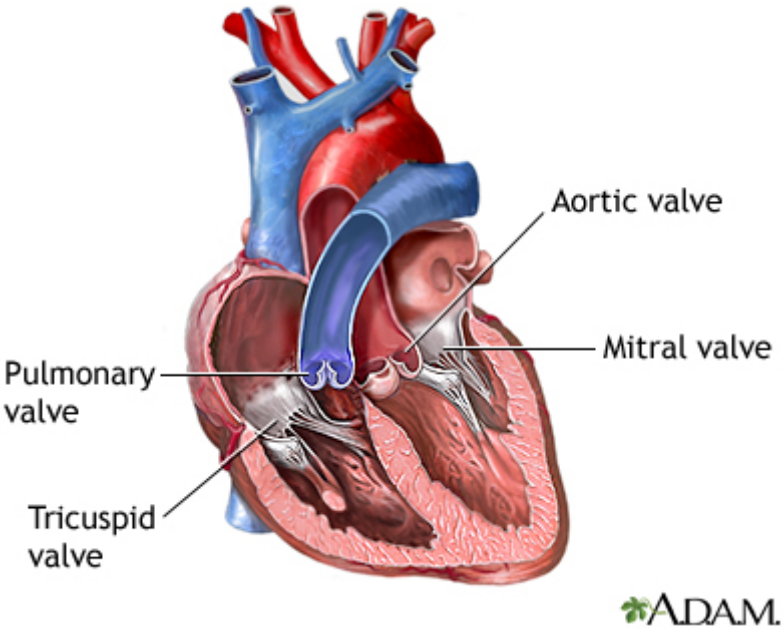
Pulmonary valve stenosis

Definition



Pulmonary valve stenosis is a heart valve disorder that involves the pulmonary valve.

This is the valve separating the right ventricle (one of the chambers in the heart) and the pulmonary artery. The pulmonary artery carries oxygen-poor blood to the lungs.

Stenosis, or narrowing, occurs when the valve cannot open wide enough. As a result, less blood flows to the lungs.

Alternative Names

Valvular pulmonary stenosis; Heart valve pulmonary stenosis; Pulmonary stenosis; Stenosis - pulmonary valve; Balloon valvuloplasty - pulmonary

Causes

Narrowing of the pulmonary valve is most often present at birth (congenital). It is caused by a problem that occurs as the baby develops in the womb before birth. The cause is unknown, but genes may play a role.

Narrowing that occurs in the valve itself is called pulmonary valve stenosis. There may also be narrowing just before or after the valve.

The defect may occur alone or with other heart defects that are present at birth. The condition can be mild or severe.

Pulmonary valve stenosis is a rare disorder. In some cases, the problem runs in families.

Symptoms

Many cases of pulmonary valve stenosis are mild and do not cause symptoms. The problem is most often found in infants when a heart murmur is heard during a routine heart exam.

When the valve narrowing (stenosis) is moderate to severe, the symptoms include:

- Abdominal distention
- Bluish color to the skin (cyanosis) in some people
- Poor appetite
- Chest pain
- Fainting
- Fatigue
- Poor weight gain or failure to thrive in infants with a severe blockage
- Shortness of breath
- Sudden death

Symptoms may get worse with exercise or activity.

Exams and Tests

The health care provider may hear a heart murmur when listening to the heart using a stethoscope. Murmurs are blowing, whooshing, or rasping sounds heard during a heartbeat.

Tests used to diagnose pulmonary stenosis may include:

- Cardiac catheterization
- Chest x-ray
- ECG
- Echocardiogram
- MRI of the heart

The provider will grade the severity of the valve stenosis to plan treatment.

Treatment

Sometimes, treatment may not be needed if the disorder is mild.

When there are also other heart defects, medicines may be used to:

- Help blood flow through the heart (prostaglandins)
- Help the heart beat stronger
- Prevent clots (blood thinners)
- Remove excess fluid (water pills)
- Treat abnormal heartbeats and rhythms

Percutaneous balloon pulmonary dilation (valvuloplasty) may be performed when no other heart defects are present.

- This procedure is done through an artery in the groin.
- The doctor sends a flexible tube (catheter) with a balloon attached to the end up to the heart. Special x-rays are used to help guide the catheter.
- The balloon stretches the opening of the valve.

Some people may need heart surgery to repair or replace the pulmonary valve. The new valve can be made from different materials. If the valve cannot be repaired or replaced, other procedures may be needed.

Outlook (Prognosis)

People with mild disease rarely get worse. However, those with moderate to severe disease will get worse. The outcome is often very good when surgery or balloon dilation is successful. Other congenital heart defects may be a factor in the outlook.

Most often, the new valves can last for decades. However, some will wear out and need to be replaced.

Possible Complications

Complications may include:

- Abnormal heartbeats (arrhythmias)
- Death
- Heart failure and enlargement of the right side of the heart
- Leaking of blood back into the right ventricle (pulmonary regurgitation) after repair

When to Contact a Medical Professional

Call your provider if:

- You have symptoms of pulmonary valve stenosis.

- You have been treated or have untreated pulmonary valve stenosis and have developed swelling (of the ankles, legs, or abdomen), difficulty breathing, or other new symptoms.

References

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