

Understanding Breathing Muscle Weakness

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Legal Disclaimer

- This presentation is based on patient experience and reading medical literature. I am not a medical professional.
- Please don't use this presentation as a substitute for medical care.
- Please seek medical care for breathing muscle weakness from a licensed professional who specializes in patients with neuromuscular disease.

If you cannot breathe, you cannot do ANYTHING.

"Breathing and thinking are two of the most important processes, one for sustaining life and the other for giving it a purpose."
 ---Tasneem Hameed

What we will learn:

- What is breathing muscle weakness ?
- What about neuromuscular disease causes it?
- What does it affect?
- How is it treated?
- What is non-invasive ventilation (NIV)?
- What is invasive ventilation?
- What is mechanical insufflation exsufflation (MIE) or mechanically assisted coughing (MAC)?
- What is normal Oxygen saturation?
- What is the normal level of Carbon Dioxide in blood?

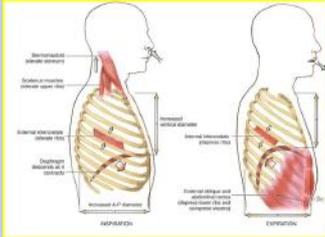


What is breathing muscle weakness?

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- Weakness in the diaphragm and accessory or intercostal muscles...



What is breathing muscle weakness?

- “...researchers found that abdominal muscles may be severely involved in the muscular dystrophy process. The abdominal muscles are important to provide respiratory support when the diaphragm muscle has been damaged by the disease, so that additional abdominal muscle involvement can worsen the respiratory situation considerably.” ---*Cardiac and respiratory function supported by abdominal muscles in muscular dystrophy*; <http://medicalxpress.com/news/2015-02-cardiac-respiratory-function-abdominal-muscles.html>

What about NMD causes it?

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What about NMD causes it?

- Smaller lung volume where the chest has less room to expand
- Bulbar muscle (swallowing) problems due to weak throat muscles
- A physical change to the skeletal structure causing scoliosis (a progressive curving of the spine) that has limited the chest cavity size and shape, meaning the lungs can't grow and/or expand normally

What does it affect?

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What does it affect?

- Cough strength (a weak and/or ineffective cough)
 - Cough requires taking a deep breath, pressure building in your lungs and air flowing forcefully out of the lungs.
 - When we cannot cough well, it can be exhausting.
 - Mucus can collect and bacteria can quickly multiply.
 - Mucus can fill the space in our lungs, crowding out room for Oxygen.
 - We are at greater risk for developing chest infections that can escalate to become pneumonia.

How is it treated?

- Weak and/or ineffective cough –
 - Medical literature and outcome studies tell us Mechanically Assisted Cough (MAC) in the form of Mechanical Insufflation and Exsufflation (MIE) is the best way to treat (CoughAssist by Philips Respironics does this)





Weak and/or ineffective cough

- "Mechanical insufflator-exsufflators simulate a cough by providing a positive pressure breath followed by a negative pressure exsufflation (27, 28). Comparison of peak cough expiratory flow rates by mechanical insufflation-exsufflation were shown to be superior to those generated either by breath stacking or manual cough assistance (29)."
- Use of mechanical insufflation-exsufflation was found to be particularly important in preventing hospitalization or need for tracheostomy in patients with DMD with peak cough expiratory flows around 160 L/min, especially when scoliosis prevented optimal use of manual assisted cough (8)."
- "...peak cough expiratory flow rate of 270 has been used to identify patients who would benefit from assisted cough techniques."

Reference: "Respiratory Care of the Patient with Duchenne Muscular Dystrophy: American Thoracic Society Consensus Statement" March 2004

How is it treated?

- Weak and/or ineffective cough –
 - Benefits of using Mechanically Assisted Cough (MAC) in the form of Mechanical Insufflation and Exsufflation (MIE):
 - ✦ Keeps the breathing muscles "stretchy" and more responsive to use of non-invasive ventilation (NIV); is a safe workout for the breathing muscles
 - ✦ When used daily, prevents mucus from settling deeply into the lungs
 - ✦ Patients report a boost of energy after use
 - ✦ Avoidance of hospitalizations, respiratory infections, pneumonias, episodes of respiratory failure, and tracheotomy

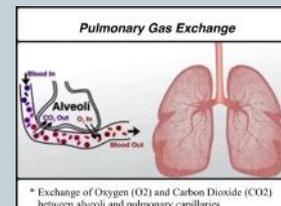
What does it affect?

- Ventilation (moving air into and out of the lungs)
 - We take shallower breaths and fill our lungs with less air.
 - During sleep:
 - ✦ We breathe even slower in our sleep.
 - ✦ Chest wall expansion and diaphragm movement is harder while lying down.



What does it affect?

- Pulmonary gas exchange (the process of exchanging Oxygen for Carbon Dioxide in the blood)



Pulmonary Gas Exchange 101:

- As we inhale, we take in room air (Oxygen).
- It moves into the lungs and is absorbed into our blood.
- Carbon Dioxide is a waste product of Oxygen and is absorbed into the alveoli of the lungs and moves out of the bronchi and into the trachea and out of the nose and/or mouth.

Pulmonary Gas Exchange 101:

- Two issues that can occur with pulmonary gas exchange in NMD:
 - ✦ Hypoxemia
 - ✦ Hypercapnia

Remember:
Hypo is low
Hyper is high

Pulmonary Gas Exchange 101:

- Hypoxemia is low Oxygen (O₂)
 - Often it's secondary to pneumonia or mucus plugging where the space in the lungs for Oxygen is crowded out by mucus.



Pulmonary Gas Exchange 101:

- Hypercapnia is retention of Carbon Dioxide (CO₂)
 - It's secondary to under-ventilation (not getting enough air into the lungs), so there is not enough O₂ to balance out the increasing amount of CO₂ that is being held in the lungs.
 - Something called the "Oxygen Dissociation Curve" allows our body to keep our Oxygen saturation relatively high, even though CO₂ is increasing. Eventually, with prolonged increases in CO₂, the O₂ level in the blood will drop.

Pulmonary Gas Exchange 101:

- Symptoms of Hypercapnia:
 - Difficulty sleeping:
 - Feeling anxiety when lying flat
 - Feeling as if you cannot breathe when lying down
 - Awakening multiple times in the night; having vivid nightmares
 - Morning headaches that usually go away after getting up
 - Daytime sleepiness
 - Fatigue
 - Difficulty concentrating
 - Memory problems
 - Confusion
 - Loss of appetite (which can lead to weight loss)
 - Altered mental status (loss of consciousness/blacking out)

How is it treated?

- Under ventilation –
 - Treated with mechanical ventilation (moving air into and out of the lungs), **not with supplemental Oxygen or CPAP**
 - **Why not Oxygen?** "Shallow breathing can provide the body with adequate oxygen supply and adequate removal of carbon dioxide. That delicate balance of oxygen and carbon dioxide allows breathing to continue. When extra or supplemental oxygen is given, this delicate balance is disturbed. The respiratory center in the brain may get the false impression that the body has enough oxygen and no longer needs to breathe. Without breathing, carbon dioxide can build to dangerous levels (called hypercapnia) that can result in death."

*"Giving Oxygen is like applying a band aid to Cancer."
– Dr. John R. Bach*



Under Ventilation

- **Why not CPAP?** CPAP offers only one level of positive pressure for both inhaling and exhaling against.
 - "CPAP helps to keep the airways in the lungs open, acting like an air splint, but not directly assisting respiratory muscle function. When there is respiratory failure with increased CO₂ in the blood, the use of CPAP alone is usually inadequate. A CPAP machine requires fairly strong respiratory muscles and could aggravate your ability... to breathe."

http://www.alsa.org/assets/pdfs/brochures/alsa_manual6.pdf

"Using CPAP is as effective as sticking your head out the window of a car while going 60 mph." – Dr. John R. Bach

Under Ventilation

- **Why BiPAP or Bi-level?** Bi-level offers two different levels of ventilation:
 - Higher inspiratory level known as IPAP
 - Much lower expiratory level known as EPAP (sometimes set to zero for maximum respiratory muscle rest)
 - Difference between the two is known as "span" i.e. IPAP of 18 and EPAP of 5.

Under Ventilation

- Non-invasive ventilation is started during sleep with a nasal mask, full face mask or nasal pillows mask and a bi-level device (sometimes a basic BiPAP with a back-up breathing rate or a multi-mode ventilator with pressure and volume support such as a Trilogy).

Under Ventilation

- People with neuromuscular disease should use a bi-level ventilator with a backup rate that can initiate breaths, particularly at night.
- The advantages of bi-level machines when compared to multi-mode and/or dual prescription ventilators are small size; light weight; lower cost and compensation for interface (mask) leaks. The disadvantages include lack of internal batteries; fewer alarms; inadequate pressures for some users; use of more electricity to operate; and discomfort from EPAP.”

Reference: International Ventilator Users Network (IVUN)
<http://www.ventnews.org/#lhmv-the-basics/c15mi>

Under Ventilation

- Some patients eventually need ventilation assistance during the day to support adequate gas exchange.
 - If Oxygen saturation drops below 95% during the day and/or the Carbon Dioxide level has risen above 45mmHg, some daytime ventilation is needed.
 - *Symptoms: Increased fatigue and sometimes shortness of breath.*

Under Ventilation

- Mouthpiece Ventilation –
 - Volume ventilation by way of mouthpiece (MPV), or “sip” ventilation allows a user who can form a lipseal to sip small breaths as needed (process is sometimes known as breath-stacking).



Mouthpiece Ventilation

- Different sized angled mouth pieces or a straw can be used.
- It can be totally mobile and connected to your chair with or without gooseneck tubing.



Mouthpiece Ventilation

- Allows for 24/7 non-invasive ventilation and is less medical in appearance than going out in public with a nasal mask
- Reduces fatigue and can allow continued employment and participation in one’s hobbies and interests outside of the home
- Must haves for Mouthpiece Ventilation:
 - *Mobile ventilator with battery/batteries with volume ventilation; basic bi-level machine/BiPAP WON'T have this.*
 - *User must form a lipseal on mouthpiece or straw*

Benefits of NIV:

- Interrelated benefits include:
 - » maintaining and/or improving the oxygen/carbon dioxide levels in the blood;
 - » resting the respiratory muscles;
 - » decreasing the work of breathing;
 - » inflating the lungs more fully;
 - » improving sleep quality and breathing during sleep;
 - » preventing/avoiding hospitalization for respiratory complications;
 - » improving quality of life, and;
 - » improving survival.

How is it treated?

- Invasive ventilation:
 - » Ventilation through a tracheostomy tube and connected ventilator
 - » According to Dr. Bach, the only form of NMD that would ever require invasive ventilation is the most severe form of ALS when it progresses to involve severe bulbar muscle wasting.
 - » Requires surgery, recovery and daily maintenance and suctioning by a trained family member or professional
 - » Requires more “consumable” supplies
 - » Increased risk for infection (varies from patient to patient)
 - » Some do choose this method of ventilation due to feelings of claustrophobia with non-invasive masks and/or inability to get a good seal with mouthpieces for daytime ventilation.

How is it diagnosed?

- Pulmonary Function Testing (PFT):
 - » “Some important measures are *Forced Vital Capacity (FVC)*, *respiratory muscle strength* and *Cough Peak Flow (CPF)*.”
 - » FVC measures the amount of air expelled during a forceful exhalation after filling the lungs with as much air as possible.
 - » Respiratory muscle strength tests measure the maximal inspiratory or expiratory pressures that an individual can generate.
 - » CPF measures the strength of an individual’s cough.
 - » These measures provide most useful information when tracked over time for any individual, allowing better determination of the course of progression, and allowing anticipation of future difficulties.
- It is ideal to perform PFTs both sitting and lying down, although not all centers can readily accommodate this. The involvement of the diaphragm will be indicated in the difference in FVC between the sitting and supine positions.”

Reference: Cure CMD <http://curecmd.org/cmd-care/breathing-issues>

How is it diagnosed?

- Arterial Blood Gas (ABG) Sampling – Blood draw from an artery, not a vein
 - » Notorious for being inaccurate based on time of day and fear causing hyperventilation during arterial “stick”
- End Tidal CO₂ via Capnograph – A device measures the CO₂ in your exhaled breath
 - » Not readily available as it requires no medical expertise to use and cannot be billed to insurance at the rate of ABG
- Polysomnogram (sleep study)
 - » You may be told you have obstructive sleep apnea and that your MD is not the issue; this test is not designed to measure sleep breathing for the NMD patient.



BE PROACTIVE!

- Get a baseline PFT.
- Get follow-up testing at least annually.
- Listen to your body; don't delay reporting changes.

The best outcomes happen when we don't allow our function to reduce to the point where we have a failure event and end up in the ER. At that point, you may not be in control of the decision-making, and the healthcare professionals may not know how to treat your condition.

Closing

To know even one life has
breathed easier because
you have lived.

This is to have succeeded.

Ralph Waldo Emerson