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Can echo diagnosis be wrong?

Yes. The echo estimates the pulmonary artery pressure, and can either underestimate or overestimate the true pressure. The gold standard is the right heart catheterization. However when echos are done carefully by experience echo cardiographers the pressures estimated by echo usually correlate fairly closely to those obtained during cardiac catheterization. Echos can also provide some additional useful information regarding size and function of the right ventricle, severity of leakage of the tricuspid valve, and extent of distension of the inferior vena cava (large vein in the abdomen that returns blood to the heart).

Does Idiopathic Pulmonary Arterial Hypertension (IPAH) as opposed to PAH associated with other diseases (APAH) have better prognosis/life expectancy?

The prognosis in APAH depends on the specific cause and the severity. Some studies suggest that APAH due to collagen vascular diseases like scleroderma has a worse prognosis than IPAH, but this depends on severity when it is detected. PAH due to congenital heart disease tends to have a better prognosis than idiopathic IPAH. Portopulmonary hypertension and HIV-related PAH have prognoses similar to that of idiopathic. Although not technically an “associated” PAH because it is classified in a

different group, PH due to chronic pulmonary embolism (blood clots in the lungs), known as Chronic Thromboembolic PH (CTEPH) has an excellent prognosis when the clots can be removed surgically.

Explain cardiac output measurements.

Cardiac output is the amount of blood the heart pumps in one minute. In a healthy individual this amounts to about 4-6 liters per minute. It is also expressed as cardiac index, which corrects the output for body surface area and is reported in liters per minute per meter squared. A cardiac index of less than 2.0 liters per minute per meter squared is recognized as a risk factor for less favorable outcome if not improved with appropriate PAH therapy. The most common way this measurement is made is by placing a catheter with a temperature-sensitive tip in the pulmonary artery (right heart catheterization) and measuring the change in temperature of the blood during the injection of a small (1/3 oz.) amount of cold sterile water into a central vein. This measurement can be inaccurate if the tricuspid valve is very leaky; in which case measuring the output by the Fick method (done by measuring oxygen saturation in the pulmonary artery and a peripheral artery and measuring oxygen consumption with a special analyzer and mask placed on the face) is another option.

My echo shows pressures of 50, but the right heart cath showed normal. Could I still have PAH?

The right heart catheterization is the most accurate way to measure pulmonary artery pressure. However, an isolated measurement by either method may not give a true representation of the pressure over a 24 hour period during a variety of activities since pressure may fluctuate. Measuring pressures during exercise can sometimes be useful if there is concern about the possibility of abnormal rise in pressure with activity, but interpreting results of such testing remains a matter of some debate. In some situations repeating a right heart catheterization following a year can be useful to double check on the pressures if uncertainty about the diagnosis persists.

I was just diagnosed and am scared. How long do I have to live?

No physician has a crystal ball and therefore can not accurately predict how long you will live. Pulmonary hypertension is a serious disease that can shorten life expectancy and as with all medical conditions, prognosis also depends on the severity of the problem when it is diagnosed. The severity of pulmonary hypertension found at the time of initial diagnosis has a wide range, and how a given patient responds to treatment can greatly alter the prognosis. There are effective treatments that prolong life and improve symptoms. New treatments are currently under evaluation and may further improve the outlook for this disease. Many patients with PAH live for many years; developing coping strategies that help a person to live well despite their PAH is an important aspect of moving forward.

If I have PAH associated with another disease, should I still see a PH specialist?

Many of the treatments for PAH are effective both in idiopathic PAH and PAH associated with other diseases. Therefore patients often receive optimal benefit when treated by physicians with experience with these treatments.

Could I have PPH which is mistakenly diagnosed as asthma?

The symptoms of PAH such as shortness of breath are largely non-specific, i.e. they can be associated with a number of lung and heart diseases. If there is a question regarding whether you could have PAH, you should discuss additional testing with your doctor. Echocardiography can be helpful as an additional step in looking for the possibility of pulmonary hypertension.

Does PH run in families?

Yes pulmonary hypertension runs in families. Approximately 6-10% of cases of primary pulmonary hypertension are familial. Therefore whenever a patient is diagnosed with PPH all first order relatives (siblings, children and parents) should be screened for the disease. The best screening test is an echocardiogram.

Is my doctor right when he tells me I would get better if I lost weight?

If you are overweight, losing weight is likely to make you feel better whatever the cause of your symptoms. So the recommendation to lose weight is almost always a good one. One can not comment on the cause of your symptoms without knowing all the details of your case. In the earlier stages of pulmonary hypertension, the symptoms can be non-specific and the findings on physical examination subtle. Therefore in some cases they may be attributed to anxiety or depression. Obesity itself may contribute to elevated pulmonary pressures, so it is to one's benefit to achieve optimum weight.

My echo shows pressures of 50-60, but the doctor says I do not have PH, can they be wrong?

The echo measurement is only an approximation of pulmonary artery pressure, however if correct an echo measurement estimating a pulmonary artery pressure of 50-60 mm Hg is far above normal. Therefore further evaluation is probably appropriate.

Why is it important for me to see a PH specialist?

Pulmonary hypertension is a very uncommon but serious disease and most generalists have little training or experience in treating the disease.

What causes PH?

Pulmonary hypertension is caused by certain forms of congenital heart disease, lung disease and blood clots in the lung arteries. It is also associated with collagen vascular disease, portal hypertension (usually caused by liver disease), diet drugs,

HIV infection and some other rare diseases. In some cases no cause can be identified and these cases are called primary pulmonary hypertension (PPH).

What is the difference between mean PA pressure and PA pressures measured by an echo?

The mean PA pressure is an average of the pulmonary pressure during one heart cycle. See response 14. The echo estimates the systolic PA pressure which is the highest pressure measured during the heart cycle. Therefore systolic PA pressure is always higher than the mean PA pressure. The systolic pressure is often 30 - 50% higher than the mean PA pressure.

What is the mean pulmonary artery pressure and how do you get the number?

Pressure in the pulmonary (lung) arteries, like blood pressure in the body arteries rises when the heart "beats" and ejects blood into the pulmonary artery and then falls during the period of time before the next beat. Therefore the pressure is always changing. The mean pulmonary artery pressure is the arithmetic average of the pressure during one cardiac cycle which is the time from the beginning of one heart beat until the beginning of the next. However, it is not as simple as taking the top number and bottom number and taking the average of the two. It is a continuous average of the pressure throughout one cardiac cycle. The mean pressure is always closer to the bottom number than the top.

What is the life expectancy with PAH?

There is no simple answer to this question. Life expectancy depends on many factors. Without knowing the specifics of each case it is impossible to even hazard a guess. Even when the specifics are known, survival is very hard to predict. Even though patients with advanced pulmonary hypertension may have a substantially shortened life-expectancy, current treatments appear to help most patients live longer with significant improvements in function and quality of life.

What tests are done to determine PH?

The gold standard test for diagnosing PH is a right heart catheterization which directly measures the pressures in the pulmonary arteries. This is recommended for everyone prior to receiving treatment for pulmonary hypertension. Prior to performing this test a number of non-invasive tests may suggest the diagnosis, including the electrocardiogram, chest x-ray and echocardiogram.

What tests are done to determine PAH over other causes of elevated pulmonary pressures?

Testing to establish the diagnosis of PAH typically involves 3 broad considerations:

1. Unequivocally establishing the level and amount of abnormal pressures in the lung.

2. Evaluating patients for other causes of increased pulmonary pressures that could account for the current clinical picture.
3. Evaluating patients for systemic conditions that are associated with PAH but may be subtle or unsuspected.

These tests usually include a thorough evaluation of the function of the heart and lungs as well as blood testing. If these studies identify no alternate cardiovascular or pulmonary disease that could account for the pulmonary hypertension, the diagnosis is PAH.

When should I start treatment if I have been diagnosed with mild PH?

This is a difficult question to answer without knowing all of the facts in your case, but in general most cases of pulmonary hypertension receive some form of treatment. In mild cases it is usually a pill form of pulmonary hypertension drug and some doctors will add a blood thinner, as well. Physicians still do not have a clear picture of what happens to someone with "mild" pulmonary hypertension, so careful follow-up is required. A number of research trials are currently trying to help answer this question.

Why do I have chest pains with PPH?

Chest pain is a fairly common symptom in patients with PPH, occurring in at least 1/3 of patients. The cause is unknown, although many experts in the field suspect it is because the muscle wall of the right-sided pumping chamber of the heart, the right ventricle, is not getting enough blood supply to meet the needs of the increased work it has to do because of the high pulmonary artery pressure. Some also believe that stretching of the pulmonary blood vessels in response to high pressures can cause pain.

My doctor is either a cardiologist or a pulmonologist. Why do I need to see a pulmonary hypertension specialist?

They are the proper specialists. However some of these specialists have very little training or experience in treating patients with pulmonary hypertension because it is an uncommon disease. If a heart or lung specialist has little experience in treating this disease, it is often helpful to have the patient consult with an experienced specialist. The pulmonary hypertension specialist can communicate with the local specialist to insure that the patient receives the best possible care.

What medications are most commonly used for treatment?

Currently, 5 drugs are specifically approved by the US Food and Drug Administration specifically for the treatment of PAH. They include (alphabetically): ambrisentan, bosentan, epoprostenol sodium, iloprost, sildenafil and treprostinil. Additionally, calcium channel antagonists may be useful in a small group of patients with PAH.

Diuretics, digoxin and oxygen are also helpful in advanced cases. Most patients are also treated with the anticoagulant, warfarin. Several research protocols exist to

assess the efficacy of new agents in the treatment of PAH at a number of PAH specialty centers.