First you tell me I have lung disease….
Now you tell me I have Pulmonary Hypertension!

By John R. Goodman BS RRT

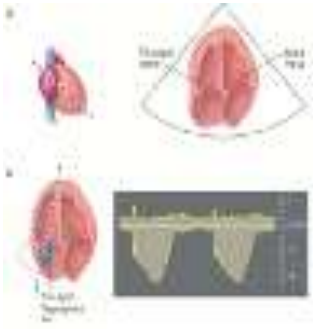
Pulmonary Hypertension (PH) may be defined as high blood pressure in the arteries that go *from* the heart *to* the lungs. It is interesting to note that the definition of an artery is a "blood vessel that carries blood away from the heart." Generally, arteries are high in oxygen content and the blood therein is bright red. Blood in the pulmonary "artery" however, is very low in oxygen, as it has just been pumped into the pulmonary artery by the right ventricle. The right ventricle receives all the venous blood from the lower extremities as well as venous blood draining in from the head neck, and shoulders. Venous blood returning from all parts of the body is a darker red color as the blood has released its oxygen to the billions of cells of the body. This is perfectly normal and is reflected in the normal saturation values associated with arterial and venous blood. If we consider a normal oxygen saturation (SaO_2) at sea level as right around 97%, most people would be shocked to learn that the "normal" saturation of venous blood (PvO_2) averages about 75%.

Unlike the left ventricle which must generate much higher pressures to pump oxygenated blood throughout the whole body, the right ventricle normally does not need to squeeze very hard to move the venous blood over to the lungs to pick up that all important oxygen. As an example we can use just the upper pressure measurements to give an idea of the difference between the right side of the heart (pulmonic) and the left side of the heart (systemic).

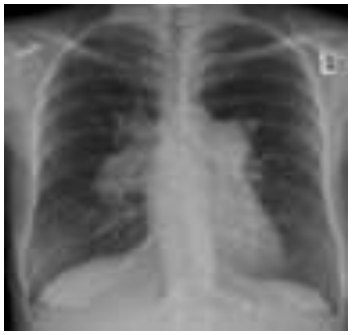
When a physician, or nurse takes your blood pressure with a blood pressure cuff, the result is normally reported out as (for example) 120/80. The top number is called the systolic pressure and in the normal adult it *does* average about 120 mm of pressure. (We will not worry about the bottom number at this time.) This is how much pressure or force is necessary to keep the blood circulating through the millions and millions of blood vessels (including capillaries) throughout the body. Since this is a reflection of the function of the left ventricle, we simplify things to say this represents left heart function.

Going over to the right ventricle, and remembering the right ventricle only has to pump blood over to, and though both lungs, it take much less pressure. Normal right ventricular pressure is reported as 25/15. So simple math shows us that normally, pressures on the left side of the heart are almost 5 times higher than on

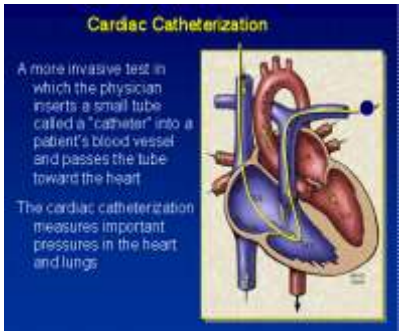
the right! (120 vs. 25) Unfortunately, there is no test as simple or as easy as taking your blood pressure that can measure your right sided heart pressures. Most commonly the information is obtained non-invasively though the use of an echo cardiogram, standard chest x-ray, or CAT scan. Positive confirmation is made by doing a catheterization of your right heart and measuring pressures directly. This is obviously an invasive procedure, and is normally performed in a cardiac catheterization lab specifically designed for this procedure.



Echocardiogram



CXR showing enlarged right heart.



Cardiac catheterization gives exact measurements of heart chamber pressures.

There are many, many **non-pulmonary** causes of Pulmonary Hypertension. If you have a specific disease or condition that you think may be related to your PH, I strongly encourage you to begin with a simple Google search. But, due to space limitations, I *must* limit our discussion to the known relationship between pulmonary disease and the development of PH. While I normally don't like to generalize the term "lung disease," it is possible for me to do just that due to the one common denominator of all chronic lung disease. That common denominator is chronically low levels of oxygen in the blood, also known as *hypoxemia*. But, more on this later. As a quick note, you may see the terms Pulmonary Hypertension (PH) and Pulmonary Arterial Hypertension (PAH) used interchangeably. To slightly add to the confusion, both PH and PAH, have been known in the past as "secondary pulmonary hypertension." That is, heart disease that is secondary to lung disease. For all intents and purposes this is just nomenclature ***unless*** you see the use of the following…***PPH***. PPH has been used for many years as an abbreviation for Primary Pulmonary Hypertension. This is a rare disorder (perhaps 4-6 cases per million) where the patient is born with the disease which may show up shortly after birth, or lay lurking in the shadows to pop up later in the patient's life. Since no one really knows what causes PPH, it is better known today as Idiopathic (unknown cause) pulmonary arterial hypertension (IPAH). Again, I must keep this discussion limited to PH that develops secondary to chronic lung disease.

Now that we have been introduced to the disease or condition known as PH, we can discuss how it develops. There is an old slogan in medicine that happens to be very, very true. That slogan is "the body in its infinite wisdom." What this means is that the human body has the marvelous ability to compensate for physiologic alterations that may be happening internally. A corollary to that statement might be…"the body never overcompensates." As I stated earlier, PH develops as a result of chronically low blood oxygen levels. Over the past 20 years other factors have been found that hasten the development of PH, or even make it worse.

Here is where the body in its infinite wisdom comes into play. If blood oxygen levels stay low enough…long enough…the body attempts to "compensate" for this deficiency. It does so by constricting blood flow though the pulmonary arterial system. Here is what the body is thinking. Okay, I've got less oxygen in the blood flowing through both the whole body and the lungs themselves. For whatever reason (pulmonary disease) there are less oxygen molecules being made available to the capillaries responsible for transporting that oxygen throughout the body. So the body compensates by constricting (actually shrinking) these tiny blood vessels so that there is a better matching of oxygen breathed in to the blood still circulating through the lungs. The body in effect says, "Well, there is less oxygen available to pick up, so let's do a better job at matching up those sections of the lungs that are still working pretty well, by re-routing blood flow preferentially to those units." Truly, this is an example of the "body in its infinite wisdom" at work.

From the body's perspective, this will have the effect of making sure the highest percentage of lung units will be functioning at their optimal best. Makes sense doesn't it? However, as we all know, there are no free lunches in life. In this case making things better for the lungs can make things worse for the heart. We start by asking an obvious question. What is the difference between a drinking straw and a garden hose?

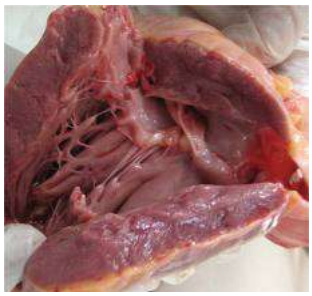
The answer to all of us is both simple and obvious. It is, of course, the diameter. You don't have to be a hydraulic engineer to figure out that it is a heckuva lot

easier to pump a fluid (like blood) through a series of garden hoses than it is through a bunch of drinking straws. This can be complicated by the fact that chronic hypoxemia can also cause the blood to become thicker than normal. In another example of the body compensating for a chronic condition, more red blood cells are released into the bloodstream in order to "deliver" more oxygen to the cells of the body. If this condition goes on long enough the viscosity of the blood goes from something like tomato juice to ketchup! This is reflected in your blood work as elevated Hemoglobin and Hematocrit levels.

Now is a good time to remember that the right side of the heart is the low pressure side. Since less pressure has to be generated, the muscular wall of the right ventricle is thinner and does not pump with as much force as the left ventricle. This means the right ventricle is much more subject to resistance downstream. Well, where is downstream from the right ventricle? That's right, the lungs. So first we have the constriction of the pulmonary arteries due to chronic hypoxemia, and then there may be the double whammy of having to pump thicker blood as well.

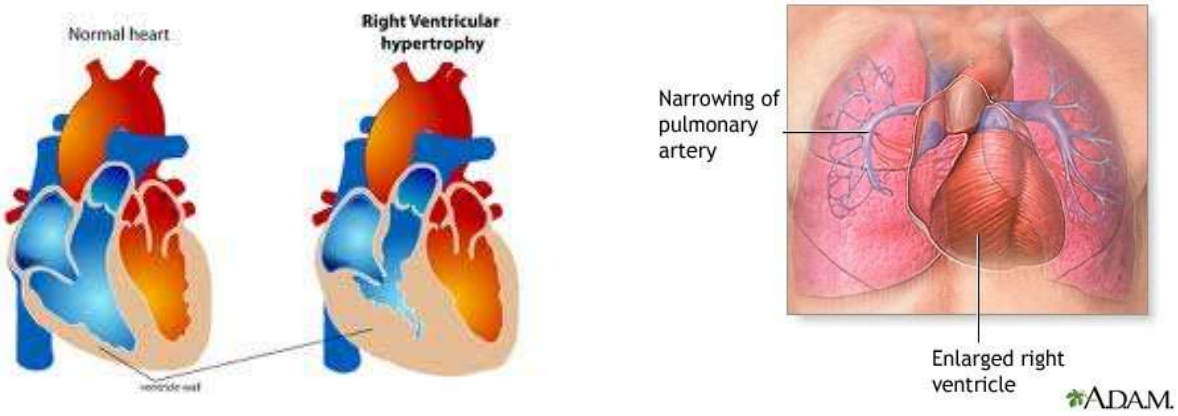
Even the body in its infinite wisdom has limits to how much it can compensate for a chronic condition. At first the muscular wall of the right ventricle tries to keep up with the rising pulmonary pressures. Like any muscle that is exercised, it actually gets larger and for a period of time, pumps with more force.

This picture illustrates just how enlarged the right ventricle can become. Normally the muscle wall is just 2-3 mm thick. Here we see it is 3-4 times thicker than normal.



The right side of the heart does make a valiant effort to keep up with the rising pressure. To quickly review, the normal pressure in the main pulmonary artery is

most commonly given as 25/15. There may be some quibbling as to the exact numbers, but overall we can use this figure.



If you look closely at these two illustrations you can see what the end result will predictably be as the narrowing of the pulmonary artery(s) continues, the right ventricle enlarges to a point where it starts to become dysfunctional. If this "back pressure" continues unabated, it can cause fluid to back up throughout the entire circulatory system. This puts further strains on both the left and right side of the heart, and you can see how a vicious cycle is created….and this cycle can lead to some very serious consequences….including heart failure.

Like many diseases, PH is classified according to the Pulmonary Artery Pressure (PAP) measurement as follows:

Mild PH = a PAP of 26-34 mm of pressure.

Moderate PH = a PAP of 35-44 mm of pressure.

Severe PH = a PAP of 45 mm or greater.

Common signs and symptoms of PH include:

Dyspnea (shortness of breath), both at rest and seen especially with exertion. This usually starts slowly and gets worse over time.

Dizziness. Perhaps suddenly passing out.

Lethargy or fatigue.

Chest pain. Cough. Swelling of the ankles or legs.

With this checklist of signs and symptoms, it is easy to see why your doctor must order a number of tests to either rule in PH, or rule PH out and some other condition in as the cause of the symptoms. Even then, your doctor must have a pretty high suspicion of PH in order to both make the correct diagnosis and not miss some other "sneaky" conditions such as pulmonary emboli (blood clots) in the lung, interstitial lung disease, certain forms of heart or heart valve disease, connective tissue disease, and even sleep disordered breathing.

Once all of the blood work, echocardiograms, radiologic testing, ECG's, and exercise testing is performed, your doctor will have a pretty good idea if you indeed have PH. These are all non-invasive tests that get a patient into the "ballpark." Your doctor may well want to be positively sure of their diagnosis by having you undergo a cardiac catheterization. This is an invasive procedure where a small catheter is inserted into one of your larger veins, and then advanced into the right side of your heart. Measurements made here are exact, and you can be quickly classified as mild, moderate, or severe.

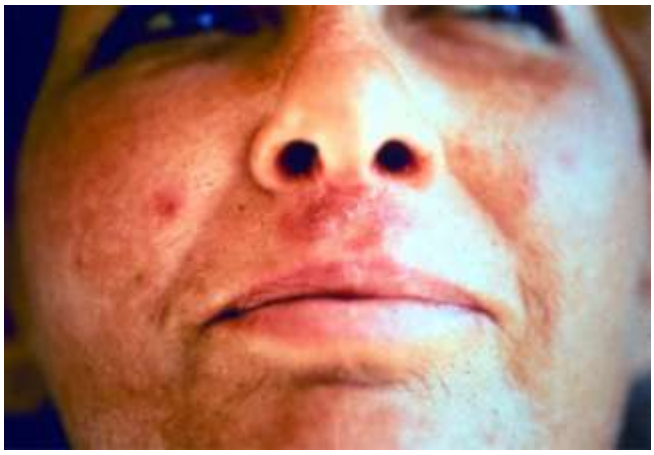
Your pressures will help dictate your treatment plan. For some patients, certain medications known to directly lower the pressure in the lungs may be given. For some patients blood thinners may be prescribed. In others, Calcium channel blocking agents may improve symptoms. Ultimately, for some patients with very severe PH a lung transplant may be the only hope.

But there IS hope…

The vast majority of PH in patients with COPD are in the mild to moderate category. Severe PH is seen in less than 5% of patients with COPD. Currently, there is no clinical test or examination finding that accurately identifies PH in patients with COPD. All of the treatment options mentioned above are used to treat a particular component of PH. There is ONE therapy however that has been shown to decrease, and even reverse the progression of PH in many patients. What is this therapy…it's as plain as the nose on your face…or at least the nasal cannula in that nose! Long term oxygen therapy works at reversing the very problem we described in the beginning of this article. Remember, the very small pulmonary arteries constrict due to the body compensating for low blood oxygen levels right? So by administering supplemental oxygen to patients with PH, we can begin to reverse the mechanism responsible for all that follows. Not only can oxygen therapy slow down or even reverse the progression of PH…oxygen has been shown to be the only drug (yes I said drug) ever scientifically proven to increase

survival! That means if you wear your oxygen as prescribed by your doctor, you will live longer than patients who either can't or won't wear their oxygen.
The CMMS estimates that there are roughly 1,200,000 patients who (should) use oxygen 24 hours a day at home. By far the most common device for administering oxygen is the nasal cannula. The nasal cannula is simple and inexpensive, but it is also very uncomfortable and inefficient.

This 26 yr old patient (authors neice) had PPH and received 2 different lung transplants in an 8 year period.



This patient would take her nasal cannula off due to extreme discomfort and constant irritation.

So in reality the *problem* with oxygen therapy is somewhat complex. We know beyond any shadow of a doubt about the survival benefits of oxygen therapy, especially when combined with an organized exercise program such as seen with Pulmonary Rehabilitation programs. Neuropyschiatric function is also improved with oxygen therapy. When I am teaching new patients about the benefits of oxygen therapy, I will usually tell them that oxygen itself is not life support…*but it sure as hell is supporting life!*

A quick review of where this little discusion has taken us is in order before we can move on. The story of PH goes something like this. Lung disease (or some other entity) causes a chronic low blood oxygen scenario (hypoxemia) to develop. Through a variety of compensatory mechanisms the body attempts to "fix" the

problem by constricting the small blood vessels in the lungs, and perhaps causing changes in the viscosity (thickness) of the blood by producing and releasing more red blood cells into the blood. The combined effect causes the right side of the heart to have to work harder and harder. If left untreated, this can lead to right ventricular hypertrophy (enlargement), and finally right heart failure.

Depending on the specific cause of the hypoxemia, a wide variety of drugs with different actions may be prescribed. Ultimately if the PH is servere and persistent enough, a lung tranplant may be the only treatment left to the patient. Of all the "drugs" that might be prescribed by the doctor, the most important is undoubtably oxygen. But, oxygen delivered via standard nasal cannula is difficult to wear on a 24/7 basis as ordered by the physician. A reasonable question therefore would be: Are there other delivery devices for oxygen therapy? And the answer is…. of *course* there are. Please remember the end-point we all seek as clinicians is true compliance with the oxygen prescription. The original plastic nasal cannula was patented in 1956, although some primitive cannulas were available as early as the 1930's. Oxygen masks have been available since before the turn of the century.


Standard nasal cannula


Nasal cannula pt.


Pt. wearing Oxy-arm

But this is 2011. Certainly oxyen delivery devices have improved and evolved over the past half century…..haven't they?? Well we know the NOSE hasn't changed in the past 55 years, and there are just so many ways you can skin a cat. Over this time period a pretty good number of new generation nasal cannulas have been introduced to the market. Different types of plastics, better anatomic designs, lighter weights, different colors and a number of devices such as the Oxy-arm have been introduced to patients in an attempt to improve comfort and compliance.

But the very bottom line is that with all the best efforts of the oxyen industry, compliance with oxygen via nasal cannula is still sub-optimal. Very classic studies on compliance have proven beyond a doubt that the vast majority of patients who are on oxygen 24/7 are only willing or able to wear their nasal cannula for about 18 hours per day. In effect, losing the benefits of their therapy for 6 or so hours per day. Is there a way to insure 24 hour per day compliance? Probably not with conventional non-invasive oxygen delivery devices. If however, you can make a

patient either more comfortable, or less self-conscious about going out in public wearing their oxygen, you should be able to improve compliance, and as an extra bonus, improve quality of life by getting pateints out of the house, and into the *mainstream* of life again.

We have already discussed the discomforts associated with the use of a nasal cannula. Much of this due is to the fact that the cannula must be worn with the prongs in the nose, and the tubing draped (lariat style) over the ears. Now imagine you also must wear glasses to see, read, and just get around in general. Wearing both glasses and a nasal cannula at the same time, is VERY uncomfortable for patients. In fact, we know many patients will take their nasal cannula off to give their nose and ears a rest.

A few years ago, an ingenious new method of delivering oxygen was developed that found a way to combine the dual necessities of *needing* to wear oxygen with *needing* to wear glasses. This product is called Oxy-View eyeglass wear. You can see from the following pictures that the frames are hollow. Oxygen up to 5 liters per minute can flow through the frame and into the nose via two, small, discreet prongs or "J-hooks." Connecting tubing is usually connected from behind, so it can be almost completely camouflaged.

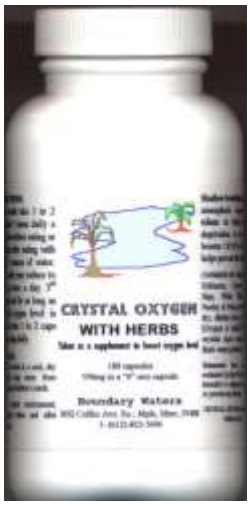
Oxy-View glasses allowed this patient the mobility she sought, significantly improving her quality of life.

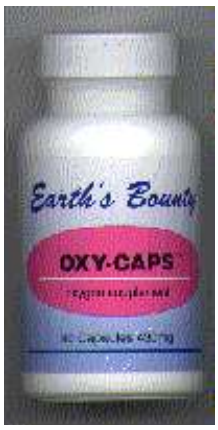
Oxy-View eyeglasses are available in a variety of colors and styles.

Finally, we can briefly discuss the MOST efficient method for oxygen delivery. This of course is transtracheal oxygen therapy or TTOT. TTOT involves the insertion of a very small flexible catheter directly into the windpipe or trachea.

There are many, many benefits associated with TTOT. You may want to talk to your pulmonolgist about TTOT to see if it may be a good option for you. You can certainly visit the website at www.tto2.com. Remember oxygen is a DRUG. It is almost certainly the most important drug you are taking. It will be one of the most important therapies your doctor employs to treat pulmonary hypertension should it develop.

Many bogus oxygen therapies can be found on line. Think not??? Think again…





This article was written to give you a better understanding of Pulmonary Hypertension and how it develops in patients with chronic lung disease. An educated patient is an empowered patient. Use the power wisely.