Conversion of complex neonatal Ebstein’s anomaly into functional tricuspid or pulmonary atresia

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Abstract

Background: Ebstein’s anomaly, due to failure of delamination of one or more leaflets of the tricuspid valve (TV), is associated with varying degrees of tricuspid regurgitation (TR) and dysplasia of the right ventricle (RV). Although refinement of tricuspid valvuloplasty and plication techniques have opened the way to a satisfactory outlook for the majority of older children and adults, Ebstein’s anomaly presenting at neonatal age, secondary to ineffective forward flow into the pulmonary and systemic circulation, has a reported mortality rate of as high as 75%. In order to improve the dismal outcome in neonatal Ebstein’s anomaly, we have strived for early univentricular palliation.

Patients and methods: Univentricular repair was performed in five neonates (median age 5 days; range 2–14 days) with Ebstein’s anomaly, ductal dependent pulmonary blood flow, severe TR, absence of forward flow across the pulmonary valve, and small left ventricular (LV) area due to right-to-left bowing of the ventricular septum and ineffective LV loading (median indexed LV area 10.5 cm²/m²). In addition, two neonates had moderate pulmonary regurgitation (PR), one with additional pulmonary stenosis. In all patients, the indexed area of the combined right atrium and atrialized RV was greater than that of the combined functional RV, left atrium, and left ventricle (median 22.0 and 20.8 cm²/m², respectively). The median preoperative systemic oxygen tension was 35 mmHg and the median pH 7.28. Repair consisted of TV closure with a pericardial patch (with the coronary sinus draining into the RV) (n = 3) or, in the presence of PR, resection of the dysplastic TV and division and oversewing of the main pulmonary artery (n = 2), as well as excision of the atrial septum, resection of redundant right atrial wall, and construction of an aortopulmonary shunt (n = 5). Results: The median indexed LV area increased from 10.5 to 18.8 cm²/m² as a result of more effective loading of the left ventricle. There was no intraoperative or late mortality. The patients were extubated at a median of 7 days postoperatively. At discharge, the median systemic oxygen tension was 46 mmHg. In all five patients, at 6, 7, 10, 12 and 16 weeks of age, a bidirectional cavopulmonary anastomosis has been constructed.

Conclusions: In neonates with Ebstein’s anomaly and ductal dependent pulmonary blood flow, rational palliation consists of the surgical creation of tricuspid atresia or, in the additional presence of PR or pulmonary stenosis, the creation of pulmonary atresia. These procedures may result in effective LV decompression and more effective volume loading of the left ventricle with increase of systemic output and improved clinical outcome. © 1998 Elsevier Science B.V. All rights reserved.

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1. Introduction

Ebstein’s anomaly is characterized by various degrees of inferior displacement of the proximal attachments of the tricuspid valve (TV) ring, TV dysplasia, right ventricular dysplasia, and abnormalities of the distal attachments of the TV. These morphologic deformities, in widely different degrees of severity, are associated with
a variety of hemodynamic alterations leading to cyanosis, congestive heart failure, and arrhythmias. Refinement of tricuspid valvuloplasty and plication techniques [1–5] has opened the way to a satisfactory long-term outlook for the majority of older children and adults, who generally are only mildly or moderately symptomatic. Ebstein’s anomaly presenting in neonates and young infants, however, has a considerably less favorable natural history with a reported mortality rate of as high as 75% [6–11]. Recent experience with modifications of univentricular surgical palliation in neonates with Ebstein’s anomaly forms the basis of this report.

2. Material and methods

Between January 1995 and February 1997, univentricular repair was performed in five full-term neonates (median age 5 days; range 2 to 14 days). One neonate who was in extremis on admission (pH 7.10 and systemic arterial oxygen tension of 25 mmHg) was not operated on and died 1 day after admission. The median weight of the operated patients was 3.3 kg (range 2.6–3.5 kg). All patients had echocardiographically documented Ebstein’s anomaly, ductal dependent pulmonary blood flow, severe tricuspid regurgitation, absence of forward flow across the pulmonary valve, and small left ventricular (LV) area due to bowing of the ventricular septum from right-to-left and ineffective LV loading (median indexed LV area 10.5 cm²/m²) (Table 1) (Fig. 1). In addition, two neonates had moderate pulmonary regurgitation, one with additional pulmonary stenosis (Fig. 2). All patients were treated with prostaglandin E₁ to maintain ductal patency. In all neonates, the septal and posterior leaflets of the TV were displaced into the right ventricle; the anterior leaflet to various degrees was tethered by accessory attachments of the leaflet to the right ventricular wall, causing restriction of motion of the valve leaflet. In all patients, the right ventricle was almost completely atrialized with the exception of a small trabecular component and the infundibulum. Echocardiographically, using a four-chamber view at end-diastole, the indexed area of the combined right atrium and atrialized right ventricle was greater than that of the combined functional RV, left atrium, and left ventricle (median 22.0 and 20.8 cm²/m², respectively). All neonates had cyanosis with a median systemic arterial oxygen tension of 35 mmHg (range 28–45 mmHg) and a median pH of 7.28 (range 7.24–7.36).

In the three patients with absence of pulmonary regurgitation or stenosis, using continuous hypothermic low-flow perfusion, repair consisted of resection of the dysmorphic TV, closure of the communication between the right atrium and right ventricle with a pericardial patch, thereby placing the coronary sinus on the ventricular side of the patch, and resection of the atrial septum (Fig. 3). Finally, a 3.5 mm (n = 2) and 4.0 mm (n = 1) central aortopulmonary shunt consisting of polytetrafluoroethylene (Gore-Tex, W.L. Gore and Associates, Elkton, Maryland) was constructed. In the two patients with additional pulmonary regurgitation and pulmonary regurgitation and stenosis, repair consisted of resection of the dysmorphic TV, excision of the atrial septum, resection of redundant right atrial wall, oversewing of the main pulmonary artery, and construction of a 4.0 mm aortopulmonary shunt (Fig. 4). All patients were weaned from cardiopulmonary bypass with inotropic medication to support the dysplastic right ventricle. Postoperatively, the patients were managed by hyperventilation and administration of prostaglandin E₁ to maximize pulmonary vasodilation.

3. Results

The median indexed LV area increased from 10.5 to 18.8 cm²/m² as a result of more effective loading of the left ventricle and reversal of septal bowing (Table 1).
Fig. 1. Preoperative echocardiographic views of Ebstein’s anomaly. (A) Patient with marked atrialization of the right ventricle (RV), small functional RV, and relatively small left ventricle (LV) due to right-to-left septal bowing and ineffective loading of the LV. (B) Continuous wave Doppler echocardiogram demonstrates severe tricuspid regurgitation.

Fig. 2. Preoperative echocardiographic views of patient with Ebstein’s anomaly. (Left) The functional right ventricle (RV) (arrows) is relatively small. (Right) Doppler echocardiogram demonstrates marked tricuspid regurgitation (TR). In addition, there is pulmonary valve stenosis (PS) with regurgitation. AO, aorta; LA, left atrium; PA, pulmonary artery; RA, right atrium.

There was no intraoperative or late mortality. The patients were extubated at a median of 7 days postoperatively (range 4–11 days). In one patient, a temporary atrioventricular block converted to sinus rhythm on the fifth postoperative day. In another patient, a paralyzed left diaphragm was successfully plicated. At discharge, the median systemic oxygen tension was 46 mm Hg (range 42–51 mmHg). In all five patients, at 6, 7, 10, 12 and 16 weeks of age, a bidirectional cavopulmonary anastomosis was successfully constructed with a resulting median systemic oxygen saturation of 89%. In the patients with surgically created tricuspid atresia, the pulmonary valve was left in communication with the right ventricle to allow egress of the coronary sinus blood. Range of follow-up has been from 7 to 24 months. All patients have been free of symptoms.

4. Discussion

Ebstein’s anomaly is a congenital defect of the tricuspid valve in which the origins of the septal and posterior leaflets are both displaced downward into the right ventricle with varying degrees of deformation of the leaflets. Right ventricular dysplasia, marked right atrial and atrialized right ventricular enlargement, left ventricular compression, and tethering of the anterior leaflet commonly coexist. The most common associated anomaly is pulmonary stenosis or atresia, which may be present in up to one-third of autopsied hearts [12]. In addition, an increased fibrous tissue content of both the right and left ventricles has been reported [13]. This constellation of morphologic features portends an extremely poor prognosis, with reported mortality rates of as high as 75% [6–11,13]. Corresponding clinical risk factors for a poor outcome in neonates and infants with
Fig. 3. Conversion of Ebstein’s anomaly into tricuspid atresia. (A) The atrial septum is excised and the dysmorphic tricuspid valve is resected. A pericardial patch is sewn into place, leaving the coronary sinus (dashed line) beneath the patch. (B) Redundant right atrial wall is resected, the right atrium is closed, and a central aortopulmonary shunt is constructed.

Ebstein’s anomaly are the need for mechanical ventilation, dependence on PGE1, congestive heart failure and the absence of adequate forward flow from the right ventricle to the pulmonary artery (i.e. physiologic pulmonary atresia).

The management of Ebstein’s anomaly depends on the age of presentation, anatomic severity of the lesion, presence of associated defects and clinical features. Fetal Ebstein’s anomaly traditionally has an appalling outlook and prenatal counseling should take this into account. Previous attempts at palliative surgical treat-

Fig. 4. Conversion of Ebstein’s anomaly into pulmonary atresia. After resection of the dysmorphic tricuspid valve and excision of the atrial septum, the pulmonary trunk is divided and both ends are oversewn. Finally, a central aortopulmonary shunt is constructed.

Fig. 5. Echocardiogram after conversion of Ebstein’s anomaly into tricuspid atresia demonstrating the pericardial patch (arrows) between right atrium (RA) and right ventricle (RV) and the unrestricted communication at atrial level. Note that the left ventricle (LV) has assumed a relatively normal size due to increased volume loading and reversal of septal bowing. LA, left atrium.
ment of severe Ebstein’s anomaly at neonatal age have produced dismal results [1,7,14]. We believe that in such situations, rational palliation consists of the surgical creation of tricuspid atresia, as first propagated by Starnes and colleagues [15]. In the additional presence of marked pulmonary regurgitation or stenosis, the creation of pulmonary atresia may be preferable to right ventricular volume or pressure overload with secondary right-to-left bulging of the ventricular septum. These procedures allow egress of coronary sinus blood into the pulmonary circulation or right atrium, respectively. In our experience, the aortopulmonary shunt should be relatively large (4.0 mm in neonates with a weight of 3.0 kg or more and 3.5 mm in smaller neonates) to accomplish adequate left ventricular filling. The ultimate beneficial effects of the creation of univentricular physiology consist of: (1) The establishment of a reliable source of pulmonary blood flow; and (2) decompression of the left ventricle by reversal of septal bowing and more effective volume loading of the left ventricle with resulting increase of systemic output and improved clinical outcome (in analogy to the situation after biventricular repair of unbalanced atroventricular canal [16]). In all five patients in this report, the creation of a functionally single ventricle resulted in a satisfactory hemodynamic situation and adequate systemic oxygen saturation. The anatomy was subsequently suitable for the construction of a bidirectional cavopulmonary anastomosis. The long-term prognosis of patients who undergo univentricular palliation for Ebstein’s anomaly is unknown and needs to be carefully studied. Increased fibrous content of the left ventricular wall may contribute to a persistent risk of late hemodynamic deterioration and sudden death.

References


Appendix A. Conference discussion

Dr Willem J. Daenen (Leuven, Belgium): I’m not sure that the creation of pulmonary atresia is always necessary, because we had a patient who fulfilled all of your criteria and he presented with a pulmonary atresia anatomically where we did the operation you described today. It went all right. But postoperatively the child went into left ventricular failure because the right ventricle distended, pushed the septum away, and gave rise to poor left ventricular ejection. Therefore, I’m not so sure that the creation of pulmonary atresia is the right approach because there is a lot of blood still draining into the right ventricle, it cannot get out, and you can end up with an extremely distented right ventricle.

Dr van Son: Your point is well taken. It is indeed possible that the right ventricle distends after the operation with resulting leftward shift of the septum and left ventricular out-flow obstruction. A relatively large aortopulmonary shunt promotes left ventricular filling and therefore counteracts right-to-left septal bowing. With a protocol of inotropic support, pulmonary vasodilation, hyperventilation, and paralyzing the neonate for 24 h or more if needed, we have been successful in managing these cases.

Dr Stellin (Padova, Italy): The drawing that you showed in your slide is the one of Dr Vaughn Starnes’ original article of 4–5 years ago. My criticism towards this operation (which implies the construction of a central shunt) is that in univentricular heart malformations, it is always difficult to balance pulmonary and systemic resistance. Furthermore, in this malformation, the blind RV may interfere with the LV contractility. I wonder whether you have encountered any problem after this procedure and what do you think will be the future of the RV camber that in this malformation is always of a consistent size?

Dr van Son: You are right that a modified Blalock–Taussig shunt may be the better physiologic option. Advantages of a central shunt are the ease of its construction and therefore reduced operative time.
In addition, there is no distortion of the branch pulmonary arteries.
In our experience, in ductal dependent Ebstein’s anomaly, the central shunt should be relatively large to accomplish adequate left ventricular filling. Even if the pulmonary blood flow is somewhat luxurious there will hardly be detrimental sequelae physiologically because we construct the Glenn shunt at an age of only several weeks. The postoperative management protocol that I alluded to helps in the prevention of right-to-left septal bowing and left ventricular outflow obstruction. In the long-term, the right ventricle adapts to its decreased volume and pressure load and atrophies to a certain degree.

Dr Stellin: In other words you have never had any problem with a dysplastic and dilated RV interfering with the LV contraction, after TV closure?

Dr van Son: I must admit that these patients can be quite sick postoperatively, but with the proper operation and careful postoperative management their clinical status usually gradually improves.

Dr Christopher Knott-Craig (Oklahoma City, Oklahoma): In my own institution I have taken a different approach and for the last 4 years have repaired all of the symptomatic neonates with severe Ebstein’s anomaly without mortality. I think there is a group of patients in whom both the tricuspid valve is amenable to repair, and the right ventricle may function quite adequately as a pulmonary ventricle. So, I would caution against dispensing all the right ventricles in this group of patients.

I have one comment: in your sketches you have shown that you patched the coronary sinus into the blind right ventricle. Do you think this is going to be a problem for the patient or do you think that the coronary sinus acts as a release valve and that some of the blood that is in the right ventricle drains backwards into the coronary sinus and then into the right atrium after your repair?

Dr van Son: Of course, as in any congenital cardiac anomaly, in Ebstein’s anomaly there is a spectrum with regard to dysmorphic features of the tricuspid valve and right ventricular dysplasia. Especially tethering of the anterior leaflet to the right ventricular wall and a small functional right ventricle, as present in all of our patients, makes a successful conventional biventricular repair unlikely. From natural history studies we know that the mortality rate in this category of patients is ≈ 75% at the age of 3 months. These sick patients with ductal dependent circulation and limited forward flow from the dysplastic right ventricle into the pulmonary circulation physiologically already behave as having a poor Fontan circulation. In this category of patients in my opinion you have to commit early on to a protocol of univentricular repair. In milder forms of Ebstein’s anomaly to which you probably alluded, biventricular repair of course remains the goal. In intermediate cases where the right ventricle is expected to be able to carry at least 50% of the preload, a supplemental bidirectional cavopulmonary anastomosis, in addition to repair of the tricuspid valve, may be a valuable alternative option.

With regard to your second question, if functional tricuspid atresia is created with the coronary sinus draining into the right ventricle, there usually is not a problem because the right ventricle in this setting is a low pressure compartment. In addition, the valve guarding the coronary sinus will prevent retrograde flow into the coronary sinus during ventricular systole. In our cases there was good egress of blood from the coronary sinus and right ventricle into the pulmonary circulation.

Dr Gosta Pettersson (Copenhagen, Denmark): I would just like to ask you, why do anything more than just a shunt?

Dr van Son: The purpose of creating functional tricuspid or pulmonary atresia in neonatal Ebstein’s anomaly is not only to provide a reliable source of pulmonary blood flow, but also to reduce massive right ventricular cardiomegaly, to abolish tricuspid regurgitation, and to create a bridge toward successful univentricular palliation. In order to create an ultimately successful Fontan circulation, it is subsequently mandatory that the volume loading of the future single (left) ventricle is reduced drastically and early in life by a bidirectional cavopulmonary anastomosis.