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
72 on CNVS mean age 86.1 (range 13–196) 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.
Respiratory Failure

- Failure of Ventilation
  \[ \text{PaCO}_2 \uparrow \ (\text{pH} \downarrow) \]

- Failure of Oxygenation
  \[ \text{PaO}_2 \downarrow \]
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
Outcomes of O2 Therapy are worse than NO THERAPY

Home mechanical ventilator use vs. O₂ on pneumonia and hospitalization rates²

Periods:
Oxygen therapy higher P&H rate (p<0.001) vs. untreated pts after initial respir distress (p<0.001) vs. tracheostomy or noninvasive IPPV
Full-time noninvasive IPPV users had the lowest P&H rates (p<0.001)

O₂ therapy is not an effective substitute for IPPV
Noninvasive IPPV can be used safely full-time
Supplemental Oxygen Impairs Detection of Hypoventilation by Pulse Oximetry

• Fu ES, Downs JB, Schweiger JW, Miguel RV, Smith RA. Chest 2004;126:1552-1558.

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
TRACHEOSTOMY
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.

plot of non-invasive vs. TIPPV by time

- Non-invasive (n=17)
- TIPPV (n=33)

Sitting PCO2 mm Hg

Time in years

0 1 2 3 4
Mechanical Ventilation via a Tube decreases diaphragm contractile properties

Le Bourdelles et al. Am J Respir Crit 1994;149.
COMPLICATIONS OF TRACHEOSTOMY

Placement: arrhythmia, hemorrhage endobronchial intubation mediastinal, subcutaneous emphysema

Acute and long term: nosocomial pneumonia bacterial colonization with chronic purulent bronchitis mucous plugging, oxygen toxicity + water balance, GI complications septicemia, abscess, cellulitis pneumothorax, granuloma, hemorrhage tracheal stenosis, rupture need for suctioning, impaired GPB impaired communication
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
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
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
STEP 1
A mouth and throat full of air is taken depresssing the tongue, jaw, and larynx to get maximum volume. The larynx has been shut. In taking this large breath, lip shape is as if saying "ooop."

STEP 2
The lips are closed and the soft palate raised to trap the air. Saying the words "oh" or "coo" does this. No air should escape from the nose.

STEP 3
The larynx is now opened. The jaw, floor of mouth and larynx are raised. This together with progressive motion of the tongue forces air through opened larynx.

STEP 4
After as much air as possible is forced through the larynx, it is closed and ready for repetition of first step. The tongue draws up and retracts. The "ng" is like saying "up."
Fig. 7. Top: Maximal GPB minute ventilation 8.39 lt, GPB inspirations average 1.67 lt, 20 gulps, 84 cc/gulp for each breath in a patient with a vital capacity of 0 cc.

Bottom: Same patient regular GPB minute ventilation 4.76 lt/min, 12.5 breaths, average 8 gulps per breath, 47.5 cc/gulp performed over one minute
SMA1 Standard of care: death from respiratory failure, pectus excavatum and funnel shaped chest
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."
Woman campaigns to have 'cough' machine on market

By NANCY JAEKLE
The Daily Journal

A portable cough machine, no longer manufactured today, once saved Florence Lunde's life.

And now she has launched a one-woman campaign to return the device to the market.

"I was alone in the house and I was eating an apple and I choked on the fluid and the pulp and I could do nothing. Somehow, I managed to get to the machine and it saved my life," the Bronx resident said.

The inability to cough threatens the lives of about 500,000 people nationwide. Because they are unable to cough on their own, these people must rely on artificial methods to rid themselves of unwanted secretions.

Known as the Coflator, the portable cough machine overcomes bronchial non-drainage, caused by abnormalities of natural mechanisms, by means of exsufflation. It creates a suction effect which triggers a cough in the patient.

There are approximately 50,000 people with neuromuscular problems such as muscular dystrophy, multiple sclerosis and myasthenia gravis. Geriatrics also have weak coughs.

About 10,000 people use ventilators of all types; however, there are just 200 Coflators in the country.

"The Coflator was originally made in 1945 for polio patients. But thanks to the Salk vaccine, polio has been eliminated. Therefore the companies stopped making these machines. Until now, Life-Care, which is the vendor for the respiratory equipment has been repairing it, but they have stopped," Lunde said.

Lunde said the company claims it is not a profit-making venture. As of 1988, Life-Care has given Coflator units to patients without responsibility for repairs or parts.

"As of last summer there was a commitment to make the Coflator. The first prototype was unsatisfactory. The second prototype was satisfactory. However, there is difficulty because of the stringent rules of the Food and Drug Administration to develop this product due to cost and time consumption," said Dr. August Alba of the New York University Medical Center's Goldwater Hospital for long-term care.

The Coflator keeps Jimmy Starita alive. Jimmy, now 19, has Bruchnan's muscular dystrophy and was not expected to live past 15.

"I'm fortunate because I have this machine in my house now. But if someone needs the Coflator I must take it to where it is needed. Therefore, it puts my son's life in jeopardy because it is no longer in our house," said Nicholas Starita, Jimmy's father and the executive director of United Cerebral of Hudson County.

Readers should contact Starita at (201) 662-0080 for further information.
Mechanical Assisted Cough
“A historical perspective”

...Since 1954 to 2010

The cof-flator

The “Tin Can”
Insufflator-Exsufflator

The In-Exsufflator

The Cough Assist
Do Flow Study

• E70 flow study varying pressures from 30 to 60 cm H2O, via airway tubes 60 to 70 cm H2O. Perform with and without concomitant cough and abdominal thrust.
能幹的莉莉 很會‘照顧’超人
這隻導盲犬會調電視、錄影機、幫忙拿東西 是李維的患難之交

國際新聞組彭淮棟／譯

「超人」影集主角克里斯多夫李維馬上就有動物縁，乍見莉莉，眼睛一亮，輕呼名字，她也乖巧，走到他身邊輕舔他的手，靈犀相通，目前正在進一步彼此適應。導盲犬一般天性溫和，智力奇高，本能敏銳而勇於護主，而且念舊，與主人情誼終身不渝，莉莉從此將成為克里斯多夫李維的良助益友。

同時，他已經與愛妻計畫著，紐約州貝德福市家中該如何配合他的需要調整一番，好讓他住得方便舒服。院方透露他快要可以回家了，預計就在感恩節到耶誕節之間。（明星周刊）
CoughAssist™

• Use at 35-60 cm H2O to -35 to -60 cm H2O
• Use with exsufflation timed abdominal thrust
• 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 NIV** –
  – Paradoxical breathing
  – Symptoms of hypoventilation

Dibujo de George Cruikshank (1792-1878)
IDEAL INTERFACE

• LEAK FREE

• COMFORTABLE

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

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

96-89%
ALVEOLAR HYPOVENTILATION
COMPENSATED BY
PATIENT EFFORT
M-IPPV first reported at a conference on post-poliomyelitis

- John E Affeldt of Rancho Los Amigos Hospital in Los Angeles observed that an intermittent positive-pressure ventilation machine with a mouthpiece circuit could be used to relieve dyspnea in ventilator dependent polio patients whose negative-pressure ventilation was interrupted for transfers, nursing care, or physical therapy.


“...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 allow that to 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 patients and they grip 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....."
Dream Music Directory
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.
PNEUMOBELT: PRACTICAL ASPECTS

2) HOW TO FIT IT?

Fig. 1. - Pneumatic belt: measurements and position.

h and w (cm): measures required for orders (APARD)

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
• Since ventilatory drive prevents asphyxia, avoid sedatives and oxygen
## TABLE 5  Considerations for assessing readiness to wean

| Clinical assessment | Adequate cough  
|                    | Absence of excessive tracheobronchial secretion  
|                    | Resolution of disease acute phase for which the patient was intubated  
| Objective measurements | Adequate oxygenation  
|                    |  
|                    | $\text{Sa}_{\text{O}_2} > 90\%$ on $\leq \text{Fi}_{\text{O}_2} 0.4$ (or $\text{Pa}_{\text{O}_2}/\text{Fi}_{\text{O}_2} \geq 150$ mmHg)  
|                    | PEEP $\leq 8$ cmH$_2$O  
|                    | Adequate pulmonary function  
|                    | $fr \leq 35$ breaths·min$^{-1}$  
|                    | MIP $\leq -20$ to $-25$ cmH$_2$O  
|                    | $\text{VT} > 5$ mL·kg$^{-1}$  
|                    | VC $>10$ mL·kg$^{-1}$  
|                    | $fr/VT < 105$ breaths·min$^{-1}$·L$^{-1}$  
|                    | No significant respiratory acidosis  
|                    | Adequate mentation  
|                    | No sedation or adequate mentation on sedation (or stable neurologic patient)  

Neuromuscular Disease Causing Acute Respiratory Failure

Sangeeta Mehta MD FRCPC

Respiratory Care • September 2006 Vol 51 No 9

Weaning and Extubation

In patients with respiratory failure due to neuromuscular disease, the decision to extubate can be a challenging one, and no studies have specifically addressed weaning and extubation in this patient population. In general, patients should have adequate cough, few secretions, and should tolerate a low level of pressure support for a prolonged period without signs of fatigue.\textsuperscript{11} > 10 mL/kg for > 4 h and tolerance of T-piece breathing for > 4 h prior to extubation.\textsuperscript{11}
Conventional Extubation

1. Oxygen administrated arbitrarily in concentrations that maintain SpO2 well above 95%.
2. Frequent airway suctioning via the tube.
3. Supplemental oxygen increased when desaturations occur.
4. Ventilator weaning attempted at the expense of hypercapnia.
• 5. Extubation not attempted unless the patient appears to be ventilator weaned.

• 6. Extubation to CPAP or low span bi-level positive airway pressure and continued oxygen therapy.

• 7. Deep airway suctioning by catheterizing the upper airway along with postural drainage and chest physical therapy.
• 8. With increasing CO2 retention or hypoxia supplemental oxygen is increased and ultimately the patient is reintubated.

• 9. Following re-intubation tracheostomy is thought to be the only long-term option ...or following successful extubation bronchodilators and ongoing routine chest physical therapy are used.

• 10. Eventually discharged home with a tracheostomy, often following a rehabilitation stay for family training.
Risk Factors for Extubation Failure in Patients Following a Successful Spontaneous Breathing Trial*

Fernando Frutos-Vivar, MD; Niall D. Ferguson, MD, MSc; Andrés Esteban, MD, PhD; Scott K. Epstein, MD, FCCP, Yaseen Arabi, MD, FCCP; Carlos Apezteguía, MD; Marco González, MD; Nicholas S. Hill, MD, FCCP; Stefano Nava, MD; Gabriel D’Empaire, MD, and Antonio Anzueto, MD

To the Editor:

John R. Bach, MD
UMDNJ-New Jersey Medical School
Newark, NJ
“…We conclude that the ability to generate CPF of at least 160L/min is necessary for the successful extubation or tracheostomy tube decannulation of patients with neuromuscular disease irrespective of ability to breathe…”
The Ventilatory Drive Controls the Ventilation if it is not Depressed by Supplemental Oxygen and Sedative Medications
### Table 1- Extubation Criteria for Continuously Ventilator Dependent Patients

- Afebrile and normal white blood cell count
- $\text{PaCO}_2$ 40 mm Hg or less
- Oxyhemoglobin saturation ($\text{SpO}_2$) $\geq 95\%$ for 12 hours or more in ambient air (may need to use mechanically assisted coughing via the tube to achieve this)
- Fully alert and cooperative, receiving no sedative medications
- Chest radiograph abnormalities cleared or clearing
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.
Extubation Procedure (cont.)

3) No supplemental oxygen, for episodes of \( \text{SpO}_2 < 95\% \) 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 \( \text{H}_2\text{O} \) to -40 to -60 cm \( \text{H}_2\text{O} \) as needed until \( \text{SpO}_2 \) 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 98 (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.

http://chestjournal.chestpubs.org/content/early/2009/12/24/chest.09-2144.
| Diagnoses (%) | ALS – 15 (10%)  
| | DMD – 15 (11%)  
| | ICUMy – 15 (11%)  
| | MD – 20 (14%)  
| | MG – 13 (9%)  
| | PPS – 10 (7%)  
| | SMA – 23 (16%)  
| | SCI – 16 (11%)  
| | oNMD – 16 (11%)  
| Use of NIV pre-intubation | noNIV – 93 (64%)  
| | C – 14 (10%)  
| | Noct – 37 (26%) |
Decanulation of Unweanable Patients

“... It was concluded that, in general because of their youth, intact mental status and bulbar musculature, and absence of obstructive lung disease, patients with traumatic high level spinal cord injury are candidates to benefit from continuous noninvasive ventilation and assisted coughing techniques even with no ventilatory autonomy...”
22 Centers in 18 countries
760 continuous NIV dependent patients with DMD, ALS, SMA type 1 for 3000 patient-years

<table>
<thead>
<tr>
<th>#</th>
<th>N</th>
<th>ptNIV</th>
<th>Age</th>
<th>Duration</th>
<th>Age</th>
<th>Duration</th>
<th>Age</th>
<th>Still</th>
<th>Ext</th>
<th>Decan Deaths</th>
<th>Tt</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>120</td>
<td>20.3±2.8 2.0±2.1</td>
<td>101</td>
<td>22.3±5.9</td>
<td>7.0±5.9</td>
<td>30.3±6.1</td>
<td>63/128</td>
<td>29</td>
<td>9</td>
<td>14/63</td>
<td>0*</td>
</tr>
<tr>
<td>2</td>
<td>10</td>
<td>22.7±3.2 1.7±1.9</td>
<td>10</td>
<td>24.3±4.1 4.6±1.6</td>
<td>28.4±4.6</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>24</td>
<td>18.0±1.6 9.9±4.5</td>
<td>24</td>
<td>28.1±4.6 4.2±2.8</td>
<td>32.3±2.8 24/24</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>9</td>
<td>18.7±5.2 4.6±2.2</td>
<td>4</td>
<td>23.3±6.5 4.0±2.8</td>
<td>27.0±5.0 4/4</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>42</td>
<td>21.9±4.1±2.5</td>
<td>42</td>
<td>26±5</td>
<td>31±3</td>
<td>32/42</td>
<td>0</td>
<td>0</td>
<td>3/10</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>6</td>
<td>22.0±2.8 3.0±1.1</td>
<td>6</td>
<td>25.0±2.4 4.9±3.2</td>
<td>29.9±3.6 4/6</td>
<td>0</td>
<td>0</td>
<td>1/2</td>
<td>0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>25</td>
<td>18.3±4.0 3.5±2.4</td>
<td>11</td>
<td>21.9±2.4 5.0±4.4</td>
<td>26.9±4.3 8/11</td>
<td>2</td>
<td>1</td>
<td>1/3</td>
<td>0</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

ALS

<table>
<thead>
<tr>
<th>#</th>
<th>N</th>
<th>ptNIV</th>
<th>Age</th>
<th>Duration</th>
<th>Age</th>
<th>Duration</th>
<th>Age</th>
<th>Still</th>
<th>Ext</th>
<th>Decan Deaths</th>
<th>Tt</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>176</td>
<td>52.5±5.6 0.9±1.1</td>
<td>109</td>
<td>53.3±5.3 0.8±2.2</td>
<td>54.6±5.7 67/109</td>
<td>15</td>
<td>6</td>
<td>42</td>
<td>44*</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>4</td>
<td>67.8±6.9 1.3±0.8</td>
<td>4</td>
<td>69.3±6.6 2.0±1.9</td>
<td>70.5±8.4</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>78</td>
<td>59.5±9.4 0.9±1.1</td>
<td>27</td>
<td>60.3±1.3 0.6±0.5</td>
<td>62.2±9.1 14/27</td>
<td>0</td>
<td>0</td>
<td>5/14</td>
<td>6</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>1</td>
<td>0/1</td>
<td>0</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>83</td>
<td>56.1±9.0 0.9±0.9</td>
<td>19</td>
<td>55.5±9.0 1.1±2.1</td>
<td>6/19</td>
<td>0</td>
<td>0</td>
<td>7</td>
<td>6</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Indications for Tracheostomy

When the Spo2 decreases below 95% and cannot be normalized by NIV or MAC
Classics of Paradigm Paralysis

• “There is no reason for any individual to have a computer in their home.” --- Kenneth Olsen, 1977 President and founder of Digital Equipment Corp.
• "Airplanes are interesting toys but of no military value." --- 1911, J. Marshal Ferdinand Foch, French Military Strategist and future World War I commander
• "Man will never reach the moon regardless of all future scientific advances." --- February 25, 1967, Dr. Lee de Forest, inventor of the Audio Tube (Television), and Father of Radio.
• "[Television] won't be able to hold on to any market it captures after the first six months. People will soon get tired of staring at a plywood box every night." --- 1946, Darryl F. Zanuck, head of 20th Century-Fox.
• "We don't like their sound. Groups of guitars are on the way out.”---1962, Decca Records rejecting the Beatles.
• "For the majority of people, the use of tobacco has a beneficial effect." --- November 18, 1969, Dr. Ian G. MacDonald, Los Angeles surgeon, as quoted in Newsweek.
• "This 'telephone' has too many shortcomings to be seriously considered as a means of communication. The device is inherently of no value to us." --- 1876, Western Union Internal memo.
• "The earth is the center of the universe." --- Popes of the Roman Catholic Church
• "Nothing of importance happened today." --- July 4, 1776, written by King George III of England
• ‘‘Everything that can be invented has been invented.’’ --- 1899, Charles H. Duell, U.S. Commissioner of patents.

To this can now be added:
• “Daytime noninvasive ventilation via a mouthpiece should not be viewed as an alternative to tracheostomy”
“Everything that can be invented has been invented.” --- 1899, Charles H. Duell, U.S. Commissioner of patents.

To this can now be added:

“Daytime noninvasive ventilation via a mouthpiece should not be viewed as an alternative to tracheostomy”


Dr. Bach obviously feels that his way of doing things is the only way and there is no room for discussion --- participant at the American Association of Neuromuscular Electrodiagnostic Medicine Course on Respiratory Problems and Solutions in Neuromuscular Disease. San Antonio, Texas, October 17, 2013.
• All truth passes through three stages.
• First it is ridiculed.
• Second, it is violently opposed.
• Third, it is accepted as being self-evident.
• Schopenhauer 1788-1860
Outcomes

- 157 unweanable NMD patients transferred intubated after failing extubation and refusing tracheotomy
- 155/157 successfully extubated, 2 bulbar ALS patients failed
- 20 were 24hr/d NIV dependent for 12 years before intubation
- 98/98 extubations with CPF > 160 L/m successful

Reasons Respiratory Failure is not Prevented

- 1. neurologists are not very interested in respiratory management
- 2. pulmonologists do not think in terms of supporting muscle function
- 3. compensation is for intensive care and interventions rather than preventive
- 4. "paradigm paralysis" or simply thinking that patients who can't breathe NEED invasive tubes
- 5. failure of clinics to have properly trained respiratory therapists
- 6. Need 24 hour attention when sick at home.
- 7. need for educated physician team
- 8. Insufficient respiratory therapist training
- 9. Inadequate equipment in hospitals
• 1. Oxygen administrated arbitrarily in concentrations that maintain SpO2 well above 95%.
• 2. Frequent airway suctioning via the tube.
• 3. Supplemental oxygen increased when desaturations occur.
• 4. Ventilator weaning attempted at the expense of hypercapnia.
• 5. Extubation not attempted unless the patient appears to be ventilator weaned.

• 6. Extubation to CPAP or low span bi-level positive airway pressure and continued oxygen therapy.

• 7. Deep airway suctioning by catheterizing the upper airway along with postural drainage and chest physical therapy.
導入開始の基準

・気管切開より早く導入して良いが、早過ぎても発達面で好ましくない（Make BJ. Neuromuscular Disorders. 1991; 1:229-230）

・M/NIPPVの歴史が本邦より10年古い欧米でも、やや早い基準から遅い基準までである

§米国（Bachら、1994年）
(1) 病気の進行のスピードが早い場合
(2) PaCO₂かEtCO₂の最高値が50mmHgを超える高CO₂血症を認める場合
(3) SaO₂平均値が95％未満になる場合
(4) CAH症状を認める場合（Bach JR. et al. Respiratory Care. 1994;39:515-531）
Comment AANEM 2013

- Dr. Bach obviously feels that his way of doing things is the only way and there is no room for discussion. He was disorganized and ranting. I think that his point that non-invasive ventilation is under used in neuromuscular disease is valid, but he was difficult to follow and did not provide useful information on how to implement his ideas.
Paradigm Paralysis?

- 1898 Nicola Tesla operated 4’ boats by radio remote control on a Central Park lake and although thought to be a magician, neither industry nor the American government was interested until the Germans reinvented remote control and used the same technology for tanks in the 1940s.
- 1680 Frenchman Denis Papin designed a steam engine without building it then 89 years later James Watt`s steam engine resulted in a paradigm shift to commercial it.
- Nikolaus Otto built his first gas engine in 1866 but, since horses were doing a good job transporting people, it was not until 1895 until the engine for first used to operate a truck and about 18 years later when automobiles began to be mass produced.
- Objects were glazed in furnaces as early as 4000 BCE, free standing glass objects appeared in Egypt and in Mesopotamia around 2500 BCE, and glass vases around 1500 BCE so why did it take another 1500 years until glass windows were produced by Rome? (Guns, germs)
Conclusion

Thus, lack of all VFBA in SCI does not mandate tracheostomy or EPP/DP. Only severe glottis dysfunction that results in aspiration of saliva and oxyhemoglobin saturation below 95%, as it can in amyotrophic lateral sclerosis, may mandate the need for tracheostomy for survival for SCI.²⁸
“[Phrenic pacing] is a drastic and dangerous procedure. The risks are enormous. Batteries fail. The procedure frees you from the ventilator, but the outcome can be fatal.”

- Christopher Reeve. Still Me