



# Cardiac Defects: Pulmonary Stenosis

When the heart squeezes, the right ventricle (the lower right chamber) contracts and pushes blood out into the pulmonary artery (the artery that takes blood to the lungs). The pulmonary, or pulmonic, valve sits on the way out of the heart between the right ventricle and the main pulmonary artery to prevent blood from leaking back into the heart between beats. A normal pulmonary valve is made up of three thin leaflets.

In pulmonary stenosis, the leaflets are fused or are too thick, or there are fewer than three. As a result, the pulmonary valve is too narrow, and the heart has to work harder to pump enough blood to the body. Pulmonary stenosis, or obstruction at the pulmonary valve, can be trivial, mild, moderate, severe, or critical. This condition is also called *pulmonic stenosis* or *pulmonary valve stenosis*.

Sometimes the stenosis is below the pulmonary valve, caused by muscular bundles. This is called *subpulmonic stenosis*. Also, the stenosis can occur above the pulmonary valve, in the pulmonary artery itself. This is called *supravalvar pulmonic stenosis*.

## What are the symptoms of pulmonary stenosis in children?

Pulmonary stenosis usually does not cause symptoms in infants or small children. As the child gets older, abnormal signs and symptoms may appear, including fatigue, a heart murmur (an extra heart sound when a healthcare provider listens with a stethoscope) and, rarely, chest pain or fainting.

## How is pulmonary stenosis diagnosed?

In rare cases, newborns have critical pulmonary stenosis, which requires immediate medical attention. Sometimes severe cases of pulmonary stenosis are diagnosed before birth.

Cardiac specialists usually diagnose pulmonary stenosis after a primary care provider detects a heart murmur and refers the child to them.

Diagnosis of pulmonary stenosis 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 ultrasound)—sound waves create an image of the heart
- electrocardiogram (ECG)—a record of the electrical activity of the heart
- cardiac MRI—a three-dimensional image shows the heart’s abnormalities
- cardiac catheterization—a thin tube is inserted into the heart through a vein or artery in either the leg or through the umbilicus (“belly button”).

Pulmonary stenosis can run in families, so be sure to tell your cardiologist if there is a history of a murmur in other close family members.

## What are the treatment options for pulmonary stenosis in children?

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

### Cardiac Catheterization

In most cases, pulmonary stenosis is treated with balloon valvuloplasty, which requires cardiac catheterization. Healthcare providers advance a thin tube (catheter) to the heart through a vein 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 may spend one night in the hospital after this procedure and will need to rest the next day but then can resume normal activity. Newborns with critical



pulmonary stenosis will stay in an intensive care unit before and after the procedure and will require some time to recover.

### **Surgery**

In rare cases, surgery will be required. Surgeons use a procedure called *valvotomy* to separate fused leaflets in the pulmonary valve. Another option includes the surgical placement of a valve called a *pulmonary homograft*, which is a donated pulmonary valve and artery. This valve may grow with the child, and blood-thinners are not required.

### **New Valve**

An exciting future option is a tissue-engineered valve that is grown with the patient's own cells on a biodegradable mesh. This may be the future of all valve replacement but is still in the research and development phase.

### **Surgery for Subpulmonic and Supravalvar**

Subpulmonic and supravalvar pulmonic stenosis do not get better with balloon dilation and will require surgery if the amount of obstruction is moderate or severe. Surgery for subpulmonic stenosis involves cutting out the muscle bundles. Surgery for supravalvar pulmonic stenosis involves enlarging the pulmonary artery with a patch.

### **Follow-Up Care Through Age 18**

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

As the child grows, blood may begin to leak through the abnormal valve. This is called pulmonary regurgitation or pulmonic insufficiency. In other children, the stenosis can reoccur. When this happens, balloon valvuloplasty can be repeated, as long as there isn't significant regurgitation. In severe cases, additional surgery may be necessary.

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

### **Into Adulthood**

Adults who were born with pulmonary stenosis must continue to see a cardiologist. Pediatric cardiologists will help with the transition to an adult cardiologist. All patients with pulmonary valve disease need some form of lifelong 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.

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