

Understanding Pulmonary Valve Stenosis and Long-Term Management

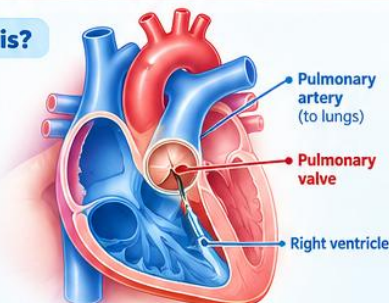


A common and usually treatable congenital heart defect with an excellent long-term outlook.

What Is Pulmonary Valve Stenosis?

Pulmonary valve stenosis (PVS) is a congenital heart defect in which the pulmonary valve – the valve between the right ventricle and the pulmonary artery – is narrowed. This makes it harder for the right ventricle to pump blood to the lungs.

PVS accounts for approximately 8–10% of congenital heart defects and can range from mild to severe.



Normal vs. Pulmonary Valve Stenosis

Normal Pulmonary Valve



Normal, wide opening allows blood to flow easily to the lungs.

Pulmonary Valve Stenosis



Narrowed valve makes it harder for blood to flow to the lungs.

Types of Pulmonary Stenosis



Valvar Pulmonary Stenosis
Narrowing at the pulmonary valve (most common)



Subvalvar Pulmonary Stenosis
Narrowing below the valve within the right ventricular outflow tract



Supravalvar Pulmonary Stenosis
Narrowing above the valve in the main or branch pulmonary arteries

Severity Classification

Based on Doppler echocardiography (peak gradient across the valve)

Severity	Peak Gradient
Mild	< 36 mmHg
Moderate	36 – 64 mmHg
Severe	> 64 mmHg

Higher gradients indicate greater obstruction and increased workload on the right ventricle.

Causes and Associated Conditions



Most cases are congenital and occur during fetal development.

Associated conditions include:

- Noonan Syndrome
- Alagille Syndrome
- Other congenital heart defects



Symptoms

Mild Stenosis



Many patients have no symptoms.

Moderate to Severe Stenosis



Shortness of breath



Fatigue



Exercise intolerance



Chest discomfort



Dizziness



Fainting (syncope)



Palpitations

Infants with critical pulmonary stenosis may develop:



- Cyanosis (low oxygen levels)
- Poor feeding
- Respiratory distress

Diagnosis



Physical Examination

- Systolic ejection murmur
- Ejection click
- Wide splitting of the second heart sound



Echocardiogram

- Confirms diagnosis
- Measures severity
- Assesses right ventricular function



Electrocardiogram (ECG)

May show right ventricular hypertrophy in severe cases.



Cardiac MRI

Used when additional anatomical detail is needed.



Cardiac Catheterization

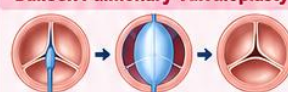
Occasionally required for intervention planning.

Treatment

Mild Pulmonary Valve Stenosis

- ✓ No medication
- ✓ No intervention
- ✓ Periodic follow-up

Balloon Pulmonary Valvuloplasty



- Catheter is advanced to the valve
- Balloon is inflated across the narrowed valve
- Valve opens, reducing obstruction

Success rates exceed 90% in appropriately selected patients.

Surgical Treatment

- Valve anatomy not suitable for balloon dilation
- Significant subvalvar obstruction
- Associated cardiac defects require repair

Long-Term Follow-Up

After successful balloon valvuloplasty, most patients do very well.

Ongoing monitoring focuses on:

- ✓ Residual stenosis
- ✓ Development of pulmonary valve leakage (regurgitation)
- ✓ Right ventricular size and function
- ✓ Exercise tolerance



Follow-up intervals depend on age and severity.

Pulmonary Regurgitation

A common long-term consequence after intervention is pulmonary valve leakage.

- Mild pulmonary regurgitation is usually well tolerated.
- Significant pulmonary regurgitation may eventually lead to:
 - Right ventricular enlargement
 - Reduced exercise capacity
 - Arrhythmias



Regular imaging helps identify patients who may require future intervention.

Exercise and Sports Participation



Mild Stenosis: Most patients can participate in all competitive and recreational sports.



Moderate Stenosis: Recommendations depend on symptoms, right ventricular function, and exercise testing results.



Severe Stenosis: Strenuous competitive sports are generally restricted until treatment is completed. After successful treatment, many can return to unrestricted activity.

Pregnancy Considerations



- Most women with mild or successfully treated pulmonary stenosis tolerate pregnancy well.
- Women with severe residual stenosis should be evaluated by an adult congenital heart disease specialist before pregnancy.

Endocarditis Prevention



Routine antibiotic prophylaxis before dental procedures is generally not required for isolated pulmonary valve stenosis.

Exceptions may apply in patients with:

- Prosthetic valves
- Prior infective endocarditis
- Certain repaired congenital heart defects

When to Seek Medical Attention



Contact your cardiologist if you develop:

- New shortness of breath
- Reduced exercise tolerance
- Chest pain
- Palpitations
- Fainting episodes
- Progressive fatigue

Prognosis



- The outlook for isolated pulmonary valve stenosis is excellent.
- Mild stenosis often never requires intervention.
- Balloon valvuloplasty provides durable relief in most patients.
- Most patients have normal growth, exercise participation, and life expectancy.
- Lifelong follow-up with a congenital heart disease cardiologist is recommended.