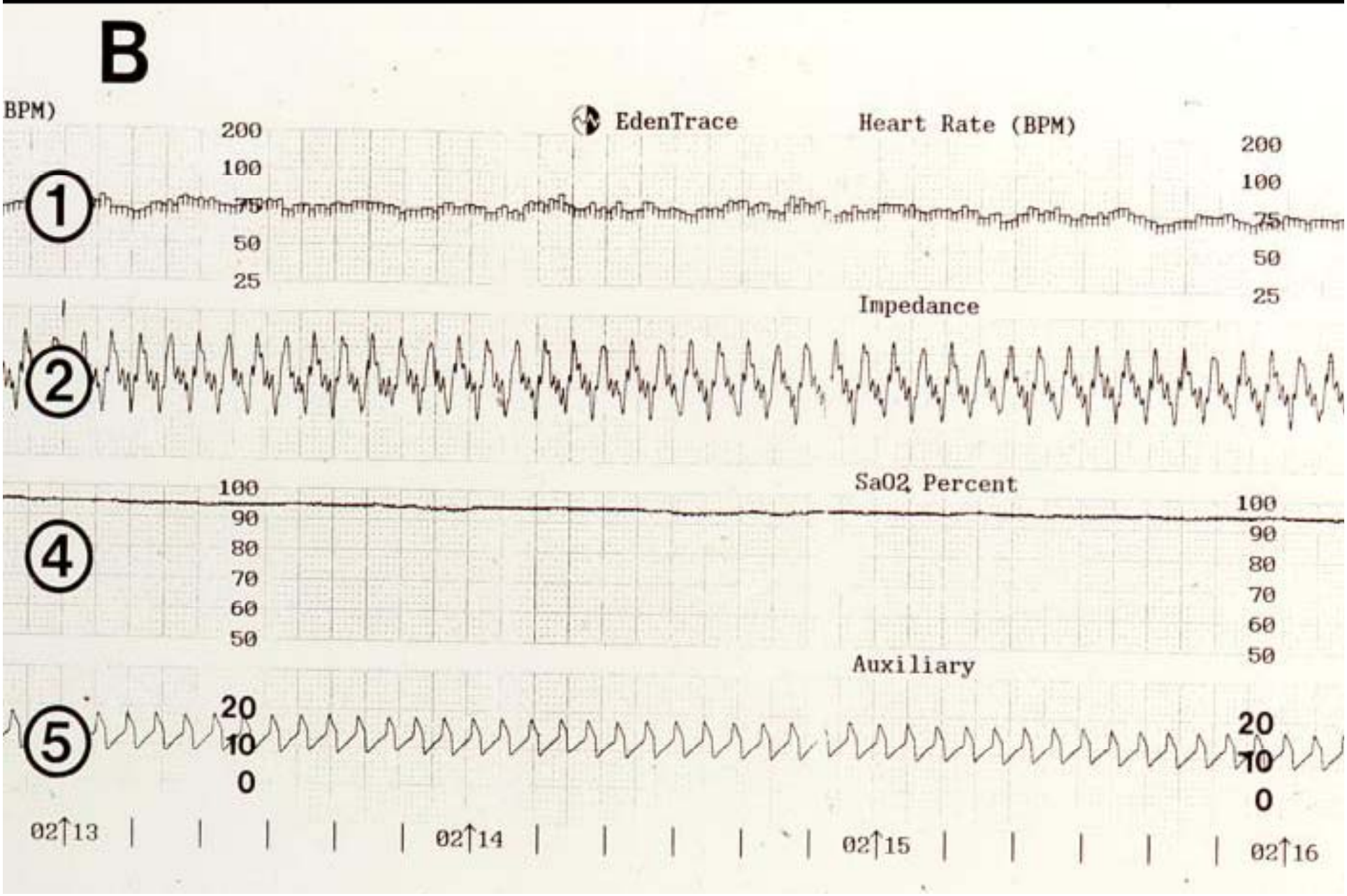
Effect of 1 week NIV Withdrawal (Hill et al, ARRD ’ 91)



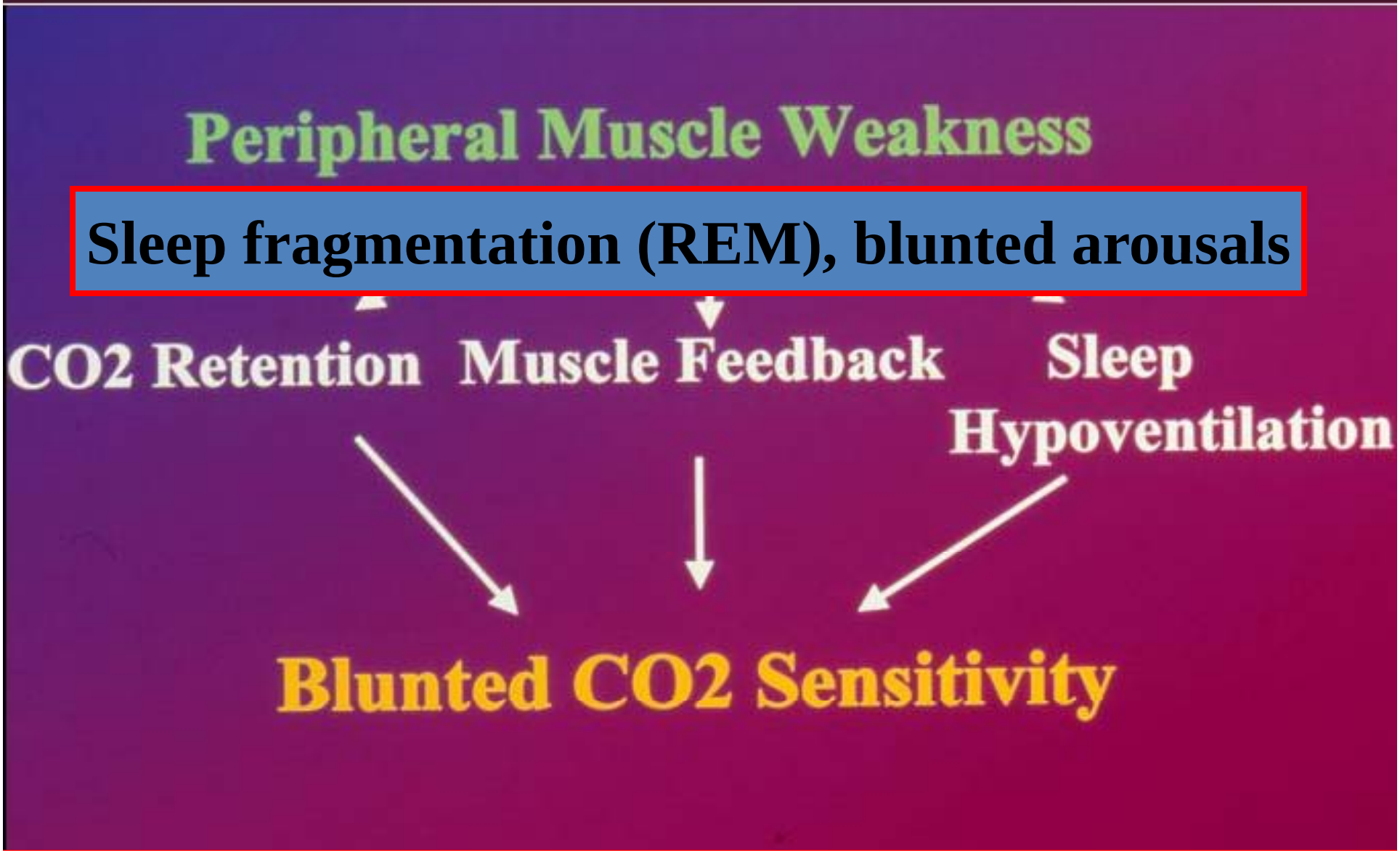
Sleep Monitoring before NPPV



Sleep Monitoring after NPPV

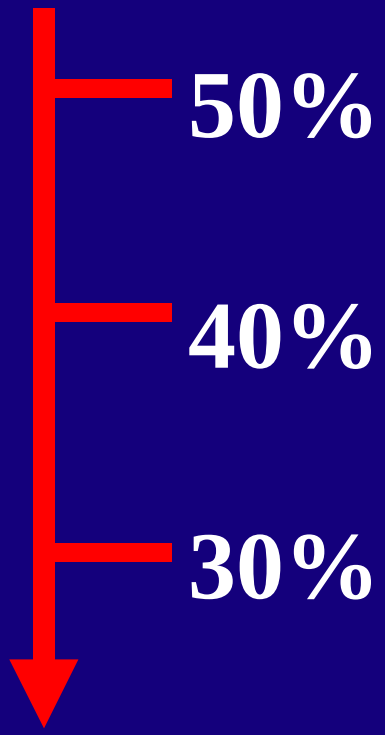


Central Fatigue



WHEN TO START NPPV?

Pulmonary Function (vital capacity)



Sleep disordered breathing

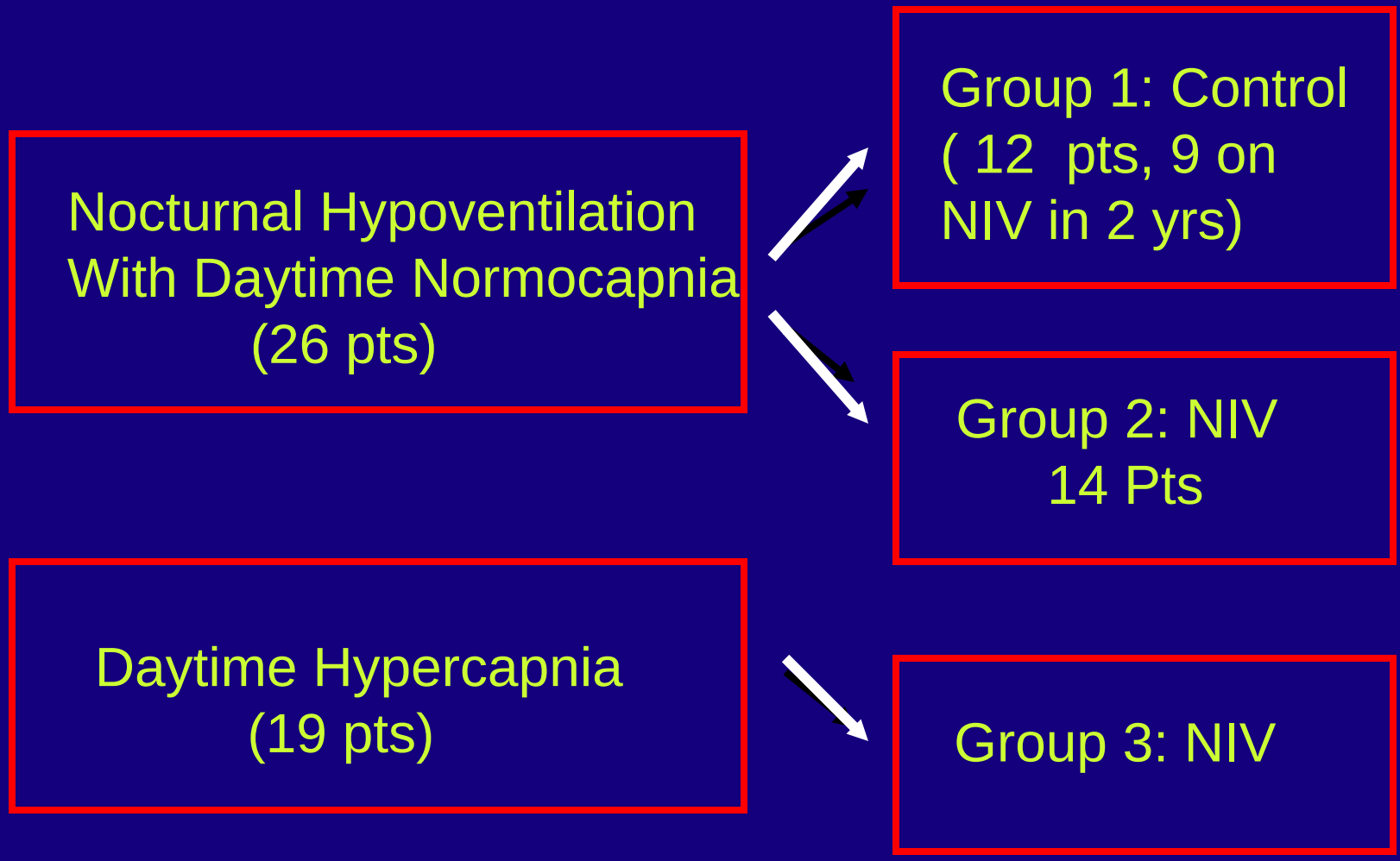
Symptoms

Nocturnal hypoventilation

Diurnal hypercarbia

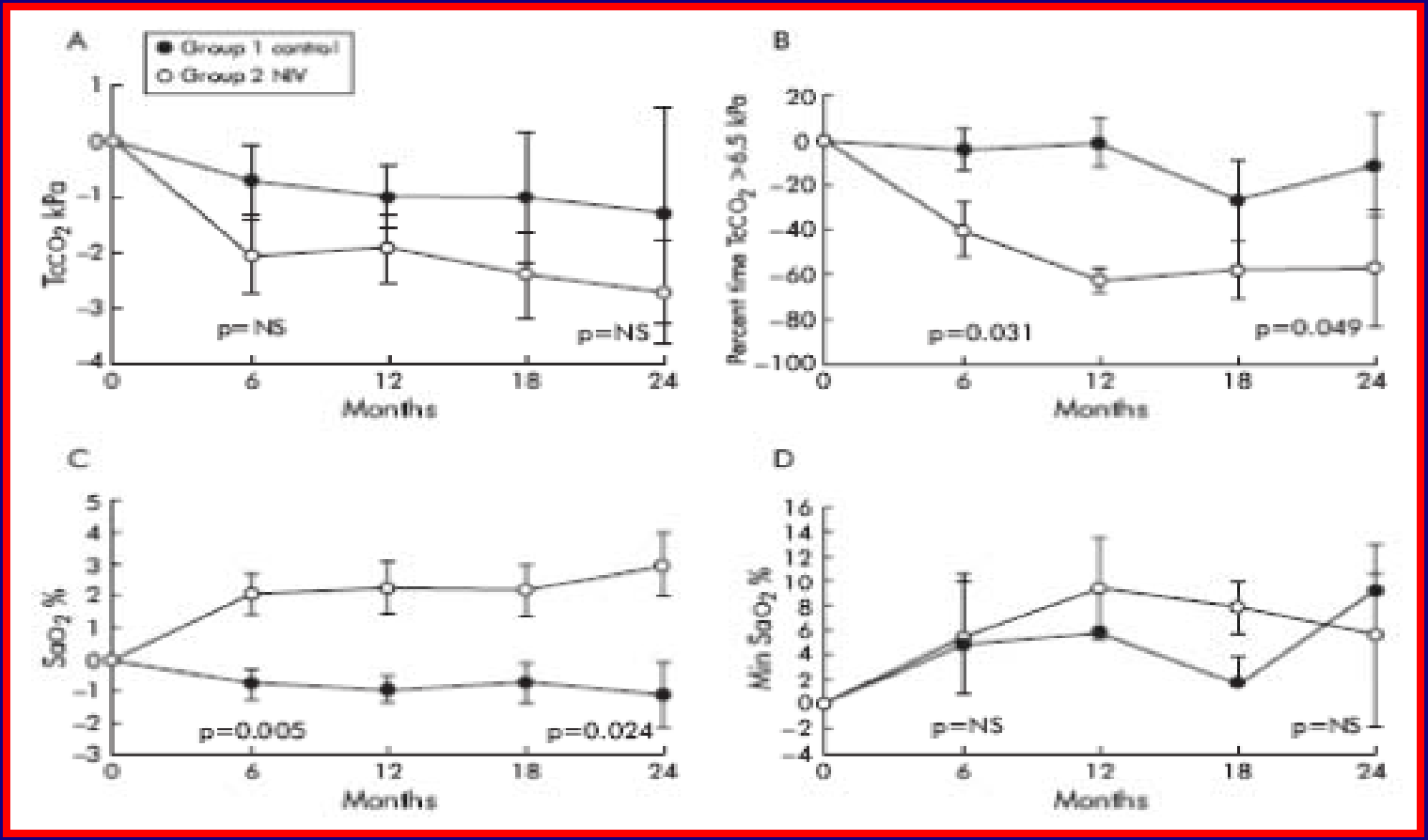
Respiratory crisis

When to Start NIV in NMD? RCT



Ward et al, Thorax 2005; 60:1019

When to Start NIV in NMD?



4.65 hrs use/night

Overall SF-36 better in Group 2

Reduce frequency of acute hypercapnic crises (trend)

Ward et al, Thorax 2005; 60:1019

How to start NIV? What settings?

- Empiric approach
  - Start low 8-10 cm H2O insp, 3-4 cm H2O exp
  - Gradually increase insp pr (IPAP)
  - Follow symptoms, daytime ABGs, noct O2 sats
  - Hospitalization confers no advantage for start
- Sleep lab titration
  - Increase lower pressure until resp events abolished, upper until ↑ arousals
  - Our pt, 11/3 was best, O2 sat 91% with 2L/min O2

Question: Which is best predictor of NIV Success in ALS?

1) Intact bulbar function
2) Orthopnea
3) High $PaCO_2$
4) All of the above

Answer D

Effect of NIV on Quality of Life in ALS

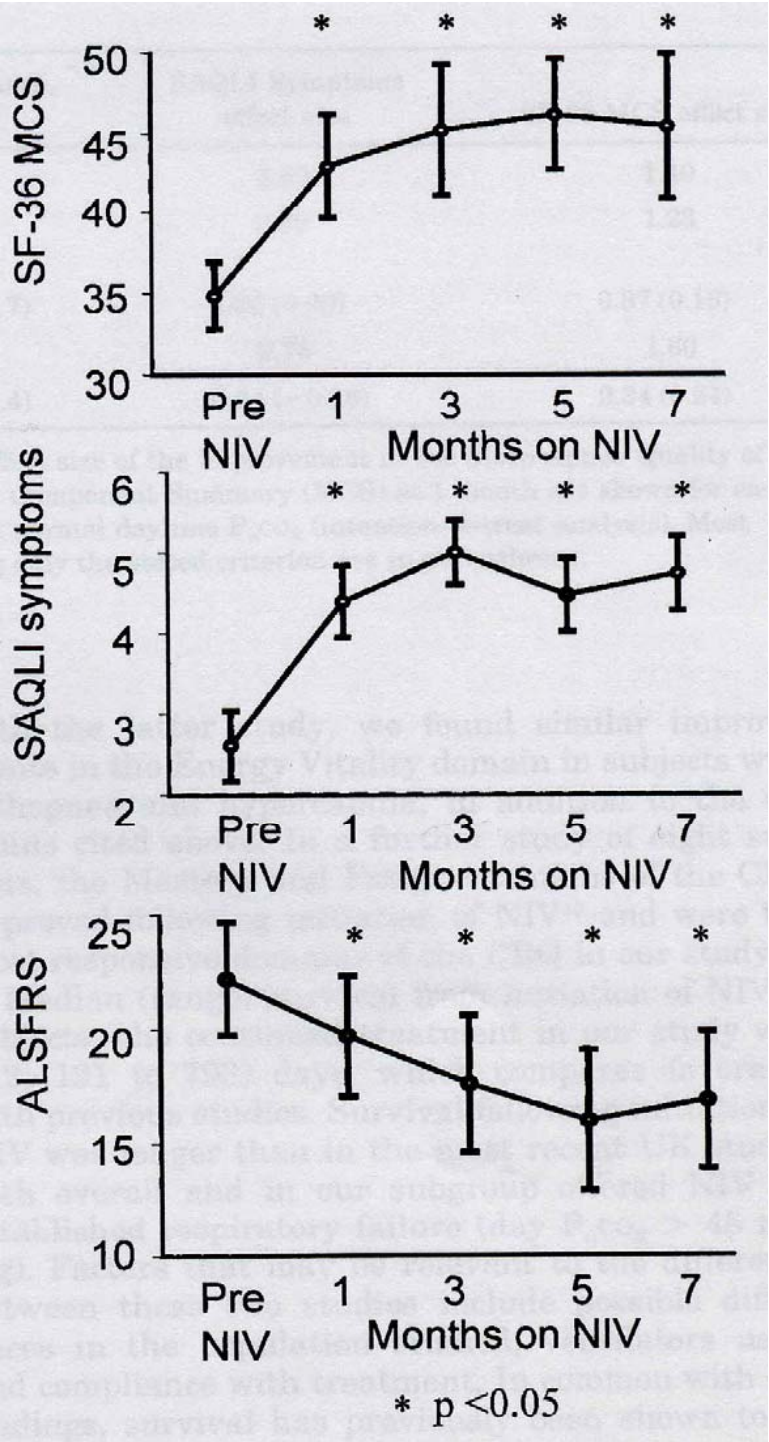
22 pts started on NIV when:

1) Orthopnea
2) Daytime sleepiness, AHI>10
3) PaCO2>45 mm Hg
4) MIP<80% predicted

Best predictors of benefit:

1) Orthopnea
2) Intact bulbar function
3) ↑ PaCO2, O2 desat

Bourke et al. Neurology 2003;61:171

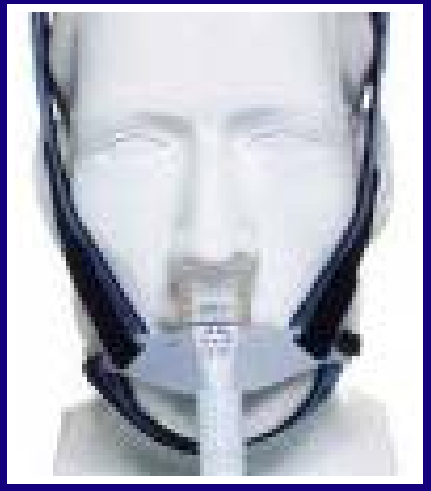


RCT for NIV in ALS

- 41 pts with orthopnea/ FVC < 60% or ↑ PaCO2
- 205 day prolongation of survival with ↑ QOL
- In bulbar, not survival but QOL↑, sleep-related symptoms improved
  - Bourke SC, *Lancet Neurol* 2006;5:140-7
- Predictors
  - survival: NIV tol, BMI, Bulbar
  - NIV tol: Orthopnea- yes, bulbar - no

Bourke SC et al. Lancet Neurol 2006; 5: 140-147

Masks for NIV




It's 5 o'clock somewhere
MAINE
It's 5 o'clock somewhere
MAINE

Ventilators for NIV

Assisting Cough in Neuromuscular Disease

- Secretion retention is a major problem in advanced disease
- Salivary inhibition, oral hygeine
- Chest physiotherapy
- Manually assisted or quad coughing
- The “Vest” – chest oscillator
- Cough inexsufflator (Cough Assist™)

Cough Assist – T70

- Delivers deep insufflations (+30-40 X 2 sec) followed immediately by deep exsufflations (-30-40).
- Simulates the physiologic mechanism of cough.

Why Long-term Noninvasive vs Invasive Ventilation?

- Less airway trauma
- Airway defense intact
- Simplifies management
- Lower cost; but

- Less secure, sleep quality less
- Necessitates ability to protect airway

Tracheostomy Ventilation

- Indicated for patients unable to protect their airway/have excessive secretions
- Upper airway obstruction
- Unable to manage NIV for extensive periods (>16hrs/day)
- But greatly increases the complexity of care
- Adds to caregiver burden
- More complications, pneumonias
- May necessitate admission to chronic care facility

Summary: Vent Insuff in Neuromusc Dz

- Progressive resp insufficiency is major cause of mortality in NMD
- Pattern of resp muscle involvement determines specific manifestations
- NIV is mainstay of rx, started after symptoms but before daytime hypercarbia
- Successful initiation requires patience, skill, comfortable and appropriate technology

Summary: Vent insuff in Neuromusc Dz

- Important to assist cough when impaired
- Discuss possible eventual need for invasive ventilation early and avoid unanticipated respiratory crises