Ancient Chinese Proverb: "The first step to wisdom is to call something by what it really is."

What is "NIV"
- Continuous positive airway pressure
- Low span (drive pressure) positive airway pressure ("BiPAP")

What is NVS?
- "NONINVASIVE VENTILATORY SUPPORT"
- Intermittent positive pressure ventilatory support at > full ventilatory support settings 800 to 1500 ml, rate 12; pressure preset 18-20 cm H2O
- High span bi-level positive airway pressure at spans >15 cm H2O

3 Aspects to Management
- Long-term noninvasive care (NVS & MIE)
- Extubation of ventilator "unweanable" patients back to NVS and MIE
- Decanulation of to NVS

Upper vs. Lower Motor Neuron Disease (MND)
- Myopathies and spinal muscular atrophy (lower MND) do not need trach tubes to prolong survival
- Amyotrophic lateral sclerosis patients usually require trach tubes to prolong survival
72 on CNVS mean age 86.1 (range 13–110) months; 13 died at 52.3 (range 13–111) months: Bach JR, Gupta K, Reyna M, Hon A. Spinal muscular atrophy type 1: prolongation of survival by noninvasive respiratory aids. Pediatric Asthma, Allergy & Immunology 2009, 22(4):151–162.
Duchenne muscular dystrophy

- 125 patients used NIV, MAC/oximetry for 10.5 ± 6.1 years
- 108 nocturnal-only NIV users extended to continuous NIV for 8.6 ± 6.1 years to age 31.8 ± 6.1
- 56 patients still alive, life expectancy 39.6.
- 32 of the 108 were not hospitalized
- 8 tracheostomy continuous users were decanulated to NIV
- 45 unweanable intubated patients were extubated to NIV/MAC.
- Of 57 deaths, 26 (46%) were probably cardiac, 14 (34%) probably respiratory, and 17 (30%) of other etiology with 8 patients dying from CHF before vent use.
- 8 of 12 deaths of TIV users were tube related.

How Long Can NVS Be Effective?

- SMA1 - > 25 years (indefinitely)
- DMD - > 30 years (indefinitely)
- Post-polio - > 60 years (indefinitely)
- ALS – up to 10 years
- All myopathic/lower NMD (indefinitely)


(42%) for 12.8 months
Outcomes of O2 Therapy are worse than NO THERAPY

Supplemental Oxygen Impairs Detection of Hypoventilation by Pulse Oximetry


It also “covers up” airway secretions

COPD/Sleep disordered breathing

- PFTs – diffusion, ABGs, plethysmography, forced expiratory volumes/ polysomnography
- Bronchodilators and oxygen therapy/CPAP or low span BiPAP
• Conclusions: Stable patients receiving prolonged mechanical ventilation (PMV) without clinical pneumonia have a high alveolar burden of bacteria. The bacterial burden in most patients exceeds the commonly accepted threshold for diagnosing ventilator associated pneumonia. The utility of quantitative bronchoscopic culture in the diagnosis of ventilator associated pneumonia in this patient population requires further study.


Mechanical Ventilation via a Tube decreases diaphragm contractile properties
Le Bourdelles et al. Am J Respir Crit 1994;149.

Survival with Tracheostomy Tubes
• 100 ALS patients, 40 died in 5 years, about 32/40 due to the tube and the others died suddenly

Duchenne Muscular Dystrophy
• Untreated 56 died at 18.6 ± 2.9 years
• 21 TMV died at 28.1 ± 8.3 years of age with three still alive. Kaplan-Meier predicted 29.1 years
• 88 CNVS dependent 5.8 ± 3.4 years had 50% survival to 39.6 years, p < 0.001.

The Respiratory Muscles
• Inspiratory
• Expiratory
• Bulbar-innervated

• The inspiratory and expiratory muscle aids
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)


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

CoughAssist™

• Use at 40-60 cm H2O to -40 to -60 cm H2O
• Use with exsufflation timed abdominal thrust if that further increases the flows.
• Use via mouth piece, oro-nasal interface, translaryngeal tube, or tracheostomy tube
• Time it to a baby’s breathing

Maintain normal alveolar ventilation around the clock

Symptoms

• Morning headaches
• Fatigue
• Hypersomnolence
• Decreased appetite
• Depression
• Impaired concentration
How to Begin

• If symptoms are questionable:
  – 1. Sleep SpO2 and EtCO2 monitoring
  If symptoms and VC and CO2 are consistent then
  offer sleep trial of nasal ventilation on
  assist/control mode 800 to 1500 ml, physiologic
  rate.
  As patient weakens are needs daytime aid, switch
  nasal interface for mouth piece
  - If inconvenience greater than benefit, reevaluate
  in 3-6 months

• Indications for NVS –
  – Paradoxical breathing
  – Symptoms of hypoventilation

  – Bach JR. Prevention of pectus excavatum for children
    with spinal muscular atrophy type 1. Am J Phys Med
IDEAL INTERFACE

- LEAK FREE
- COMFORTABLE
- MAINTENANCE FREE
"...some of our physical therapists, in struggling with (iron lung) patients, noticed that they could simply take the positive pressure attachment, apply a small plastic mouthpiece, and let that hang in the patient's mouth...We even had one patient who has no breathing ability who has fallen asleep and been adequately ventilated by this procedure, so that it appears to work very well, and I think does away with a lot of complications of difficulty of using (invasive) positive pressure. You just hang it by the mouth and the patients grab it with their lips, when they want it, and when they don't want it, they let go of it. It is just too simple...."

The Trilogy “Kiss” Trigger

The Trilogy Mouth Piece Ventilation Mode detects when a patient touches a mouthpiece to deliver on-demand positive pressure ventilation without counterproductive expiratory pressure, the foundation of noninvasive intermittent positive pressure ventilatory support.
How To Avoid Respiratory Failure and Tracheotomies

- Maintain O2 saturation >94% without O2 especially during colds
- How? By using Mechanical insufflation-exsufflator (MIE) and noninvasive intermittent positive pressure ventilation
- If SpO2 < 95%, you have either hypoventilation, mucus, or pneumonia
- Since ventilatory drive prevents asphyxia, avoid sedatives and oxygen

The Ventilatory Drive Controls the Ventilation if it is not Depressed by Supplemental Oxygen and Sedative Medications

Extubation and Decanulation

**Extubation and Noninvasive Ventilatory Management of Unweanable Patients with Neuromuscular Weakness**

**Extubation Procedure**

1) Nasogastric tube removed if present.

2) Noninvasive (nasal/oronasal/mouthpiece) interface placed for immediate post-extubation NIV at rate 10-12/min, pressure support of 18-20 cm H2O, or assist-control volumes of 700 to 1500 ml, using portable volume ventilator or other ventilator on noninvasive mode.

### PNEUMOBELT: PRACTICAL ASPECTS

2) HOW TO FIT IT?

- Upper horizontal border: at xypoid process level
- Lower curved border: well over epigastic region

3) No supplemental oxygen, for episodes of SpO₂ < 95% the ventilator positive inspiratory pressure (PIP) is checked and the interface observed for leak. CO₂ is checked, ventilator settings are checked, mechanically assisted coughing is used at pressures 40 to 60 cm H₂O to -40 to -60 cm H₂O as needed until SpO₂ returns to greater than or equal to 95% via the airway tube and subsequently via the oro-nasal interface.
Extubation After Weaning Failure

- 157 intubated patients failed SBTs both before and after extubation to full-setting NIV/MAC
- Before hospitalization 90 (61%) had no experience with NIV, 39 (26%) used it nocturnally, and 20 (15%) were continuously NIV-dependent.
- First attempt protocol extubation success 96% (147 patients).
- All 98 extubation attempts on patients with assisted CPF ≥ 160 L/m were successful.
- Continuous NIV dependence and duration of NIV dependence prior to intubation correlated with extubation success (p<0.005).
- Six of 7 patients who initially failed extubation succeeded on subsequent attempts, so 1 underwent tracheotomy despite continuous post-extubation ventilator dependence.

Decanulation of Unweanable Patients


VC increased by 270% (P < .001) from admission to post-extubation. Weaning from CNVS to part-time NVS was achieved by all 52 subjects who had not been CNVS-dependent before intubation. One subject with ALS underwent tracheotomy.


NIV Support Consensus, April 2010

22 Centers in 18 countries
760 continuous NIV dependent patients with DMD, ALS, SMA type 1 for 3000 patient-years

When Do Patients Need Tracheostomy Tubes?

When Mechanical in-exsufflation (MIE) expiratory flows do not exceed 100 L/m and airway secretions cause O2 sat to remain below 95% despite MIE and NVS

No one with any myopathic or lower motor neuron neuromuscular disorder needs a tracheostomy tube for even continuous ventilatory support. Only patients with CNS and/or upper motor neuron disease whose MIE-EF diminish to about 100 L/m and who aspirate airway secretions such that O2 sat baseline remains below 95% need tracheostomy tubes to prolong survival. MIE is safe to use at 60-70 cm H2O via invasive airway tubes and safe at 40-60 cm H2O via the upper airways. Definitive CNVS rather than trach ventilation requires rapid access to MIE and at times extubation or decannulation to CNVS even when “unweanable”

Money – Duchenne alone

- 101 on 24 hr NIV for a mean 8.6 years, 26 without hospitalization, cost for respiratory and other equipment $800/month
- 25 on 24 hr TMV cost for nursing/institutionalization $22,000/month
- Cost savings $21,400/month or 94% or $236,400/year or 101x8.6x$236,000=$205 million assuming that they would survive as long using TMV

WHO Rankings, Per Capita Expenditure, Life Expectancy

1 – France 4 82 (8th)
2 – Italy 11 83 (2nd)
6 – Singapore 38 83 (2nd)
37 – U.S.A. 1 79 (34th)

- In 2017 the US is spending $3.6 trillion (including >$500 billion for administrative costs) on health care ($3 trillion in 2014), more than twice our budget deficit of almost $2 trillion, compared with $770 billion for our military, 160 billion for higher education. CMS now over $1.3 trillion.

- If we had the same health care system as Singapore, we would be in the black by $1.8 trillion per year.

The Institute of Medicine estimates that unnecessary and inefficient medical services cost almost $1 trillion per year. By some estimates, it is $2.8 trillion.

Over Treatment
- Primary Care Medicine: Shannon Brownlee, Overrated: Why Too Much Medicine Is Making Us Sicker and Poorer.
- David Goldhill, Catastrophic Care: How Everything We Think About Health Care Is Wrong. 2007.

Respiratory “overtreatment”?
- Tracheotomy is the most frequent operation in every trauma hospital in the United States yielding $10s millions in revenues for hospitals and the majority of income for many surgeons.
- Tracheotomies are performed when patients are intubated and fail ventilator weaning parameters and spontaneous breathing trials.

www.breatheBB.com
www.breatheNVS.com