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Ebstein's Malformation

20.1 Morphology & Pathophysiology

It is characterized by a downward displacement of the septal and posterior leaflets of the tricuspid valve that are often hypoplastic, with an enlarged anterior leaflet. The tricuspid annulus is dilated. The normally functioning right ventricular cavity is reduced as the ventricle between the true atrioventricular junction and the attachments of the displaced leaflets functions like atrium, i.e., dilates during ventricular systole and contracts in diastole, resulting in wasted right ventricular stroke work. The right ventricular myocardium may show impaired contractility and histological dysplasia in severe cases.

The consequences of this lesion are related to severity of the leaflet displacement and valvular regurgitation. In severe cases, the pulmonary blood flow is decreased, the right atrium becomes dilated, blood is shunted right to left across an ASD or PFO, and the patient may become cyanotic. Congestive heart failure may also develop secondary to a small functional right ventricle, ventricular dysfunction, and decreased right ventricular compliance. In addition, paroxysmal supraventricular tachycardia (PSVT) occurs in 25-50%, and Wolff-Parkinson-White (WPW) syndrome is noted in 5-10% of these patients.

20.2 Perioperative Management

20.2.1 Neonatal Ebstein's Disease

Neonates with Ebstein's are often present with cyanosis, severe ventricular dysfunction, congestive heart failure, and metabolic acidosis.

1. Patient should be sedated, intubated, paralyzed, and mechanically ventilated.

2. Prostaglandin E_1: Start the infusion to maintain patency of the ductus arteriosus and to improve pulmonary blood flow. If cyanosis improves, then PGE_1 infusion can be weaned.
3. Following weaning from PGE_1 therapy, close observation is required to assess whether the neonate oxygenates adequately after closure of the ductus arteriosus.
4. Inhaled nitric oxide: Therapy may be required in severe disease to lower pulmonary vascular resistance and improve pulmonary blood flow.
5. Metabolic acidosis: Corrected by sodium bicarbonate infusion and for improving the ventricular function.
6. Inotropic support: It is often is required to improve biventricular function.
7. Optimize: Serum electrolyte balance, oxygen carrying capacity, and blood volume status prior to surgery.

20.2.2 Older Children and Adults with Ebstein's Disease

The mainstay of initial treatment is medical to address symptomatic relief of congestive heart failure and management of arrhythmias. Surgical therapy is warranted in any patient with progressive symptoms, exercise intolerance, or frequent arrhythmia. Most patients with Ebstein's disease require surgical correction, at some time, though patients who are mildly symptomatic may never require surgery.

20.3 Surgical Treatment

The aim of surgical treatment is to minimize tricuspid insufficiency, increase pulmonary blood flow, reduce or eliminate right-to-left shunting, optimize right ventricular function, and reduce or eliminate arrhythmia. Ideally, if the tricuspid

valve can be repaired, it avoids the need for future valve replacement. If valve repair is not feasible, valve replacement with a bioprosthetic valve is needed.

Most operations are performed using CPB (cardiopulmonary bypass), bicaval cannulation, moderate systemic hypothermia, and cardioplegic arrest. In neonates, a single venous cannulation and a period of circulatory arrest may also be used to complete the repair.

20.3.1 Single Ventricular Palliation

In a sick neonate with severe Ebstein's disease, create functional tricuspid atresia as palliation (Starnes procedure) to a subsequent single ventricle repair with a Fontan procedure. The atrial septal defect is enlarged by excising the remaining septum. The tricuspid valve orifice is closed with a Gore-Tex patch. A Gore-Tex shunt is placed to connect the innominate artery to the right pulmonary artery.

20.3.2 Valve Repair Techniques

Most of these techniques aim to achieve tricuspid valve competence by creating a monocusp valve based on the anterior leaflet. The atrialized ventricle is plicated (i.e., horizontal plication described by Danielson, vertical plication described by Carpentier). Tricuspid valve annuloplasty is performed (posterior annuloplasty or a ring tricuspid annuloplasty), the atrial septal defect is closed, and the right atrial reduction may be performed. The Hetzer repair consists of the reduction in the size of the tricuspid orifice to achieve adequate coaptation of the most mobile leaflet tissue, closure of the atrial septal defect, and the right atrial reduction. Newer repair techniques involve either septal leaflet augmentation or detachment of both the abnormal septal and posterior leaflets and their respective chordae and papillary muscles, and reimplantation to allow complete relocation of the leaflet tissue to the level of the true annulus.

Alternatively, the anterior and posterior tricuspid valve leaflets are mobilized from their anomalous attachments in the right ventricle, and the free edge of this complex is rotated clockwise to be sutured to the septal border of the anterior leaflet, thus creating a cone the vertex of which remains fixed at the right ventricular apex and the base of which is sutured to the true tricuspid valve annulus level. Additionally, the septal leaflet is incorporated into the cone wall if possible.

The bidirectional superior vena cava to pulmonary artery shunt (bidirectional Glenn shunt) may be added to intracardiac repair to reduce volume load of the ventricle and tricuspid regurgitation, and improve pulmonary blood flow in patients with severe Ebstein's disease and ventricular dysfunction.

20.3.3 Tricuspid Valve Replacement

If a valve repair is not feasible or successful, valve replacement with porcine bioprosthesis is required. Mechanical valve replacement in the tricuspid position is associated with a high frequency of valve malfunction and thrombotic complications. The atrialized right ventricle is plicated in a horizontal plane, then valve sutures are placed on the atrial side of the coronary sinus and atrioventricular node to avoid injury to the conduction system. Sutures are tied with the heart beating and perfused to ensure the conduction system is intact. Some replacement techniques oversize the bioprosthesis and attach the sewing annulus to the atrial wall proximal to the coronary sinus. This supra-annular implantation avoids suturing in the area of conduction tissue, but it places the coronary sinus in the right ventricle.

In Laks modification of the technique, the valve insertion is begun anterior to the coronary sinus using a continuous running suture. A glutaraldehyde-treated pericardial patch is sutured to the septal portion of the prosthetic valve sewing annulus. The free margin of the patch is then sutured to the atrial tissue beyond

the area of the conduction tissue. This technique allows placement of the valve at the true annulus, avoids suturing in the area of conduction tissue, and leaves the coronary sinus draining into the right atrium.

20.3.4 Electrophysiologic Mapping and Maze Procedure

In addition to the above, the right atrial Maze procedure or the modified Maze procedure is done to eliminate or reduce atrial arrhythmias. The accessory conduction pathways can also be ablated with a catheter-based radiofrequency ablation before or after a surgical repair.

20.3.5 Mechanical Circulatory Support & Heart Transplantation

In patients with a failed repair or with severe biventricular dysfunction and in severely symptomatic neonates with associated pulmonary artesia or other major cardiac defects, heart transplantation and mechanical support as a bridge to heart transplantation is indicated.

20.4 Postoperative Management

Postoperative course is variable due to the variety of surgical procedures performed for correction. The hospital stay averages 10 days to 2 weeks.

20.4.1 Hemodynamic Management

In critically ill neonates, therapy in the first 24 hours is aimed to reduce pulmonary vascular resistance and pulmonary artery hypertensive episodes to maintain hemodynamic and respiratory stability.

Inotropic agents are often needed to support the right ventricular function.

Central venous pressure and waveform should be monitored for any evidence of recurrent tricuspid insufficiency and right ventricular failure.

Patients undergoing valve repair may require echocardiographic evaluation in the early postoperative period to assess the tricuspid valve and right ventricular function.

Some patients with mild-to-moderate residual tricuspid incompetency progress to severe regurgitation, enough to warrant early return to an operating room for tricuspid valve replacement.

Neonates undergoing a repair to create a single ventricle physiology have shunt-dependent pulmonary circulation, and balancing of pulmonary and systemic vascular resistances is essential to maintain adequate shunt flow, oxygenation, and adequate systemic blood flow for optimal tissue perfusion.

Myocardial ischemia may occur because the right coronary artery may be compromised by a suture plication of the atrialized right ventricle.

20.4.2 Measures to Reduce Pulmonary Vascular Resistance

The following measures are often employed during the postoperative period.

1. Sedation with infusions of a fentanyl.
2. Paralysis with a neuromuscular blocking agent.
3. Mechanical ventilation and ventilator manipulation.
4. Oxygenation and hyperventilation.
5. Inhaled nitric oxide and PGE_1 infusion.

20.4.3 Invasive Pressure Monitors

Arterial, central venous, and pulmonary artery (i.e., PA) line.

An oximetric catheter placed in the PA assesses cardiac output and pulmonary artery pressure.

20.4.4 Vasoactive Drug Infusions

Dopamine or dobutamine, milrinone isoproterenol, nitroprusside, magnesium, and inhaled nitric oxide (see Section I, Chapters 4 & 16).

20.4.5 Postoperative Bleeding

It is very rarely encountered.

20.4.6 Arrhythmias & AV Conduction Abnormalities

Reentrant atrial arrhythmias and ventricular arrhythmias occur and should be treated aggressively with intravenous anti-arrhythmic drugs (see Section I Chapter 16).

Temporary overdrive pacing:

Arrhythmias, occurring in the early postoperative period, may be difficult to manage medically and may be associated with ventricular dysfunction. Temporary overdrive pacing may be needed (see Section I Chapter 4).

Pacemaker:

Complete heart block may occur with either a valve repair or a valve replacement, requiring the use of a temporary pacemaker and possibly, the implantation of a permanent pacemaker.