Chapter 26

Stage I Palliation, Hypoplastic Left Heart Syndrome

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Case Scenario

A 6-week-old infant weighing 4 kg is scheduled for laparoscopic Nissen fundoplication with gastrostomy tube placement. She was born at term with hypoplastic left heart syndrome, for which she underwent a Stage I palliation (Norwood procedure) with a Sano modification at age 5 days. Her initial postoperative course was uncomplicated, but she has had persistent difficulty feeding and has failed to gain weight. She was discharged from the hospital on furosemide and aspirin.

Transthoracic echocardiogram at the time of discharge 3 weeks earlier showed the following:

- *Mild-to-moderate tricuspid regurgitation*
- *Mildly diminished right ventricular systolic function*
- *A patent Sano (right ventricular to pulmonary artery) shunt with a 40 mm Hg peak gradient*

Despite placement of a nasogastric tube for feeding, she has gained little weight and has persistent reflux. She appears alert, although small and thin, with a heart rate of 150 beats/minute, respiratory rate of 40 breaths/minute, and SpO_2 86% on room air. Her cardiologist would like surgery to take place as soon as possible.

Key Objectives

- Describe the anatomy and physiology of hypoplastic left heart syndrome.
- Describe available options for the initial palliation of hypoplastic left heart syndrome.
- Describe the anatomy and physiology of a patient who has undergone surgical Stage I palliation.
- Describe an appropriate plan for perioperative anesthetic management of this infant.
- Identify markers of inadequate pulmonary or systemic blood flow in infants after Stage I palliation.
- Describe the physiology of laparoscopy as it pertains to this population.
- Discuss perioperative complications related to this population.

Pathophysiology

What is hypoplastic left heart syndrome?

The term "hypoplastic left heart syndrome" (HLHS) is used to describe a spectrum of congenital cardiac abnormalities involving underdevelopment of left-sided heart structures. Findings may include mitral valve stenosis or atresia, aortic stenosis or atresia, hypoplasia or absence of the left ventricle (LV), and hypoplasia of the ascending aorta and aortic arch. Hypoplastic left heart syndrome occurs relatively commonly (approximately 1 in 4000 births).

The combination of these defects results in single-ventricle physiology, wherein a single ventricle, in this case the right ventricle (RV), is required to support both pulmonary and systemic circulations. Single-ventricle physiology requires complete intracardiac mixing of pulmonary venous and systemic venous blood that is then supplied to parallel pulmonary and systemic circuits.

Clinical Pearl

Hypoplastic left heart syndrome is a spectrum of congenital cardiac abnormalities involving underdevelopment of left-sided heart structures and resulting in physiology wherein a single ventricle, in this case the RV, is required to support both pulmonary and systemic circulations.

What is the circulatory pattern in patients with HLHS after birth and what does "ductal-dependent" mean?

In patients with HLHS, due to the inadequacy of the left-sided heart structures, blood flow through the native LV outflow tract is limited and therefore flow to the systemic circulation after birth is dependent on flow provided via the patent ductus arteriosus (PDA). In patients with aortic atresia, the child is completely dependent on the PDA for blood flow to the coronary and cerebral circulations and