

What is subaortic stenosis?

Subaortic stenosis is the second most common form of left ventricular outflow obstruction. It occurs when the flow of blood from the heart's pumping chamber or left ventricle is restricted. This obstruction is in the area of the heart under the aortic valve.

There are three types of subaortic stenosis. Two of them are associated with congenital heart disease (CHD).

Subaortic membrane is the most common type. It is a shelf-like membrane that forms under the aortic valve. It causes obstruction to the flow of blood from the left ventricle into the aorta. This results in blood going through turbulence. A doctor hears this as a heart murmur.

About 50% of patients with a subaortic membrane also have leakage of the aortic valve.

The subaortic membrane is twice as common in males. It has been reported in family clusters. It is more severe when diagnosed in children, rather than in adults. When detected in childhood it commonly worsens over time. It can occur alone as part of another CHD, such as Shones complex, septal defects or tetralogy of Fallot.

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Another type of subaortic stenosis is a subaortic tunnel. It is not as common as the subaortic membrane. In this defect, the whole area of the heart from the base of the left ventricle to the aortic valve is narrowed—very much like a tunnel. The aortic valve is often smaller than normal as well.

What are the symptoms of subaortic stenosis?

The symptoms of subaortic stenosis depend on how severe the obstruction is.

- Some patients will not have symptoms until they do activities that cause physical stress. These include exercise or pregnancy. Symptoms may include light-headedness, shortness of breath or fatigue.
- As the obstruction worsens, some patients may have chest pain or pass out during exercise or exertion. Others may develop fast heart beats or, rarely, congestive heart failure as fluid builds up in the heart.
- Patients with severe or untreated subaortic stenosis may be at risk for sudden cardiac death.

How is subaortic stenosis diagnosed?

A heart murmur is a key sign in suspecting and diagnosing subaortic stenosis. Your doctor may hear a rumbling sound when he listens to your heart. This occurs as the blood goes through the obstructed area under the aortic valve. Lab tests are usually normal. An echocardiogram or heart ultrasound will show the level and severity of the obstruction. It will also show if the left ventricle is thickened or enlarged. A magnetic resonance imaging (MRI) study or cardiac catheterization may also be done. These will help confirm the diagnosis and assess the severity.

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

As a person gets older, the condition can get worse. The progression is often very slow. It can take years before it causes major symptoms or interferes with a person's life. This is especially true in people whose obstructions are not detected until they are adults.

How is subaortic stenosis treated?

Surgery may be necessary to stop the progression of subaortic stenosis. Not everyone needs surgery. Your adult congenital heart disease (ACHD) doctor may recommend surgery if you have one or more of these:

- Severe obstruction or associated symptoms
- Increasing aortic valve leakage (regurgitation)
- A left ventricle that is getting thicker (hypertrophy)
- An abnormal stress test.

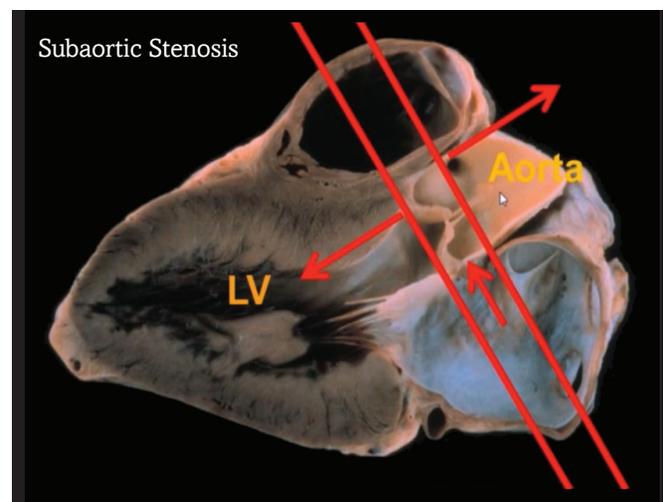


Illustration courtesy of Dr. Naser Ammash and the Mayo Clinic.

You should talk to your ACHD doctor about whether surgery is right for you.

If you have surgery, the surgeon removes the membrane or muscle causing the obstruction. In about one-third of patients, the aortic valve can be repaired. Between 6% and 29% of patients require valve replacement.

Surgery does not cure subaortic stenosis. It reduces the obstruction and improves symptoms. Thus, a person's quality of life is better. Up to 65% of people who have surgery do not have problems at 10 years.

In centers with high level of experience, the risk of surgery is between 0% and 3.8%. The risk is higher if the aortic valve needs to be replaced. It is also higher in patients with other serious medical problems.

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surgery do not have problems at 10 years. Reoperation may be needed in 25% of these patients if the obstruction comes back or the leaking of the valve worsens in 5 years.

Can women with subaortic stenosis have children?

If you have subaortic stenosis, it is very important that you talk to your ACHD heart doctor before you get pregnant. If you have severe obstruction or symptoms like difficulty breathing, you may need to have surgery before you have a baby. Your doctors can develop a plan to care for and protect your heart during your pregnancy and delivery.

What kind of heart care is recommended for adults with repaired and unrepaired subaortic stenosis?

Patients with subaortic stenosis should receive treatment in ACHD centers. These programs have the experience and expertise in treating adults with CHD. Experts recommend that adults with repaired or unrepaired subaortic stenosis be seen every two years or more frequently depending on severity. Specific recommendations for follow-up will be determined by the ACHD cardiologist.

For his work on this article, ACHA thanks **Naser M. Ammash, MD**, Mayo Clinic, Rochester, MN.