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How I Manage Neonatal Ebstein's Anomaly

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Ebstein's anomaly of the tricuspid valve is characterized by a wide spectrum of severity. The condition involves more than a malformation of the tricuspid valve alone; a substantial portion of the right ventricle may be involved as well. Those patients presenting with symptoms in the first month of life represent a challenging group with a high mortality and uncertainty as to the best treatment options. In this review, the practical decision making process and the outcomes for neonates at one center are discussed.
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Morphologic Features and Clinical Presentation

Ebstein's malformation was first described by Wilhelm Ebstein in 1866 as a constellation of clinical findings resulting from an abnormality of the tricuspid valve.[1] However, it is much more than a valve issue because the condition involves the structure and function of the entirety of the right heart. Ebstein's anomaly involves a spectrum of anatomic abnormalities of variable severity, including apical displacement of the septal and mural leaflets of the tricuspid valve, which have failed to delaminate from the underlying myocardium; thinning or atrialization of the inlet component of the right ventricle with a variable degree of dilatation; and malformation of the antero-superior leaflet with anomalous attachments, redundancy, and fenestrations. Several other cardiac anomalies are often associated with Ebstein's anomaly, including atrial and ventricular septal defects, right ventricular outflow tract obstruction, and Wolff-Parkinson-White syndrome.

The malformation is rare, accounting for no more than 1% of all congenital cardiac anomalies. Because of the significant anatomic variability in the abnormalities of the tricuspid valve and right ventricle, the age at presentation and severity of symptoms can be highly variable. The presentation of patients with Ebstein's anomaly generally takes two forms, neonates and older children or adults. It is the symptomatic neonate, however, where management has been controversial and the risk considerable. There is a high rate of fetal demise, and symptomatic newborns will often present with a combination of cyanosis and congestive heart failure. An element of pulmonary hypoplasia resulting from the massively dilated heart in utero may contribute to the symptoms and affect outcomes.

The symptomatic neonate will generally present with cyanosis, and pulmonary blood flow may be duct dependent. The chest radiogram often demonstrates an enormously dilated heart filling the entire chest cavity. An infusion of prostaglandin E_1 is begun to maintain ductal patency and assure adequate pulmonary blood flow. At this stage, complete anatomic evaluation by Doppler/echocardiography is performed to assess the specifics, particularly the presence of right ventricular outflow tract obstruction. However, even this may be difficult to document because the elevated pulmonary vascular resistance (PVR) of the newborn may inhibit forward flow across the pulmonary valve, particularly when tricuspid valve regurgitation is severe and right ventricular function poor, resulting in uncertainty whether the outflow tract obstruction is functional or anatomic. If it is anatomic, a systemic to pulmonary artery shunt or right ventricular outflow tract reconstruction will be needed to maintain pulmonary blood flow. However, if it is functional, a trial of weaning the prostaglandin as the PVR falls is indicated. The administration of inhaled nitric oxide may also be useful in this setting. If the wean is successful without congestive heart failure, no further intervention may be needed. An unsuccessful wean, however, may result in two different scenarios. If cyanosis alone is noted, a systemic to pulmonary artery shunt is

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generally all that is needed. On the other hand, if cyanosis is accompanied by congestive heart failure due to significant tricuspid regurgitation and right ventricular dysfunction, a shunt alone will only mimic the condition seen on prostaglandin. The tricuspid valve itself must then be addressed. In this situation, either closure of the tricuspid valve annulus with a patch or tricuspid valve repair can be performed. The patch may or may not be perforated, a necessary procedure with associated right ventricular outflow tract obstruction.

The decision to repair the valve rather than simply closing it depends on the anatomy and likelihood that a repair will be successful. However, repairing a valve in the setting of a gravely ill newborn with poor right ventricular function is generally unwise given the variability of anatomy and unpredictable result. In either situation, the atrial septal defect must be kept open to allow a controlled degree of right-to-left shunt and avoid right heart failure. When repair is undertaken, a variety of surgical options exist to address the malformed tricuspid valve. Most repair techniques are designed to mobilize the leading edge of the antero-superior leaflet to create a competent monocusp valve with or without plication of the atrialized portion of the right ventricle. There is no clear consensus as to the necessity of obliterating the atrialized portion of the right ventricle. Historically, plication of this portion of the ventricle has been an integral part of most repairs, even though no clear physiologic benefit with regards to improved ventricular function has been demonstrated. In addition, the potential for injury to the right coronary artery and its branches as a result of the plication, which may adversely impact late outcomes and contribute to ventricular arrhythmias, must be considered. For these reasons, it has been our policy to avoid plication.

The technique reported by Dearani and Danielson[2] involves the horizontal plication of the atrialized portion of the right ventricle and mobilization of the antero-superior leaflet to create a monocusp valve. A modified technique developed by Carpentier et al[3] involves the mobilization of the antero-superior leaflet followed by the detachment of that leaflet from the annulus. The atrialized ventricle is then plicated longitudinally and the anterior leaflet is reattached at the level of the true annulus. The technique promoted by Ullmann and Sebening[4] does not involve exclusion of the atrialized ventricle. With this technique, the septal leaflet is mobilized and reattached to the true annulus. The secondary attachments of the antero-superior leaflet are also mobilized to allow the free edge to coapt with the newly liberated septal leaflet. The valvuloplasty technique pioneered by Sebening[5] also does not involve the exclusion of the atrialized ventricle. The repair creates a monocusp valve using a single mattress suture reinforced with a pledget placed from the mid-portion of the free edge of the antero-superior leaflet to the atrialized wall of the right ventricle directly opposite. The antero-inferior commissure is closed and a De Vega[6] annuloplasty is performed as necessary to achieve competence.

The preferred technique at the University of Michigan was popularized by da Silva and colleagues,[7] and is referred to as the "cone" procedure. It represents a modification of the basic principle reported by Carpentier.[3] In this technique, the leading edge of the mobilized anterior and posterior leaflets are rotated clockwise and sutured to the septal surface of the anterior leaflet to form a cone within the right ventricle.

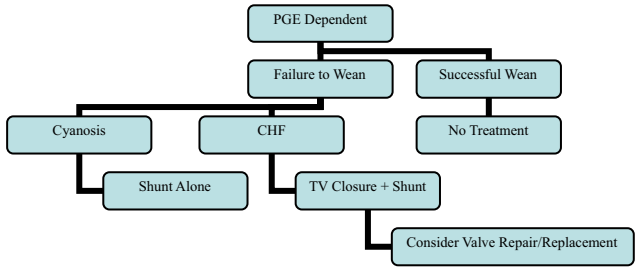

Figure 1 Treatment protocol for Ebstein's anomaly in the neonate.

University of Michigan Neonatal Experience

Between January 1988 and June 2008, 40 consecutive neonates with Ebstein's anomaly presented to the University of Michigan C.S. Mott Children's Hospital (Ann Arbor, MI). Sixteen of the patients required no surgical intervention and the remaining 24 patients underwent surgery during the neonatal period according to the criteria described above (Fig. 1). Among those requiring surgical intervention, median age at the time of operation was 6 days (range, 1 to 17 days) and median weight was 3.2 kg (range, 2.5 to 4.1 kg). Eleven out of the 24 patients underwent tricuspid valve closure with a systemic to pulmonary artery shunt, nine underwent a shunt alone, and four patients underwent a tricuspid valve repair. Two patients required tricuspid valve closure after an initial shunt, one was closed 2 days after the shunt procedure, and the other was performed at the time of additional surgical palliation (hemi-Fontan procedure).[8]

Among the 16 patients who received no intervention, 15 survived the neonatal period, two patients were lost to follow-up, three patients required tricuspid valve repair later in life (4, 8, and 13 years of age), and one patient required coarctation of the aorta repair at 1 year of age. Among the 24 neonates undergoing operation, there were six hospital deaths, three who underwent closure of the tricuspid valve and three who underwent a tricuspid valve repair. There were four late deaths among the 18 early survivors at a mean follow-up of 7.2 $\pm$ 5.1 years. All long-term survivors are in NYHA Class I or II. Subdividing those patients requiring intervention by procedure revealed that among the 11 patients who underwent RV exclusion, eight patients (73%) survived to discharge. One patient died suddenly at home 3 months after operation from an unknown etiology and seven patients (63%) successfully completed a Fontan procedure, although one patient died 11 years later from systemic infection.

All nine patients who underwent a systemic to pulmonary artery shunt survived to discharge. One patient required extracorporeal membrane oxygenation support and tricuspid valve closure early postoperatively. Two patients have completed a Fontan procedure, one patient underwent tricuspid

valve replacement and RV-PA conduit, one patient underwent 1 and ½ ventricle repair, and one patient is awaiting a Fontan procedure following tricuspid valve closure at the time of a stage II procedure. There were two late deaths; one patient died at the time of stage II palliation because of hypoxemia and low cardiac output syndrome and one patient died at the time of Fontan procedure because of severe ventricular dysfunction. Two patients were lost to follow-up.

Of four patients who underwent tricuspid valve repair, there were three hospital deaths. One patient survived and is doing well without re-operation for 12.5 years.

The overall survival estimate for all surgical intervention patients was 66.7% at 1 year, 62.2% at 5 and 10 years, and 51.9% at 15 years. Survival estimates for patients who underwent a RV exclusion procedure was 63.6% at 1, 5, and 10 years, and 47.7% at 15 years. For patients who underwent a systemic to pulmonary shunt, the actuarial survival was 88.9% at 1 year and 76.2% at 5 and 10 years. For patients who underwent a tricuspid valve repair, the actuarial survival was 25% at 1, 5, and 10 years. There was a significant statistical difference in the survival estimates between the patients with a systemic to pulmonary artery shunt and those undergoing tricuspid valve repair ($P = .019$).

Discussion

Until fairly recently, the outcome for the symptomatic neonate with Ebstein's anomaly was extremely poor. Lack of complete understanding of the morphology and pathophysiology contributed to these results, in addition to ill-conceived surgical approaches. The introduction of the concept of excluding the right ventricle by. Reemtsen and colleagues[9] was an important step in recognizing that the poorly functioning right heart must be dealt with to achieve optimal outcomes. Surgical procedures aimed at repairing or replacing the valve alone are unlikely to be successful in the majority of these patients.

At the University of Michigan, surgery is offered only for symptomatic neonates who fail a trial of weaning prostaglandin. Most patients with Ebstein's anomaly presenting in the neonatal period may be managed satisfactorily without intervention and may require repair later in life. The neonate who remains symptomatic, however, will require operative intervention. If a trial of prostaglandin results in a satisfactory clinical condition with acceptable systemic oxygen saturation in the absence of clinical congestive heart failure, a systemic to pulmonary artery shunt is generally all that is needed. If the prostaglandin results in satisfactory oxygen saturation but also produces congestive heart failure, the tricuspid valve and the right ventricle need to be addressed either by tricuspid valve closure, repair, or replacement. Even a successful repair or replacement may fail to address the inability of the right ventricle to support sufficient pulmonary blood flow. Although transplantation remains an option in severe cases, standard approaches are usually associated with satisfactory outcomes.

The treatment algorithm outlined in this review has been successful in improving outcomes for this otherwise highly lethal condition. Although any single institutional experience will be limited because of the rarity of this condition, our results suggest that a systemic to pulmonary shunt alone produced the best outcomes. However, these data must be viewed in the appropriate context. As a retrospective, nonrandomized review, patients were selected for a particular treatment protocol based on anatomy and response to the administration of prostaglandin. It became evident to us that those neonates who improved their systemic oxygen levels at the expense of increasing congestive heart failure were unlikely to survive without addressing the major underlying problem, namely, the severe tricuspid regurgitation in association with poor right ventricular function. Once right ventricular exclusion became a part of the management, outcomes improved dramatically. The superior results in those patients undergoing a shunt alone is likely due to more favorable right ventricular function in those patients, not to an inherent superiority of that procedure.

In summary, the symptomatic neonate with Ebstein's anomaly is optimally stabilized on prostaglandin to support pulmonary blood flow while the specific cardiac morphology is determined. As PVR falls, a trial of weaning the prostaglandin is performed and both the success and the clinical pattern of response to re-initiation of the drug, if needed, are assessed. If cyanosis alone is the dominant symptom, a systemic to pulmonary artery shunt alone is performed. However, when poor right ventricular function in association with severe tricuspid regurgitation results, right ventricular exclusion by patching the tricuspid valve is the best option. In our opinion, repair or replacement of the valve is best avoided in the neonate but may be utilized selectively when the morphology of the valve and the RV function are favorable. The decision for a two-ventricle, 1 ½ ventricle, or total cavopulmonary connection approach can be made at a later date.

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