













## 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

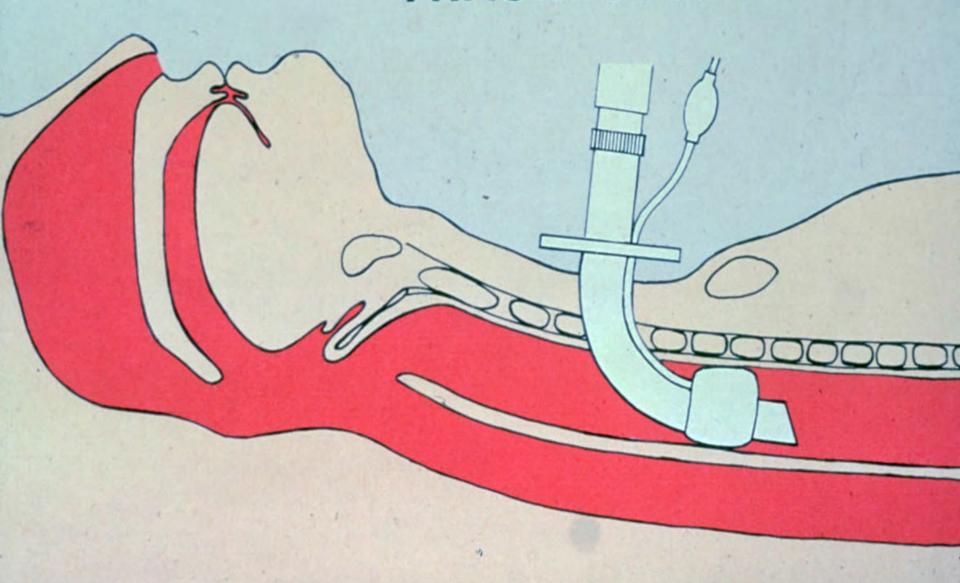
### SYMPTOMS OF CHRONIC HYPOXIA

Dyspnea
Cognitive changes
Anxiety
Personality changes
Sequelae of hypertension

#### Symptoms of Chronic Alveolar Hypoventilation

frequent hospital admissions for SOB, atelectasis, pneumonia 02, CPAP treatments without benefit morning headaches, fatigue, SOB daytime drowsiness (hypersomnolence) frequent awakening at night, esp. with SOB, tachycardia, frequency, enuresis difficulty falling asleep and awaking 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

## TRACHEOSTOMY



# Pulmonary Function and Aging Normal: VC 30cc/yr after 19 FEV1 30cc/yr after 19 (1-1.2%) MVV 0.8%/yr after 30 Pa02 = 109 - 0.43 (age)

Loss of Volumes doubled in COPD Variable in neuromuscular disease

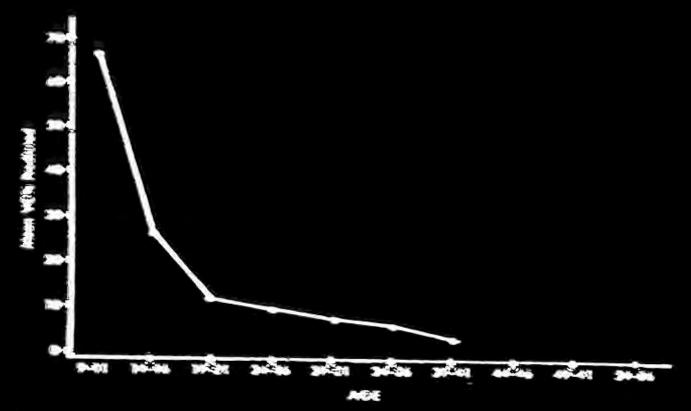
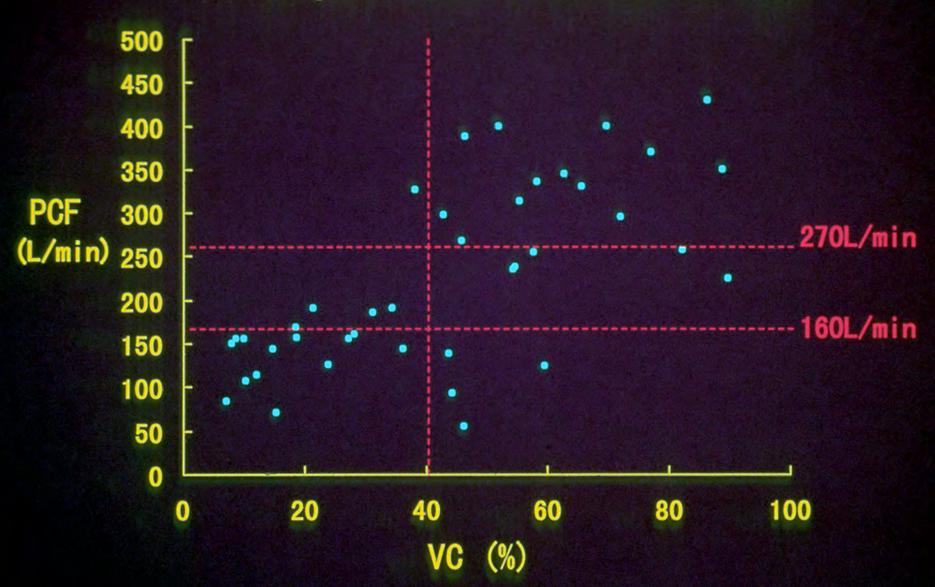
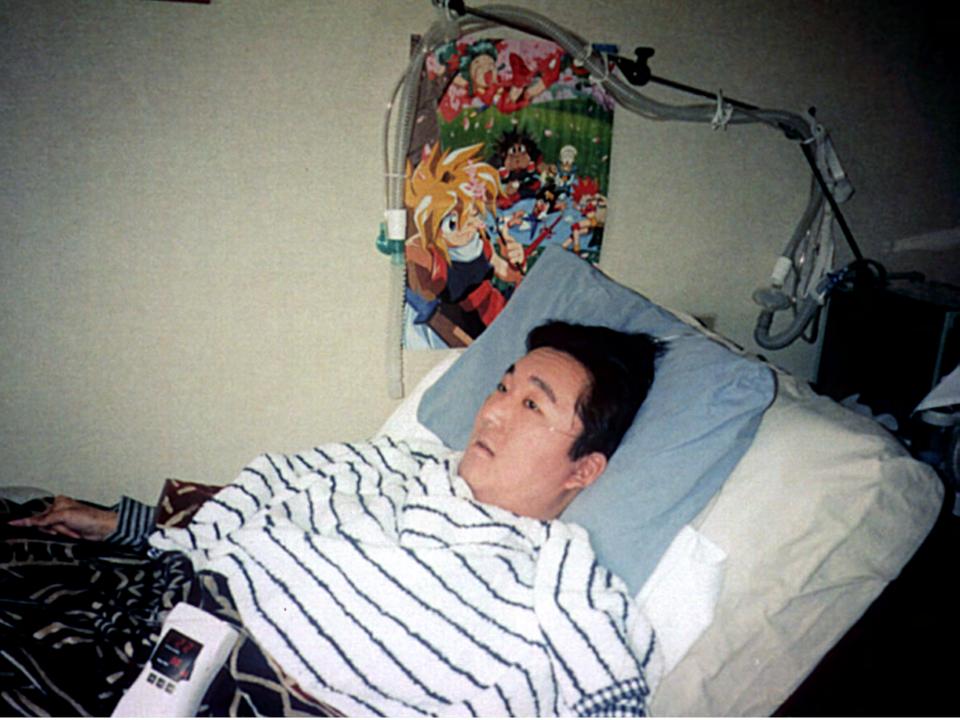


Fig 2-Deterioration of vital capacity as a function of age.

Bach J, Alba A, Pilkington LA, Lee M, Longterm Rehab. In Advanced Staged Childhood Onset, Rapidly Pro. Mus. Dys, Arch Phys Med Rehabil Vol 62, July 1981

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









#### Treatment Goals

•Optimize chest wall/lung ROM and growth

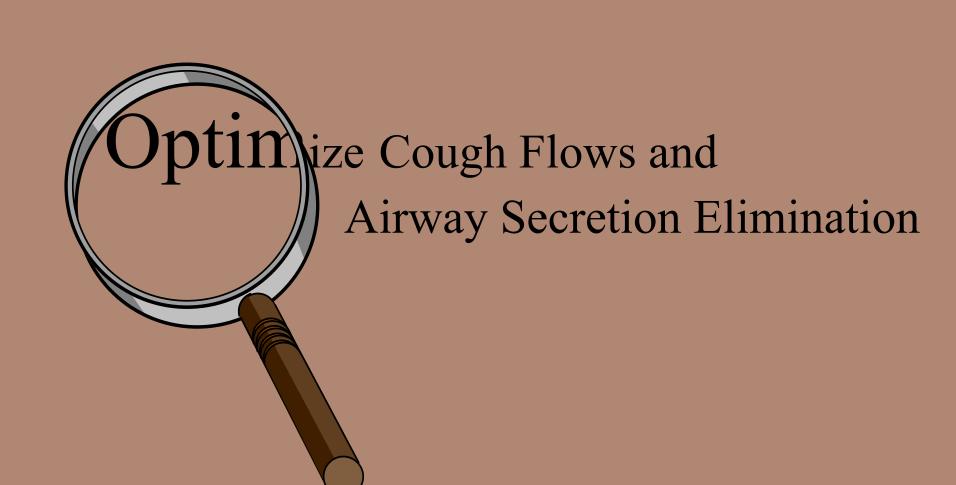
Optimize cough flows

Maintain normal ventilation



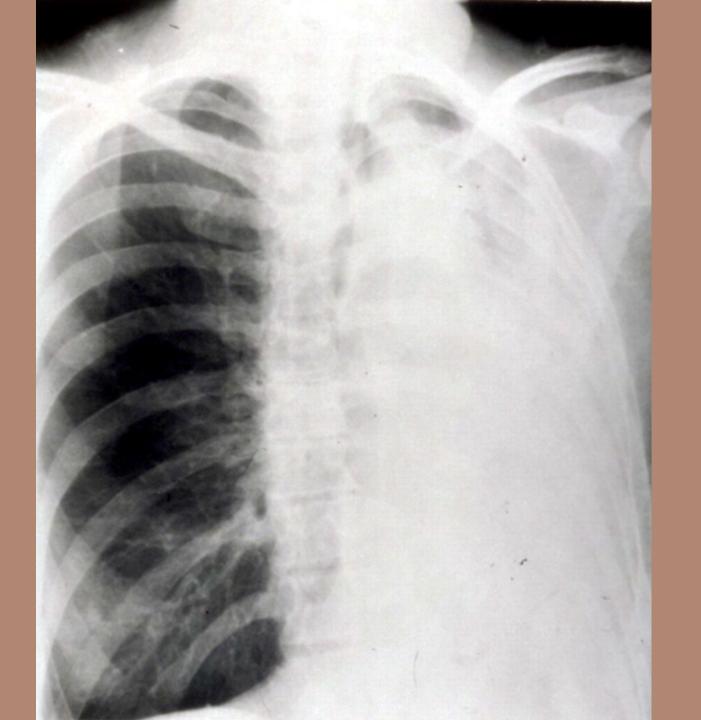






Mucus plugging is the primary cause of pneumonia, lung collapse, failure to wean, obstructive emphysema.(1)
It simulates pulmonary embolism.(2)

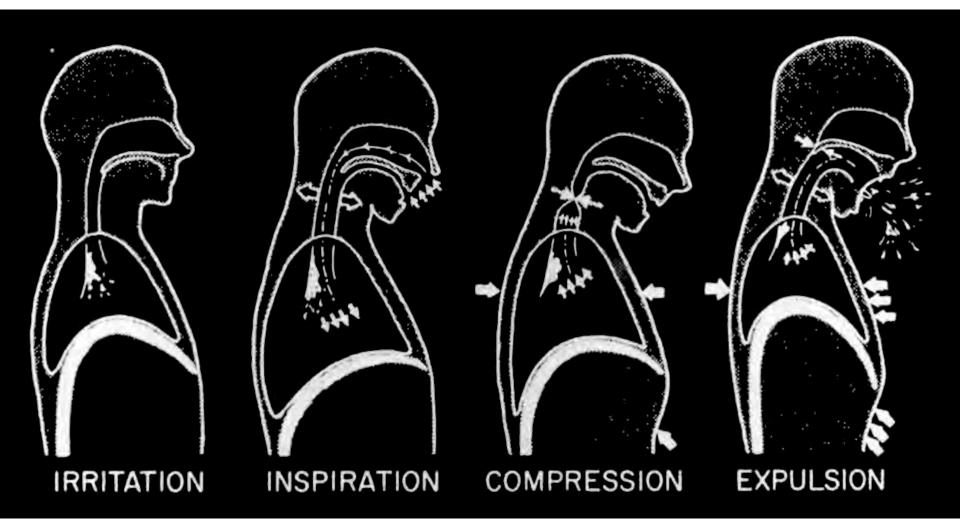
Cohn et al. Arch Phys Med Rehabil 1987:68. Dee et al. Chest 1984:85.

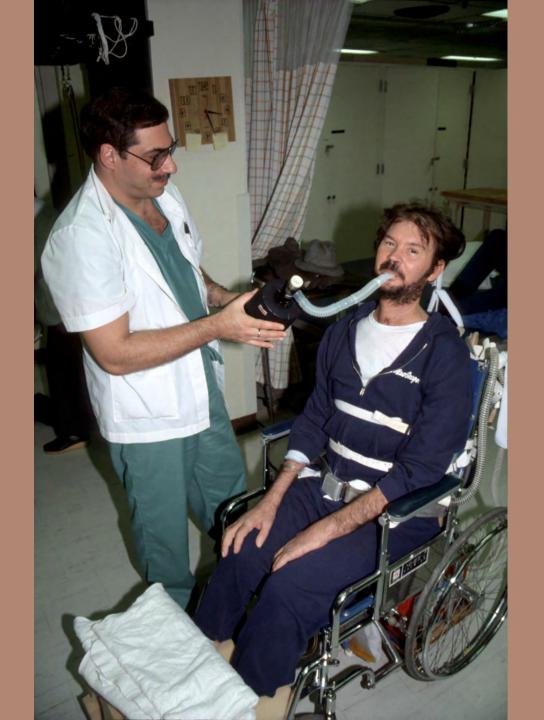


Up to 90% of mortality in Duchenne muscular dystrophy and episodes of respiratory failure are triggered by chest colds

Bach JR et. Neuromuscular ventilatory insufficiency: the effect of home mechanical ventilation vs. oxygen therapy on pneumonia and hospitalization rates. Am J Phys Med

Rehabil 1998;77:8-19.





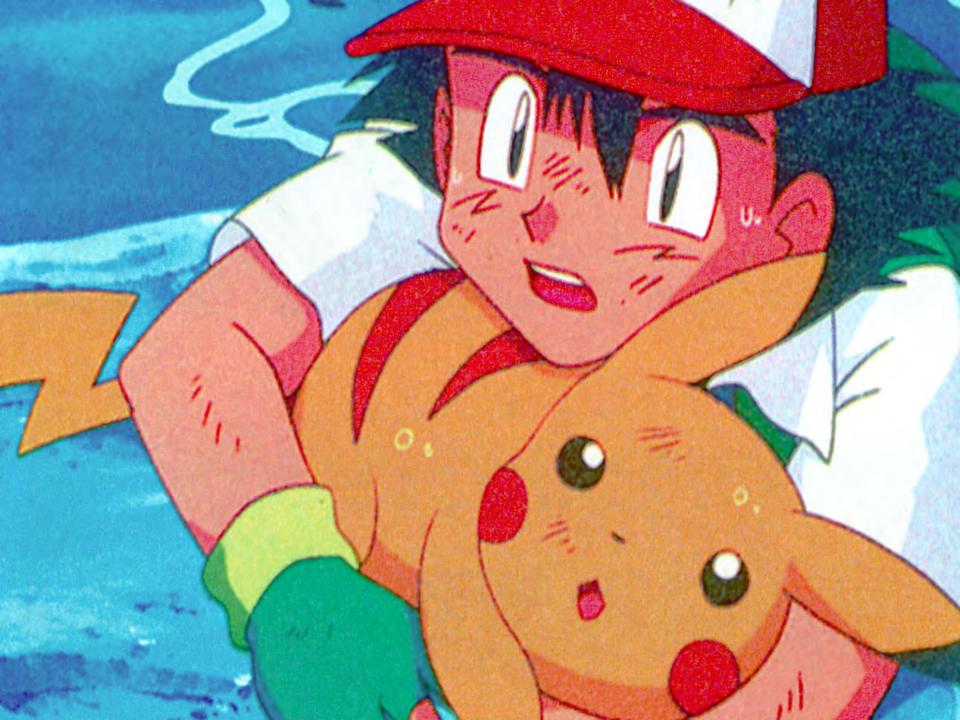






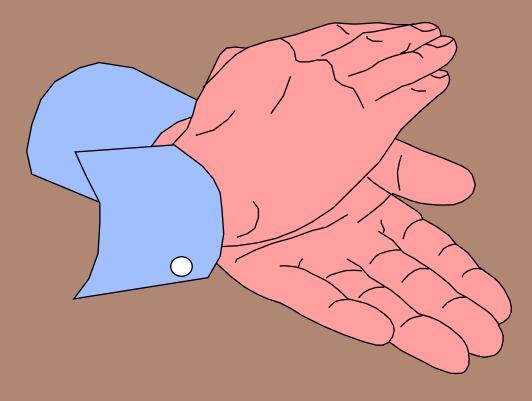








Fortunately for Sparky, Zeke knew the famous "Rex maneuver."

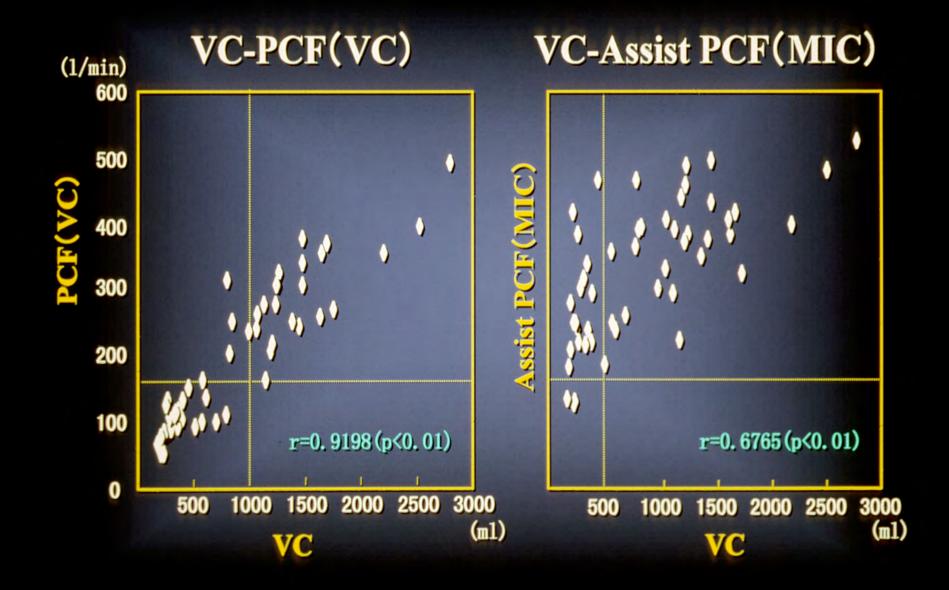


**BRAVO**!





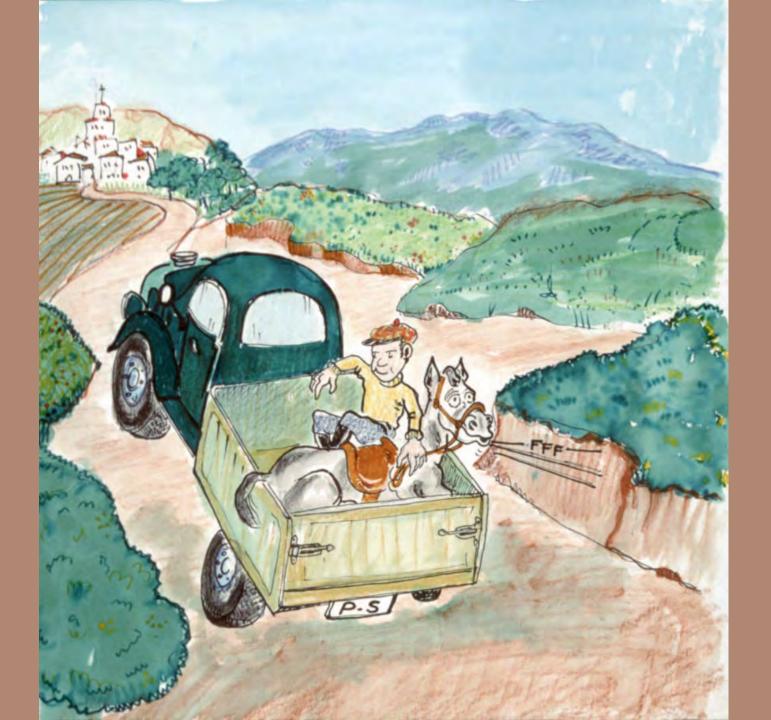




## Maintain normal alveolar ventilation around the clock



















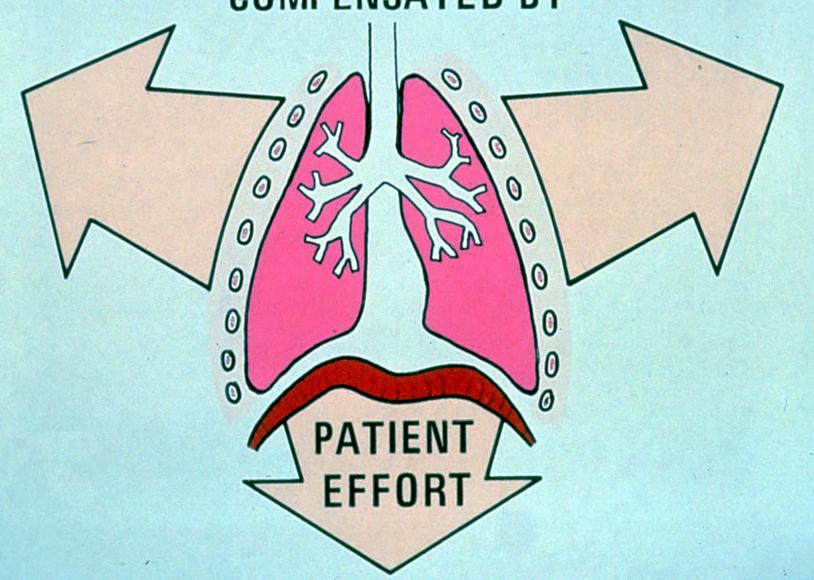


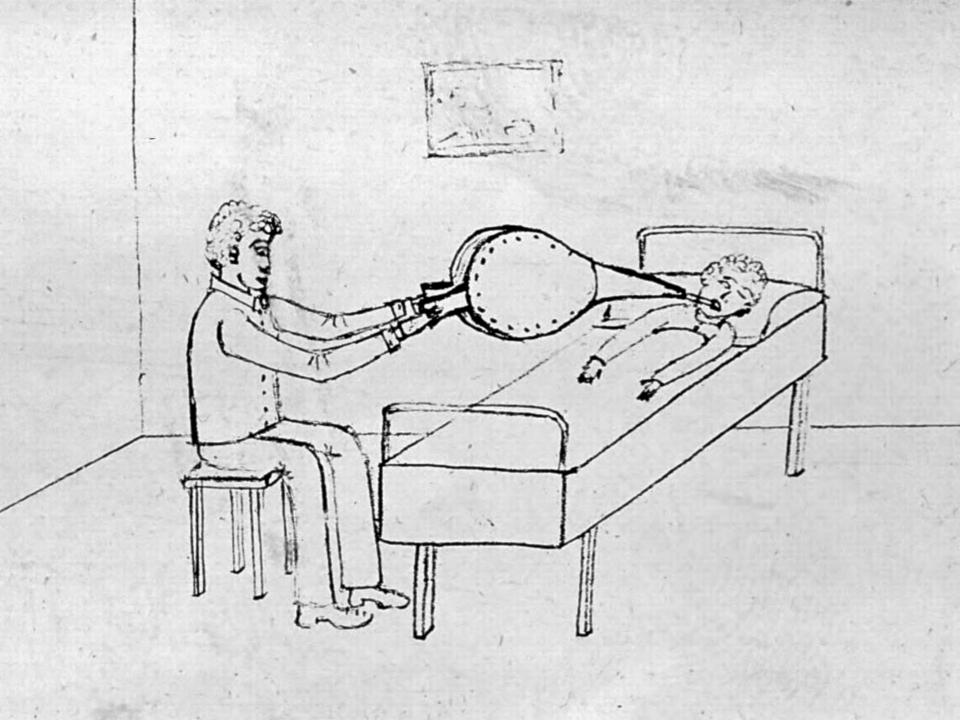




## ALVEOLAR HYPOVENTILATION

**COMPENSATED BY** 







Pour miguel de Colion mars 2009 Sparta

























## IDEAL INTERFACE

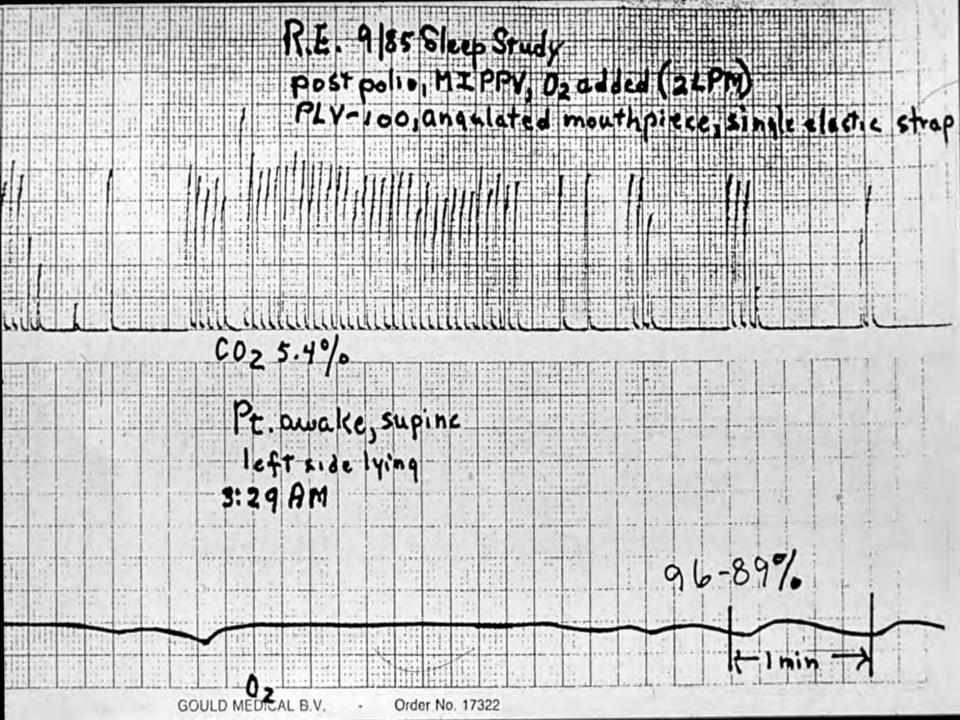
LEAK FREE

COMFORTABLE

MAINTENANCE FREE









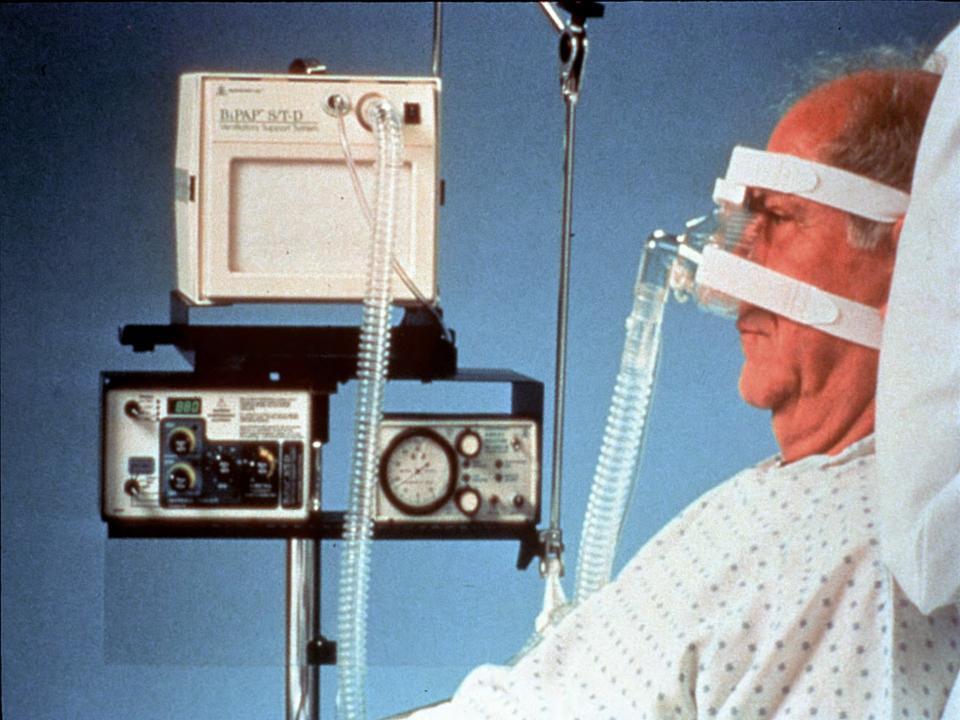


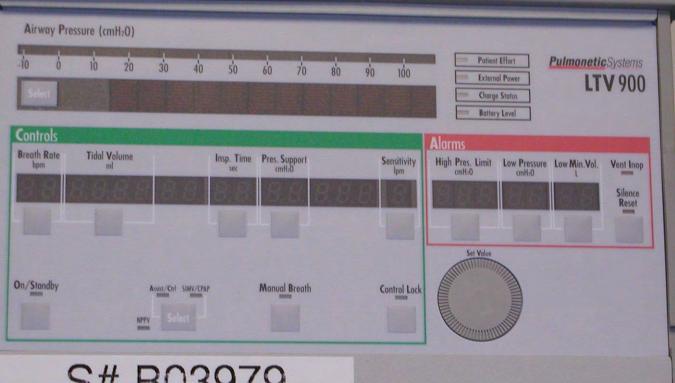












#### S# B03979



Millennium Respi		1-800-269-9		
Volume Ventilato	r Prescription			
Patient:			Date:	
Mode:	Tidal Vol.:			
FIO <sub>2</sub> :	Flow Rate:		RR: Insp. Time:	
Sensitivity:	High Press.:		Low Press.:	
Hours of Use:		Interface:		
Humidifier:				
Comments:				Initials



### **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<sub>2</sub>.
- 2. Mechanical insufflation-exsufflation used via the tube at 25 to 40 cm  $\rm H_2O$  to -25 to -40 cm  $\rm H_2O$  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 SpO2 >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  $CO_2$  retention or ventilator synchronization difficulties nasal interface leaks were eliminated, pressure support and ventilator rate increased or the patient switched from BiPAP-ST<sup>TM</sup> 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  $SpO_2$  remained within normal limits for 2 days and when assisted coughing was needed less than 4 times per day.





#### Noninvasive ventilation can not be used when Spo2 is <94% with normocapni

Bach JR, Baird JS, Plosky D, Nevado J, Weaver B. Spinal muscular atrophy type 1: management and outcomes. Pediatr Pulmonol 2002;34:16-22.

Bach JR. Amyotrophic lateral sclerosis: prolongation of life by noninvasive respiratory aids. Chest 2002;122:92-98.

Gomez-Merino E, Sancho J, Marin J, Servera E, Blasco ML, Bach JR, Baird JS, Plosky D, Nevado J, Weaver B. Spinal muscular atrophy type 1: management and outcomes. Pediatr Pulmonol 2002;34:16-22.

Bach JR (ed). Noninvasive Mechanical Ventilation. Philadelphia: Hanley & Belfus, 2002, 348 pages.

## 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<sup>1</sup>,<sup>2</sup>

<sup>1</sup>Bach JR. Arch Phys Med Rehabil 1995;76:828-832.

<sup>2</sup>Bach JR, Saporito LR. Chest 1996;110:1566-1571.







## NONINVASIVE MECHANICAL VENTILATION











JOHN R. BACH, MD

# Management of Patients with Neuromuscular Disease





John R. Bach, MD

