



# Cardiac Defects: Aortic Stenosis

When the heart squeezes, the left ventricle (the lower left chamber) contracts, pushing blood out into the aorta, the main artery that takes blood to the body. The aortic valve is located on the way out of the heart, to prevent blood from leaking back into the heart between beats. A normal aortic valve is made up of three thin leaflets.

In aortic stenosis, either the annulus is too small or the leaflets are fused or too thick, or there are fewer than three. As a result, the valve is too narrow, and the heart has to work harder to pump enough blood to the body. Aortic stenosis, or obstruction at the aortic valve, can be trivial, mild, moderate, severe, or critical.

Sometimes the stenosis is below the valve, caused by a fibrous membrane or a muscular ridge; this is called *sub-aortic stenosis*. The stenosis also can occur above the valve, in the aorta itself; this is called *supravalvar aortic stenosis*.

## Symptoms

Aortic stenosis usually won't cause symptoms in infants or small children. As the child gets older, signs and symptoms of aortic stenosis may appear, including fatigue; a heart murmur (an extra heart sound when a doctor listens with a stethoscope); or rarely, chest pain, fainting, or arrhythmias (abnormal heart rhythm).

## How is aortic stenosis in children diagnosed?

In rare cases, newborns have critical aortic stenosis, which requires immediate medical attention. Sometimes these severe cases are diagnosed before birth through a fetal heart program.

In most cases, cardiologists diagnose aortic stenosis after a primary care doctor detects a heart murmur and refers the child to a cardiologist.

Diagnosis may require some or all of these tests:

- pulse oximetry—a painless way to monitor the oxygen content of the blood

- chest X ray
- echocardiogram (also called echo or cardiac ultrasound)—sound waves create an image of the heart
- electrocardiogram (ECG)—a record of the electrical activity of the heart
- cardiac magnetic resonance imaging (MRI)—a three-dimensional image shows the heart's abnormalities
- cardiac catheterization—a thin tube (catheter) is inserted into the heart through a vein or artery in either the leg or through the umbilicus ("belly button").

Aortic stenosis can run in families, so be sure to tell your cardiologist if there is a history of a heart murmur in other close family members to allow for testing of the fetus before birth.

## What are the treatment options for aortic stenosis?

The exact treatment required for aortic stenosis depends on each child's heart anatomy. Trivial and mild aortic stenosis typically require no treatment. However, moderate, severe, and critical aortic stenosis require treatment.

## Cardiac Catheterization

In most cases, aortic stenosis is treated with balloon valvuloplasty, which requires cardiac catheterization. Doctors advance a thin tube (catheter) to the heart through an artery in the leg. The catheter has a balloon on the end of it. To open up the narrow valve, the balloon is briefly inflated, deflated, and withdrawn. Sometimes, two catheters and balloons are used. Sometimes, in newborns, the blood vessels in the umbilical cord are used as the site where the catheters are inserted and advanced toward the heart.

Older children will spend one night in the hospital after this procedure. They will need to rest the next day but then can resume normal activity. Newborns with critical aortic stenosis will usually stay in an intensive care unit before and after the procedure and will require some time to recover.



## Valvuloplasty Surgery

Surgery to repair or to replace the valve often is necessary in severe cases. Depending on the age, gender, and particular needs of your child, as well as the valve anatomy, surgeons may attempt to repair the valve, or at least improve its function, with a surgery called a valvuloplasty. Some research suggests that a surgical approach rather than a balloon approach has a slightly higher chance of avoiding aortic regurgitation in the future, yet the nature of the procedure poses a more immediate risk to the neonate (Benson, 2016).

## Artificial Valves

Another option to treat aortic stenosis includes the use of mechanical (artificial) valves as replacement valves. If this is the case, your child may need to stay on blood-thinning medicines for the rest of his or her life.

## Ross Procedure

Yet another option to treat aortic stenosis is the Ross Procedure. In this operation, the aortic valve is replaced with the patient's pulmonary valve. The pulmonary valve is then replaced with one from a donated heart. This procedure allows the patient's own pulmonary valve (now in the aortic position) to grow with the child.

## Subaortic and Supravalvar Stenosis Treatment

Subaortic and supravalvar stenosis do not get better with balloon dilation and will require surgery if the amount of obstruction is moderate or severe or, with subaortic stenosis, the aortic valve begins to leak significantly. Surgery for subaortic stenosis involves cutting out the ridge. Surgery for supravalvar aortic stenosis involves enlarging the aorta with a patch.

## Follow-Up Care Through Age 18

Children with aortic stenosis require regular check-ups with a pediatric cardiologist. Some children must remain on medicine and limit physical activity.

As a child with aortic stenosis grows, blood may begin to leak through the abnormal valve. This is called aortic regurgitation or aortic insufficiency (AI). In other children, the stenosis can reoccur. Neonates have an increased risk of AI and re-intervention, which may be related to their valve morphology prior to treatment (Petit et al., 2016). When stenosis happens, balloon valvuloplasty can be repeated, as long as there isn't significant aortic regurgitation. In severe cases, additional surgery may be necessary. The incidence of AI and repeat surgeries contributes to the debate of whether a balloon or surgical valvuloplasty leads to better future health.

Pediatric cardiologists follow patients until they are young adults, coordinating care with the primary care providers.

## Into Adulthood

Adults with aortic stenosis must continue to see a cardiologist regularly. Your child's pediatric cardiologist will help with the transition to an adult cardiologist. All patients with aortic valve disease need some form of life-long follow-up with a cardiologist. Because of enormous strides in medicine and technology, today most children with heart conditions go on to lead healthy, productive lives as adults.

## References

- Benson, L. (2016). Neonatal aortic stenosis is a surgical disease: An interventional cardiologist view. *Seminars in Thoracic and Cardiovascular Surgery: Pediatric Cardiac Surgery Annual*, 19(1), 6–9.
- Petit, C., Gao, K., Goldstein, B., Lang, S., Gillespie, S., Kim, S., Sachdeva, R. (2016). Relation of aortic valve morphologic characteristics to aortic valve insufficiency and residual stenosis in children with congenital aortic stenosis undergoing balloon valvuloplasty. *The American Journal of Cardiology*, 117(6), 972–979.

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