

## **Aortic Valve Stenosis (AS) and Aortic Insufficiency (AI)**

### **What is it?**

The aortic valve opens to let blood flow from the main left pumping chamber (left ventricle) to the main body artery (aorta).

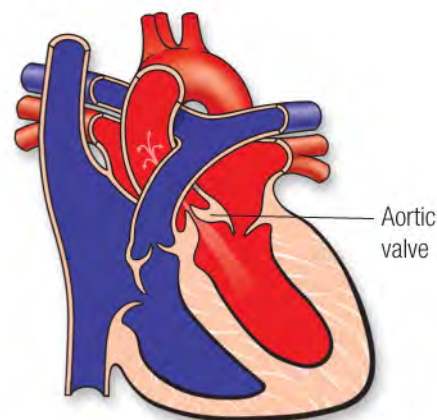
Stenosis (narrowing or obstruction) of the aortic valve makes the left ventricle pump harder to get blood past the blockage.

Insufficiency (also called regurgitation) is when blood that's just been pumped through the valve leaks backwards into the pumping chamber between heartbeats.

Some children can have mostly obstruction; others mostly insufficiency. Some children have a valve with both problems.

Aortic stenosis (AS) occurs when the aortic valve didn't form properly. A normal valve has three parts (leaflets or cusps), but a stenotic valve may have only one cusp (unicuspid) or two cusps (bicuspid), which are thick and stiff, rather than thin and flexible.

Stenotic Aortic Valve



### **What causes it?**

In most children, the cause isn't known. It's a common type of heart defect. Some children can have other heart defects along with AS.

### **How does it affect the heart?**

In a child with AS, the pressure is much higher than normal in the left pumping chamber (left ventricle) and the heart must work harder to pump blood out into the body arteries. Over time this can cause thickening (hypertrophy) and damage to the overworked heart muscle. In a child with AS, the heart also works harder to pump the normal amount of blood required by the body, and also all the blood that has leaked back into the left ventricle through the valve in between heartbeats. This can cause the left ventricle to be enlarged (dilated) and also may cause damage to the heart muscle.

### **How does the abnormal aortic valve affect my child?**

If the obstruction and leak are mild, the heart won't be overworked and symptoms don't occur. Sometimes stenosis is severe and symptoms occur in infancy. In some children chest pain, unusual tiring, dizziness or fainting may occur.

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Otherwise, most children with aortic stenosis have no symptoms, and special tests may be needed to determine the severity of the problem.

### **What can be done about the aortic valve?**

The valve can be treated to improve the obstruction and leak, but the valve can't be made normal.

Children with aortic stenosis will need treatment when the pressure in the left ventricle is high (even though there may be no symptoms). In most children the obstruction can be relieved during cardiac catheterization by balloon valvotomy. In this procedure, a special tool, a catheter containing a balloon, is placed across the aortic valve. The balloon is inflated for a short time to stretch open the valve (called a valvotomy).

Some children with stenosis may need surgery. The surgeon may be able to enlarge the valve opening if it's too small. Some valve leakage is likely to develop or increase after a balloon or surgical treatment for obstruction.

If your child's aortic valve no longer responds to valvotomy or has become severely insufficient (leaky), it will probably need to be replaced. The aortic valve can be surgically replaced in three ways:

- (1) The Ross procedure, a surgery in which the abnormal aortic valve is removed and replaced by the child's own pulmonary valve. Then the pulmonary valve is replaced with a preserved donor pulmonary valve.
- (2) Aortic valve replacement with a preserved donor valve.
- (3) Aortic valve replacement with a mechanical valve.

Each option has advantages and disadvantages. Discuss them with your child's pediatric cardiologist, cardiac surgeon or both.

### **What activities can my child do?**

If the aortic valve is abnormally formed but has no important obstruction or leak, your child may not need any special precautions regarding physical activities and may be able to participate in normal activities without increased risk. Some children with obstruction, leak or heart muscle abnormalities may have to limit how much they do some kinds of exercise. Check with your child's pediatric cardiologist about this.

### **What will my child need in the future?**

Children with aortic stenosis need lifelong medical follow-up. Your child's pediatric cardiologist will examine periodically to look for problems such as worsening of the obstruction or leak. Even mild stenosis may worsen over time. Also, balloon or surgical relief of a blockage is sometimes incomplete. After treatment the valve keeps working in a mildly abnormal way.

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### **What about preventing endocarditis?**

Children with AS and AI risk developing endocarditis. Children who have had their aortic valve replaced will need to take antibiotics before certain dental procedures.