Chapter

12

Ebstein Anomaly

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Case Scenario

A 4-year-old boy weighing 16 kg with known Ebstein anomaly presents from the emergency department for urgent upper endoscopy and foreign body removal. His mother found him choking and coughing at home several hours ago and chest radiography reveals a radiopaque toy in the lower esophagus.

The mother states that the child has a "heart problem" and that he sees a cardiologist. She also adds that he often seems more easily tired than his siblings. The most recent clinic note from nearly a year ago states that the patient has Ebstein anomaly. He has not had any previous surgeries, and the cardiologist recommended yearly follow-up and imaging.

Currently the child looks uncomfortable and is drooling, but he is without any respiratory distress. His vital signs are blood pressure 85/52, heart rate 94 beats/minute, respiratory rate 22 breaths/minute, room air SpO_2 94%, and temperature 36.8°C. A right-sided cardiac murmur and mild liver enlargement are noted.

The most recent echocardiogram from 11 months ago demonstrated:

- *An inferiorly displaced tricuspid valve with mild-to-moderate tricuspid regurgitation*
- *Mildly reduced right ventricular cavity size*
- *Mildly diminished right ventricular function*
- *Normal left ventricular function*
- *Presence of an atrial connection with bidirectional flow*

Key Objectives

- Describe the anatomy and spectrum of disease in Ebstein anomaly.
- Define the term "atrialized ventricle."
- Describe perioperative management considerations for a patient with Ebstein anomaly.

Pathophysiology

What is Ebstein anomaly?

Ebstein anomaly (EA) is a rare heart defect affecting the tricuspid valve (TV) and right ventricle (RV). The TV is dysplastic, with downward displacement of the septal and posterior leaflets inferiorly into the RV. The anterior leaflet is elongated and frequently has fenestrations and abnormal chordal attachments. As a result, there is dilation of the TV annulus resulting in tricuspid regurgitation (TR) and, depending on the degree of TV displacement, loss of RV cavity volume. (See Figure 12.1.) Right atrial enlargement frequently exists, and arrhythmias may occur related to right atrial dilation and abnormalities of the conduction system. Ebstein anomaly represents a wide spectrum of anatomic and clinical presentations, ranging from minimally symptomatic disease presenting in late childhood or early adolescence that requires medical management over time to critical disease requiring neonatal surgical intervention.

While the mildest forms of EA include minimal TV regurgitation and TV displacement, more severely affected patients will have greater TV displacement with resultant loss of RV volume and RV dysfunction. Varying degrees of

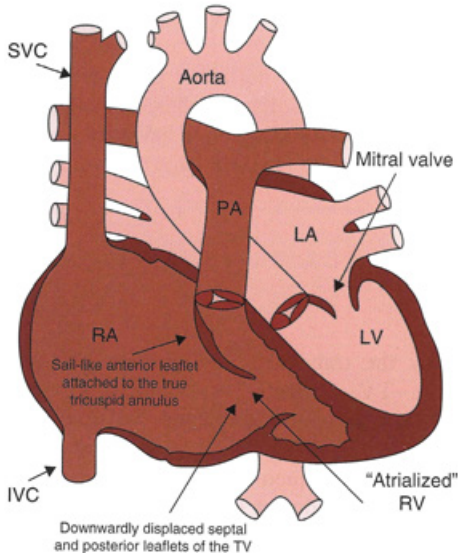

Figure 12.1 Ebstein anomaly of the tricuspid valve. Ao, aorta; LA, left atria; LV, left ventricle; PA, pulmonary artery; RA, right atria; RV, right ventricle; TV, tricuspid valve. From Nasr V. G. and DiNardo J. A. *The Pediatric Cardiac Anesthesia Handbook*, 1st ed. John Wiley & Sons, 2017; 239–42. With permission.

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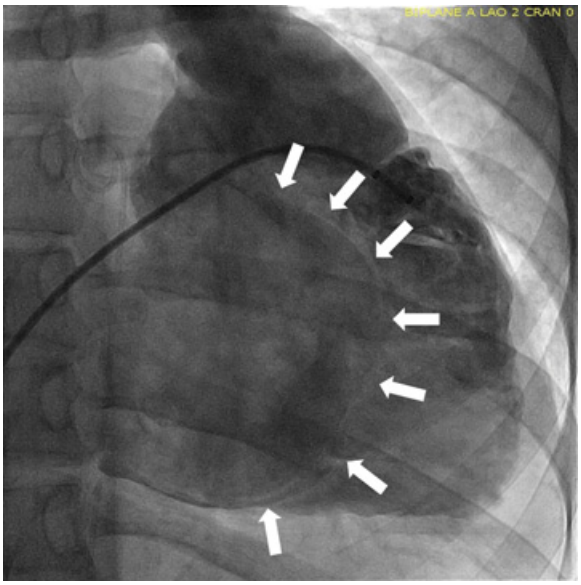

Figure 12.2 Right ventricular injection. An angiogram is performed in the right ventricle in the AP projection. The displacement of the tricuspid valve leaflets into the right ventricular cavity is indicated by the arrows. Courtesy of Russel Hirsch, MD.

cyanosis via right-to-left (R-to-L) shunting at the atrial level via either an atrial septal defect (ASD) or patent foramen ovale (PFO) can occur. The most severely affected patients may require single ventricle palliation, as the RV is diminutive and insufficient to support adequate antegrade pulmonary blood flow (PBF). (See Figure 12.2.)

Clinical Pearl

Ebstein anomaly represents a wide spectrum of anatomic and clinical presentations, ranging from minimally symptomatic disease that requires medical management over time to critically ill neonates with severe disease requiring neonatal surgical intervention.

How common is Ebstein anomaly?

While EA is the most common congenital malformation of the TV (1 in 20,000 live births), it is extremely rare, accounting for fewer than 1% of congenital cardiac defects. The true cause of EA is unknown but there are associations with usage of benzodiazepines in the first trimester of pregnancy and lithium usage.

What are the key anatomic characteristics of Ebstein anomaly?

Ebstein anomaly includes abnormalities of the TV and the RV. The hinge points of the septal and posterior leaflets of the TV are downwardly displaced from the atrioventricular junction into the RV itself, while the anterior leaflet is elongated with a hinge point at the true TV annulus. Depending on the severity of the TV abnormalities the resultant degree of "atrialization" of the RV varies, and in the most severe forms of EA the RV may have only trabecular and outflow portions. The atrialized portion of the RV is thin and dilated; it has ventricular morphology but functionally serves as part of the right atrium. An atrial level communication, either ASD or PFO, also exists in nearly all patients. In more severely affected patients R-to-L shunting at the atrial level results in varying degrees of cyanosis.

What are typical symptoms of Ebstein anomaly?

Ebstein anomaly has a wide spectrum of presentations. The clinical presentation will depend on the extent of TV displacement, RV size and function, right atrial pressure, and the degree of R-to-L shunting. Patients with EA are at particular risk for arrhythmias related to abnormal conduction patterns in the atrialized RV and right atrial dilation due to TR. Many patients with EA display findings on electrocardiogram (ECG) of preexcitation and/or Wolff–Parkinson–White syndrome (WPW).

Mildly affected patients may display mild exercise intolerance and there may initially be minimal symptomatology. The diagnosis may not be established until early adolescence or adulthood with the advent of diminished RV function and/or atrial arrhythmias.

Moderately affected patients may have cyanosis related to inadequate RV size, moderate TV malformation, TR, and atrial level R-to-L shunting. Symptoms may be exacerbated by exercise. There may be progressive dilation and dysfunction of the RV related to the degree of TR.

The most ***severely affected patients*** are often recognized in utero or present early in the neonatal period with refractory cyanosis (due to obligatory R-to-L shunting at the atrial level), extreme cardiomegaly, and arrhythmias. Inadequate blood flow through the pulmonary valve can result in functional or anatomic pulmonary stenosis or atresia. Infants with less severe forms of EA may be mildly cyanotic and will continue to improve as pulmonary vascular resistance (PVR) falls after birth, allowing increased antegrade PBF. Neonates with severe TR and a diminutive, poorly contractile RV with little to no antegrade PBF may be dependent on a prostaglandin E_1 infusion to maintain ductal patency and assure adequate PBF until palliative surgery can take place.

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What are surgical options for EA and when would they be considered?

Surgical intervention for EA is prompted by severity of symptomatology, with multiple options existing depending on the patient's specific anatomy and physiology. The morphology and size of the RV are important in determining the patient's suitability for either a two-ventricle, one-and-a-half-ventricle, or single-ventricle approach.

- A two-ventricle repair, with TV repair and plication of the atrialized portion of the RV, is proposed for patients with an adequate RV and antegrade PBF. Several methods of TV repair have been described. An annuloplasty ring may be added if necessary. In patients with compromised RV function a small atrial fenestration may be left to augment cardiac output. Although a last resort in children, TV replacement may be required in adults.
- A one-and-a-half-ventricle palliation can be considered for patients with insufficient RV size or function. This operation consists of TV repair, a superior cavopulmonary anastomosis (bidirectional Glenn), and a fenestrated ASD. In this scenario blood from the inferior vena cava would traverse the TV and RV while blood from the superior vena cava would passively flow directly to the pulmonary vascular bed.
- In the most severe cases of EA, when anatomic pulmonary atresia exists as a result of right ventricular outflow tract obstruction, a Starnes procedure can be performed, consisting of oversewing or pericardial patch closure of the TV, an atrial septectomy, and construction of a modified Blalock–Taussig (mBT) shunt to provide PBF. If significant pulmonary insufficiency exists, the pulmonary valve may be oversewn and the main pulmonary artery divided. Typically, patients who have undergone the Starnes procedure will then proceed with single-ventricle palliative procedures (bidirectional Glenn followed by completion Fontan). (See Chapters 10, 27–30.)

Anesthetic Implications

What findings should be considered in the preoperative evaluation of a patient with unrepaired EA?

In addition to a comprehensive preoperative history and physical, special considerations for patients with EA include an understanding of the patient-specific anatomy, physiology, and rhythm. The history should include a review of symptoms, with special attention to any history of cyanosis, increasing exercise intolerance, arrhythmias, and/or failure to thrive. Consultation with the patient's cardiologist may be beneficial if there has been a significant change in his cardiac symptoms since the last office visit. If available, findings from the most recent cardiology visit, echocardiogram, and ECG should be reviewed.

Other concerns include any history of a recent upper respiratory infection or other recent illnesses that might adversely affect PVR. Information should be sought regarding the patient's last oral intake and whether he has taken anything by mouth since the ingestion.

While a patient with only mild TR may present with no signs or symptoms, those with more significant pathology may present with signs and symptoms related to TR and/or RV dysfunction. Cardiac auscultation may reveal a widely split first heart sound and a soft pansystolic murmur of TR. In the presence of significant RV dysfunction, a cardiac gallop may be appreciated.

In this patient, the preoperative hemoglobin–oxygen saturation of 94% on room air that was noted in the emergency department is significant. If possible, it would be useful to find previous measurements from cardiology office visits to determine whether a saturation of 94% is chronically seen in this patient (due to shunting at the atrial level) or whether it is due in part to the current foreign body ingestion.

Clinical Pearl

In patients with EA, exercise tolerance is an indicator of the severity of the disease and ability to tolerate the stress of anesthesia.

Is review of imaging necessary in a patient with unrepaired EA?

Echocardiography is used to assess size of the tricuspid annulus and severity of TR, the size of the RV, and degree of pulmonary stenosis and RV dysfunction. In addition, echocardiography may reveal other associated defects; although rare, they can include ventricular septal defects, congenitally corrected transposition of the great arteries, and atrioventricular canal defects. Atrial septal defects are nearly always present, allowing R-to-L shunting to occur when right atrial pressures are high, contributing to cyanosis. Additionally, left heart function can be compromised due to abnormal positioning of the interventricular septum and decreased left ventricular chamber size.

Cardiac magnetic resonance imaging (MRI) is also useful for ongoing assessment of patients with Ebstein

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anomaly. Recent studies have shown this imaging modality to be useful for defining the volume of RV atrialization and ratio of right to left cardiac volumes, providing information about exercise capacity and heart failure in patients with Ebstein anomaly.

Preoperative imaging choices should be made with consideration of the history and physical examination findings and the urgency of the planned procedure. Echocardiography is generally most accessible and provides useful information. When the most recent echocardiographic findings are not available or it is not feasible to obtain an echocardiogram due to procedural urgency, physical examination and history may provide clues to right heart function and degree of TR. Recent changes in exercise capacity should be noted with concern. Without recent echocardiographic data, it is wise to proceed with caution and have emergency plans and resuscitation medications available.

Is premedication appropriate for this patient?

Midazolam is one of the most common preoperative sedatives used to ease separation anxiety. Though frequently used as an oral premedication, in this case administration of an oral sedative to a patient with an esophageal foreign body would not be advisable. Because this is an urgent procedure, an intravenous line should be placed prior to the procedure, if not already done in the emergency department. Depending on the child's level of anxiety and symptomatology administration of a small dose of midazolam via IV could be considered prior to separation and would likely prove beneficial in decreasing agitation and anxiety.

What are the predominant concerns during induction of anesthesia?

In this case predominant concerns involve the presence of an esophageal foreign body in a patient with preexisting TR and mildly depressed RV function. Patients with esophageal foreign bodies often have hypersalivation, pooled secretions, and difficulty swallowing. This patient has had difficulty managing secretions since the aspiration event and is at risk for aspiration during induction due to his lack of fasting as well as his ingestion of a foreign body. Additionally, foreign body migration from the esophagus to airway during induction is a concern. If not already present, IV access should be placed prior to a planned intravenous rapid sequence induction and intubation.

In this patient with mildly depressed RV function, IV induction of anesthesia may be achieved with either propofol (1–2 mg/kg) or ketamine (2–4 mg/kg) depending on the child's functional status. Etomidate can be utilized for patients with severe RV dysfunction. Patients with Ebstein anomaly are dependent on preload and will respond poorly to anesthetic-induced vasodilation, therefore IV fluid replacement may be necessary.

The potential impact of anesthetic agents and positive pressure ventilation (PPV) on RV performance should be considered, and peak airway pressures minimized. Appropriate steps should also be taken to keep PVR normal, with avoidance of hypercarbia, hypoxemia, acidosis, and hypothermia. Owing to the propensity for atrial arrhythmias close attention should be paid to the underlying rhythm and any changes that occur intraoperatively.

In patients with significant RV dysfunction, RV failure may present with hypotension, hypoxemia, and bradycardia. When an intraatrial connection is present, hypoxia may be prominent while hemodynamics are preserved as elevated right atrial pressures exceed left atrial pressures, causing R-to-L shunting. Left ventricular preload is preserved by systemic venous return shunted from the right to left atrium.

Clinical Pearl

Induction of anesthesia and the transition from negative to positive pressure ventilation increases demand on the RV.

In addition to standard monitoring, is an arterial line indicated?

Standard monitoring as recommended by the American Society of Anesthesiologists should be utilized for this procedure. If possible, a 5-lead ECG should be employed, as patients with EA are at increased risk for atrial arrhythmias. The planned case will have minimal blood loss and fluid shifts and therefore arterial line placement would not be warranted in this case. The risk for hemodynamic instability for this patient is likely greatest during induction of anesthesia.

What are the major considerations in patients with EA who require mechanical ventilation?

The main consideration in mechanical ventilation of the patient with EA and presumed RV dysfunction is minimizing RV afterload. This is achieved through careful ventilation strategies and avoidance of increases in PVR.

As increases in intrathoracic pressure result in increased RV afterload, ventilation utilizing minimal mean airway pressures is advisable. A low tidal volume and high respiratory rate strategy will achieve adequate ventilation while maintaining low mean airway pressures.

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As increases in PVR will increase RV work and perhaps decrease PBF, care should be taken to avoid those factors that can result in increasing PVR such as hypercarbia, hypoxemia, acidosis, and hypothermia. Atelectasis also increases PVR; a successful ventilation strategy should balance the need to minimize mean airway pressure with maintaining functional residual capacity.

What are the anesthetic management considerations for patients with EA?

Tricuspid regurgitation causes RV volume and pressure overload and may contribute to RV dysfunction. Intraoperative management of TR involves reducing RV afterload to encourage forward flow and maintenance of RV function. A normal or slightly elevated heart rate may be required to support RV forward flow to the pulmonary circulation. Bradycardia in patients with EA and TR may not be well tolerated due to an increase in regurgitation with longer diastolic times and decrease in cardiac output. Vasoactive infusions or inhaled nitric oxide may be required to support RV function in severe cases.

The nurse in the post-anesthesia care unit is concerned about a sudden change in the ECG tracing. What specific concerns exist for this patient?

An abnormal ECG in a patient with EA is not uncommon. If a 12-lead ECG was obtained prior to the case, it may have identified baseline abnormalities. An elevation in P wave morphology is often seen due to right atrial enlargement. Other baseline findings may include right-sided bundle branch block or first-degree heart block.

Patients with EA are also at particular risk for arrhythmias. Up to 20% of patients will have an accessory conduction pathway, most often recognized as Wolff–Parkinson–White (WPW). Characteristic ECG findings in WPW include a short PR interval, a wide QRS complex, and a delta wave. Right atrial enlargement predisposes the patient to atrial fibrillation and atrial flutter. Atrialization of the RV can also create a dysynchrony between the left and right sides of the heart, increasing the likelihood of ventricular arrhythmias.

Atrial and ventricular arrhythmias should be triaged and treated as per American Heart Association Pediatric Advanced Life Support Guidelines. Atrial arrhythmias are most common, including supraventricular tachycardia (SVT) due to WPW, atrial flutter, and atrial fibrillation. If the patient is unstable as a result of the arrhythmia, rapid treatment may be indicated. Urgent SVT treatment may include vasovagal maneuvers (if hemodynamically stable), adenosine, or cardioversion. Given the patient's underlying diagnosis of EA, management of atrial arrhythmias is best done in consultation with a pediatric cardiologist.

Clinical Pearl

Patients with EA are at risk for arrhythmias, particularly if they have had a history of arrhythmias in the past. The preoperative history should include any history of arrhythmias and a baseline ECG to assist with perioperative ECG interpretation.

Should the patient be admitted after the procedure?

The decision whether to admit the patient for overnight observation depends on the child's baseline disease and perioperative course. In patients with minimal cardiac symptoms and uneventful perioperative course, discharge after extended post-anesthesia care unit stay can be considered. If the child has significant cardiac symptoms or a complicated operative course, consultation with pediatric cardiology and admission may be indicated.

Suggested Reading

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