

Pulmonary valve disease

The pulmonary valve normally has three cusps and is responsible for regulating the flow of deoxygenated blood from the right ventricle to the lungs.

What is pulmonary valve disease?

Pulmonary valve disease is very uncommon and can be due either to stenosis or to insufficiency. The majority of pulmonary valve problems occur as the result of congenital heart disease but the pulmonary valve may rarely be damaged as result of infection (eg, [infective endocarditis](#)) or as a result of disease (eg, [carcinoid disease](#), [Marfan's syndrome](#) or [pulmonary hypertension](#)).

An obstructed right ventricle outflow tract causes a pressure overload in the right ventricle. Pulmonary regurgitation leads to a volume overload and therefore to a dilation of the right ventricle.

Both obstruction and regurgitation may be combined. Both pulmonary stenosis (PS) and pulmonary regurgitation may be tolerated for long periods but the overload of the right ventricle has a progressively detrimental effect on right ventricular function.^[1]

How common is pulmonary valve disease? (Epidemiology)

- Pulmonary valve disease is rare.
- Congenital disease represents the majority of all pulmonary valve problems.

Pulmonary stenosis^[2]

PS can be due to isolated valvular (90%), subvalvular or supra-valvular obstruction, or it may be found in association with more complicated congenital heart disorders.

Presentation

See also the separate [Heart Auscultation](#) and [Heart Murmurs in Children](#) articles.

- PS may be diagnosed in utero or shortly after birth. Critical PS causes cyanosis and is potentially lethal in the neonate. See also the article on [Congenital Heart Disease in Children](#).
- Ultrasound images of the four chambers of the heart in utero may demonstrate isolated PS due to an abnormally thickened or bicuspid valve, or PS in association with other congenital anomalies - eg, Fallot's tetralogy, Noonan's syndrome, or as a result of intrauterine infection with rubella.
- In infancy the condition is usually diagnosed by auscultation of the heart and the presence of a murmur.

Symptoms

The presentation can be isolated or in association with other congenital heart disease including [patent foramen ovale](#), [ventricular septal defect](#), [patent ductus arteriosus](#) and [atrial septal defect](#).

The symptoms of PS will vary with the severity of the stenosis. Mild PS may be asymptomatic.

- Shortness of breath.
- Chest pain.
- Fainting or exertional syncope.
- Sudden death.

Signs

- Soft pulmonary systolic murmurs are more easily heard with the patient lying down. They are often heard in healthy individuals and may be due to physiological changes associated with respiration.
- Ejection systolic murmur along the left upper edge of the sternum.
- Pulmonary ejection click.
- Delayed second heart sound with severe stenosis.
- Parasternal thrill and heave.
- 'A waves' in the JVP.

Investigations

- Echocardiography: will confirm the valve defect and measure the flow across the valve.
- ECG: may show right ventricular hypertrophy, right atrial hypertrophy and right axis deviation.
- Angiography: may be required in children with multiple cardiac abnormalities. Pulmonary angiography may be needed to establish the diagnosis of peripheral pulmonary stenosis.
- CXR: may show a prominent pulmonary artery and possibly enlargement of the right atrium and ventricle.

Treatment and management

Medical

- Initial treatment of critical PS in a neonate includes general resuscitation and infusion of prostaglandin E1 to dilate the ductus arteriosus.
- If the patient is asymptomatic and the right ventricular pressure is less than 60 mm Hg, the patient can be monitored by a cardiologist who will perform ECG and echocardiography and may perform CXR and an exercise ECG.
- Management of [infective endocarditis](#) may be required.

Surgical

- If the patient is symptomatic, and/or has a right ventricular pressure greater than 60 mm Hg, the National Institute for Health and Care Excellence (NICE) recommends balloon valvotomy via cardiac catheterisation as effective treatment.^[3]
- Percutaneous balloon pulmonary valvuloplasty is generally safe and effective but a small proportion of patients require a second percutaneous balloon pulmonary valvuloplasty.^[4]
- In children with moderate, severe, and critical pulmonary stenosis, pulmonary balloon valvuloplasty is recommended as the treatment of choice, and is safe and effective treatment.
- Percutaneous pulmonary valve implantation is an alternative to surgical valve repair or replacement for right ventricular outflow tract dysfunction.^[5]
- Pulmonary artery balloon angioplasty, with or without placement of an expandable metal stent, can be used to treat supra- and peripheral PS.

Prognosis

- The prognosis will depend on the severity of the PS, and any damage to the right ventricle and right atrium.

Pulmonary regurgitation^[6]

- Pulmonary regurgitation occurs only very rarely as a congenital anomaly.
- However, it is a common complication after surgical or percutaneous relief of PS and following repair of Fallot's tetralogy.^[3]
- Pulmonary regurgitation may also occur secondary to a dilated pulmonary valve ring due to pulmonary hypertension or Marfan's syndrome.

- Significant pulmonary regurgitation may also be caused by primary pulmonary hypertension, secondary pulmonary hypertension, infective endocarditis (rare, but may occur in an intravenous drug user or with an atrial septal defect and a large left-to-right intracardiac shunt), rheumatic heart disease, carcinoid heart disease, Marfan's syndrome and a few medications (eg, methysergide, pergolide).

Presentation

See also the separate [Heart Auscultation](#) and [Heart Murmurs in Children](#) articles.

- Pulmonary regurgitation is usually asymptomatic unless severe, when it may lead to signs of right heart failure.
- Soft diastolic murmur at the left upper sternal edge.
- Right ventricular hypertrophy.
- Loud P2 (pulmonary component of the second heart sound).
- May be differentiated from aortic regurgitation by lack of collapsing pulse.

Investigations

- ECG.
- CXR.
- Echocardiography: colour flow Doppler echocardiography is the mainstay for recognising pulmonary regurgitation.
- Cardiac catheterisation is usually not necessary but may help to determine the underlying cause and any co-existing abnormalities.

Treatment and management

- Patients should be advised of the risk of endocarditis. See the separate [Infective Endocarditis](#) article.
- Treatment of any underlying cause.

- Pulmonary regurgitation usually doesn't require any specific intervention other than follow-up by a cardiologist every 1–3 years depending on severity and cause.
- If symptomatic or right ventricular dilatation, pulmonary valve replacement should be considered.
- If there is severe right heart failure and pulmonary hypertension, a heart-lung transplant may be required.

Prognosis

- Pulmonary regurgitation is usually well tolerated in childhood.
- Long-term studies have demonstrated that pulmonary regurgitation may lead to progressive right ventricular dilatation, right ventricular dysfunction, exercise intolerance, ventricular tachycardia, and sudden cardiac death.^[7]

Referral^[8]

In the guideline for heart valve disease presenting in adults, NICE recommends:

- Consider an echocardiogram for adults with a murmur and no other signs or symptoms if valve disease is suspected based on the nature of the murmur, family history, age (especially if over 75), or medical history (eg, atrial fibrillation).
- Offer an echocardiogram to adults with a murmur if valve disease is suspected (based on the nature of the murmur, family history, age or medical history) and they have:
 - Signs (such as peripheral oedema) or symptoms (such as angina or breathlessness) or an abnormal ECG; or
 - An ejection systolic murmur with a reduced second heart sound but no other signs or symptoms.

Referral for urgent specialist assessment or urgent echocardiography

If valve disease is suspected (based on the nature of the murmur, family history, age or medical history):

- Offer urgent (within two weeks) specialist assessment that includes echocardiogram or if not available an urgent echocardiogram alone to adults with a systolic murmur and exertional syncope.
- Consider urgent (within two weeks) specialist assessment that includes echocardiogram for adults with a murmur and severe symptoms (angina or breathlessness on minimal exertion or at rest) thought to be related to valvular heart disease.

Refer urgently for cardiovascular assessment, with the referral reviewed and prioritised by an appropriate specialist within 24 hours, anyone with transient loss of consciousness and a heart murmur. [9]

Further reading

- [Balloon dilatation with or without stenting for pulmonary artery or non-valvar right ventricular outflow tract obstruction in children](#); NICE Interventional procedures guidance, July 2004
- [Qureshi AM, Prieto LR; Percutaneous pulmonary valve placement. Tex Heart Inst J. 2015 Jun 1;42\(3\):195-201. doi: 10.14503/THIJ-14-4276. eCollection 2015 Jun.](#)
- [British Heart Foundation](#)

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