

What is Aortic Stenosis?

Aortic Stenosis is a condition where the valve between the left ventricle and the aorta (main artery leading out of the heart) is not shaped properly and does not open all the way. This can vary quite a bit. The most common form is called a **bicuspid aortic valve** where the valve has two cusps (or parts) instead of three. In this condition there is no pressure difference across the valve and the only finding is an extra noise called a click. Many times, one or more parts (or cusps) of the aortic valve is thickened and stiff. Usually, when there is an abnormal valve there is a pressure difference across the valve forcing the left ventricle to work harder than normal. Aortic stenosis can be seen with other cardiac defects such as Coarctation of the Aorta and Mitral valve abnormalities.

We grade the amount of narrowing of the valve by estimating the difference in pressure across the valve. Normally there should be no difference. For example, normally if the pressure in the aorta was 120 mmHg (units of blood pressure) then the left ventricular pressure should also be 120 mmHg. If a person had a pressure of 120 mmHg in the aorta and 150 mmHg in the left ventricle we would say he had a gradient of 30 mmHg. The degree of stenosis is broken down into three groups - mild (0 to 30 mmHg gradient), moderate (30 to 60 mmHg gradient), and severe (over 60 mmHg gradient).

Children with mild stenosis have no symptoms and normal exercise tolerance. They have physical findings of a click and murmur. Children with moderate stenosis usually have no symptoms and the murmur is louder. Those with severe stenosis have, by definition, some type of symptoms or significant thickening of the left ventricular walls or enlargement of the heart. Children with severe aortic stenosis can develop chest pain with exercise, palpitations, fainting, shortness of breath, and even sudden death.

Newborns with aortic stenosis can be difficult to predict in terms of how they will do. The newborn heart muscle does not have as much strength as that of an older child and sometimes they will not tolerate as much stenosis. They require careful monitoring and can suddenly develop signs of heart failure. Often when the aortic valve is not shaped properly it will also leak. This is called **aortic insufficiency**. This leakage can cause the left ventricle to enlarge. About 15% of patients with abnormal aortic valves will have abnormal ascending Aortas that can potentially enlarge and may need to be repaired later in life.

Management of these patients has changed a lot over the past 10 years. The usual first line treatment for severe aortic stenosis is called balloon valvuloplasty. This is where a balloon is delivered by catheter (no surgery) across the valve and rapidly inflated. This causes the valve tissue to tear and the opening of the valve is enlarged thereby decreasing the gradient. Occasionally when the valve is torn it will leak. This is usually a safe procedure, however in the younger child and particularly babies the risk of damage to the arteries in the legs is increased. Sometimes it is necessary to surgically open the valve. Many times if the balloon procedure is not enough or if the valve leaks too much afterwards it is necessary to replace the valve with either a mechanical valve or one harvested from a cadaver (homograph). The disadvantage to the mechanical valves that the patient will require a blood thinning medicine for life. A new technique called the "Ross™ procedure" has recently become the surgery of choice for many patients. In this surgery, the patient's own pulmonic valve is used to replace the aortic valve and a homograph valve is placed in the pulmonic position. The advantage of this is that the "new" aortic valve is natural, living tissue that will grow with the patient. The results in the infants and younger children are not as good as in older kids. Some of these patients may need a reoperation later in life. The decision to do surgery is based on how the patient is doing, his weight and the size and function of the left ventricle. Children with moderate to severe aortic stenosis require a lifetime of monitoring by a physician.

All children with Aortic Stenosis are at higher risk of infection of the heart, and require SBE Prophylaxis with antibiotics when they go to the Dentist or have any kind of surgery.

If you have questions, please ask one of the doctors.

What is Aortic Stenosis ?

