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

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Symptoms of Hypoventilation

- Faitgue
- Shortness of breath
- Morning or continuous headache
- Sleep awakenings with SOB or heart aching
- Poor concentration
- Frequent nightmare
- Sxs & signs of heart failure due to breathing impairement
- Decreased libido

- Lower limb swelling
- Irritability, anxiety
- Frequent arousal from sleep to urinate
- Impaired intellectual function
- Depression
- Excessive weigh loss
- Muscle ache
- Memory impairment
- Obesity
- Muscle ache

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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

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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

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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

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Screening Tests for Respiratory Muscle Dysfunction

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

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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

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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

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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

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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

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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

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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

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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

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

- It clears retained bronco-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.*

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Cough Assist-cont'd

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

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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

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End tidal CO_2 ($EtCO_2$)

- 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 $EtCO_2$ in regular & maximal breathing
  - Hypercapnia when VC < 55% of normal

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Oxygen Saturation (SpO2)

- May be normal despite $EtCO_2$ < low 60's
- If hypercapnia causes SpO2<95%, patients develop symptoms of alveolar hypoventilation
- Should maintain > 94% : if not, start nocturnal monitoring

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Definition of Non-invasive Mechanical Ventilatory Care

- Ventilatory support without an endotracheal airway – Hillberg, N Eng J Med, 1997
- 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

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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

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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)

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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)

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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)

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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)

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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

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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)

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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

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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)

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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

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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.

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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)

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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

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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

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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

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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

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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

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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

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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<40cmH2O despite aggressive MAC (pressure: -40 to +40 cmH2O)

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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 cleard
  - Afebrile, normal CBC

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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

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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

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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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