ACHA Adult Congenital Heart Association

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ACHA Q and A: Pulmonary Hypertension

What is pulmonary hypertension?
If you have pulmonary hypertension (PH), it means that the blood pressure in your lungs is higher than normal. The heart has two sides that send blood in two directions: into the lungs and into the body. When your caregiver uses a blood pressure cuff on your arm, s/he is measuring the pressure of blood in your body. Blood pressure is always measured at two points: when the heart beats (systole) and when the heart relaxes (diastole). Normal blood pressure in the body should be less than 120mm/80mm. In the lungs, the pressure should be less than 40mm/18 mm, or under 25mm overall. If the pressure in the lungs is higher than 25mm, you have PH.

How do you measure lung pressure?
The most direct way to measure lung pressure is by catheterization. A small tube is placed in your heart and a direct pressure measurement is taken. A less invasive way to measure lung pressure is through an ultrasound of the heart (echocardiogram or echo). Through echo, your caregiver can measure the speed of your blood flow throughout the heart. It also allows your caregiver to determine whether you have any narrowing or thickening which would increase resistance. This allows him/her to estimate your lung pressure.

What causes pulmonary hypertension?
Science tells us that pressure equals flow times resistance. If you have ever played with a hose, you know that there are two ways to increase pressure. You can turn the spigot and increase the amount of water. You can also bend or block the hose and make the water flow through a smaller opening. PH can be caused by problems with too much blood flow to the heart. It can also be caused by narrowing or stiffening in the heart and blood vessels, which increases resistance.

Can congenital heart disease (CHD) cause PH?
There are many problems in the heart that can cause PH. If you have a connection (shunt) in your heart that creates extra blood flow to the lungs, this can cause PH. The most common defects that cause PH are holes in the heart such as atrial septal defects (ASD), ventricular septal defects (VSD), and patent ductus arteriosus (PDA). Other defects that can cause PH include truncus arteriosus, double-outlet right ventricle (DORV) and any defect in which there is one working ventricle.

If the filling pressure on the left side of your heart gets too high, it can also cause PH. The pressure can rise due to a thickening of the heart's wall. Blood flows into your ventricle like air flows into a balloon. You might have noticed that a thin, stretched balloon is easier to blow up than a thick, stiff balloon. If your heart wall gets thickened and/or stiffened, resistance increases. This can also cause your blood to get backed up in the lungs and cause PH.

What are the other causes of PH?
Beyond the heart, there are many problems that can cause or worsen PH. These include problems in the lungs and problems elsewhere in the body.

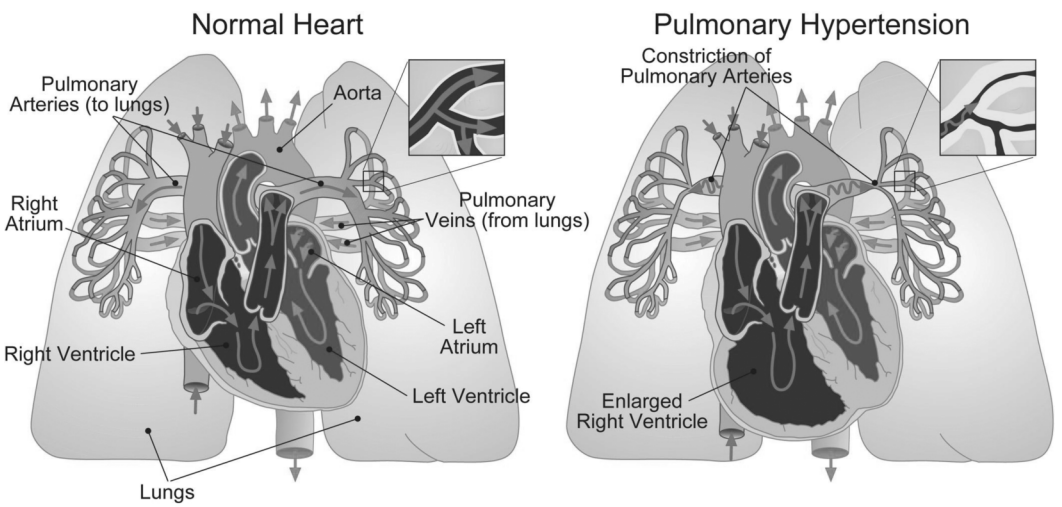

Image courtesy of the Columbus Ohio Adult Congenital Heart Disease Program at Nationwide Children's Hospital Heart Center, Columbus, Ohio

Problems in the lungs: There are many lung issues common in CHD that can cause or worsen PH. Because heart and lung development are connected, CHD patients can have inborn problems in their lung structures. Your lungs may not have developed enough oxygen-processing areas. Certain heart defects can cause the lung's blood vessels to become blocked or compressed. Your lung tissue may also be damaged by scarring from heart surgeries. If your CHD caused spine or ribcage problems, your lungs may not be able to expand normally. If your heart is too big, this can also compress your lungs. CHD patients can sometimes develop blood clots in the lungs. All of these things can decrease your heart's ability to process oxygen. When this happens, your lung pressure can increase as your heart tries to increase the body's oxygen supply.

If you have congenital heart disease and pulmonary hypertension, it is important that your caregiver look beyond your heart and check for all possible causes.

In rare cases, PH starts with problems in the lung cells themselves. Sometimes lung cells become inflamed for no known reason. These changes lead to a decrease in oxygen uptake and an increase in pressure. This form of PH is called "idiopathic" PH, meaning that there is no known cause. Most people with CHD develop "secondary" PH. This means that their lung problems were caused by ("secondary" to) their heart problems and/or another problem in their body.

Problems outside the heart and lungs: When we look beyond the heart and lungs, the most common cause of PH is obesity. If you are obese, your heart has to work much harder to pump your blood throughout your body. This can cause the heart to thicken and PH to develop. In some people, losing weight and controlling blood pressure can resolve or improve their PH. If you have PH, it is especially important that you maintain a heart-healthy weight and habits.

If you have a health problem that causes your blood pressure to rise throughout your body, this can also cause PH. These include thyroid problems, immune system problems, infections, anemia, parasites, liver disease, and severe vitamin B deficiency. Certain drugs, such as amphetamines, can make lung pressure rise. Some people also have a metabolism that tends toward higher blood pressure.

If you have CHD and PH, it is important that your caregiver look beyond your heart and check for all possible causes. Many CHD patients find that there are several factors involved in their lung problems.

What are the treatments for PH?
PH is like a fever: it is a sign that there is problem somewhere that is causing lung pressure to rise. The first step in treatment is making sure that the cause of the rise is found. Different kinds of causes require different kinds of treatment.

Heart structure problems: If a heart structure problem is causing your PH, your caregivers may be able to correct it. For example, lung pressure might be lowered by closing a hole and/or opening blockages that increase pressure. These interventions might happen through surgery or through catheter-based interventions. The same procedures that are used to correct blood flow through the heart can often lower lung pressure. If you had previous heart repair, and you now have PH, it is very important that an ACHD specialty center assess your anatomy and determine if additional heart repair is needed.

Lung damage problems: When heart defects cause high pressure in the lungs over a long time period, it can result in permanent damage. The lungs can become scarred and inflamed, leaving the cells less able to transport oxygen efficiently. Once this happens, PH will continue even if the heart is repaired.

Medication therapies: There are many new medications now that can improve lung function in patients who have permanent PH caused by their CHD. There are four main types of medications that work to increase the lung cell's ability to provide oxygen to your body:

- Calcium channel blockers (nifedipine, amlodipine)
- Endothelin receptor antagonists (bosentan, ambrisentan)
- Phosphodiesterase inhibitors (sildenafil, tadalafil)
- Prostacyclins (Flolan, remodulin, iloprost, tyvaso)

Different medications affect different cell functions. All medications prescribed for PH require careful monitoring by experts in CHD and PH. While these medications do not cure PH, they can significantly improve health and well-being. Medication therapies are often combined for best results, and many new drugs are being developed to help patients with PH.

The good news about PH and CHD is that in the last 10 years there has been a big increase in both knowledge of the problem and effective PH treatments. If you are an ACHD patient and have been told that there are no treatments for your PH, we encourage you to seek care at an ACHD program offering PH expertise. This way you can ensure you learn about any new options available to help you feel better and live longer.

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