Neonatal Ebstein's Anomaly: Surgical Decision Making

Awori M, Mutwiri RK, Mutunga WM, Jowi SO, Peninan KN, Mang'usu OE
School of Medicine University of Nairobi

Correspondence to: Dr Mark Awori, P. O Box 14677-00800, Nairobi, Kenya. Email: mnawori@yahoo.com

Summary

Ebstein's Anomaly (EA) is a rare form of congenital heart disease. Surgical decision-making in neonates is controversial. In developing countries, neonates with Ebstein's anomaly requiring surgical intervention rarely present to health institutions capable of providing intervention. We present a 2.2 kg term female neonate who presented with Ebstein's anomaly. Data was obtained retrospectively from clinical case notes. We describe our surgical decision-making process and outcome in relation to prevailing thought regarding the management of this condition and we make a recommendation on what we consider to be the best surgical option for these patients.

Key words: Ebstein's anomaly, Neonatal, Treatment

Ann Afr Surg. 2018; 15(1):40-43
DOI:http://dx.doi.org/10.4314/aas.v15i1.9
© 2018 Author. This work is licensed under the Creative Commons Attribution 4.0 International License

Introduction

Ebstein's anomaly is a rare form of congenital heart disease with and estimated prevalence of 1:20,000 live births (1). It has 3 characteristic features: (i) a downward displacement of the septal and posterior leaflets into the right ventricle; (ii) an atrialized region of the right ventricle between the tricuspid annulus and the attachment of septal and posterior leaflets; and (iii) a malformation of the right ventricle (RV) where it is small, thin-walled and often has abnormal systolic function. There may be right ventricular outflow tract obstruction (RVOTO). The key surgical decisions to be made are: (i) whether a two ventricle repair is feasible and (ii) where it is not, whether a Starne's procedure performed on cardiopulmonary bypass is required to facilitate successful single ventricle repair. We present our surgical experience in 2.2 kg neonate presenting with Ebstein's anomaly.

Case Report

A 2.2 kg term female neonate was referred to the Kenyatta National Hospital (the main teaching and referral hospital in Kenya) on the 8th day of life with a diagnosis of 'complex congenital heart disease'; this had been confirmed by echocardiography on the 2nd day of life. She was born via emergency caesarean-section (CS) for fetal distress. At CS there was meconium stained liquor; the Apgar score was '4'at 1 minute, '6' at 5 minutes and '8' at 10 minutes. On admission (to our institution) she was 'sick-looking'; in respiratory distress (respiratory rate of 42 breaths/minute) and was afebrile. She had both peripheral and central cyanosis; her heart rate was 154 beats each minute and she had a pan systolic murmur. The rest of her physical examination was unremarkable. The patient was tolerating full nasogastric feeds of expressed breast milk. A Prostin (Prostaglandin E_2) intravenous infusion was running at 0.05mcg/kg/minute and intravenous flucloxacillin and amikacin had been started at the referring institution for presumed neonatal sepsis (we changed the flucloxacillin to ceftazidime). A chest radiograph showed gross cardiomegaly (cardiothoracic ratio 0.8) and a repeat transthoracic echocardiogram confirmed Ebstein's anomaly (EA). The septal leaflet of the tricuspid valve was moderately displaced inferiorly and the anterior leaflet was long and sail like. The right ventricular volume was approximately ¼ that of the left ventricle and there was severe tricuspid regurgitation associated with a 6mm (diameter) secundum atrial

septal defect shunting right-to-left. The interventricular septum was intact. The pulmonary valve was atretic, the branch pulmonary arteries were confluent; the right pulmonary artery was 3mm (diameter) and the left was 4.5mm (diameter). There was a moderate sized patent ductus arteriosus that was tapering at the pulmonary end. Arterial blood gas analysis (on admission) revealed: a pH of 7.083 (7.35-7.45), a pCO2 of 10.21(4.2-6.00kpa), a pO2of 4.44 (10.00-13.33kpa) and an HCO3 of 22.4mmol/l (22-26mmol/l). We suspected that this was a venous sample; unfortunately another sample was not taken for a repeat analysis. Additional pre-operative blood works were performed on the 10th day of life and the renal function test and the full hemogram results were essentially normal for an 8 day old neonate.

After adequate resuscitation, and following a Cardiac team meeting, a right modified Blalock-Taussig shunt (3.5mm diameter ePTFE; figure 1) was created via a right thoracotomy on the 10th day of life. An intravenous infusion of heparin was started on return to the neonatal intensive care unit on the day of surgery. An echocardiogram performed on the 1st postoperative day (POD 1) showed a functioning shunt and liver function tests revealed the following (significant derangement):

- Albumin= 32g/l (35-49g/l)
- Total bilirubin= 42.5µmol/l (2.0-21.0µmol/l)
- Indirect bilirubin= 20.5µmol/l
- Direct bilirubin= 22.0µmol/l (0.0-5.1µmol/l)
- Alanine transaminase= 37U/l (0-34U/l)
- Aspartate transaminase=115U/l (0-31U/l)
- Gamma Glutamyl transferase= 138U/l (0-32U/l)
- Alkaline phosphatase=209U/l (40-125U/L)

The patient was extubated on POD 2; the haemoglobin (Hb) was 7g/dL and the arterio-venous oxygen saturation difference was 42% (the liver was not palpable). The patient was transfused (whole blood) to a Hb of 13g/dL. On POD 3 the patient was re-intubated because of worsening respiratory acidosis; this resolved soon after positive pressure ventilation was initiated. There were no postoperative radiographic features of pneumothorax, pulmonary edema or congenital lung abnormality prior to this re-intubation. On POD 6 the FiO2 was reduced to 40% (the arterial O2 saturation was 80%) and then to 30% later the same day. Trophic feeds had been started on POD 3 but were later stopped because of 'coffee ground' naso-gastric tube aspirates associated with an activated partial thromboplastin time (aPTT) that was greater than 120 seconds (The heparin infusion was adjusted as per protocol). On POD 7 a left internal jugular central venous catheter (CVC) was inserted in anticipation of starting total parenteral nutrition. A pneumothorax was noted on a post CVC insertion chest radiograph and a chest drain was inserted. On POD 8, the patient desaturated suddenly and went into bradycardia; resuscitation was unsuccessful.

Discussion

Ebstein's anomaly can be classified into 4 types depending on the severity of these malformations (2):

• Type a. Large, mobile anterior leaflet with minimal displacement of the septal and posterior leaflets (the volume of the true RV is adequate)

• Type b. Large, mobile anterior leaflet with moderate displacement of the septal and posterior leaflets (the true RV is small).

• Type c. Movement of the anterior leaflet is restricted with severe displacement of the septal and posterior leaflets (the true RV is small and there is RVOTO).

• Type d. The leaflets are sacular and attached to the ventricular wall; they have no movement.

Newborns with this anomaly may be cyanotic or acyanotic; cyanosis tends to improve as the pulmonary vascular resistance (PVR) falls with age. Generally speaking hemodynamic deterioration with increasing cyanosis, congestive heart failure (CHF) and left ventricle (LV) dysfunction occurs later in life. Supraventricular tachycardia (SVT) occurs in about 20% of all patients (3). In developed nations, approximately half of the neonates with Ebstein's anomaly present with cyanosis and a cardiac murmur in the 1st week of life(4,5) and some 18% of symptomatic newborns die in the neonatal period; 30% of patients die before 10 years of age (usually from CHF) and the median age at death is 20 years(3).

Symptomatic newborns receive intensive treatment with PGE1, inotropes and mechanical ventilation as required. Acute episodes of SVT are treated with adenosine; beta

blockers are first-line prevent therapy (3). Symptomatic newborns who have mild EA and who respond to intensive treatment will have their PGE1 and inotropes gradually removed to see the effect of ductus closure. Asymptomatic children with mild EA will only require regular observation; if CHF develops; anti-failure medication (digoxin and diuretics) will be required.

Failed intensive treatment (i.e. reliance on mechanical ventilation and persistent cardiomegally) in the neonatal period is an indication for surgical intervention; the nature of surgical intervention is controversial. The goal of surgery is to ultimately give the child a two ventricular system; if this is not possible, an efficient single ventricle system (Fontan circulation; figure 2) is the alternative. Some authors believe that total correction in the neonatal period should be avoided because of poor results (6). These authors recommend RV exclusion and the creation of a single ventricle physiology with the Fontan procedure being the ultimate destination. These authorities feel that if EA is associated with anatomical pulmonary atresia (APA), even RV exclusion should be avoided and the only option left for such a patient is heart-lung transplantation. This position implies that only candidates who are asymptomatic and have a good-sized RV should have a two ventricular repair (3).

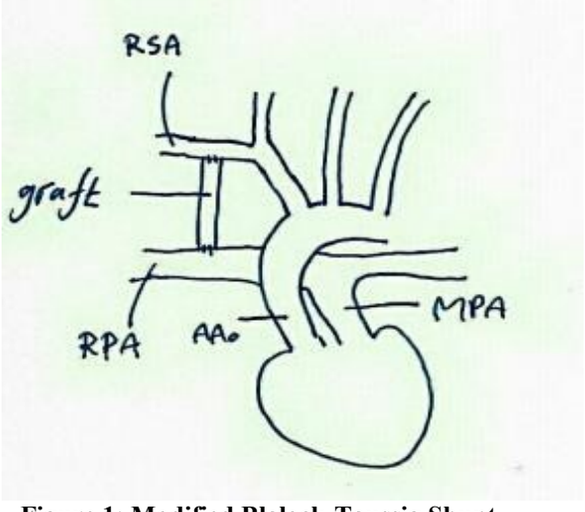

Figure 1: Modified Blalock-Taussig Shunt
RSA= right subclavian artery; RPA= right pulmonary artery
AAo = ascending aorta; MPA= main pulmonary artery

Creation of a modified Blalock-Taussig shunt (MBTS) and enlargement of the atrial septal defect (ASD) has been reported to be life saving if the LV is not 'pancaked' by a grossly enlarged RA and RV (3). The

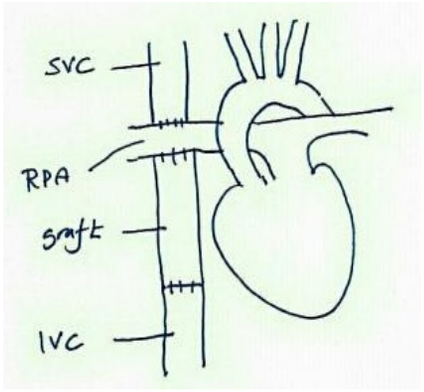

Figure 2: The Fontan Circulation
SVC = superior vena cava; IVC = inferior vena cava; RPA = right pulmonary artery

largest study (7) on the surgical management of neonatal EA recommended a two ventricular repair even in cyanotic neonates as long as the ventricles are of adequate size [NB: a RV-to-Pulmonary artery (PA) conduit would be required if anatomical pulmonary atresia exists]. This study also advocates the Starnes procedure if there is APA, moderate /severe tricuspid regurgitation and a small RV. In addition, if the RV is small but there is only mild TR, the Starnes procedure is not indicated; a BTS and reduction atrioplasty is sufficient. Other studies have found operative mortalities as high as 75% for palliative procedures such as BT shunts (8).

Our patient had APA, a small RV and severe TR. As the LV was not "pancaked" by the RV and as there was some evidence that a BTS alone might be adequate (3), we thought that the most pragmatic option would be to perform a BTS only. The post-operative period was marked by persistent respiratory failure secondary to compression if the lungs by a grossly enlarged heart. When mechanical ventilation was recommenced, respiratory function and cardiac output were sufficient. We feel, in retrospect, that had we performed a procedure that would have resulted in a decrease in the size of the heart (Starnes with atrioplasty); we may have been able to wean the neonate off the ventilator. In this regard we are now in agreement with the recommendation by Knottscraige (7); a Starnes procedure with a right reduction atrioplasty would have been the optimal procedure for this patient. A rapid two stage Starnes procedure has been reported to be successful. This method involves performing an BTS

and right reduction atrioplasty at the first operation followed by a Starnes procedure on cardiopulmonary bypass a few days later(9). This strategy may allow transfer of a reasonably stable patient to an area that has bypass capabilities or better experience with neonatal surgery on cardiopulmonary bypass. Our patient desaturated suddenly on POD 8; developed bradycardia and could not be resuscitated. We think that the desaturation was most likely due to a compromised airway, perhaps a blocked endotracheal tube or a trachea/bronchus obstructed by blood (patient had a raised aPTT) or mucous.

In conclusion, two ventricular repair of EA should be attempted in the neonatal period if both ventricles are of adequate size. If the RV is small and there is moderate or severe TR, a Starnes procedure with right reduction atrioplasty should be performed; failure to do this (i.e., by performing a BTS alone) may result in compression of the lungs by a grossly enlarged heart and subsequent failure to wean from mechanical ventilation. We now surmise that a Starnes procedure with reduction atrioplasty is required in patients such as ours. Patients diagnosed in Kenya can be referred to Kenyatta National Hospital in a timely manner for appropriate management by the resident multidisciplinary team.

References

1. Siebert JR, Barr M Jr, Jackson JC, et al: Ebstein's Anomaly and Extracardiac Defects. Am J Dis Child 1989; 143: 570-2.
2. Carpentier A, Chauvaud S, Mace L, et al. A New Reconstructive Operation for Ebstein's Anomaly of the Tricuspid Valve. J Thorac Cardiovasc Surg.1988; 96:92-101.
3. Park MK. Cyanotic Congenital Heart Defects. Pediatric Cardiology for Practitioners.5th Edition. Philadelphia: Mosby Elsevier 2008. 273-9.
4. Giuliani ER, Fuster V, Brandenburg RO. Et Al. Ebstein's Anomaly: The Clinical Features and Natural History of Ebstein's Anomaly of the Tricuspid Valve. Mayo ClinProc1979; 54: 163-73.
5. Radford DJ, Graff RF, Neilson GH. Diagnosis and Natural History of Ebstein's Anomaly. Br Heart J 1985; 54: 517-22.
6. Nichols DG, Ungerleider RM, Spevak PJ, et al. Ebstein's Malformation. Critical Heart Disease in Infants and Children. 2nd Edition. Philadelphia. Mosby Elsevier 2006. 777-87.Print.
7. Knott-Craig CJ, Goldberg SP, Ballweg JA, et al. Surgical Decision Making in Neonatal Ebstein's Anomaly: An Algorithmic Approach Based on 48 Consecutive Neonates. World Journal for Pediatric and Congenital Heart Surgery 2012; 3:16-20.
8. Sarris GE, Giannopoulos NM, Tsoutsinos AJ, et al. Results of Surgery for Ebstein's Anomaly: A Multicentre Study from the European Congenital Heart Surgeons Association. J Thorac Cardiovascular Surgery 2006; 132: 50-7.
9. Kajihara N, Asou T, Takeda Y, et al. Rapid Two-Stage Starnes Procedure for a Symptomatic Neonate with Ebstein's Anomaly. Ann Thorac Surg. 2010; 90: 2073-5.