Chapter 13

# Ebstein Anomaly, Palliated

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**Case Scenario**

A 2-year-old boy, weighing 11.5 kg, is seen in the preoperative clinic prior to planned outpatient ptosis repair. He is currently growing and developing well despite a complex cardiac history. The patient was born at 32 weeks gestational age with an antenatal diagnosis of severe Ebstein anomaly. He was intubated shortly after birth for hypoxia and required an emergent Starnes procedure (oversewing of the tricuspid valve and placement of a systemic-to-pulmonary artery shunt) at day 2 of life. His postoperative course was complicated by cardiac dysfunction and need for extracorporeal membrane oxygenation for 3 days. He remained stable following decannulation and was discharged home at 6 weeks of age. At age 5 months he underwent an uneventful bidirectional Glenn shunt (superior vena cava to right pulmonary artery anastomosis) and ligation of previous systemic-to-pulmonary artery shunt. His only current medication is aspirin.

## Key Objectives

- Describe the presentation, anatomy, and decision making for newborns with severe Ebstein anomaly.
- Describe cardiac surgical options for patients with severe Ebstein anomaly.
- Describe pulmonary blood flow in patients after bidirectional Glenn procedure and expected hemoglobin–oxygen saturations.
- Discuss anesthetic management for patients having noncardiac surgery after bidirectional Glenn procedure, including perioperative risk stratification and planning.

## Pathophysiology

### What is Ebstein anomaly?

Ebstein anomaly is a rare and highly variable congenital abnormality, comprising fewer than 1% of all congenital heart disease (CHD), affecting the tricuspid valve (TV) and adjacent right ventricular myocardium. The TV is dysplastic and displaced inferiorly into the RV. Valve leaflets may be larger or smaller than normal, resulting in failure of coaptation with resultant tricuspid regurgitation (TR) of varying degrees. Depending on the degree of apical TV displacement, there is loss of effective right ventricular volume and size, with "atrialization" of the right ventricle (RV). Right ventricular outflow tract obstruction can exist, limiting functional ejection and resulting in decreased pulmonary blood flow (PBF). Ebstein anomaly represents a wide spectrum of anatomic and clinical presentations, ranging from minimally symptomatic patients who require medical management over time to critically ill neonates with severe cyanosis who require intensive medical and surgical intervention. (See Chapter 12 and Figures 13.1 and 13.2.)

**Clinical Pearl**

*Ebstein anomaly is a highly variable lesion in which forward flow through the right heart can be limited because of an apically displaced, dysplastic, and incompetent TV with resultant functional impairment of the "atrialized" RV.*

### What are the anatomic characteristics of Ebstein anomaly?

Newborns with severe Ebstein anomaly present with significant apical displacement of the TV, leading to severe TR and an ineffective RV. Nearly all patients have an atrial communication [either an atrial septal defect (ASD) or patent foramen ovale (PFO)], and they are therefore cyanotic, as most systemic venous return is shunted right-to-left (R-to-L) across the atrial communication. There is often insufficient antegrade flow through the RV to the pulmonary circulation, resulting in inadequate PBF. Particularly in the neonatal period, pulmonary vascular resistance (PVR) is a major determinant of RV antegrade flow.



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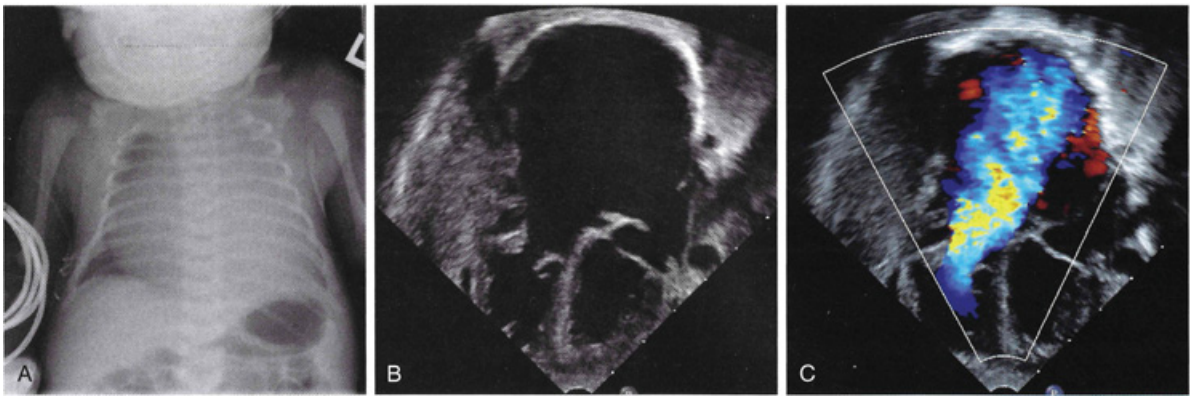


**Figure 13.1 Ebstein anomaly in a neonate**. **(A)** Chest plain film showing marked cardiomegaly. **(B, C)** Echocardiogram four-chamber view showing a severely dilated right atrium, a secundum atrial septal defect, and severe tricuspid regurgitation. From Kussman B., et al. Congenital cardiac anesthesia. Non bypass procedures. In Davis P. J. and Cladis F. P., eds. *Smith's Anesthesia for Infants and Children*, 9th ed. Elsevier; 2017: 699–743. With permission.

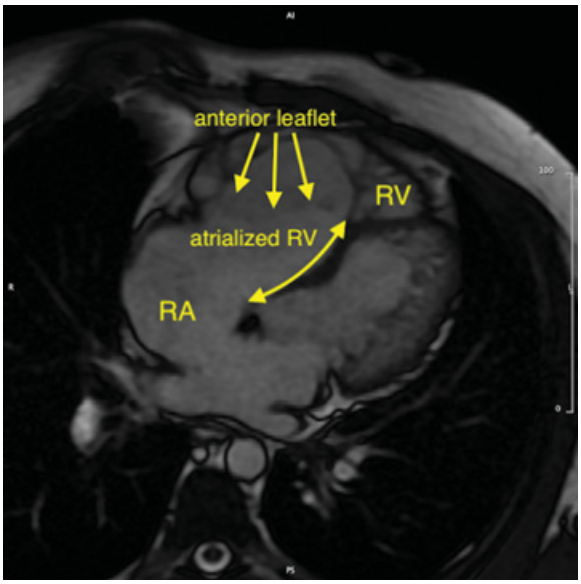


**Figure 13.2 Ebstein anomaly.** Four-chamber magnetic resonance imaging of severe Ebstein anomaly. There is marked apical displacement of the tricuspid septal leaflet creating a very small right ventricle and a large atrialized right ventricle. Courtesy of Michael Taylor, MD.

In the presence of anatomic RV outflow tract obstruction (RVOTO) or pathophysiologic pulmonary atresia, PBF is dependent on left-to-right (L-to-R) flow via a patent ductus arteriosus (PDA) from the aorta to the pulmonary artery. When the disease is less severe, the natural progressive reduction in PVR will improve RV to pulmonary artery antegrade flow, leading to clinical improvement.

Severe cardiomegaly is also present in Ebstein anomaly, typically due to the dilated right atrium. The right atrial dilation and stretch on the conduction system makes these patients prone to atrial tachyarrhythmias.

**Clinical Pearl**

*Newborns with severe Ebstein anomaly present with significant apical displacement of the TV, leading to severe TR and an ineffective RV. As nearly all patients have an atrial communication, they are cyanotic, as most systemic venous return is shunted right-to-left across the atrial communication.*

## Is the left ventricle affected in Ebstein anomaly?

In the presence of right atrial and RV enlargement, the ventricular septum may bulge to the left, reducing the size of the left ventricle (LV). When severe, this may reduce LV filling and ability to generate appropriate cardiac output. The LV myocardium may also show variable degrees of fibrosis, usually beyond childhood.

## What is the natural course and survival of patients with Ebstein anomaly?

In neonates with severe TR or cardiomegaly who are otherwise asymptomatic and who do not have medical or surgical intervention the associated mortality rate is 45% within the first year of life. In the presence of severe and symptomatic Ebstein anomaly, nearly all neonates will die without surgical intervention. However, patients who survive early childhood can expect reasonable longevity, while those undergoing single-ventricle palliation will have survival rates comparable to other single-ventricle patients.



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## What is the medical management and decision making for a newborn with severe Ebstein anomaly?

Initial management of the newborn with severe Ebstein anomaly is focused on maintaining effective PBF and oxygen saturation. Tricuspid valve and RV anatomy and amount of antegrade PBF are assessed at birth via echocardiography, oxygen saturation, and physical examination. Neonates who are stable are treated with supplemental oxygen and prostaglandin $E_1$ ($PGE_1$) to maintain PBF through the PDA.

Daily echocardiograms are obtained to assess antegrade PBF via the native RV outflow tract while the patient is slowly weaned off $PGE_1$, the PDA closes, and PVR decreases. Approximately 50% of neonates with severe Ebstein anomaly will stabilize, with sufficient antegrade PBF to maintain oxygen saturations of >75%–80%, while the other 50% will require neonatal surgical intervention.

Patients are closely monitored in an intensive care setting during this time to allow rapid assessment and treatment of decreases in cardiac output or oxygen desaturations. Therapy may include restarting or increasing $PGE_1$ to reopen the PDA, and, if the neonate is unstable, intubation, sedation, and paralysis may be required. Once the patient is intubated, high tidal volumes (10–12 mL/kg) may be needed due to the deleterious effect of cardiomegaly on lung expansion. Initiation of inotropic support may be necessary, including epinephrine, milrinone, and calcium infusions.

**Clinical Pearl**

*In neonates with severe Ebstein anomaly, early decision making about surgical intervention is dependent on the amount of effective PBF as PVR falls and the PDA closes.*

## When would a patient require a surgical intervention?

Neonates with persistent deterioration (oxygen saturation <75%) despite adequate medical management or who are unable to wean from $PGE_1$ will require surgical intervention. Additionally, patients who successfully wean from $PGE_1$ but cannot tolerate feeding or fail to thrive may also require early intervention.

## What are the major pathways for a patient with severe Ebstein anomaly requiring surgical intervention?

Surgical intervention depends on the individual patient's anatomic and pathophysiologic characteristics. Initial decision making depends on the amount of antegrade PBF and oxygen saturation. Longer term surgical decision making is directed toward either a single-ventricle palliation pathway, a one-and-a-half ventricle repair, or a two-ventricle repair, based on the size and function of the RV. When the size and function of the TV and RV are adequate, a two-ventricle repair with creation of an RV-pulmonary artery conduit (if needed) may be performed.

**Clinical Pearl**

*Depending on the individual patient's anatomy and pathophysiology, either a single-ventricle palliative pathway or a two-ventricle repair may be chosen.*

## What are the surgical options for a patient who cannot be weaned from $PGE_1$?

For the neonate with inadequate antegrade PBF who is unable to be weaned from $PGE_1$, the initial surgical procedure will include creation of a modified Blalock–Taussig (mBT) shunt, typically a GORE-TEX® shunt from the subclavian or innominate artery to the pulmonary artery, to establish another source of PBF. Determining whether the infant's pulmonary atresia is anatomic or functional will determine the surgical approach taken in addition to the mBT shunt. (See Figure 13.3.)

**Anatomic Pulmonary Atresia with Moderate to Severe TR** The Starnes procedure may be utilized for anatomic pulmonary atresia with moderate to severe TR. As the Starnes procedure excludes the RV, this plan most often commits the patient to the single-ventricle palliation pathway, including a bidirectional Glenn procedure and, eventually, Fontan completion.

The Starnes procedure consists of the following:

- Oversewing/pericardial patch closure of the TV
- Atrial septectomy (allowing mixing of systemic and pulmonary venous return)
- Creation of a mBT shunt for pulmonary blood flow

Systemic venous return now flows to the common atrium, through the mitral valve to the left ventricle, and exits via the aorta. Pulmonary blood flow is provided via the mBT shunt.

**Functional Pulmonary Atresia** The size of the RV and severity of the TR will determine the most appropriate surgical option. If the RV is adequately sized or may grow to allow for two-ventricle repair and the TV function is



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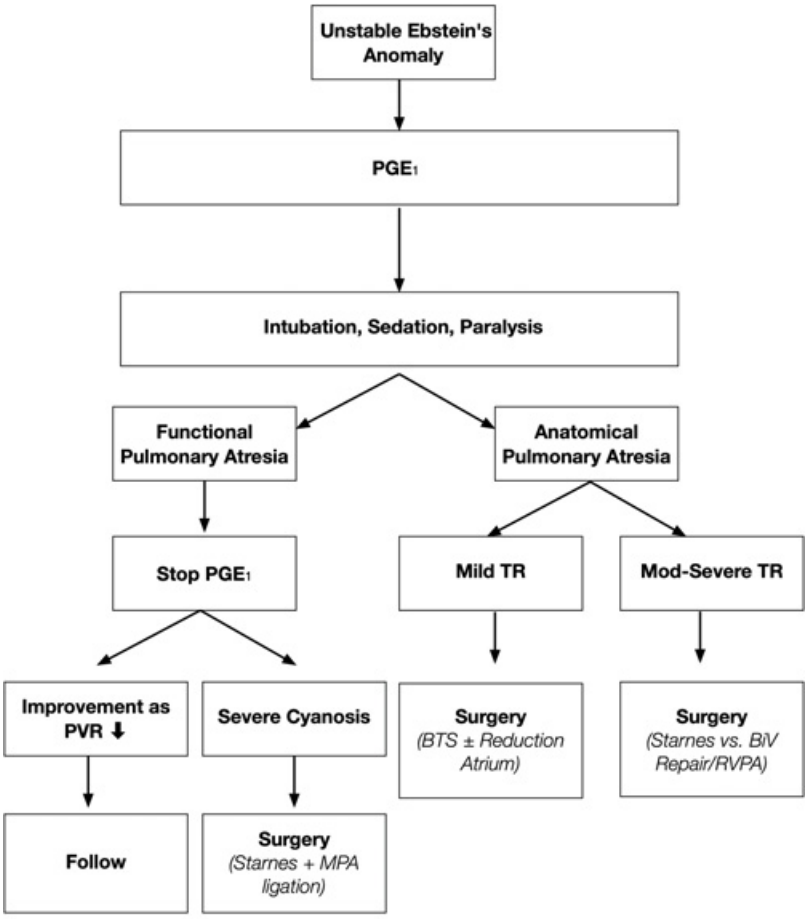


**Figure 13.3** Simplified algorithm for neonatal decision making in severe Ebstein anomaly.

*Starnes, Starnes Procedure; BiV, Biventricular; PVR, pulmonary vascular resistance; PGE1, Prostaglandin E1; BTS, Blalock-Taussig Shunt; RVPA, Right Ventricle to Pulmonary Artery Shunt)*

sufficient, the initial surgical approach will be directed at establishing PBF, perhaps through a mBT shunt, while maintaining RV antegrade flow. This approach leaves the option for either one- or two-ventricle surgical pathways in the future. When the size and function of the TV and RV are adequate, a two-ventricle repair with creation of an RV–pulmonary artery conduit (if needed) may be performed.

**Clinical Pearl**

*Initial decision making depends on the amount of antegrade PBF and oxygen saturations. Longer term surgical decision making is directed toward either a single-ventricle palliation or a two-ventricle repair, based on the size and function of the RV.*

## What is the Starnes procedure and what is the long-term strategy for patients in this pathway?

In 1991, Starnes et al. reported a single-ventricle palliation for critically ill neonates with Ebstein anomaly. He described a procedure including a pericardial patch closure of the TV orifice, atrial septectomy, and creation of an mBT shunt. To address the issue of pulmonary regurgitation, ligation of the main pulmonary artery may also be performed. Starnes et al. later described two important modifications to their technique: coronary sinus retained in the right atrial side of the TV patch to prevent drainage into the excluded RV and fenestration of the TV patch with a 4 mm opening to prevent RV distension.



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Patients who undergo the Starnes palliation are generally obligated to the single-ventricle palliative pathway and will require a Glenn procedure at the age of 3–9 months and Fontan completion around the age of 2–4 years.

**Clinical Pearl**

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## What is a bidirectional Glenn procedure?

The bidirectional Glenn (BDG) procedure is frequently performed as part of staged surgical palliation of single-ventricle patients, including severe Ebstein anomaly. The BDG shunt procedure involves rerouting upper body systemic venous return directly to the lungs by anastomosing the superior vena cava (SVC) to the right pulmonary artery. This anastomosis provides a source of PBF and reduces ventricular work, as upper body systemic venous return does not return to the heart. Blood flows passively from the SVC to the pulmonary artery, without a pump. As originally described, the Glenn procedure provided blood flow to one lung only; modern modification has blood flowing from the SVC into both right and left lungs, thus the term *bidirectional* Glenn shunt or procedure. (See Chapter 27.)

## What are the sources of PBF and venous return to the single ventricle after BDG?

Pulmonary blood flow in a BDG shunt is dependent on SVC flow. Superior vena cava flow accounts for approximately 50% of cardiac output in newborn infants, reaching a maximum of 55% at 2.5 years old and then gradually decreasing to the adult value of 35% by 6 years of age.

After BDG, blood enters the single ventricle from two sources: oxygenated blood enters the heart from the pulmonary veins and deoxygenated blood from the lower body enters via the inferior vena cava (IVC). Because patients have had a complete atrial septectomy as part of their first-stage procedure, flow from the IVC mixes with flow from the pulmonary veins in the atrium and then proceeds to the single ventricle before being ejected into the systemic circulation. Due to this mixing, expected oxygen saturations in patients with BDG physiology are 75%–85%.

# Anesthetic Implications

## What preoperative assessment should be done in a patient with a BDG?

Prior to providing an elective anesthetic in a patient with a BDG, a complete preoperative assessment should include a review of the most recent cardiology visit, including echocardiographic, catheterization, and recent laboratory data. Information regarding current ventricular function, the presence of valvular regurgitation, PVR, ventricular end-diastolic pressure, and the presence of any collateral circulation should be reviewed. Questions during the preoperative interview should be directed at eliciting information regarding exercise tolerance, syncope or other symptoms of arrhythmias, and growth and development. Symptoms of ventricular dysfunction, severe valvular regurgitation, and/or a history concerning for heart failure or arrhythmias may prompt further discussion and optimization. Laboratory tests such as complete blood count, electrolytes, urea nitrogen, creatinine, coagulation profile, and liver function may be considered depending on the patient's history, medication profile, and scope of the anticipated procedure.

## What should the parents be told regarding anesthetic and surgical risk in this patient?

Children with congenital heart disease (CHD) undergoing noncardiac surgery are known to be at higher risk for morbidity and mortality than patients without cardiac disease. Among patients with CHD, those with single-ventricle physiology or ventricular dysfunction are at particular risk. The patient's functional capacity should be carefully reviewed. Recent studies support a lack of correlation between intrinsic surgical risk and the risk of complications in children with CHD undergoing noncardiac surgery, suggesting that the severity of the cardiac lesion and the patient's functional status are more important in risk assessment.

## What are the guidelines for endocarditis antibiotic prophylaxis?

Preoperative antibiotic prophylaxis is necessary for all procedures likely to produce bacteremia (e.g., dental extraction) in patients with unrepaired CHD, including palliative shunts and conduits such as the BDG. In this particular case, no endocarditis prophylaxis would be required.



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## What are the key points regarding BDG physiology?

Bidirectional Glenn flow is passive; there is no pump between the SVC and the pulmonary artery. Pulmonary blood flow is therefore dependent on SVC pressure, pulmonary artery pressure/PVR, intrathoracic pressure, and volume status. Reductions in PBF with resultant oxygen desaturation can occur with increased PVR, hypovolemia, or increased intrathoracic pressure due to coughing, laryngospasm, excessive positive pressure ventilation (PPV), or breath holding. Desaturations due to reduced PBF in BDG physiology can be significant, but saturations will typically return to baseline when the cause is addressed.

**Clinical Pearl**

*Pulmonary blood flow in BDG physiology is passive as there is no ventricular pump. Pulmonary blood flow will decrease with increases in either intrathoracic pressure (due to coughing, laryngospasm, or positive pressure ventilation) or increases in pulmonary vascular resistance, resulting in hypoxemia.*

With Glenn physiology, hemodynamic performance tends to be resilient because the single ventricle is less volume loaded than it was prior to BDG, when the presence of parallel circulations required the single ventricle to handle both pulmonary and systemic volumes. Cardiac output after BDG is not entirely dependent on PBF because the IVC remains directly connected to the heart, providing venous return even when pulmonary blood flow is decreased. Therefore, should PBF be compromised by an increase in PVR or intrathoracic pressure, cardiac output can be maintained via IVC flow.

**Clinical Pearl**

*Cardiac output after a Glenn shunt is not entirely dependent on PBF because the IVC remains directly connected to the heart, providing venous return even when PBF is decreased. Therefore, should PBF be compromised by an increase in PVR or intrathoracic pressure, cardiac output can be maintained via IVC flow.*

## What oxygen saturations are expected for a child with a BDG?

Due to intracardiac mixing in the common atrium of deoxygenated blood (from the IVC) with oxygenated blood (following the path from the SVC/Glenn anastomosis → pulmonary arteries → lungs → pulmonary veins), the typical oxygen saturations for a patient with Glenn physiology are between 75% and 85% on room air.

**Clinical Pearl**

*After a BDG, mixing in the common atrium of deoxygenated blood from the IVC with oxygenated blood from the pulmonary veins means that typical oxygen saturations for a patient with Glenn physiology are between 75% and 85% on room air.*

## What considerations exist for a BDG patient with a recent or current respiratory infection?

Recent respiratory illness or recent changes in oxygen saturation merit careful consideration in patients with Glenn physiology. In addition to the increased risk of pulmonary complications following recent infection that is common to all patients, the increased PVR and airway hyperreactivity that can accompany respiratory infections may result in increased cyanosis and hypoxemia in patients with passive PBF. Therefore, the risk–benefit analysis of anesthesia for BDG patients in the presence of a recent or current respiratory tract infection should be carefully assessed.

## What are the anesthetic considerations for a BDG patient presenting for noncardiac surgery?

In patients with Glenn physiology and preserved ventricular function, the mild to moderate fluid or pressure shifts associated with surgery and general anesthesia are typically well tolerated. Inhalation induction may take slightly longer than in a patient with biventricular physiology because of the decreased ratio between pulmonary and systemic blood flow. Anesthetic induction can also be associated with peripheral vasodilatation, which may require volume administration and sometimes vasoconstrictor agents such as phenylephrine to maintain mean arterial pressure. Anesthetic management is directed to preserving cardiac output by supporting ventricular function and promoting PBF. Hypovolemia is disadvantageous because it significantly reduces PBF, ventricular preload, and output. The most appropriate ventilation strategy should be considered in the context of the planned procedure with the understanding that positive pressure ventilation (PPV) may negatively impact PBF.



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**Clinical Pearl**

*Inhalation induction may take slightly longer than in a patient with biventricular physiology because of the decreased ratio between pulmonary and systemic blood flow. Anesthetic management is directed to preserving cardiac output by supporting ventricular function and promoting PBF.*

## What ventilation strategy is most appropriate?

Ventilatory management should aim to promote PBF by minimizing mean airway pressure. Maintenance of low to baseline PVR is essential and can be accomplished by a ventilation strategy that maintains adequate tidal volume and utilizes a short inspiratory time, minimizing mean airway pressure. While significant hypercarbia increases PVR, mild hypercarbia may improve PBF and oxygen saturations by increasing cerebral blood flow and SVC flow to the pulmonary bed. Hyperventilation should be avoided due to its association with a decrease in cerebral blood flow and consequently PBF. Although the use of a laryngeal mask airway may be preferred over intubation when possible, as it encourages the use of spontaneous ventilation and lower mean airway pressures, securing the airway with an endotracheal tube may be preferable based on patient or procedural considerations.

**Clinical Pearl**

*While significant hypercarbia increases PVR, mild hypercarbia may improve PBF and oxygen saturations by increasing cerebral blood flow and SVC flow to the pulmonary bed.*

## What fluid management and transfusion strategy should be applied?

Patients with BDG physiology require adequate fluid status (normovolemia) to maintain passive PBF while avoiding fluid overload for the single ventricle. Crystalloid fluids may be utilized to account for fasting deficits and any decrease in SVR associated with general anesthesia. A bolus of 5–10 mL/kg is sometimes useful during or immediately following induction if desaturation or hypotension due to PPV and decreased PBF occurs. However, fluid management should be carefully weighed in patients with decreased cardiac function. Although the surgical procedure planned in this scenario presents a relatively low bleeding risk, patients with single-ventricle physiology are at risk for increased bleeding due to therapeutic use of anticoagulant medications and intrinsic coagulopathy. Patients with cyanotic heart disease require a higher hemoglobin for improved oxygen delivery; the preoperative hemoglobin should therefore be assessed, and patient contextualized transfusion triggers considered. At a minimum, an appropriate hemoglobin level of 9–10 g/dL or higher should be targeted; most practitioners advocate a hemoglobin level of 13–15 for a patient with single-ventricle physiology.

## Is a patient with severe Ebstein anomaly at increased risk for arrhythmias?

Conduction system abnormalities may be present in children with Ebstein anomaly. The right bundle may be fibrotic and, occasionally, accessory pathways have been noted with pre-excitation syndrome. Supraventricular tachycardias due to right atrial dilation are challenging and sometimes refractory to therapy in patients with Ebstein anomaly. Care of patients with Ebstein anomaly who have a history of arrhythmias should be coordinated in consultation with cardiologists and electrophysiologists to assist in perioperative medication administration and planning for potential intraoperative rhythm disturbances.

## What are the major anesthesia concerns during emergence and the postoperative period?

As PBF is passive in BDG patients, avoiding significant increases in PVR or intrathoracic pressure should be the goal during emergence and postoperatively. Adequate pain control is necessary. Dexmedetomidine is useful to decrease the incidence of postoperative agitation and coughing without adversely affecting spontaneous respiratory drive. Antiemesis prophylaxis is also recommended, as vomiting may cause dehydration and impair respiratory mechanics, leading to cyanosis.

## Where should this patient recover?

Clear guidelines regarding postoperative destination do not exist for patients with BDG physiology. However, the decision to recover the patient in the postoperative anesthesia care unit versus intensive care unit should be based on the patient's functional status and perioperative hemodynamic stability as well as local practice patterns. If the patient has good functional status, is well palliated, and the case is uneventful, post-anesthesia care unit recovery may be appropriate. Patients with single-ventricle physiology should be recovered in an environment where nurses and physicians are familiar and comfortable with the pathophysiology.



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## Can this procedure be performed on an outpatient basis?

Single-ventricle patients may not be suitable candidates for surgery in centers that do not have a cardiac program and cardiac-trained anesthesiologists. However, patients with good functional status for low-risk procedures may be treated in a same-day surgery unit of a tertiary care hospital following the collaborative decision-making of cardiologist, surgeon, and anesthesiologist.

## Suggested Reading

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