

What is pulmonary stenosis?

The pulmonary artery carries blood to the lungs where it is oxygenated. It then returns the blood to the heart. Pulmonary stenosis (PS) is an obstruction of the blood flow from the right ventricle to the pulmonary artery. It results from a narrowing (stenosis) at several points on or near the pulmonary valve.

Pulmonary stenosis is a common form of congenital heart disease (CHD). It occurs in 7-10 percent of all CHD.

What are the different types of pulmonary stenosis?

Pulmonary stenosis is classified according to location and severity of the defect.

There are 3 types:

- **Valvar pulmonary stenosis** is the most common. In this, the pulmonary valve is dome-shaped and the opening is narrow (B). The leaflets are fused together. If the leaflets are thick, it is called a dysplastic valve. Pulmonary valvar stenosis can occur alone or with other congenital heart defects. These include tetralogy of Fallot or congenitally corrected transposition of the great arteries. It may also be part of a genetic syndrome, such as Noonan's syndrome.
- When the muscle under the pulmonary valve is thickened (C), it is called **subvalvar pulmonary stenosis**. This is caused by additional muscle bundles in the right ventricle.
- **Supravalvar stenosis** occurs when there is narrowing in the pulmonary artery above the pulmonary valve (A). There can be one or more than one areas of narrowing. This may be seen with genetic syndromes such as Williams or Alagille syndrome.

Pulmonary stenosis is also classified as mild, moderate, severe or critical. Mild and sometimes moderate stenosis does not progress. Severe and critical PS do. Knowing how severe your PS is will help your doctor know how to treat it. To figure this out, he/she will do an echocardiogram. The higher the gradient from the right ventricle to the pulmonary artery, the more severe the disease.

How common is pulmonary stenosis?

Pulmonary stenosis is a common form of congenital heart disease (CHD). It occurs in 7-10 percent of all CHD.

What happens in pulmonary stenosis?

When blood flow from your heart to your lungs is obstructed, the right ventricle has to work harder. This causes the heart muscle to get thicker. This is known as hypertrophy. The greater the obstruction, the thicker the right ventricle becomes. Mild hypertrophy may be acceptable. Too much can be harmful.

If severe obstruction is not treated, the right ventricle can begin to function poorly. This might cause arrhythmias (irregular heartbeats), loss of energy and fluid retention. If the pressure in the right heart is high enough, unoxygenated or "blue" blood can cross over into the left atrium. It then mixes with red or oxygenated blood. This results in cyanosis or blueness.

Pulmonary Stenosis

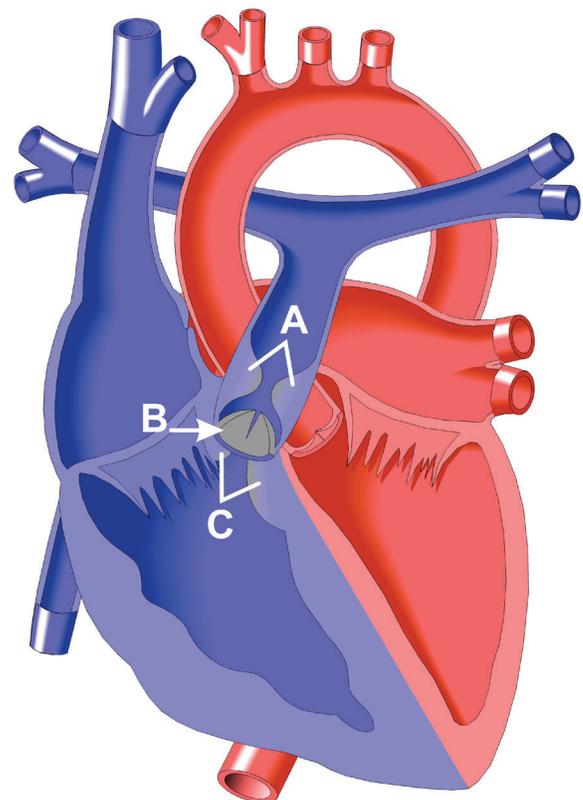


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What causes pulmonary stenosis?

Pulmonary stenosis occurs when the pulmonary valve does not form correctly. This happens in the first 8 weeks of fetal development. We don't know exactly why. Most of the time it occurs by chance.

What type of symptoms should I look for?

Signs and symptoms vary depending on how much obstruction you have. People with mild PS may have few or no signs and symptoms. If you develop any of the following symptoms, you should contact your adult congenital heart disease (ACHD) cardiologist for evaluation:

1. Shortness of breath with exercise and/or not being able to exercise as much as you have in the past
2. Heart racing
3. Dizziness
4. Fainting
5. Chest pain
6. Swelling in your legs or abdomen
7. Cyanosis or high red blood cell count.

Diagnosis

The most common diagnostic test used to confirm pulmonary stenosis is an echocardiogram. Cardiac MRI is becoming more important.

People with pulmonary stenosis can develop rhythm issues in the heart (arrhythmias). These can be in either the upper (atrial) or lower (ventricle) chambers of the heart. They may require treatment.

How is pulmonary stenosis treated?

Mild pulmonary stenosis and some moderate disease may not require treatment. Severe and critical levels always do. Critical pulmonary stenosis in the newborn is life threatening. If you were born with this, you had immediate treatment.

Surgery for valvar pulmonary stenosis was developed and performed in 1948 by Dr. Brock. Since that time, treatment, survival, and the quality of life for patients has improved. Today balloon valvuloplasty done in the cardiac catheterization lab is used to open the obstruction in most valvar pulmonary stenosis. Surgery is still the treatment of choice for dysplastic valves and patients who need surgery for other cardiac abnormalities. Both are successful in relieving the blockage. Surgery should be done by a congenital heart surgeon.

If I have PS, what is the risk that my children will have congenital heart disease?

Adults with CHDs are more likely than the general population to have a child with a heart defect. The risk is higher if the mother is the parent with a CHD and if there is a sibling with CHD. The defects may be the same or different than the parent's defect.

What are the long term outcomes for adults with pulmonary stenosis?

After surgery, there is the possibility of a leak developing in the pulmonary valve. This is called pulmonary insufficiency (PI). It means that blood being pumped from the right ventricle into the pulmonary artery leaks back into the right ventricle. When this happens, the right ventricle has to work harder. Over time it may thicken (hypertrophy). PI can be well tolerated for several decades. Many patients will need pulmonary valve replacement (PVR).

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Can I exercise?

It is important to ask your ACHD doctor before starting any new physical activity routine. Restrictions are individualized. They will vary according to the amount of obstruction you have. Patients who have had the valve repaired usually do not have exercise limits.

Can I have a baby?

All women with PS should consult with an ACHD cardiologist before getting pregnant. This will help identify any heart problems that should be addressed before pregnancy occurs. Those with mild or moderate pulmonary stenosis can often tolerate pregnancy. Those with more severe pulmonary stenosis may experience heart complications during pregnancy. They should have the obstruction repaired before becoming pregnant. Once it is, most women do well in pregnancy.

What type of follow up do I need?

Follow up with an ACHD specialist is recommended for any patient with repaired or unrepaired pulmonary stenosis. Your ACHD heart doctor will decide how often you need to be seen. By taking good care of your heart and getting recommended care, you will help ensure that you and your heart continue to thrive.

For his contribution to this article, the Adult Congenital Heart Association (ACHA) thanks Marlon Rosenbaum, MD, Director, Schneeweiss Adult Congenital Heart Center, Columbia University Medical Center.