PULMONARY VALVE STENOSIS

Synonyms:
- Congenital pulmonary stenosis

Definition:
In this disease, the pulmonary valve is pathological and creates an obstruction to blood flow from the right ventricle to the pulmonary artery.
This is a relatively common disease, representing 80-90% of all right ventricular outflow tract obstructions and 8% of all congenital heart diseases.
Basically, the valve may have 3 different morphologies. The most common one is the **dome-shaped valve**, with a central narrow hole without recognizable leaflets.

The next most frequent one is the **dysplastic valve** with 3 thickened leaflets with fused comisures. This is the most frequent type in Noonan syndrome, in which 50% of the patients has a congenital heart disease, this being the most frequent.
**Monocuspid** and **bicuspid** pulmonary valves belong to the third group. Both are rare.
Valvular stenosis generates reactive infundibular hypertrophy (abnormal muscle growth in the right ventricular outflow tract) and dilation of the main pulmonary artery and its left branch, especially when the valve is dome-shaped.

Clinical Picture:
Patients with mild or moderate stenosis are usually asymptomatic for many years. Diagnosis is suspected during auscultation of a murmur accompanied sometimes by a thrill (vibration felt in the chest) in a routine check. Patients with severe stenosis may present with dyspnea (shortness of breath), especially during exercise. Some patients can
also experience syncope (fainting) and chest pain (See Clinical Picture in Diagnosis and Treatment). When an atrial septal defect coexists, the patient may become cyanotic (bluish).

Some newborns may have severe stenosis, the so-called critical pulmonary stenosis of the newborn. Cyanosis is important from birth, as it is almost always associated with a large atrial septal defect and varying degrees of hypoplasia (under development) of the right ventricle.

Mild valvular pulmonary stenosis in adults tends not to progress. The moderate degree may progress to valvular calcification or generate reactive infundibular hypertrophy.

**Diagnosis:**

As mentioned before, diagnosis is usually made during the auscultation of a murmur during medical check. Chest x-ray shows a convexity on the left edge of the heart by dilation of the pulmonary artery trunk and its left branch.

Transthoracic echocardiography is the best diagnostic method. It shows the area of obstruction, the thickening of the valve and the pulmonary artery dilation. The severity of the obstruction can be categorized by measuring right ventricular pressure and the gradient through the valve. In milder forms, the right ventricular pressure is less than 50% of the pressure of the left ventricle and aorta, and the gradient is less than 35-40 mmHg. In moderate types, pressure is between 50 and 75%, and the gradient between 40 and 60-70 mmHg. In severe ones, pressure exceeds 75% and the gradient is greater than 60-70 mmHg (See Diagnosis in Diagnosis and Treatment).

While catheterization as a diagnostic method is usually not necessary, it is performed frequently, because the treatment of choice in this disease is balloon dilation. It gives the same information as the echocardiogram, and measures pressures and gradients accurately (see Hemodynamics in Diagnosis and Treatment).
Treatment:
Patients with mild stenosis do not require treatment. When stenosis is moderate, and the patient has symptoms or cardiomegaly (enlarged heart), some intervention shall be performed, as in the case of severe stenosis. The treatment of choice is percutaneous balloon dilation. Guides and catheters are inserted through a vein in the groin. As the guide reaches the heart, it is passed through the pulmonary valve and the balloon is placed inside. It is later inflated, tearing the valve to enlarge its opening and relieve the obstruction.
In patients for whom balloon dilation has proved ineffective, surgery should be indicated (See Surgery in Diagnosis and Treatment). With the aid of cardiopulmonary bypass, surgical valvuloplasty is performed by opening the valve with a scalpel and relieving the obstruction. In dysplastic valves, the comisures connecting the leaflets are opened, occasionally removing one or more of them in order to relieve the obstruction.

Treatment results are excellent, both by catheterization and surgery, relieving the stenosis in most of the cases. The postoperative period is usually simple, with little risk of complications and early discharge (See Postoperative in Diagnosis and Treatment).

Prognosis:
Quality of life after treatment is usually excellent, with normal aerobic capacity. Balloon dilation and surgical valvuloplasty might progress to pulmonary regurgitation (blood back flow from the pulmonary artery into the right ventricle). If this is of significant magnitude, it might require a pulmonary valve replacement years later (See Follow-up and Control in Diagnosis and Treatment).