Noninvasive Mechanical Ventilation of Neuromuscular disorders

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Respiratory Considerations
- Impairment of Oxygenation due to lung disease
- Impairment of Ventilation due to weak respiratory muscles

Symptoms of Hypoventilation
- Fatigue
- Shortness of breath
- Morning or continuous headache
- Sleep awakenges with SOB or heart aching
- Poor concentration
- Frequent nightmare
- Sxs & signs of heart failure due to breathing impairment
- Decreased libido
- Lower limb swelling
- Irritability, anxiety
- Frequent arousal from sleep to urinate
- Impaired intellectual function
- Depression
- Excessive weight loss
- Muscle ache
- Memory impairment
- Obesity
- Muscle ache

Scoliosis in NMD
- When the curve reaches 40 degrees, Vital Capacity (VC, breathing capacity) may be below 23% of expected normal. They become ventilator dependent after operation if VC < 23%
- If FVC < 40% of predicted normal, -- contraindicate surgical spinal arthrodesis

Indications for Scoliosis Surgery
- Absolute FVC > 2,000 cc tended not to show severe progressive scoliosis
- VC plateau: between 1,500-1,800 cc
- VC < 30% of predicted normal
- Scoliosis curve > 35 degrees
- VC < 1,500 cc in W/C dependent + pelvic obliquity

Pulmonary Rehab.
- Screening: EtCO2, O2 saturation, Maximum Insufflation Capacity (MIC), Peak Cough Flow (PCF), Vital Capacity (VC)
- Prevention: Air stacking exercise with range of motion of lung- by glossopharyngeal breathing, with manual insufflator, or volume cycled ventilator, assisted cough
- Maintenance: Non-invasive ventilator (BIPAP, IPPV), Cough Assist (In/Ex-sufflator, Cofflator)
- Education
Screening Tests for Respiratory Muscle Dysfunction

Vital Capacity
Maximum Insufflation Capacity
Cough Flow
End tidal CO2
O2 Saturation

Vital Capacity: by Spirometer

VC in normal plateaus at 19 y/o (4-5 liter ↓ by 1% to 1.2%/year)
VC in DMD plateaus between 10-15 yrs old

Maximum Insufflatin Capacity (MIC)

By Spirometer in sitting and lying
- The maximum quantity of air that can be held with a closed glottis.
- Correlates with bulbar muscle function & pulmonary compliance
- For patients with VC 30% less than predicted levels

Maintenance of Pulmonary Compliance

- Deep insufflations
  - prevent atelectasis,
  - improve lung compliance (ex; VC 300cc but Maximum Insufflation Capacity can be 1,200 cc)
  - Can be achieved by mouth piece attached manual resuscitator with air-stacking ex. (3x/day): air-stacking ex.
  - Should be introduced before VC decreases to 50-60% of normal

Glossopharyngeal Breathing (GPB)

- Introduced as an aid for coughing
- Good bulbar muscle function needed
- The tongue & throat muscles project (gulp) boluses of air into the lungs.
- Both inspiratory & indirectly expiratory muscle function are assisted by GPB
- GPB allows ventilator free breathing for ventilator dependent patients

Cough Flows

- A normal cough requires a precough inspiration or insufflation to about 85-90% of total lung capacity
- Normal peak cough flows (PCF) attain 6000-17,000cc/sec
- Assisted PCF below 4.5 liters/second: high risk of pneumonia and respiratory failure due to inability to clear airway mucus
- In case unassistd cough flows are not enough to eliminate airway secretion, use expiratory & inspiratory muscle aids can increase cough flows
Cough Flows-cont’d

- If VC < 1000cc (1 liter), assisted cough after maximally insufflating the lungs
- Normal PCF: 6 L/sec, unable to cough with PCF < 2.7 L/sec (intubation needed)
- Assisted PCF < 4.5 L/sec: manually assisted or mechanically assisted coughing to prevent pneumonia
- Failure of assisted PCF to attain at least 2.7 L/sec: severe bulbar m. weakness, scarring of vocal cords, narrowing of airways, severe coincident airway disease
- Assisted PCF can be improved 3 times than unassisted cough

Mechanical In-Exsufflation (Cofflator/Cough Assist)

- Insufflation: forced inhalation to volumes greater than one can achieve with inspiratory muscles
- Exsufflation: forced exhalation of greater volumes than can be generated with the expiratory muscles.
- Cough Assist inflated the lungs at positive pressures up to 40 mmHg over 2 seconds → immediately switch to negative pressures for 3 seconds
- Cofflator at 1953 → discontinued due to tracheostomy, tracheal suctioning & bronchoscopy

In/Ex-sufflator (Cofflator) or CoughAssist MI-E Cough Machine

- It clears retained broncho-pulmonary secretion by gradually applying positive pressure to airway, & then rapidly shifts to negative pressure. The rapid shift produces a high expiratory flow from the lungs, simulating a cough.

Cough Assist-cont’d

- Indications: pts w/ PCF 2-3 Liter/second, high SCI, NMD, severe fatigue associated with intrinsic lung disease.
- Contraindications: Bullous emphysema, pneumothorax, pneumo-mediastinum, recent barotrauma, impaired consciousness/inability to communicate

Cough Assist-When/How?

- 4 session /day
  - 1 session: 3-5x/day
  - 1 treatment: 3-5 cycles
  - 1 cycle: 1 insufflation & 1 exsufflation
- Best before meals and at bedtime
- Combine w/ assisted cough 2 times a day or PRN if secretion present

End tidal CO₂ (EtCO₂)

- Correlates with lung underventilation, cardiac output, lung perfusion pressure
- Correlates closely with PaCO₂ except pt w/ severe lung & vascular disease, congestive heart failure or impaired lung diffusion or perfusion
- Measure EtCO₂ in regular & maximal breathing - Hypercapnia when VC < 55% of normal
Oxygen Saturation (SpO2)

- May be normal despite EtCO₂ < low 60's
- If hypercapnia causes SpO₂ < 95%, patients develop symptoms of alveolar hypoventilation
- Should maintain > 94%: if not, start nocturnal monitoring

Definition of Non-invasive Mechanical Ventilatory Care

- The use of nasal, oronasal, & oral interfaces for the delivery of positive-pressure ventilation or the use of body ventilators that do not provoke the gag or cough reflex due to a direct mechanical stimulus

Invasive Ventilation

- Tracheostomy
  - for ongoing ventilator use
  - for effective airway secretions
- Complications
  - Subglottic stenosis, innominate artery hemorrhage
  - Tube & cuff disrupt mucociliary transport.
  - Increase infection due to decreased ciliary destruction, bacterial colonization of the airway (Sinusitis occur)
  - Vocal cord paralysis

Noninvasive Ventilation: TYPES

- Continuous Positive Airway Pressure (CPAP)
- Intermittent Positive Pressure Ventilation (IPPV)
- Combination of Positive Inspiratory Pressure + Positive End-Expiratory Pressure (PIP+PEEP): BiPAP
- Negative Pressure Body Ventilators (NPBVs)

E. Maintenance of Normal Lung Ventilation Around the Clock

- Daytime Ventilator: mouthpiece IPPV or use intermittent abdominal pressure ventilator
- Nighttime Ventilator: nasal IPPV
- Pressure cycled ventilator (BiPAP): for small children with neuromuscular disease, for treating sleep-disordered breathing
- Volume-cycled ventilators: for older patients (for air-stacking)

Indication of nocturnal noninvasive ventilation in DMD

- Hypercapnia (EtCO₂ >50 mmHg) during sleep with VC < 50%
- A mean SpO₂ < 95% for >1hr of sleep
- Symptoms of ventilatory insufficiency
- Multiple episodes of oxyhemoglobin desaturation of > 4%/hour during sleep
- PaCO₂ >45mmHg and/or Pao₂ <60mmHg in DMD
(Purpose of use of nocturnal noninvasive IPPV during sleep in the absence of respiratory muscle function: to keep the central ventilatory drive intact)
Selection of ventilator

- Patients who need full-time ventilatory support prefer noninvasive IPPV. (Negative Pressure Body Ventilators will be used for during tracheostomy site closure for transition from tracheostomy to noninvasive IPPV)
- No clear advantage could be appreciated using either volume-cycled ventilators or pressure-cycled ventilators.
- More clinicians preference
- Pressure cycled ventilators: PIP + PEEP, BiPAP
- LP-10, LTV-900 (volume or pressure-cycled ventilator)

Volume Ventilators

- Advantages
  - Deliver higher volume
  - Adjust flow rates for comfort
  - Low electricity
  - Less mean thoracic pr. -> less untoward hemodynamic effects on cardiac preload
  - Able to do air-stack
- Disadvantages
  - Heavier
  - Annoying alarms
  - Complicated

Pressure Ventilator-BiPAP

- Advantages
  - No annoying alarms
  - Light weight
  - Lower cost
  - Can compensate to some extent for small insufflation leaks
- Disadvantages
  - Inability to air-stack
  - Mouth drying, gagging, insufflation leakage, arousal from sleep
  - Discomfort & increased thoracic pr. due to unnecessary EPAP
  - No alarms, Noisier
  - CO2 rebreathing (corrected by non-breathing valve)

Pressure Support Ventilator

- The ventilator delivers a preset inspiratory pressure to assist spontaneous breathing efforts.
- Preset time-cycled inspiratory & expiratory pressures are delivered at a controlled rate with adjustable inspiratory-to-expiratory ratios.
- BiPAP

Continuous Positive Airway Pressure (CPAP)

- Delivery of a continuous flow of air into the airways via nose
- CPAP keep the airway open (splints it), but does not directly assist inspiratory muscle activity & does not help to ventilate the lungs
- Invented to treat sleep-disordered breathing (While airways are open by CPAP during sleep, inspiratory muscles are used for ventilating the lungs. If their inspiratory muscles are weak, hypercapnia will occur due to hypoventilation)

Bilevel Positive Airway Pressure (BiPAP)

- Was developed because of the frequent ineffectiveness of CPAP and the difficulties of tolerating high CPAP.
- Deliver air continuously under positive pressure like CPAP. However positive pressures delivered during inspiration separately from those delivered during expiration.
- Become preferable due to light weight, inspiratory pressure >20cmH2O, PEEP (=EPAP) capabilities, similarity to CPAP (CPAP+IPAP), low cost
BiPAP-Cont’d

“Span” = Inspiratory positive airway pressure (IPAP) – Expiratory PAP (EPAP) : the amount of inspiratory muscle assistance that the patient receive
Low spans (less than 10 cmH2O) provides a small pressure boost to assist inspiratory effort. Low span can only allow inadequate rest of respiratory muscles & insufficient assistance to inspiratory muscles.

Nocturnal-Only Nasal Ventilation

Nocturnal low-span BiPAP for mildly affected patients
High-span BiPAP or volume-cycled ventilators when low-span pressure assistance is no longer adequate
Intubation or tracheostomy tube connected to ventilator provide appropriate tidal volumes at adequate peak inspiratory pressure (20-25 cmH2O)

Portable Volume-Cycled Ventilators

PLV-100 (Respironics, Inc) LP-10, LP-20, LTV-900 can deliver volumes of 2500 cc
The most commonly used mode for home: assist-control, which delivers set-volumes of air, triggered by patient’s inspiratory effort
Provide regular deep insufflation, the capability of air stacking safety alarm
For typical volume for adult and adolescent: 800-1200 cc (can be >2000cc): a normal breath requires only 500-700cc., Rate: 10-12 cycles/min

Negative Pressure Body Ventilators (NPBVs)

Intermittently lower pressure around the chest & abdomen for air to go through the nose & mouth and into the lungs to equalize lung pressure.
Iron lung, chest-shell ventilators, PortaLung, Wrap-style pneumowarp

What to monitor for the effect of noninvasive ventilator?

SpO2, EtCO2,
Ventilator pressure gauge for adequate airway pressures
Monitoring expiratory volumes by attaching a spirometer to the expiratory valve when volume cycle ventilation used
Blood gas monitoring—not necessary unless oxygenation problems

Signs of successful use of non-invasive ventilation:

Decreased in respiratory rate & accessory respiratory muscle use
Increased chest expansion
Breathing synchrony with pressure-cycled ventilators or ventilator gauge pressures of 18-25 cmH2O with exhalation via exhalation valve of volume-cycled ventilators
Normalization of EtCO2 & SpO2
Relief of dyspnea
**Education for home use**

- **Pulse oximeter**
- **Cough Assist**

In case of chest cold ➔ cold & secretion cause fatigue & additional muscle weakness ➔ worsening hypercapnia & mucus plug ➔ SpO2 < 80%, VC decreased by 50-80% from baseline ➔ Pneumonia

**Oximetry Feedback-Respiratory Muscle Aid Protocol**

- For respiratory infection: continuous non-invasive IPPV to maintain alveolar ventilation, to increase PCF by air-stacking
- SpO2 < 95% despite ventilatory support & aggressive assisted cough ➔ need formal evaluation such as CXR, CBC to r/o chest infection, microscopic atelectasis
- SpO2 become normal: wean to nocturnal ventilator
- During hospitalization: use Mechanical Assist Cough (MAC) to remove airway secretion. Avoid sedatives, excessive oxygen

**Intubation**

- A clinical judgment
- Antibiotics, hydration, nutrition, chest PT, Intrapercussive ventilator, Cough Assist
- Need to maintain PaCO2 between 30 and 40 cmH2O.
- Supplemental oxygen can be given for SpO2 < 95%, PaCO2 < 40 cmH2O despite aggressive MAC (pressure: -40 to +40 cmH2O)

**Extubation**

- Criteria for extubation
  - No supplemental oxygen needed to maintain SpO2 > 94%
  - Chest X-Ray cleared or clearing
  - Respiratory depressant removed
  - Airway secretion less than on admission
  - Nasal congestion cleared
  - Afebrile, normal CBC

**Post-extubation Care**

- Wean: for some one who was already non-invasive pulmonary care prior to intubation ➔ directry to full time noninvasive IPPV
- Extubation succeeds when postextubation PCF can reach 160 liter/min (Bach & Saporito, Chest, 1996).
- Provide MAC via an oral-nasal interface with oximetry feedback to keep SpO2 > 94%
- Feeding with Pureed diet dink fluid through straw
- Episodes of desaturations for 1-2 hours after a meal are managed by MI-E without abdominal thrust

**Decannulation & Conversion to Noninvasive Respiratory Aids**

- Candidates for decannulation:
  - understandable speech with deflated cuff
  - good bulbar function,
  - the ability to clear airway secretions noninvasively
  - assisted PCF of about 160 L/min (Assisted PCF can be maximized with Cough Assit, GPB)
- Method: Initially use a noninvasive IPPV ➔ deflated cuff for 24 hrs ➔ tracheostomy button for few days ➔ occlusive tracheostomy site dressing with silicon donut for 24-72 hrs
Vitamin “O”

- One of the most common errors for people with neuromuscular disorder (NMD) is oxygen therapy- “Putting a Band-Aid on a cancer”
- Oxygen turn off the brain’s drive to breathe and greatly increases the ventilatory failures.
- Common cause for the desaturation in patients in NMD is mucus plug due to airway secretion, not due to pneumonia.

Thank You!!!!

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