CAUSES & PATHOLOGY

- 1st cause: **congenital** (7-10% congenital patients) – fusion of the valve leaflets
  - Associated with ASD, peripheral pulmonary artery stenosis, Noonan, Williams, Alagille syndromes (inheritance is low)
  - Valve is **dome shaped with narrow opening**, fusion of leaflets and calcifications (adults)
  - 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 persistant
- S4
- Systolic ejection click upper LSB that is louder during **EXPIRATION**
  - 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 of 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)

<table>
<thead>
<tr>
<th></th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
</tr>
</thead>
<tbody>
<tr>
<td>Peak velocity (m/s)</td>
<td>&lt; 3</td>
<td>3 - 4</td>
<td>&gt; 4</td>
</tr>
<tr>
<td>Peak gradient (mm Hg)</td>
<td>&lt; 36</td>
<td>36 - 64</td>
<td>&gt; 64</td>
</tr>
<tr>
<td>RV systolic pressure</td>
<td>25 - 49</td>
<td>50 - 79</td>
<td>&gt; 80</td>
</tr>
<tr>
<td>Transvalvular pressure gradient (mm Hg)</td>
<td>50 - 74</td>
<td>75 - 100</td>
<td>&gt; 100</td>
</tr>
</tbody>
</table>


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 heart 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


**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

<table>
<thead>
<tr>
<th>Content of this summary from these references:</th>
</tr>
</thead>
</table>