Chapter 14

Critical Aortic Stenosis

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Case Scenario

A 2-day-old male neonate, diagnosed with critical aortic stenosis, is scheduled for emergent balloon valvuloplasty in the cardiac catherization laboratory. He was born at 38 weeks gestational age via spontaneous vaginal delivery. His Apgar scores were 9 at 1 and 5 minutes, and he was admitted to the neonatal intensive care unit due to mild tachypnea and a murmur on auscultation. No obvious dysmorphic features were noted. On day 2 of life he was noted to be more tachypneic and tachycardic, with diminished pulses, cool extremities, and poor capillary refill. A chest radiograph revealed cardiomegaly and pulmonary edema.

Venous lactate is 4 with a blood pH of 7.1. A prostaglandin E_1 infusion and low-dose inotropic support were started.

Transthoracic echocardiogram was remarkable for the following:

- *A dysplastic aortic valve with a mean gradient of 45 mm Hg*
- *A dilated, thick left ventricle with qualitatively moderately reduced function*
- *No patent ductus arteriosus*

Key Objectives

- Define critical aortic stenosis.
- Discuss the options for treatment of a patient with ductal-dependent aortic stenosis.
- Describe the preoperative assessment and intraoperative management of this patient.
- Discuss the potential complications and expected outcomes of balloon valvuloplasty.
- Discuss the postprocedural management and disposition.

Pathophysiology

What is critical aortic stenosis?

Critical aortic stenosis (AS) is defined as the presence of severe aortic valve stenosis with systemic perfusion that is dependent on right ventricular (RV) output through a patent ductus arteriosus (PDA). Mean gradients of >40 mm Hg or peak-to-peak gradients >50 mm Hg are usually reported. As the PDA begins to close, left ventricular function deteriorates, with signs of decreased perfusion to end organs such as the kidneys, gastrointestinal tract, and brain, manifesting as renal failure, necrotizing enterocolitis, and intracerebral bleeds, respectively. Critical AS is not defined by an absolute valve area or gradient because in patients with ventricular dysfunction (either systolic or diastolic) critical AS may exist with larger valve areas and gradients may be underestimated. (See Figure 14.1.)

Clinical Pearl

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What is the pathophysiology of AS in a neonate?

Aortic stenosis in older infants, children and adults leads to left ventricular (LV) pressure overload and progressive LV concentric hypertrophy and failure, with resultant elevation in end-diastolic pressure and pulmonary edema. The hypertrophied LV thus becomes susceptible to subendocardial hypoperfusion and ischemia.

In neonates with critical AS, a PDA provides systemic cardiac output via flow from the RV to the descending aorta via right-to-left (R-to-L) shunting, and thus systemic perfusion is preserved. The degree of LV hypertrophy in these neonates, though not as severe, still exists, and the ventricle may be dilated and poorly contractile. This heart failure is associated with increases in heart rate and LV end-diastolic pressure which can lead to inadequate coronary blood flow and the risk of ischemia. The elevated left atrial (LA) pressure leads to pulmonary edema and pulmonary hypertension with dilation of the RV; in addition,