

PULMONARY VALVE DISEASE

Pulmonary Valve Regurgitation

Aetiology

Mild pulmonary regurgitation (PR) may be a normal finding on Doppler echocardiography. The most common causes of pathologic PR in adults are prior interventions for congenital heart disease such as tetralogy of Fallot repair or surgical valvotomy for congenital pulmonary stenosis. Other rare causes of PR are congenital pulmonary annular dilation, pulmonary hypertension, rheumatic or carcinoid heart disease, endocarditis, and trauma.¹

Pathophysiology

Long-standing severe PR results in progressive RV dilation and reduced RV function. RV dilatation is reflected on QRS duration and is associated with ventricular arrhythmias, and sudden death. This mechanical-electrical association has been described in patients with repaired tetralogy of Fallot. Significant PR is the main predictor of sudden death in these patients (see Chapter 9).

Physical Examination

RV parasternal heave in severe PR with enlarged RV.

Systolic ejection murmur increased by inspiration.

Soft, diastolic, decrescendo murmur best heard in the left upper sternal border.

An increase in intensity of the murmur may be noted during inspiration. The murmur is easily audible and occasionally palpable in patients with PHT.

ECG findings are nonspecific or reflect RV enlargement.

Chest radiography may demonstrate cardiomegaly and pulmonary artery enlargement.

The diagnosis is made by **echocardiography**.² Severe PR is indicated by a color jet filling the outflow tract and a dense continuous wave Doppler signal with a steep deceleration slope.

Cardiac magnetic resonance is the imaging modality of choice to assess RV size and function in asymptomatic patients and guide therapy.

Therapy

Medical therapy is not effective in reducing the degree of PR or affecting the impact of PR on the RV. PV replacement is recommended in symptomatic patients (NYHA II or III) and severe PR. The management of asymptomatic patients is controversial. Most would agree that PVR is indicated in: decreased RV systolic function (ejection fraction <40% by cardiac magnetic resonance imaging), progressive RV dilation (cardiac magnetic resonance imaging RV end-diastolic volume 160 mL/m² or 82 mL/m² for RV end-systolic volume) or TR related to progressive annular dilatation, severe PR in a patient requiring another cardiac operation, and

QRS duration 180 milliseconds or QRS duration increase >3.5 ms/y).¹ Recommendations on specific entities associated with PR have been provided in the 2018 AHA/ACC Guideline on ACHD, and relevant previous statements.³ Transcatheter pulmonary valve replacement (Melody or Sapien XT and 3) has become the standard therapy for postoperative right ventricular outflow tract obstruction or pulmonary regurgitation in patients with an anatomically suitable prosthetic pulmonary conduit or valve.⁴

Pulmonary Valve Stenosis

Carcinoid syndrome and rheumatic valve disease may cause pulmonary stenosis but essentially always occur in conjunction with other valve disease. The majority ($>95\%$) of pulmonary stenosis cases are related to congenital or genetic disorders and are discussed in the relevant Chapter on ACHD.

References

1. Bruce CJ and Connolly HM. Right-sided valve disease deserves a little more respect. *Circulation*. 2009;119:2726-34.
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3. Bhatt AB, et al. Congenital heart disease in the older adult: a scientific statement from the American Heart Association. *Circulation*. 2015;131:1884-931.
4. Doff B, McElhinney DB and al. e. Reintervention and Survival After Transcatheter Pulmonary Valve Replacement. *JACC*. 2022.