When respiratory weakness from neuromuscular disease occurs, the muscles involved are those used for breathing, coughing and the bulbar innervated muscles we use for speaking, swallowing, and protecting our airways.

**The Case Against Tracheostomy:**

- Dr. Bach was careful to point out, "I know there are some adults in your group that do have trach tubes. And I know that you get used to it, and you do fine and have it for years, and that's okay. But the fact is, no one with a neuromuscular disease of any kind except for maybe someone with Lou Gehrig's Disease (ALS) ever needs a trach."

- He explained the spasticity in people with ALS can result in what is called “stridor,” and the airway is partially closed. When the airway is closed, secretions cannot be brought out, and that causes pneumonia. Trach becomes the only option for these patients to prevent pneumonia and death.

- ALS is the only disease that when the patient’s disease starts with the throat muscles (bulbar weakness), a trach is always needed to survive. The non-bulbar ALS patients can live many years without having to get a trach tube.

- Mean survival for ALS patients on trach ventilation is 6-7 years.

- Ninety percent of SMA Type 1 patients can use non invasive ventilation (NIV).

- Patients with trach tubes have as much bad bacteria colonization in their lungs and airway as people with pneumonia.

- Patients with trachs become more ventilator-dependent than those on non-invasive ventilation. They don’t need as much ventilation when the trach tube comes out. Bach explained that when you have a trach, the diaphragm withers away.

- Bleeding and mucus plugging which blocks the airways are the biggest risks two to three months after tracheostomy.

- In 17-65% of patients, trachs develop tracheal stenosis (trach perforates the airway).

- Most patients who survive the first six months after tracheostomy survive long-term.

- From 1983 to 1993, all Duchenne patients got trached; all died by age 29. From 1993 to today, a Bach-trained doctor in Japan has trached zero Duchenne patients, leaving their life expectancy to be 40 years.

- Thirty percent of trach users will aspirate food and liquids. Of those who have the trach tube removed, 7% will aspirate for the rest of their lives.
There are patients that survive 20 to 30 years on trach ventilation, but that's an exception.

**Weak or non-functional cough:**

- Bach told us "Often people with neuromuscular weakness do not need to use ventilators, but their cough is weak or non-functional."

It's not just because the expiratory muscles are weak. Bach gave an example of a patient with 800 milliliters of vital capacity who had normal ventilation and did not need assisted ventilation. He only had a very weak cough. As the vital capacity goes down, the cough gets weaker. Generally both inspiratory and expiratory muscles get weak at the same time, he explained.

- When peak cough flows go below 160 or so, every significant respiratory condition like a cold will cause bacteria to multiply, and the patient will go into pneumonia and respiratory failure unless he or she can cough out sputum.

- Anything below 300 milliliters per minute for cough flow puts you at risk from having an otherwise benign respiratory infection turn into pneumonia. This cough flow level qualifies patients for use of the Cough Assist to stimulate a larger cough.

**Surviving on Non-invasive Ventilation:**

- Many of Dr. Bach’s patients with Duchenne and Spinal Muscular Atrophy Type 1 are 24 hour ventilator dependent on non-invasive ventilation and don’t get sick.

- Dr. Bach showed a slide with a 48 year-old Duchenne patient who had lived many years on non-invasive ventilation but had a very low ejection fraction with his heart. It was suspected he died of sudden cardiac arrhythmia, based on the fact that he was left very briefly by a family member and then found to be gone with no signs of distress.

- Dr. Bach still has patients on non-invasive ventilation for 61 years with 24 hour ventilator support who have never been hospitalized and have never been sick with respiratory infections or complications.

**Intubation and Extubation to Non-invasive Ventilation:**

- For patients who get intubated because of a surgical procedure or get an acute infection and pneumonia, Dr. Bach’s team routinely extubates those who were considered unweanable at other hospitals.

  ICU doctors have ventilator weaning parameters, and if you pass these weaning parameters, they extubate you. Dr. Bach’s team does not wean; they extubate to non-invasive ventilatory support, not with CPAP or BiPAP but with a ventilator (which they have been doing for decades). They use a device called the Cough Assist to clear the airways during intubation to get the lungs healthy before they extubate.

These are available from Breathe with MD on the private, members only Breathe with MD Support Group on Facebook or by e-mailing breathewithmd@charter.net with the subject “extubation articles.”

- If you are ever in a situation where you or a loved one needs to be extubated to full ventilatory support, Dr. Bach is happy to talk to the doctors and shares that “If they follow what's in the articles, they will succeed.”

**Trach Tube Removal:**

- If you have a trach tube, Dr. Bach’s team can take it out. The criteria for removal is that you must have a flow of up to 120-160 liters per minute through the upper airway. He shared, “How this is done is we put the vent dependent person on mouthpiece ventilation, put our finger over the ostomy, take the tube out, pump them up with air, press on their belly and measure that flow. If that flow is at least 120 liters per minute, that patient doesn't need a trach tube, so it's removed.”

This can be done for both children and adults, but Bach’s team does not like taking trach tubes out of ventilator dependent children, because they are frightened and don't understand. They wait until they're old enough to cooperate, before they attempt removing the tube.

**Supplemental Oxygen, Sedatives and Narcotic Pain Medication:**

- Bach explained that if you don't have lung disease, you don't need supplemental oxygen. If your CO2 is elevated, you definitely should not be given supplemental O2.

- He continued that if your vital capacity is 70% of normal and you have no problem breathing while lying down, supplemental oxygen during a procedure won't be a problem. People who have low vital capacities and elevated CO2 would be ones that need to stay away from supplemental oxygen.

- Bach explained if you are intubated and they are controlling your breathing, it is okay to take supplemental oxygen; it's not going to stop you from breathing.
• Bach further explained that if you are not intubated and cannot breathe while lying flat, it means you have low vital capacity, your diaphragm is extremely weak and you would be in danger of oxygen causing your CO2 to rise, putting you at risk for a respiratory crisis.

• Dr. Bach’s team teaches patients to keep their oxygen saturation up without needing supplemental oxygen. He said, "No one with a neuromuscular disease should ever get oxygen at home unless they want to die."

• Dr. Bach explained that if a patient wants to die, Hospice can have their doctor order administration of morphine and supplemental oxygen. When combined, these two turn off the central nervous system, causing the carbon dioxide (CO2) level to rise, placing the patient into a coma and causing death.

• Supplemental oxygen causes the brain to think the body doesn't need to breathe because the ventilatory drive in the brain stem is suppressed.

• No one with a trach tube or on non-invasive ventilation needs oxygen. “All the oxygen is doing is like putting a band aid on cancer,” said Bach.

• No one ever goes below 95% oxygen saturation during the day unless there is a problem. If you have a weak cough and you go down to 94% that’s a crisis. In this case, there are only three things that could be happening.
  1.) Your CO2 is high.
  2.) You are not ventilated enough and/or the ventilator settings are wrong.
  3.) Airways are blocked by secretions, and if you don't clear them, pneumonia and lung collapse will occur.

• “If you give unventilated supplemental oxygen to people with neuromuscular weakness, they end up having respiratory failure quicker. You are better off doing nothing,” said Bach. “Supplemental Oxygen makes the oximeter useless.”

• Bach instructed that if you are taking oxygen and your saturation shows 99%, your carbon dioxide could still be very high (high CO2 is anything above 45). Your airways could be full of secretions to the point you could have a lung collapse, and the doctors would not know based on your high oxygen level.

• If a patient is on non-invasive ventilation (NIV) and taking sedatives, Bach explained that air will leak out of the mouth, CO2 will increase, and the NIV won't work.

• If narcotic pain meds are required while the patient is on non-invasive ventilation (i.e. for post surgical pain), Bach explained the patient has to have a closed method of non invasive ventilation. They can't use a nasal mask or a simple mouthpiece during sleep; they must have a full face mask to prevent too much mouth leakage, to keep air in and to prevent the CO2 from going up. If this isn't done, the patient can become comatose from being under-ventilated.
Secretion or Mucus Clearance:

- Dr. Bach stressed the need for stronger coughing to prevent mucous plugging, because mucous plugging causes lung collapse and pneumonia.

- Ninety percent of pneumonias will be in the left lung. Today’s Cough Assist by Philips Respironics suctions from both left and right lung and does so without damaging tissues.

- Cough Assist is basically a vacuum cleaner. It gives you a big volume of air, and the pressure drops to 50-60 cm/H2O, and airway secretions come out.

- History of Cough Assist:
  
  In 1952, the predecessor to Cough Assist, called the “Coughilator,” came on the market and saved all of the Polio patients in Los Angeles and New York from needing a trach tube.

  Bach explained that people who didn't know any better agreed to getting trached back then, and the Polio patients who did get trached did not survive. The Polio patients who chose non invasive ventilation (NIV) survived. He still knows some of them today. These patients kept their Coughilators since they knew when they got sick, if they didn't use it, they would get pneumonia and go into respiratory failure.

  In 1979, two of Dr. Bach’s patients put an article in the newspaper about getting the machine back on the market. *(It went off the market in 1967, because everyone was getting trached and getting suctioned through the trach. No one ever described using the device through a trach tube up until that point.)*

  Dr. Bach got five Coughilators from a respiratory products museum in Seattle, and his patients shared them when they got sick.

  In 1988, Dr. Bach convinced Jack Emerson to put the Coughilator back on the market.

- Dr. Bach says that the Cough Assist we have today should be used at 50-60 cmH2O in the upper airway or 60-70 cmH2O for patients with a trach. Bach explained that it doesn't work at all if pressures are set lower than that. *(These pressures are for both children and adults, and he’s working to revise and older article to explain this.)*

- Everyone with a weak cough needs an oximeter, because if you’re not clearing secretions and your oxygen saturation goes below 95%, your ineffective cough and the remaining bronchial secretions will cause pneumonia.

- For Dr. Bach’s adult patients in the New Jersey area, he doesn't prescribe the Cough Assist but instead tells them they have to get the device within an hour of developing a cough. *(He has a company that assists him with this rapid delivery of the device.) Use of Cough Assist within an hour of developing a cough prevents pneumonia. *(The challenge is many live in areas where rapid delivery of Cough Assist is not possible. Many companies want approval from insurance, taking on average, four days. Many doctors, including Bach, order the Cough Assist for any adult patient in this situation so that they have in the home, preventing delay during a time of need.)*
• The new Cough Assist model T70 allows for timing the inspiration and expiration breaths which helps for babies and kids.

• Bronchodilators are not recommended as a maintenance medication for secretion clearance since they can cause the heart to beat faster. Bronchodilators may help clear secretions when a patient is sick.

• Manually assisted coughing involves pumping the patient up with air and pressing on the belly for coughing.

• Some doctors prescribe a vest that shakes up the chest. Dr. Bach says this does not bring up mucus from deep in the lung for neuromuscular disease patients. “There is no evidence in medical literature that it works for us,” he said.

**Pulmonary Function Values:**

• Every NMD patient should know what their vital capacity is. To find out, you take as deep of a breath as possible and you blow it into a spirometer, and it tells you how many milliliters you are able to blow.

When babies cry, they measure from their cry (called a cry vital capacity). For a baby, it needs 50 milliliters to not need a ventilator. For an adult, you need 250 milliliters to not need to use a ventilator during the day. But if you drop below 200, you’re going to need it 24 hours a day.

If you have only 200 milliliters and you have a trach tube, and we take the trach to tube out, you will go from 24 hour ventilator dependent to only needing a vent at night. The vital capacity goes up when the trach is removed, because the trach causes airway secretions that block the respiratory exchange membrane. Once the vital capacity is over 250 milliliters, the patient transitions to only using a vent for sleep.

• When vital capacity goes below 40%, peak cough flows go down, Bach says.

**CPAP, BiPAP and Polysomnograms:**

• Physicians don’t separate out people do are too weak to breathe from people who have lung disease. This is part of why neuromuscular disease respiratory complications are incorrectly treated by CPAP and BiPAP.

• Bach explains that people who use by CPAP and BiPAP have also been sent for polysomnograms (sleep studies). He explains that polysomnograms are programmed to interpret every shallow breath as being due to the brain not telling the patient to breathe or the throat being obstructed (apneas). The test has never been programmed to say that the problem is a weak diaphragm and that what the patient needs is positive pressure for inhalation.
• Polysomnograms cost on average $5,000- $6,000, but Dr. Bach’s team does not typically do them. They have thousands of patients on noninvasive ventilation who were never sent for this test. Sometimes Bach’s team does a sleep study, if the patient doesn't really know if they are symptomatic or not. His study is with a sleep continuous CO2 taken via capnograph, and it's done in the home.

• Dr. Bach pointed out that sometimes it's helpful to get oxygen saturation and carbon dioxide while sleeping. He does this with some of his patients and in particular when insurance requires a sleep study to approve equipment.

• Bach told us that CPAP and BiPAP give positive pressure when you inhale and when you exhale. When you exhale, the air will stop you from exhaling. He explains this is of no benefit, and no one needs the expiratory pressure (which is part of the two continuous positive pressures in BiPAP).

• The only thing CPAP does is help keep the airway open so that you can use your muscles to breathe. But if you don't have muscles to breathe in air, the CPAP does no good. "CPAP is exactly like breathing with your head out the window with your car going 60 mph. That's not going to give you a deep breath; its going to put more air in your lungs, but it stops you from getting more air out," said Bach.

• Dr. Bach said, "A weak diaphragm is like this poor horse [showing slide of horse]. If you don't rest the horse, it collapses. The ventilator is like a pickup truck but only if you use it like a ventilator not like CPAP or BiPAP."

• Bach explained that 18 cmH20 is required to rest the muscles or ventilate the lungs, and anything less is useless. He said that most doctors prescribe BiPAP at 8 and 4 or 10 and 4, and if you're very strong, it will help you. However, it does not provide rest to the breathing muscles and won't prevent respiratory failure.

• Bach says a lot of patients who use BiPAP at night get sick, go into the hospital and then end up getting trached. He says if they had been on a ventilator with the appropriate settings, they would have never gotten sick, would have never gone to the hospital and would have never gotten a trach.

• Bach says the expiratory pressure should be turned off and further explained that on the Trilogy model ventilator, you could do this through Assist Control or CMV mode with Pressure Control or Volume control set at 1,000 milliliters or at a pressure of 27cmH20.

• To further explain why Dr. Bach advises against BiPAP, there is no exhalation valve on the common BiPAP masks. It's called a passive circuit, and you cannot or it's extremely difficult to use volume cycling or pressure support or turn off the expiratory pressure with a passive circuit.

• Alternatively, he says an active circuit has an exhalation valve, so when you exhale, air exits the valve. You generally need an active circuit to do mouthpiece ventilation and pressure control ventilation with no expiratory pressure. Bach warned that many companies have no experience with active circuit.

The Solution: Inhalation Only through a Ventilator:
• In summary, Bach says CPAP is useless, but BiPAP is not totally useless; it's just uncomfortable, requires use of an expiratory pressure and prevents air stacking. A ventilator is the preferred devices for non-invasive ventilation and provides the best outcomes, according to Dr. Bach.

• Bach’s team prescribes the “other half of BiPAP, just inhalation pressure” (through a ventilator). Bach explained that we need positive pressure to give the volume of air when you inhale. But if you get positive pressure that stops you from exhaling it negates the pressure you are inhaling.

• Dr. Bach cited an example of a boy with SMA who had been living for more than 25 years on 24 hour NIV without any respiratory failure. He described a wheelchair attached arm that holds a mouthpiece for the Philips Respironics Trilogy ventilator for daytime Mouthpiece Ventilation (MPV) use that includes a “kiss trigger” that only delivers air when you touch the mouthpiece. This keeps it from constantly blowing air in the patient’s face.

_Pulmonary Function Testing:_

• Bach told the audience "Pulmonary function testing is practically useless for people with muscle weakness; it's designed for people with lung disease."

• A peak flow meter (used by asthmatics) is used in Bach’s clinic to measure cough flows and assisted cough flows. You can get one online for approximately $15.

• Bach’s team uses spirometry to measure vital capacity; (normal is about 5,000 milliliters and reaches peak at about age 19 or 20). Men lose 1%/yr and women 1.2% for the rest of their lives. Vital capacity plateaus at a much earlier age in neuromuscular disease patients.

• Bach explained that with a spirometer, they ask patients to blow into it as hard as they can, and that shows their vital capacity. Used along with air stacking, they can determine maximum insufflation capacity (MIC).

• He said the amount of air your glottis can hold, minus your vital capacity is dependent on your glottis. If the glottis is not strong, you can't close the glottis to hold stacked breaths. By closing the glottis, you don't exhale.

If vital capacity goes down, they teach patients to air stack to reach maximum insufflation and keep their lungs stretched and pliable.

If you are able to close your glottis, you are also able to protect your airway. It also means your bulbar muscle function for speaking and swallowing is good.

_Oximetry for Measuring Oxygen Saturation:_

• After 1980, everyone started getting oximeters to measure oxygen saturation. Today, you can get one for $18 from OximetersForLess.com. *(Note: Many models have no price listed, so a call may be required to identify affordable models.)* You can buy an oximeter in a local department store but may pay up to three times more.
• An oximeter with an alarm is all you need. Nine-five percent or greater oxygen saturation is normal.

*Carbon Dioxide Testing:*

- Capnography through a capnograph samples the exhaled air out of the nose to read the Carbon Dioxide (CO2) level. Bach explained it is not used often, because Arterial Blood Gas sampling (ABG) brings more money to hospitals and clinics.

- Capnography is proven to be extremely accurate. In contrast, when patients get stuck from an ABG in an artery, Bach explained they hyperventilate because of the pain, and their CO2 goes down. This means the ABG is almost never accurate.

- If bicarbonate from a venous sample of blood (what is done in pre-admission testing for surgery), is normal, your CO2 should be normal. Forty-five or greater is abnormal for CO2.

*Three Main Treatment Goals Recommended:*

1). Maintain elasticity of the lungs through air stacking. Bach explained that just like we get contractures in our knees from sitting in a wheelchair all day, our lungs and chest wall muscles can get contractures, and it can impact the amount of air we get into our lungs.

   - Air stacking can be done with a $35 ambu bag, using a nasal interface or lip seal to inflate the lungs.

   - You can only air stack with volumes, not pressure. Bach explains that you cannot air stack with a BiPAP because it uses a set pressure; ventilators can give volumes of air.

2). Optimize cough flows.

3). Maintain ventilation.

   - Bach shared that "Nobody dies because they start nasal ventilation too late."

*Frog Breathing:*

- Dr. Bach explained that frogs do not have diaphragms but breathe by using their glottis. He said 70% of us should go at least 500 milliliters past our vital capacity with this technique also known as glossopharyngeal breathing. *(Glossopharyngeal breathing is a sports medicine technique use by swimmers and deep sea divers.)*

- Some patients with the neuromuscular weakness can frog breathe during the day so that they don't have to use their ventilator or need their mouthpiece ventilation less. Frog breathing will allow you to grab your mouthpiece only once or twice a minute versus four to five times a minute.

- Frog breathing can be done without any functioning muscles below the neck.

- Thirty percent of patients with Duchenne muscular dystrophy can learn frog breathing. Almost everyone else with muscular dystrophy can learn to frog breathe as long as they have good throat/swallowing muscles. Bach explained it is only helpful when you are very impaired.

*Dr. Bach’s App:*
• Dr. Bach is writing an app to help people find respiratory centers that will help prevent them from having respiratory failure and avoid any need to get a tracheostomy.

This app will have three areas in which to direct and guide people
1.) Long term management
2.) Extubation protocol
3.) Removal of trach tubes

Conclusion:
Towards the end of Bach’s presentation, based on a patient question, he recommended that ALL with a neuromuscular disease (including those with subtypes that don’t commonly cause respiratory complications), once a year have their end tidal CO2 checked by capnograph, purchase and use an affordable oximeter with alarm, have spirometry testing done to determine your vital capacity while seated and lying down, and to get your peak cough flow measured.

Bach re-iterated that some of the first common symptoms of under ventilation are morning headaches, fatigue, daytime sleepiness, decreased appetite and depression.

*Notes prepared by Andrea L. Klein of non-profit Breathe with MD who was in attendance at The Speak Foundation’s 2015 Conference. TSF and Breathe with MD are patient-run organizations. We are not medical experts and are not personally making recommendations. Please consult a medical professional.