

Texas Children's Hospital Pulmonary Hypertension Center

Frequently asked questions about pediatric pulmonary hypertension

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What causes PH?

The normal person has a vast unused pulmonary circulation at rest. When a healthy person is sitting or resting, cardiac output is low and the pressures in the pulmonary circulation are quite low – in the order of 25/10 compared with 120/80 in the systemic circulation which is measured on the arm with a blood pressure cuff. When a healthy child or teen exercises, cardiac output can increase five to 10 times. Normally, the pulmonary artery pressures do not change at all, which allows the right ventricle not to have to work any harder than normal. This can only happen when unused pulmonary blood vessels are “recruited” and opened up.

In PH, the total amount of blood vessel area is markedly diminished and therefore, the right ventricle has to work much harder and at higher pressure to push the blood through the lungs. With exercise, a crisis can occur since there is likely in most patients with PH limited additional blood vessels to be recruited. This can cause a significant rise in the right ventricle. As the left ventricle increases the blood squeezed out of the heart, there is not enough blood coming through the lungs and the left ventricle becomes inadequately filled up. That is when individuals can get dizzy and faint.

Most adults with PH have idiopathic or primary pulmonary hypertension. A minority of children have this form of disease. More children are likely to have incompletely developed lungs without a fully developed pulmonary arterial system. This developmental problem is seen in children born with congenital diaphragmatic hernia and omphalocele as well as some children with congenital heart disease and children with certain genetic abnormalities. The single largest group

of patients in the Texas Children's Hospital Pulmonary Hypertension Clinic are infants, children and adolescents with Down syndrome.

What tests are done to diagnosis PH?

The most common tests are the electrocardiogram (EKG), the echocardiogram and the chest x-ray. At Texas Children's, we are making increasing use of a blood test called BNP or brain natriuretic peptide, which increases in value with strain within the heart muscle. Elevated BNP is an indication of strain on the heart and is NOT specific for pulmonary hypertension.

My doctor originally thought my child had asthma instead of PH. Is this common?

Because difficulty breathing with exercise is such a common symptom at the onset of PH and because asthma is such a common disease, it is not rare for PH patients to be labelled with asthma before PH is diagnosed. Fortunately, asthma medications do not complicate PH. In addition, many children with PH have evidence of what we call bronchial reactivity when pulmonary function testing is administered. Some of these children indeed are treated with asthma medications. However, very few have wheezing attacks as do most people with asthma.

What is the difference between PH, PAH and PPH?

PH is short for pulmonary hypertension. PAH is short for the most common kind of pulmonary hypertension called pulmonary arterial hypertension. PPH was the term used for PAH of unknown cause and stands for primary pulmonary hypertension. The World Health Organization reformulated names and recommends the use of the term idiopathic PH instead of PPH. Idiopathic means of unknown cause.

Should my child be seen by a pulmonary hypertension (PH) specialist if he or she already is under the care of a cardiologist or pulmonologist?

Either regular or periodic consultation with a PH specialist is mandatory, especially considering the increasing choices of therapeutic options, for individuals who live at some distance from the Houston area. For local patients, a long-term relationship with our PH specialists is what we recommend. Our PH team is ready to assist children, their families and physicians in their home communities.

As you'll come to see, pulmonary hypertension is an uncommon and severe disease. In the past few years, the number of medications available to treat PH has mushroomed to at least 6 medications in three different drug classes. There is no way that a general physician or most cardiology and pulmonology physicians can keep up with this rapidly changing field. Therefore, whenever possible, consulting with a specialist in the field is in the patient's best interests.

How dangerous is cardiac catheterization?

With Texas Children's team of specialized cardiologists and trained anesthesiologists, a right heart catheterization is quite safe in most patients and has a very low mortality (under 1%).

The prominent exception to this rule pertains to infants with well defined predisposing conditions such as extreme prematurity, Down syndrome or other genetic syndromes. Only those infants with relatively mild PH who respond well to therapy can be safely managed without catheterization.

We believe that a cardiac catheterization should be done in more than 95 percent of patients when they are newly diagnosed with PH. Cardiac catheterization allows our team to determine the severity of each patient's PH and which type of treatment may be best for the patient.

From what I've read on the Internet, PH is a fatal disease. Our family physician also has been very pessimistic about the future. Is there no hope for a child with pulmonary hypertension?

Our team believes there is always room for optimism in regards to pulmonary hypertension, especially for those patients who have not been on a trial of therapy.

At Texas Children's, we have PH patients on single-agent therapy while others are on a combination therapy using two or three agents who are reacting positively to the therapy. In addition, lung transplant may be an option for some patients because of the co-existence of our Pulmonary Hypertension Clinic and Lung Transplant Program.

Does pulmonary hypertension run in families?

Most patients with idiopathic PH do not have a clear family history of PH. However, a significant minority do.

Investigators at Vanderbilt University have been studying familial PH for decades. There is a test for the most common genetic mutation leading to PH that will be performed in selected idiopathic PH patients.

TREATMENT

When should my child start treatment after being diagnosed with mild PH?

There isn't a definitive answer to this question. We do know that individuals do not have symptoms or signs of PH until most of the pulmonary circulation is narrowed and compromised. As more clinical experience is collated by national and international patient registries, with which we enroll our patients, we will be able to answer this question.

Is oxygen supplementation therapy always needed for children with PH?

Most individuals with PH have normal or near normal lung function and do not benefit from routine oxygen supplementation therapy. If a patient has low oxygen tension inside the lung related to lung disease or sleep apnea, which can cause elevation in pulmonary artery pressures, then oxygen supplementation therapy may be recommended.

What are the pros and cons of intravenous epoprostenol (Flolan®) or treprostinil (Remodulin®)?

Flolan has been commercially available for 15 years and is the best studied and probably most potent PH therapy. It does require surgical placement of a tunneled (buried through the skin tissues) intravenous catheter (Broviac or Hickman) so that the medication can be given 24/7.

Flolan is very short acting so that a problem with the catheter requires an urgent transport to a local emergency department for resumption of the medication via a peripheral intravenous catheter. Catheters can break or an infection can develop. Infection will lead to hospitalization, antibiotic treatment, removal of the catheter and then surgical re-implantation of another catheter. There is more published experience with Flolan than any other PH therapy. Flolan has been mixed at home daily and kept cool.

Remodulin is similar chemically to Flolan but is longer acting. For that reason, it is not as urgent

to get to an emergency room if and when there is a catheter problem. It is approved for continuous infusion at a very low rate through a needle placed under the skin or via a Broviac or Hickman catheter. Remodulin comes from the national pharmacy in pre-mixed cassettes that usually last 48 hours. It does not need refrigeration. Its side-effect profile is similar to Flolan.

Patients with Broviac or Hickman catheters must bathe with caution – usually without a traditional shower – and cannot go swimming. Many individuals adapt to this therapy well while others find the burden of a continuous IV medication overly burdensome.

Will my child's weight affect his PH?

The heart has to pump blood to the entire body. The greater the body mass, the more blood and the harder the heart must pump, especially with exercise. For PH patients with excessive weight, weight loss is highly desirable.

In addition, excess weight is a risk factor for obstructive sleep apnea (OSA). During episodes of apnea during sleep, blood oxygen levels can drop below 85 percent saturation, thereby raising the pulmonary artery pressure for periods during the night, that, if prolonged enough, can worsen PH. Testing and treatment for OSA in any overweight patient with PH is strongly recommended.

How will we know if our child is improving because of PH therapy?

Close monitoring of PH patients allows us to evaluate the effectiveness of therapy. At Texas Children's, PH patients are usually seen in our clinic every three months and the patient's status is always discussed with parents.

We believe that physical examination, six-minute walk tests, blood tests and periodic echocardiograms are the most effective methods for assessing clinical status. In addition, many of our patients undergo repeat cardiac catheterization as the most accurate measure of pulmonary artery pressure. The frequency of cardiac catheterization is determined on a case-by-case basis but may be as frequent as annually or as infrequent as every five years.

Why would a child on PH medications ever need a lung transplant?

Our current PH medications generally slow down the progression of PH. PH is usually a progressive disease. Thus, if a patient is diagnosed late in the course of therapy or has a poor response to treatment, worsening strain on the heart can put an individual's life at risk. For most patients, PH remains a fatal disease – usually in the long run for most. The only therapy that cures PH in 2008 is lung transplantation. With the removal of the patient's lungs including his or her pulmonary arteries, the new lungs provide a low pressure circulation that immediately relieves the strain on the heart. The immediate post-transplant period is a critical time since the surgery and cardiopulmonary bypass can add stressors to already over-loaded heart. Our experience at Texas Children's Hospital is encouraging in that all of our PH lung transplant recipients have survived surgery and are alive and well from 1 to 5 years after lung transplantation. On the other hand, there are new problems and vulnerabilities after lung transplantation with concerns about possible rejection of the foreign tissue and increased infections due to the daily strong medications that all transplant recipients must take.