The Respiratory Muscles

- Inspiratory
- Expiratory
- Bulbar-innervated

- The inspiratory and expiratory muscle aids
COPD/Sleep disordered breathing

- PFTs – diffusion, ABGs, plethysmography, forced expiratory volumes/ polysomnography
- Bronchodilators and oxygen therapy/CPAP or low span BiPAP
MOST COMMON ERRORS
MISS INTERPRETATION OF SYMPTOMS
INADEQUATE PFTs
FAILURE TO MONITOR SLEEP
OVER RELIANCE ON ABGs
OVER RELIANCE ON TRACHEOSTOMY
OVER RELIANCE ON SUCTIONING
OXYGEN THERAPY
SYMPTOMS OF CHRONIC HYPOXIA

Dyspnea
Cognitive changes
Anxiety
Personality changes
Sequelae of hypertension
Symptoms of Chronic Alveolar Hypoventilation

frequent hospital admissions for SOB, atelectasis, pneumonia
O2, CPAP treatments without benefit
morning headaches, fatigue, SOB
daytime drowsiness (hypersomniaolence)
frequent awakening at night, esp. with
SOB, tachycardia, frequency, enuresis
difficulty falling asleep and waking in AM
nightmares (especially respiratory)
loss of concentration, memory
fall off in grades, intellect, injuries
anxiety, personality changes
weight change, difficulty with secretions
hx. of CHF, hypertension, nausea
Pulmonary Function and Aging

Normal: VC 30cc/yr after 19
FEV1 30cc/yr after 19 (1-1.2%)
MVV 0.8%/yr after 30
PaO2 = 109 - 0.43 (age)

Loss of Volumes doubled in COPD
Variable in neuromuscular disease
Fig 2—Deterioration of vital capacity as a function of age.

Peak cough flow (PCF) and %VC in 40 patients with DMD or SMA
Treatment Goals

• Optimize chest wall/lung ROM and growth
• Optimize cough flows
• Maintain normal ventilation
Optimize Cough Flows and Airway Secretion Elimination
Mucus plugging is the primary cause of pneumonia, lung collapse, failure to wean, obstructive emphysema.(1) It simulates pulmonary embolism.(2)

Up to 90% of mortality in Duchenne muscular dystrophy and episodes of respiratory failure are triggered by chest colds

Fortunately for Sparky, Zeke knew the famous "Rex maneuver."
Maintain normal alveolar ventilation around the clock
ALVEOLAR HYPOVENTILATION
COMPENSATED BY
PATIENT EFFORT
Pour Miguel de Colía
mars 2009 Sparta
Dibujo de George Cruikshank (1792-1878)
IDEAL INTERFACE

• LEAK FREE

• COMFORTABLE

• MAINTENANCE FREE
R.E. 9/85 Sleep Study
post polio, MIPPY, O₂ added (2LPM)
PLY-100, angulated mouthpiece, single elastic strap

CO₂ 5.4%  
Pt. awake, supine
left side lying
3:29 AM

96-89%
Outpatient Protocol

• Maintain Spo2 > 94% at all times, especially during colds
• How? By using Mechanically assisted coughing (MAC) and noninvasive ventilation
• If Spo2 < 95%, you have either hypoventilation, mucus, or pneumonia
PROTOCOL – Dr. Bach

1. Oxygen administration limited only to approach 95% SpO\textsubscript{2}.
2. Mechanical insufflation-exsufflation used via the tube at 25 to 40 cm H\textsubscript{2}O to -25 to -40 cm H\textsubscript{2}O pressures up to every 10 minutes as needed to reverse oxyhemoglobin desaturations due to airway mucus accumulation and when there is auscultatory evidence of secretion accumulation. Abdominal thrusts are applied during exsufflation. Tube and upper airway are suctioned following use of expiratory aids as needed.
3. Expiratory aids used when desaturations occur.
4. Ventilator weaning attempted without permitting hypercapnia.
5. Extubation attempted whether or not the patient is ventilator weaned when meeting the following:
   A. Afebrile
   B. No supplemental oxygen requirement to maintain SpO\textsubscript{2} >94%
   C. Chest radiograph abnormalities cleared or clearing
   D. Any respiratory depressants discontinued
   E. Airway suctioning required less than 1-2x/eight hours
   F. Coryza diminished sufficiently so that suctioning of the nasal orifices is required less than once every 6 hours (important to facilitate use of nasal prongs/mask for post-extubation nasal ventilation)
6. Extubation to continuous nasal ventilation and no supplemental oxygen.
7. Oximetry feedback used to guide the use of expiratory aids, postural drainage, and chest physical therapy to reverse any desaturations due to airway mucus accumulation.
8. With CO\textsubscript{2} retention or ventilator synchronization difficulties nasal interface leaks were eliminated, pressure support and ventilator rate increased or the patient switched from BiPAP-ST\textsuperscript{™} to using a volume cycled ventilator. Persistent oxyhemoglobin desaturation despite eucapnia and aggressive use of expiratory aids indicated impending respiratory distress and need to re-intubate.
9. Following re-intubation the protocol was used for a second trial of extubation to nasal ventilation ...or following successful extubation bronchodilators and chest physical therapy were discontinued and the patient weaned to nocturnal nasal ventilation.
10. Discharge home after the SpO\textsubscript{2} remained within normal limits for 2 days and when assisted coughing was needed less than 4 times per day.


Indications for Tracheostomy

When the Spo2 decreases below 95% and can not be normalized by NIV or MAC
Indications for Decanulation

Noninvasive ventilation can be used when assisted peak cough flows can exceed 160 L/m$^1$, $^2$

Management of Patients with Neuromuscular Disease

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