Chapter 6

Critical Pulmonic Stenosis

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Case Scenario

A 5-day-old male presents with tachypnea and poor feeding after being discharged home on day 2 of life. His mother had an uneventful pregnancy with routine prenatal care and testing and he was born via spontaneous vaginal delivery without complications. On day 4 of life his mother noted that his breathing was fast and he was not eating well and decided to seek care. At the pediatrician's office the patient was noted to have room air oxygen saturations of 75%.

Transthoracic echocardiography showed the following:

- *Critical valvular pulmonary stenosis, peak gradient 72 mm Hg*
- *Intact ventricular septum*
- *Mild right ventricular hypertrophy*
- *Normal left ventricular function*
- *Mild to moderately depressed right ventricular function*
- *Moderate tricuspid regurgitation*
- *Patent foramen ovale with bidirectional shunting*

He was admitted to the congenital cardiac intensive care unit and was started on prostaglandin E_1 and milrinone. He is scheduled for balloon valvuloplasty of the pulmonic valve in the cardiac catheterization laboratory.

Key Objectives

- Describe the role of the ductus arteriosus during fetal life.
- Discuss echocardiographic findings related to pulmonary stenosis.
- Describe perioperative management of a patient with critical pulmonary stenosis.

Pathophysiology

What is the pathophysiology of critical pulmonary stenosis?

During fetal development, little blood flows through the lungs because of high pulmonary vascular resistance (PVR). During fetal life, the majority of pulmonary artery blood flow is directed through the ductus arteriosus (DA) to the aorta, flowing right to left (R-to-L). After birth, PVR is initially elevated and then decreases over the first few days of life. As pulmonary pressures become lower than systemic pressures, flow in the DA reverses, becoming left-to-right (L-to-R) from the aorta to the pulmonary artery.

In normal infants the DA is not needed after birth and begins to functionally close during the first 24–72 hours after birth. It is anatomically closed between the third and fourth week of life. Persistent patency of the DA can be caused by stress, hypoxia, and acidosis.

In this infant, critical PS limited the amount of antegrade pulmonary blood flow (PBF) through the pulmonary valve. Therefore, the major source of source of PBF was provided via the DA, flowing from the aorta to pulmonary artery. (See Figure 6.1.) As the DA began to close during the first few days of life, PBF was reduced. Antegrade blood flow through the critically stenosed pulmonic valve was not sufficient and the infant became hypoxemic, causing tachypnea and poor feeding. (See Figures 6.2 and 6.3.)

How is critical pulmonary valve stenosis defined?

Intervention is recommended for patients with a gradient across the pulmonary valve >50 mm Hg.

The following echocardiographic classifications are defined:

- **Mild PS**: Peak instantaneous gradient <30 mm Hg
- **Moderate PS**: Peak instantaneous gradient between 30 and 60 mm Hg
- **Severe PS**: Peak instantaneous gradient >60 mm Hg

Clinical Pearl

Echocardiographic and direct catheter measurement of PV gradients vary, as they use different measurement techniques. The peak instantaneous Doppler gradient can overestimate the catheter gradient by 20–30 mm Hg, and the mean echocardiographic gradient may underestimate the gradient compared to catheterization. During cardiac catheterization the pressures on each side of the valve are directly transduced.