Breathing Issues: In Depth Tutorial

Hypo=low Hyper= high or more

In the normal state, when we breathe in, we take in oxygen, the oxygen goes to the lungs and gets absorbed into the circulation, in exchange for carbon dioxide. Then, we exhale the carbon dioxide. One way to think of this is that you take in oxygen and you breathe out carbon dioxide.

There are two issues that can occur with breathing problems. One is a low oxygen problem (hyoxemia). The other is retention of carbon dioxide (hypercarbia). A low oxygen and a high carbon dioxide level (in lungs and blood) can sometimes be related. Pulmonologist talk about these two issues using the following terms:

1. you can have a problem with oxygenation AND/OR
2. you can have a problem with ventilation (ventilation means the process by which your lungs expand and by taking in the air you exchange the oxygen for carbon dioxide). Ventilation is the problem for people with neuromuscular disease. Ventilation problems lead to carbon dioxide build up.

Ventilation requires you to have the muscle support to expand your lungs during inhalation (your chest wall muscles assist in this effort as does the diaphragm. Scoliosis can impair chest expansion). Depending on how weak you are, you may or not be able to take an effective large enough breath. Because you are not taking a deep enough breath in, when you exhale you have a smaller amount of air to breathe out and it is not enough volume to get rid of the carbon dioxide. The carbon dioxide slowly starts to build up over time.

You might wonder at this point if someone with NMD is having trouble getting enough air in, why doesn't their oxygen level drop as their carbon dioxide level rises? Eventually this happens. The one thing that may explain the lack of initial hypoxemia (as measured by O2 Sat probe) with hypercarbia has to do with the oxygen dissociation curve. Basically, with the high partial pressure of oxygen in the lungs, you have a high oxygen saturation reading (as measured by the pulse oximeter). The dissociation curve is nearly flat for levels above 60. So, theoretically, given that the high oxygen partial pressure in the lungs remains constant – you are still able to load up the hemoglobin molecules sufficiently that you don't initially see a drop in oxygen saturation. However, with increasing hypercarbia the curve shifts to the RIGHT (Bohr effect) – a higher carbon dioxide leads to a lower blood pH (a measure of how acid the blood is) which means that oxygen drops off hemoglobin sooner. So, you could start seeing a lower oxygen saturation as the blood carbon dioxide level increases.

Two links that review the oxygen dissociation curve:

http://www.bio.davidson.edu/Courses/anphys/1999/Dickens/Oxygendissociation.htm

http://www.ventworld.com/resources/oxydisso/dissoc.html

The only other piece that may or may not be pertinent. You can also have CENTRAL hypoventilation – meaning that your airways/muscles are actually functioning perfectly but that you lack the central drive to ventilate. It is possible that certain patients who have more central nervous system (brain) involvement with any of the CMDs may have an additional central etiology of their hypoventilation.

Once again, people with neuromuscular disease (NMD) have problems with ventilation because:

1. they are too weak to expand the chest
2. their weakness limits how much air they can take in, because of the poor chest expansion
3. because they take in a smaller amount of air, they exhale a smaller amount of air and carbon dioxide levels start to build up in the lungs and blood.

People with NMD usually do not have a problem with oxygenation (except maybe at night or when they have become so weak that they not only have a problem with carbon dioxide build up, but start to drop their oxygen level as well. This is usually in the very END stages of someone with NMD). Low oxygen is usually do to a problem with the lung tissue, such as pneumonia or asthma. People with NMD can start to have problems with oxygen when they get mucous plugging because they cannot clear their sputum (or lung secretions) or when they get pneumonia. Mucous plugging and pneumonia affect the actual lung airways or lung tissue and prevent inhaled oxygen from getting through the lung tissue into the blood stream.

Another way to think of the oxygen versus the carbon dioxide issue is that a low oxygen results from problems getting the inhaled oxygen to the blood. Reviewing how we breathe, you take in air it goes into your trachea (windpipe) and into your bronchi (branches that take off from trachea) and into your alveoli (lung tissue, air filled sacs) and then get absorbed across the alveolar sacs into arteries near the alveoli. So any obstruction to oxygen flowing freely from the mouth to the arteries, such as airway narrowing (asthma), increased mucous build up (NMD-poor cough), or pneumonia (collapse of alveoli) results in low oxygen.

Carbon dioxide or problems with ventilation result from not being able to get rid of carbon dioxide. Let's review the path of carbon dioxide out of the body. Carbon dioxide is transported to the lungs as a byproduct of our cellular energy costs by way of veins. Carbon dioxide is absorbed across the veins into the alveoli, then out the bronchi, into the trachea and out the mouth. The major determinant of how much carbon dioxide you exhale depends on how much air you have in your lungs to exhale and how strong your muscles are to give a good exhalation. This is ventilation.

Problems with ventilation are worse at night than during the day, because all human beings have slightly shallower breaths and a slower rate they are breathing at night. Lying flat on the back may also make it harder for the chest wall to expand as opposed to sitting upright. This means that people with neuromuscular weakness, usually first experience problems with ventilation during the night (hypoventilation). When people sleep at night our oxygen level decreases a little as well, in part, because our tissues in the back of our throat relax and may obstruct our breathing somewhat. For people with NMD, their main problem is not oxygenation but hypoventilation. When they go to sleep they start to breathe more shallowly, as the night goes on their carbon dioxide level rises. Eventually, their oxygen may also fall. This is usually a late sign, but can be a clue to the underlying high carbon dioxide. Once again the primary problem is not oxygen but carbon dioxide which is treated not with oxygen but with a machine to increase how much air is getting into the lungs at night (bipap or ventilator).

Symptoms of hypoventilation (build up of carbon dioxide) are difficulty sleeping, morning headaches, fatigue, and when the process goes on for long time, alteration in mental status. These symptoms can be difficult to detect in a young child. Because most of the symptoms occur at night, it is important to looks for signs of hypoventilation at night, therefore a sleep study. However, that said, sleep studies are difficult to tolerate as you are simulating in a hospital environment what occurs naturally at home. Most people don't sleep during their sleep study. And, as you pointed out, putting a mask on a kid's face is the number one way to keep them awake and fighting. The easiest way to determine if there really is a problem, is to let the child fall asleep at home with a pulse ox on (this measures oxygen levels, which will fall somewhat during everyone's sleep, but will fall more in someone with NMD). The Medicare consensus guidelines used to argue for bipap in ALS, state that if the oxygen level consistently is below 88% for 5 consecutive minutes or more, there is a need for BIPAP.

The current ATS (American Thoracic Society) guideline for Duchenne (which is the closest we are to guidelines for CMD and practically speaking do apply to CMD) show that BIPAP is necessary when:

1. daytime hypercapnia is present (means doing a blood gas during the daytime, and finding an elevated carbon dioxide). This means the child was retaining carbon dioxide all night, and this effect spilled over and was not corrected during the daytime when everyone's respiratory rate has picked up somewhat. This means that they can no longer compensate for the carbon dioxide they are retaining at night.
2. however, they acknowledge that some children are so symptomatic during the night from their hypoventilation that they may require noninvasive ventilation for a good night of sleep, even though their daytime carbon dioxide numbers are normal.
3. if the FVC <50% or the maximum inspiratory pressure is <60cm H20 or the pulse ox drops to <88% for 5 consecutive minutes, then noninvasive ventilation is recommended. FVC and MIP can be difficult to measure in a kid, given that these tests are effort dependent.

In a study from England (), they looked at two parameters overnight to decide if there was hypoventilation and retention of carbon dioxide. One, they hooked up a pulse ox, two they used a transcutaneous carbon dioxide measuring system. This is a device like a sticker that you place on a body part after it has been calibrated and it reads the carbon dioxide level without needing to do an arterial blood gas. We had one of these in the ER where I used to work. It is somewhat of a newer device and would need to be carefully calibrated to be accurate. However this would mean that you would have two noninvasive means of measuring in a child or adult whether:

1. their oxygen drops
2. their carbon dioxide builds up when they sleep. If either happens, they would benefit from noninvasive ventilation overnight.

During the sleep study, a pulse ox is placed on the child to measure their oxygen level. This is cheap, easy to measure and is therefore used as a criteria of whether someone needs bipap even though it is measuring oxygenation and not ventilation (the primary problem in neuromuscular disease). Like all people, the person with neuromuscular disease may have a drop in oxygen when they sleep. Most pulmonologists base their assessment of whether someone needs bipap on Medicare guidelines that have been established for ALS patients: an elevated carbon dioxide on a blood gas >45, a low pulse ox or oxygen level <88% for 5 consecutive minutes, and a low FVC or MIP. FVC and MIP are pulmonary function tests that get tested in a lab, not at night. In people with muscle weakness, the numbers are usually low because you are measuring how much force they can generate when they inhale and they usually have trouble with this. These tests can be difficult to do in young children as they are effort dependent.

There is a way of measuring someone's carbon dioxide (mentioned above) without an arterial blood test by using a sensor on the skin or having them breathe into a portable monitor. I am not sure if all sleep labs currently have this capability. The only other way to detect an elevated carbon dioxide is by doing an

arterial blood test. Because it involves sticking a needle into an arm, the patient may hyperventilate prior to insertion making it a less accurate test unless they are very weak and don't compensate in this way.

The mask may be uncomfortable initially. Like all things you teach children, you need a patient instructor who can make it fun and accessible and gives the child control over wearing the mask.

Information courtesy of Cure CMD ← create link to www.curecmd.org