

RIGHT VENTRICULAR OUTFLOW TRACT OBSTRUCTION

Definitions and Classification of RVOT Obstruction

Obstruction to the right ventricular outflow tract (RVOT) in the adult patient can be either congenital or acquired. Congenital obstruction can be at the pulmonary valve, below the pulmonary valve, or above the valve. Below the pulmonary valve, obstruction can be either at the infundibular (as happens in Fallot) or the subinfundibular level.¹

Valvular Pulmonary Stenosis

Obstruction of the RV outflow tract is valvular in 90% of patients. Congenital pulmonary stenosis constitutes 10% of the cases of congenital heart disease in adults.¹ Valvar pulmonary stenosis may be an isolated abnormality or in association with a VSD or Noonan syndrome (dysplastic pulmonary valve, facial dysmorphism, cardiac defects and variable cognitive deficits). The leaflets are thin and pliant. In 10-15% of patients the valve is dysplastic and the leaflets thickened and immobile, composed of myxomatous tissue.^{1,2}

Presentation

Adults may be asymptomatic. With severe stenosis, exercise intolerance, SOB, anginal pain and syncope may occur. Gradients >30 mm Hg may deteriorate with aging and result to RV hypertrophy and eventually right heart failure (elevated JVP, hepatic congestion, ascites, and peripheral edema).

Physical Examination

RV heave and thrill may be present in moderate/severe PS

S₂ widely split but moves normally with respiration

Pulmonary ejection click

Crescendo-decrescendo murmur increased by inspiration

Investigations

ECG may show right axis deviation and RV hypertrophy.

Chest radiography may show post-stenotic dilatation of the main PA and diminished vascular markings.

Echocardiography reveals RV hypertrophy and paradoxical septal motion. Doppler mean gradients are considered (Table 1).³

Computed Tomography Angiography (CTA) with optimized protocols is useful to delineate the anatomy, although assessment of smaller PV stents and subtotal-occluded vessels is challenging with conventional scanners.⁴

Magnetic Resonance Imaging (MRA) avoids ionizing radiation but is limited by lower spatial resolution, loss of signal with PV stents, and reduced access compared with CTA⁴.

Stress testing with pulse oximetry may reveal a right to left shunt in the setting of severe pulmonary stenosis and an associated patent foramen ovale.

Cardiac catheterization is not necessary for diagnosis. It may be used for accurate gradient assessment in patients with echo peak gradients >30 mm Hg for consideration of balloon valvotomy.

Therapy

Asymptomatic patients with mild stenosis do not need intervention (Figure 1, and Tables 2-4). Balloon valvotomy is the treatment of choice for symptomatic patients with less than moderate PS and peak gradient > 50 mm Hg (mean > 30 mm Hg), or asymptomatic patients with peak gradient > 60 mm Hg (mean > 40 mm Hg).^{5,6} Paclitaxel-coated balloons and the use of stents may offer additional advantages.⁴ Surgical valvotomy or replacement with a bioprosthetic valve is reserved for patients with significant PR (an ominous prognostic sign for subsequent RV dilation and failure) or dysplastic valves unsuitable for balloon valvuloplasty.⁷ Rare complications of balloon valvotomy are PR, pulmonary edema, cardiac perforation, AV block and transient reactive RVOT obstruction. Transcatheter pulmonary valve implantation is also feasible.⁸ Recommendations for PR after repair of PS are provided in Tables 5 and 6, and Figure 2.

Subvalvular Pulmonary Stenosis

Usually occurs in association with tetralogy of Fallot (before or after surgery) or double-chambered RV.² In double-chambered RV a muscular band divides the RV into a high-pressure proximal inflow portion and a low-pressure distal outflow chamber. VSD or membranous subaortic stenosis may also be present. When the obstruction is mild, these patients may present with exercise intolerance and a harsh systolic murmur as young adults. With more severe obstruction and hypertrophy, ventricular arrhythmia, syncope, and sudden cardiac death may be the first presenting sign. Surgical muscle resection with or without patching of the outflow tract is indicated in symptomatic patients with peak Doppler gradient >60 mm Hg (mean 40 mm Hg).

Supravalvar Pulmonary Stenosis

Supravalvar pulmonary stenosis can occur as an isolated abnormality or in association with complex cardiac malformations such as Williams syndrome (infantile hypercalcemia, elfin facies, and mental retardation), tetralogy of Fallot, Noonan syndromes, and rubella or toxoplasmosis infection during the first trimester of pregnancy.¹ It may also be the result of surgical scarring from operations such as pulmonary artery banding or arterial switch. Patients with exercise intolerance or significant gradients are referred for surgery. Pulmonary artery stenting can be successfully achieved in most cases, but adverse events are common.⁹

Branch Pulmonary Artery Stenosis

They may be isolated or multiple as happens in the Williams and Noonan syndromes, congenital rubella and Alagille (intrahepatic cholestasis) and Keutel (cartilage calcification and brachytelephalangia) syndromes.¹ Although uncommon, the diagnosis of branch PA stenosis should always be considered in patients with a history of congenital heart disease who present with symptoms of pulmonary embolism such as dyspnea, fatigability, and segmental lung ventilation-perfusion mismatches. These patients should be evaluated and followed-up by echocardiography- Doppler plus 1 of the following: MRI angiography, CT angiography, or contrast angiography. Balloon and stent angioplasty are used in isolated stenoses with >50% diameter stenosis and RVSP>50 mm Hg (Tables 7 and 8).

Right ventricle to Pulmonary Artery conduits

RV to PA conduits are widely used in the treatment of severe RVOT obstructive lesions including pulmonary atresia. These conduits may be homografts or prosthetic conduits with bioprosthetic valves used within the conduit. A minority of conduits may show early dysfunction because of kinking or aneurysmal dilation. The remainder will become dysfunctional over time and usually require replacement or intervention because of progressive stenosis within the conduit or at the valve, and/or valvular regurgitation, at a mean interval of 10 to 15 years from placement. Transcatheter pulmonary valve replacement is now an option in patients with an anatomically suitable prosthetic pulmonary conduit or valve.¹⁰ Recommendations are provided in Tables 9-11.

Double-chambered RV

Double-chambered right ventricle is uncommon in adults. Hypertrophied muscle bundles develop in the RV cavity, creating RVOT obstruction, commonly associated with a VSD.¹¹ Recommendations are provided in Tables 12 and 13.

Pregnancy in RVOT obstruction

Pulmonary valve stenosis is generally well tolerated. In severely symptomatic PS, which is unresponsive to medical therapy and bed rest, percutaneous valvuloplasty can be appropriate (Table 14).¹² Pregnancy after RVOT reconstruction with an allograft conduit is safe, but at the cost of higher incidence of pre-term delivery and children small for gestational age.¹³

References

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