40 - 278 Pulmonic Valve Disease

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Pulmonic Valve Disease PULMONIC STENOSIS Pulmonic valve stenosis (PS) is essentially a congenital disorder (Table 278-1). With isolated PS, the valve is typically domed. Dys plastic pulmonic valves are seen as part of the Noonan syndrome (Chap. 292), which maps to chromosome 12. Mutations in the PTPN1 gene are associated with about half of all cases of Noonan syndrome. Much less common etiologies include carcinoid and obstructing tumors or bulky vegetations. The pulmonic valve is only very rarely affected by the rheumatic process. ■ ■PATHOPHYSIOLOGY PS is defined hemodynamically by a systolic pressure gradient between the right ventricle (RV) and the main pulmonary artery (PA). RV hypertrophy (RVH) develops as a consequence of sustained obstruc tion to RV outflow, and systolic ejection is prolonged. Compared with the ability of the LV to compensate for the pressure overload imposed by aortic stenosis (AS), RV dysfunction from afterload mismatch occurs earlier in the course of PS and at lower peak systolic pressures, because the RV adapts less well to this type of hemodynamic burden. With nor mal systolic function and cardiac output (CO), severe PS is defined by a peak systolic gradient across the pulmonic valve of >64 mmHg (mean gradient >35 mmHg, Doppler jet velocity >4 m/s); moderate PS corre lates with a peak gradient of 36–64 mmHg (Doppler jet velocity 3–4 m/s). Mild PS is characterized by a jet velocity <3 m/s (peak gradient <36 mmHg). PS rarely progresses in patients with mild PS but may worsen with age in those with moderate disease due to valve thickening and calcification. The right atrial (RA) a wave elevates in relation to the higher pressures needed to fill a noncompliant, hypertrophied RV. A prominent RA v wave signifies functional tricuspid regurgitation (TR) from RV and annular dilation. The CO is maintained until late in the course of the disease. ■ ■SYMPTOMS Patients with mild or even moderate PS are usually asymptomatic and first come to medical attention because of a heart murmur (or early systolic click) that leads to echocardiography. With severe PS, patients may report exertional dyspnea or early-onset fatigue. Anginal chest pain from RV oxygen supply-demand mismatch and syncope may occur with very severe forms of obstruction, particularly in the pres ence of a destabilizing trigger such as atrial fibrillation, fever, infection, anemia, or pregnancy. ■ ■PHYSICAL FINDINGS The murmur of mild or moderate PS is mid-systolic in timing, cre scendo–decrescendo in configuration, heard best in the left second interspace, and usually introduced by an ejection sound (click) in younger adults whose valves are still pliable. The ejection sound is the only right-sided acoustic event that decreases in intensity with inspiration. This phenomenon reflects premature opening of the pulmonic valve by the elevated RV end-diastolic (post-atrial a wave) pressure. The

systolic murmur increases in intensity during inspira tion. With progressively severe PS, the ejection sound moves closer to the first heart sound and eventually becomes inaudible. A right-sided fourth heart sound may emerge. The systolic murmur peaks later and may persist through the aortic component of the second heart sound (A2). Pulmonic valve closure is delayed, and the pulmonic component of the second heart sound (P2) is reduced or absent. A prominent a wave, indicative of the higher atrial pressure necessary to fill the noncompliant RV, may be seen in the jugular venous pulse. A parasternal or RV lift can be felt with significant pressure overload. Signs of right heart failure due to RV systolic dysfunction, such as

TABLE 278-1 Causes of Pulmonic Valve Disease VALVE LESION ETIOLOGIES Pulmonic stenosis Congenital Carcinoid Tumor Endocarditis CHAPTER 278 Pulmonic regurgitation Primary valve disease Congenital Post-valvotomy Endocarditis Carcinoid Annular enlargement Pulmonary hypertension Idiopathic dilation Marfan syndrome Pulmonic Valve Disease hepatomegaly, ascites, and edema, are uncommon but may appear very late in the disease. ■ ■LABORATORY EXAMINATION The electrocardiogram (ECG) will show right axis deviation, RVH, and RA enlargement in adult patients with severe PS. Chest x-ray find ings include poststenotic dilation of the main PA in the frontal plane projection and filling of the retrosternal airspace due to RV enlarge ment on the lateral film. In some patients with RVH, the cardiac apex appears to be lifted off the left hemidiaphragm. The RA may also be enlarged. Transthoracic echocardiography (TTE) allows definitive diagnosis and characterization in most cases, with depiction of the valve and assessment of the jet velocity, gradient, RV function, PA pressures (which should be low), and any associated cardiac lesions. Transesophageal echocardiography (TEE) may be useful in some patients for improved delineation of the RV outflow tract (RVOT) and assessment of infundibular hypertrophy. Cardiac catheterization is not usually necessary for diagnostic purposes, but if performed, pressures should be obtained from just below and above the pulmonic valve with attention to the possibility that a dynamic component to the gradient may exist. The correlation between Doppler assessment of peak instantaneous gradient and catheterization-measured peak-to-peak gradient is weak. The latter may correlate better with the Doppler mean gradient. TREATMENT Pulmonic Stenosis Diuretics can be used to treat symptoms and signs of right heart failure. Provided there is less than moderate pulmonic regurgita tion (PR), percutaneous pulmonic balloon valvuloplasty is recom mended for symptomatic patients with moderate or severe PS and for asymptomatic patients with a peak gradient >64 mmHg (or mean gradient >35 mmHg). Surgery may be required when the valve is dysplastic (as seen in patients with Noonan’s syndrome and other disorders). A multidisciplinary heart team is best positioned to make treatment decisions of this nature. PULMONIC REGURGITATION PR may develop as a consequence of primary valve pathology, annu lar enlargement, or their combination; after surgical treatment of RVOT obstruction in children with such disorders as tetralogy of Fal lot; or after percutaneous pulmonic balloon valvotomy (Table 278-1). Carcinoid usually causes mixed pulmonic valve disease with PR and PS. Long-standing severe PA hypertension from any cause can result in dilation of the pulmonic valve ring and PR. Trace or mild PR of no hemodynamic or clinical consequence is frequently observed on TTE in the absence of structural pulmonic valve disease (Fig. 278-1).

PART 6 Disorders of the Cardiovascular System A B FIGURE 278-1 Pulmonic regurgitation. A. Transthoracic short axis window at the level of the aortic valve (AoV). The pulmonic valve (PV) is shown with the yellow arrow. Tricuspid valve (TV) leaflets are partially visualized (white arrows). B. Color flow Doppler image shows moderate pulmonic regurgitation (PR, white arrow). ■

■PATHOPHYSIOLOGY Severe PR results in RV chamber enlargement and eccentric hyper trophy. As is the case for aortic regurgitation (AR), PR is a state of increased preload and afterload. The diastolic pressure gradient between the PA to the RV, which drives the PR, progressively decreases throughout diastole and accounts for the decrescendo nature of the murmur. As RV diastolic pressure increases, the murmur becomes shorter in duration. The forward CO is preserved during the early stages of the disease but may not increase normally with exercise and declines over time. A reduction in RV ejection fraction may be an early indicator of hemodynamic compromise. In advanced stages, there is significant enlargement of the RV and RA with marked elevation of the jugular venous pressure. ■ ■SYMPTOMS Mild or moderate degrees of PR do not, by themselves, result in symp toms. Other issues, such as PA hypertension, may dominate the clini cal picture. With progressively severe PR and RV dysfunction, fatigue, exertional dyspnea, abdominal fullness/bloating, and lower extremity swelling may be reported. ■ ■PHYSICAL FINDINGS The physical examination hallmark of PR is a high-pitched, decre scendo diastolic murmur (Graham Steell murmur) heard along the left sternal border that can be difficult to distinguish from the more frequently appreciated murmur of AR. The Graham Steell murmur may become louder with inspiration and is usually associated with a loud and sometimes palpable P2 and an RV lift, as would be expected in patients with significant PA hypertension of any cause. Survivors of childhood surgery for tetralogy of Fallot or PS/pulmonary atresia may have an RV-PA conduit that is freely regurgitant because it does not contain a valve. PA pressures in these individuals are not elevated, and the diastolic murmur can be misleadingly low pitched and of short duration despite significant degrees of PR and RV volume overload. ■ ■LABORATORY EXAMINATION Depending on both the etiology and severity of PR, the ECG may show findings of RVH and RA enlargement. On chest x-ray, the RV and RA

may be enlarged. Pulmonic valve morphology and function can be assessed with transthoracic Doppler echocardiography. RV systolic pressure can be estimated from the tricuspid valve systolic jet veloc ity. Cardiac magnetic resonance (CMR) imaging can provide greater anatomic detail, particularly in patients with repaired congenital heart disease, and more precise assessment of RV volumes and function. Car diac catheterization is not routinely necessary but would be performed as part of a planned transcatheter PV procedure. TREATMENT Pulmonic Regurgitation In patients with significant functional PR due to PA hyperten sion and annular dilation, efforts to reduce pulmonary vascu lar resistance and pressure should be pursued. Such efforts may include pharmacologic/vasodilator and/or surgical/interventional strategies, depending on the cause of the PA hypertension (e.g., idiopathic PA hypertension, left-sided heart valve disease). Diuret ics can be used to treat the manifestations of right heart failure. Surgical valve replacement for primary, severe, pulmonic valve disease, such as carcinoid or endocarditis, is rarely undertaken. Transcatheter pulmonic valve replacement has been successfully performed in many patients with severe PR after childhood repair of tetralogy of Fallot or pulmonic valve stenosis or atresia. This procedure was introduced clinically prior to transcatheter aortic valve replacement. Earlier concerns regarding an excess hazard of infective endocarditis with first-generation valves have led to pro cedural modifications. ■ ■FURTHER READING Otto CM et al: 2020 AHA/ACC guideline for the management of patients with valvular heart disease. A report of the American Col lege of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. Circulation 143:e72, 2021. Stout KK et al: 2018 ACC/AHA guidelines for the management of adults with congenital heart disease. J Am Coll Cardiol 73:e81, 2019.

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