

PULMONARY STENOSIS

CAUSES & PATHOLOGY

- 1st cause: **congenital** (7-10% congenital patients) – fusion of the valve leaflets
 - o Associated with ASD, peripheral pulmonary artery stenosis, **Noonan, Williams, Alagille syndromes** (inheritance is low)
 - o Valve is **dome shaped with narrow opening**, fusion of leaflets and calcifications (adults)
 - o Dysplastic if leaflets are thick without fusion of cusps
- Rheumatic inflammation, carcinoid (rare)
- Obstruction of ejection from RV → ↑RV pressure → RV hypertrophy → RV dilation → ↓ CO

Congenital

- Neonates with **hypoplastic RV + shunt R-L through PFO**
- Balloon dilation is the procedure of choice to relieve obstruction
- Pulmonary valvotomy of systemic to pulmonary arterial shunt if underdevelopment of RV or in moderate to severe stenosis, even in absence of symptoms
- Most patients are **asymptomatic** and develop symptoms on exertion

PHYSICAL EXAM

Unoperated adult

- Prominent *a* wave (RV hypertrophied)
- RV lift
- Systolic thrill LSB
- Normal S₁
- **Delayed P₂ due to prolonged ejection time**
- **Delayed splitting S₂ with increased obstruction and persistent**
- S₄
- Systolic ejection **click** upper LSB that is louder during **EXPIRATION**
 - o ↓ inspiration owing to premature opening of the pulmonary valve by the atrial kick into the stiff RV
- **Harsh crescendo-decrescendo systolic ejection murmur heart best at upper LSB**, which radiates to the **back** and may be augmented with inspiration – peak later in systole with increasing obstruction

Physical signs suggestive of severe obstruction in pulmonary stenosis:

- Cyanosis and clubbing
- Widely split S₂
- Reduced or absent P₂
- Short S₁ ejection click interval
- Long systolic ejection murmur
- Peak of murmur late in systole

ELECTROCARDIOGRAPHY

- Right axis deviation
- RV hypertrophy
- RA enlargement

CHEST X-RAY

- Prominent main pulmonary artery
- Vascular fullness in the left lung base greater than the right base (**Chen's sign**)
 - o Preferential pulmonary flow to the left lung in patients with PS
- RA and RV enlargement and decreased pulmonary vascular markings in severe PS
- Pulmonary valve calcification (occasional)

ECHOCARDIOGRAPHY

$$\Delta P = 4V_{PS}^2$$

Degree of severity based on echo **peak instantaneous gradient** because of a good correlation between maximal Doppler systolic pulmonary gradient and catheter peak-to-peak gradient (lower pressures on right side)

	Mild	Moderate	Severe
Peak velocity (m/s)	< 3	3 - 4	> 4
Peak gradient (mm Hg)	< 36	36 - 64	> 64
RV systolic pressure	25 - 49	50 - 79	> 80
Transvalvular pressure gradient (mm Hg)	50 - 74	75 - 100	> 100

★ **Suggested reference:** Baumgartner H. et al. Echocardiographic assessment of valve stenosis: EAE/ASE recommendations for clinical practice. 2009 J Am Society Echocardiography;22:1-23 – **Tables 22**

MRI to assess level of obstruction, size and function of RV, size of pulmonary arteries

CATHETERIZATION

- Oxymetry run – shunts
- Severity and level of obstruction
- Tricuspid valve and pulmonary artery abnormalities
- Coronary artery anatomy

OUTCOME / COMPLICATIONS

Unoperated adult

- RV hypertrophy
- Symptoms of decrease exercise tolerance, dyspnea, fatigue, syncope, chest pain, palpitations
- Mild cyanosis and clubbing in patients with severe obstruction (ASD or R-L shunt from PFO)
- Complications: subvalvular dynamic obstruction, RV heart failure, arrhythmias, sudden death

Operated adult

- No cyanosis after successful intervention
- Soft systolic ejection murmur may be heard in 2nd intercostal space
- Surgical valvotomy
 - o RBBB

- Main and left pulmonary arteries remains dilated even after successful relief of obstruction
- Balloon pulmonary valvuloplasty
 - 7% with moderate to severe PR
 - 6,5% with immediate reduction of gradient < 36 mmHg
- Complications: pulmonary regurgitation, RV dilation, RV heart failure, arrhythmias, sudden death

CCS congenital guidelines 2009

Indications for intervention/reintervention/medical therapy in patients with valvar RVOTO :

- **Symptomatic**: peak instantaneous gradient > 50 mm Hg or mean gradient > 30 mm Hg
- **Asymptomatic**: peak instantaneous gradient > 60 mm Hg or mean gradient > 40 mm Hg
 - Consider balloon valvotomy
- Patients with arrhythmias (sustained atrial flutter), associated ASD or VSD with R-L shunt, recurrent endocarditis

★ **Suggested reference**: Silverside C. K. et al. Canadian cardiovascular Society 2009 consensus conference on the management of adults with congenital heart disease : executive summary. Can J Cardiol ;26:143-150.

FOLLOW-UP

With a congenital cardiologist :

- R-L shunt
- RVOTO
- Moderate to severe pulmonary regurgitation
- Recurrent stenosis, subvalvular stenosis (infundibulum), tricuspid regurgitation with RV failure, atrial and ventricular arrhythmias

Content of this summary from these references:
<ul style="list-style-type: none"> • Otto C & Bonow R. Valvular Heart Disease. (2012) In Bonow R. <i>et al.</i> Braunwald's Heart Disease, 9th edition, pp. 1468-1539. Philadelphia, PA: Elsevier. • Nishimura R. A. et al. 2014 ACC/AHA guidelines for the management of patients with valvular heart disease. J Am Coll Cardiol 2014;63:e57-185. • Baumgartner H. et al. Echocardiographic assessment of valve stenosis: EAE/ASE recommendations for clinical practice. 2009 J Am Society Echocardiography;22:1-23. • Silverside C. K. et al. Canadian cardiovascular Society 2009 consensus conference on the management of adults with congenital heart disease : executive summary. Can J Cardiol ;26:143-159.