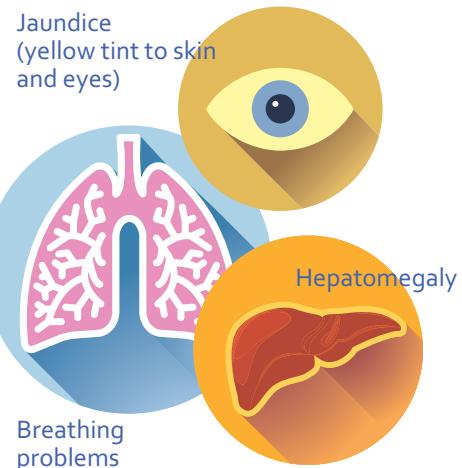


PATIENT EDUCATION | INFORMATION SERIES

Pulmonary Vascular Complications of Liver Disease

People who have advanced liver disease can have complications that affect the heart and lungs. It is not unusual for a person with severe liver disease to have shortness of breath. Breathing problems can occur because the person can't take as big a breath due to large amounts of ascites (fluid in the abdomen) or pleural effusions (fluid build-up between the tissues that line the lung and chest) or a very large spleen and liver that pushes the diaphragm up. Breathing problems can also occur with liver disease from changes in the blood vessels and blood flow in the lungs. There are two well-recognized conditions that can result from liver disease: hepatopulmonary syndrome and portopulmonary hypertension. This fact sheet will review these two conditions and how they relate to liver disease.



What is liver disease?

The liver is the second largest organ in the body and has many important roles within the body including helping with digestion, metabolizing drugs, and storing nutrients. Its main job is to filter blood coming from the digestive tract and remove harmful substances from it before passing it to the rest of the body. Liver disease can occur from several causes, including infections (hepatitis), genetic diseases, alcoholism, and other toxins.

One problem that can develop with liver disease is portal hypertension. Portal hypertension refers to increased pressure in the vein that enters the liver. In certain cases, portal hypertension can develop without advanced liver disease. Risk factors for this can include: schistosomiasis (a disease caused by parasitic worms that is more common in developing countries), certain medications, or clots that prevent drainage of blood from the liver and intestines.

How does liver disease cause changes in the blood vessels and blood flow in the lungs?

Liver disease and portal hypertension can cause problems in the pulmonary vasculature, which are the blood vessels in the lungs. Your pulmonary vasculature is vital to effective breathing (it is the site of gas exchange of carbon dioxide for oxygen) and for the normal flow of blood from the right to left side of your heart. One factor that causes problems in the blood vessels of the lungs is the diseased liver's inability to remove harmful substances (toxins) from your digestive tract before they enter

the rest of your body. These toxins can damage blood vessels in your lungs leading to dilated (enlarged) or constricted (narrowed) vessels. Two different conditions can be seen in the lungs that arise from liver disease: hepatopulmonary syndrome and portopulmonary hypertension:

Hepatopulmonary syndrome is a condition where the blood vessels in your lungs become enlarged, leading to low oxygen levels.

Portopulmonary hypertension is a condition where the blood vessels in your lungs become narrowed, leading to elevated blood pressure in the pulmonary vasculature which obstructs the free flow of blood through the lungs.

What are the symptoms of hepatopulmonary syndrome and portopulmonary hypertension?

Symptoms of hepatopulmonary syndrome stem from low oxygen levels in your blood. Early on, there may be no signs or symptoms. As the disease progresses, you may find yourself experiencing the following signs or symptoms:

- Becoming short of breath easily
- Feeling breathless in the sitting or standing position and better when lying flat
- Having higher oxygen saturation when lying flat as opposed to a sitting or standing position
- Fingers/feet/lips turning blue (called cyanosis)
- Stroke, brain abscess, and brain hemorrhage are serious complications of hepatopulmonary syndrome

Symptoms of portopulmonary hypertension are similar to other types of pulmonary hypertension. In the early stages, there may be no signs or symptoms. As the disease progresses, you may find yourself experiencing the following signs or symptoms:

- Becoming short of breath or tired easily
- Chest pain, which can be mistaken for a heart attack
- Feel your heart pounding or racing
- Feel lightheaded or even pass out
- Swelling (edema) of your feet and ankles

Many of the symptoms mentioned above can commonly occur in people with advanced liver disease without these conditions. There are many different causes of breathlessness with liver disease, and your healthcare provider can help you figure out what is causing problems if you are having symptoms.

How do I know if I have liver disease?

Your healthcare provider can do blood tests and other studies to check for liver disease if you have symptoms or a condition that makes you at risk for liver problems. To evaluate for liver disease or portal hypertension, your healthcare provider may order an ultrasound of your abdomen or recommend a procedure to directly measure the pressure in the vein that enters your liver (portal vein).

How are hepatopulmonary syndrome and portopulmonary hypertension diagnosed?

There are three basic features required to make a diagnosis of hepatopulmonary syndrome:

- low blood oxygen levels (hypoxemia)
- enlarged blood vessels in the lung
- liver disease

You must have all three features to be diagnosed with hepatopulmonary syndrome. Your healthcare provider will usually order an arterial blood gas (a sample of blood from an artery) test to evaluate your blood oxygen levels. Your healthcare provider will also order tests to look for enlarged blood vessels in your lung. The test of choice is usually a contrast echocardiogram (ultrasound of your heart), where agitated saline is injected to look for bubbles going from the right to left side of the heart because of dilated blood vessels in the lung. An alternative test is the technetium macroaggregated albumin perfusion scan, where radiolabeled human serum albumin is injected to look for uptake in the brain or spleen.

The diagnosis of portopulmonary hypertension is made with the presence of elevated pulmonary pressures associated with liver disease. If you are being evaluated for liver transplantation, you should be screened for portopulmonary hypertension. Your healthcare provider will order an echocardiogram to evaluate the right side of your heart. If the echocardiogram is abnormal, then they may order a cardiac catheterization. During this procedure, a catheter (long rubber tube) is placed in your blood vessel and through the chambers of your heart to measure pressures in the right side of your heart.

How are hepatopulmonary syndrome and portopulmonary hypertension treated?

The only definitive treatment for hepatopulmonary syndrome is liver transplant. While transplant is a cure for most people with hepatopulmonary syndrome, some people are not good candidates for a variety of reasons. Whether you are a transplant candidate is determined by a liver transplant team at an expert center. For people in whom transplant is not an option, there are limited medical therapies available. Currently, there are no medications that have proven to be effective. While oxygen therapy can be offered to help with low blood oxygen levels, it may not be fully correctable in severe cases.

Treatment of portopulmonary hypertension depends on the severity of your disease. Therapies available include: medical therapy to dilate the pulmonary arteries and liver transplant. While liver transplant can be a cure for portopulmonary hypertension in some people, in others it can persist or even worsen over time. As liver transplant is only beneficial in a carefully selected group of people, you should be evaluated by both a liver transplant team and pulmonary hypertension specialist.

Can I have both hepatopulmonary syndrome and portopulmonary hypertension?

Yes! These disorders were originally thought to be two separate conditions, as they cause different changes in the blood vessels. However, in recent years, studies have shown that these two complications of liver disease may be on a spectrum, either occurring together or one after another. More research is underway now to clarify what this means in terms of treatment and prognosis.

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Treatment Action Steps:

- ✓ If you have liver disease and are having difficulty breathing, let your healthcare provider know
- ✓ Talk with your liver specialist about tests you may need to look for complications of liver disease

Additional Patient Resources:

Pulmonary Hypertension Association

- <http://www.phassociation.org>

PH and Liver Disease

- <https://phassociation.org/patients/aboutph/diseases-and-conditions-associated-with-ph/liver-disease/>

The Canadian Hepatopulmonary Syndrome Program

- <http://hpscicare.com>

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