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Single-Ventricle Lesions

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Introduction

The term single ventricle or univentricular heart applies to a wide variety of anatomic lesions (Table 12.1) and their subsequent postoperative anatomy. Single-ventricle physiology presents unique challenges to the intensive care physician, as these children often respond to common interventions such as supplemental oxygen, mechanical ventilation, and vasoactive infusions differently than children with more conventional circulatory physiology. Infants and children with single-ventricle physiology undergo multiple cardiac operations throughout the course of their lives and may be more adversely affected by intercurrent illnesses than other children with congenital heart disease. As such, these patients are commonly encountered in pediatric critical care medicine and frequently have chronic cardiac problems that often require intensive care. This unique physiology represents the sine qua non of pediatric cardiac intensive care, and it is imperative that pediatric intensivists have a thorough understanding of the nuances of single-ventricle physiology. This chapter addresses the important physiologic issues that arise in the care of infants and children with single-ventricle physiology.

The Neonate with Single-Ventricle Physiology

Although virtually all newborns with single-ventricle physiology have mixing of pulmonary and systemic venous return, the relative amounts of each vary substantially depending on the underlying anatomy. The most important anatomic issue that dictates management is the outflow to and from the systemic ventricle and lungs. The neonate with single-ventricle anatomy may have (1) unobstructed pulmonary blood flow and obstructed systemic blood flow; (2) unobstructed systemic blood flow and obstructed pulmonary blood flow; or (3) bilaterally unobstructed outflow. Additionally, either systemic or pulmonary venous return may also be obstructed in the newborn single-ventricle circulation. Early survival is largely dependent on achieving a balanced circulation without excessive pulmonary blood flow, yet with enough pulmonary blood flow to prevent severe cyanosis.

Systemic Outflow Obstruction

Systemic outflow obstruction is characteristic of hypoplastic left heart syndrome (HLHS) (Figure 12.1) (see later), tricuspid atresia with transposed great arteries [1–4], double-inlet left ventricle (DILV) [1–8], and other less common anatomic variations. Single-ventricle physiology with systemic outflow obstruction also pertains to newborns with critical aortic stenosis, severe coarctation of the aorta, or interrupted aortic arch (discussed in Chapter 10). The important features of this type of anatomy are complete mixing of systemic and pulmonary venous return and ventricular outflow directed primarily to the pulmonary artery. Systemic blood flow (Qs) is provided largely by right-to-left shunting across the patent ductus arteriosus (PDA) and is dependent on the relative pulmonary and systemic vascular resistances. In general, systemic outflow obstruction is poorly tolerated and in the face of single-ventricle anatomy is usually accompanied by signs or symptoms of profound shock.

Hypoplastic left heart syndrome encompasses a continuum of congenital heart lesions producing left-sided obstruction and hypoplasia and generally ranges from severe, with mitral and aortic atresia with a diminutive left ventricle to mitral and/or aortic stenosis with a nonapex-forming left ventricle. Although biventricular repair may be possible for those infants at the more favorable end of the spectrum (i.e., mitral stenosis, aortic stenosis) [9–11], the severe end of the spectrum is universally fatal, with an average life expectancy of around 5 days (untreated) [12]. In contrast to most neonates born with complex congenital heart disease, these children usually are otherwise normal with no abnormalities of other organ systems. Even with a staged surgical approach to palliation (discussed later), HLHS carries a substantial risk of morbidity and mortality, with hospital survival following Norwood stage I palliation as low as 47% [13]. The most optimistic rates of mortality in the current era range from 14% to 25% with a staged surgical approach to palliation [14–20], while the mortality rate in centers that favor heart transplantation is approximately 30%, including those infants who die while waiting for transplant [21,22]. A multicenter review of 323 neonates with HLHS reported an overall

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30-day mortality rate of 33% for either approach (transplant or staged palliation) [23]. There are many excellent reviews and chapters that discuss the myriad issues pertaining to the evaluation and management of infants with HLHS in great detail—these would be difficult to improve on and to attempt to do so is well beyond the intended scope of the present discussion—the reader is therefore directed to other reviews for additional information [24–34].

### Pulmonary Outflow Obstruction

Single-ventricle physiology with pulmonary outflow obstruction is characteristic of lesions such as tricuspid atresia, pulmonary atresia, and severe Ebstein's anomaly of the tricuspid valve. The salient anatomic features are complete mixing of systemic and pulmonary venous return and ventricular outflow predominantly directed out the aorta. Low pulmonary blood flow (Qp) in these patients implies an obligate right-to-left shunt, generally at the atrial level, and results in deoxygenated blood reaching the systemic circulation and, hence, cyanosis. The clinical consequences of low Qp are variable and depend on the degree of pulmonary outflow obstruction. Mild obstruction may permit an inordinate amount of the total cardiac output to go to the pulmonary circulation, sometimes at the expense of systemic cardiac output. Treatment is therefore directed at limiting, rather than increasing, Qp. Infants with this type of anatomy may be only minimally cyanotic and can have signs and symptoms of congestive heart failure. At the other end of the spectrum are those infants with severe pulmonary outflow obstruction or even atresia. These patients are profoundly cyanotic unless an alternate source of Qp is quickly established.

### Obstructed Venous Return

Unobstructed pulmonary or systemic venous return in infants with single-ventricle anatomy frequently depends upon an unrestrictive interatrial communication. When one of the atroventricular (AV) valves is severely stenotic or atretic, as occurs in HLHS, tricuspid atresia, or pulmonary atresia with intact ventricular septum, a large atrial septal defect is mandatory for decompression of the atrium with the inadequate AV valve. Obstruction of the systemic venous atrium causes increased central venous pressures and third spacing of fluid, eventually limiting systemic cardiac output and producing signs and symptoms of shock. Although a patent foramen ovale allows for some right-to-left shunting of blood across the atrial septum, it may be inadequate to permit unobstructed flow of all systemic venous return.

Obstruction of the pulmonary venous return causes elevated pulmonary venous pressures and pulmonary hypertension. This phenomenon may be helpful in the immediate neonatal period, as it can limit Qp and enhance systemic flow, thereby increasing systemic oxygen delivery (DO₂), even if at the expense of arterial oxygen saturation (SaO₂). Nevertheless, the atrial septum must be opened at the time of the first palliative operation to avoid the long-term consequences of elevated pulmonary vascular resistance. A severely restrictive or intact atrial septum with pulmonary venous hypertension usually requires emergent creation of an atrial level shunt because of profound cyanosis. These procedures carry a high risk of morbidity and may imply a worse prognosis for further palliative surgery [35–38].

### Postoperative Anatomy

The goal to any palliative surgery is to establish (1) unobstructed pulmonary and systemic venous return, (2) unobstructed systemic outflow, and (3) a regulated source of pulmonary blood flow. Typically, this is accomplished via a stage 1 Norwood type procedure (Figures 12.2 and 12.3), modified systemic-to-pulmonary artery (Blalock-Taussig) shunt, or pulmonary artery band (the latter two