Surgical treatment for neonatal Ebstein’s anomaly with circular shunting

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Abstract

We report a case of surgical treatment for neonatal Ebstein’s anomaly with circular shunting. A prenatal diagnosis of Ebstein’s anomaly was made at 30 weeks of gestation. In addition to severe tricuspid regurgitation, severe pulmonary regurgitation was also noted, resulting in a ‘circular shunt’. At 38 weeks and 2 days of gestation, the baby was delivered by Caesarean section. A two-stage palliative operation was planned; the first palliative operation was performed as the second-stage palliation. The tricuspid valve was closed by an ePTFE patch with 3-mm fenestration. Pulmonary blood flow was supplied by a right modified Blalock-Taussig shunt with a 3.5-mm diameter ePTFE graft. The infant’s postoperative haemodynamic status was stable. The patient is being followed up carefully and is scheduled to have a univentricular repair in the future.

Keywords: Ebstein’s anomaly • Circular shunt • Congenital heart disease

INTRODUCTION

Ebstein’s anomaly is a malformation of the tricuspid valve. The clinical symptoms vary in severity depending on the anatomical malformation; severe tricuspid regurgitation (TR) and right ventricular hypoplasia produce a critical state in the neonate. The presence of both severe pulmonary regurgitation (PR) and tricuspid regurgitation is rare, with reversal of blood flow to the right ventricle, resulting in a ‘circular shunt’ [1] (Fig. 1). A rare case that underwent surgical treatment for severe neonatal Ebstein’s anomaly with circular shunting is described.

CASE REPORT

At a routine prenatal check-up at 30 weeks of pregnancy, foetal cardiomegaly was noted. Foetal echocardiography demonstrated severe TR, and Ebstein’s anomaly was diagnosed. PR was also present. Aortic flow passed through the ductus arteriosus and regurgitated to the left atrium. The right atrial pressure was normal and the aortic pressure was increased. The ductus arteriosus was patent. After birth, high right atrial pressure and pulmonary regurgitation were noted. At 38 weeks and 2 days of gestation, the baby was delivered by Caesarean section. The Apgar score was 8/10, height was 51.0 cm and weight was 2.7 kg. Soon after birth, the baby was intubated, and oxygen saturation was 80%. The chest X-ray showed cardiomegaly (cardiothoracic ratio (CTR), 72%). Lipo-PGE1 was administered continuously to keep the ductus arteriosus patent. Echocardiography showed that PR was moderate and trans-valved flow was to and fro. TR was severe and interatrial communication was adequate. The neonate was transferred to another operating theatre soon after birth for the first-stage palliative operation. After confirming the safety of the haemodynamics with pulmonary artery test clamping, the pulmonary artery was ligated. Postoperatively, the neonate was managed in an intensive care unit. Postoperative echocardiography showed that circular shunting was eliminated, TR was severe and TR pressure gradient was 6.7 mmHg. Four days after birth, symptoms of high pulmonary blood flow (dyspnoea, pulmonary artery congestion) appeared. Thus, the neonatal pulmonary hypertension appeared to have improved, and modified Starnes operation was performed. After re-median full sternotomy, cardiopulmonary bypass was established with direct cannulation to the ascending aorta, superior vena cava and inferior vena cava. After induction of cardioplegic arrest, the ligated pulmonary artery was transected. The dilated right atrium was opened, and the atrial septal defect was enlarged. The atrialized right ventricle was plicated. The tricuspid valve was closed with Gore-TEX® Cardiovascular Patch (W. L. Gore and Associates, Flagstaff, AZ, USA). A 3-mm fenestration was created in the centre of the patch using an aortic puncher. After...
declamping of the aorta, a spontaneous regular sinus rhythm was obtained. Then, a modified Blalock-Taussig shunt was created between innominate artery and right pulmonary artery with a GORE-TEX® Vascular Graft (3.5 mm, W. L. Gore and Associates). The patient was weaned off cardiopulmonary bypass easily. The sternum was closed, and the operation was completed. The baby was extubated on postoperative day 6. Postoperative echocardiography showed neither overdistension nor thrombus in the right ventricle. The patient is being followed up and scheduled to have a bidirectional Glenn and Fontan procedure in near future.

DISCUSSION

Ebstein’s anomaly sometimes presents with functional pulmonary atresia because the pulmonary artery pressure is greater than the Ebsteinoid right ventricular pressure. In the present case, pulmonary regurgitation was also present, and blood flow was reversed into the right ventricle. Circular shunting results in blood flow that does not contribute to the systemic circulation, causing low output syndrome and multiple organ failure. Surgical treatment must be performed to salvage this fatal state. Surgical treatments for neonatal Ebstein’s anomaly have two strategies, two ventricle repair [2] or univentricular repair. In the present case, the tricuspid valves were plastered onto the right ventricle wall and extremely thin. The right ventricle was hypoplastic, and pulmonary regurgitation was severe. These anatomical malformations led to us to perform univentricular repair.

A two-stage palliation operation that involved pulmonary artery ligation and a Starnes operation was planned. In our case, the neonate with circular shunting had a severe heart failure. In this poor condition, we were concerned about performing one-stage Starenes procedure for the following reasons. (i) The Starnes procedure dramatically changes cardiac haemodynamics and we are not sure that the baby can adapt to it. (ii) Neonatal pulmonary hypertension may cause failure of a systemic-to-pulmonary shunt. (iii) Cardiopulmonary bypass may cause low output syndrome. So, we performed pulmonary artery ligation as a first stage operation for closing circular shunt and improving heart failure. Actually, the CTR was improved from 74 to 67% after pulmonary artery ligation. The modified Starnes operation was then performed after the neonatal pulmonary hypertension improved. According to the operative strategy for Starnes operation, Kajiwara et al. [3] also reported a successful case of two-stage Starnes procedure for neonatal Ebstein’s anomaly. Although two-stage Starnes procedure may be a conservative strategy, we think that it is a good strategy for a seriously symptomatic neonate with Ebstein’s anomaly.

The Starnes operation was reported by Starnes in 1993 [4]. It is a useful palliation for Ebstein’s anomaly that is unsuitable for two-ventricle repair, and several cases have achieved functional complete repair [5–7] after the Starnes operation. In this procedure, some blood flow from the Thebesian vein was returned to the right ventricle, a drainage route from the right ventricle (RV) to the right atrium (RA) was necessary to prevent thrombus formation in RV [8]. In our hospital, a 3-mm fenestration is created in the centre of the patch to maintain RV–RA flow communication. Postoperative echocardiography showed that there was
neither overdistension nor thrombus in the RV. Blood drainage from the RV appeared to be adequate.

CONCLUSION
A rare case of neonatal Ebstein’s anomaly with circular shunting was reported. A two-stage surgical procedure was performed, and the outcome was good.

Conflict of interest: none declared.

REFERENCES

eComment. Two-stage repair of Ebstein’s anomaly in a neonate

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Ebstein’s anomaly is a congenital heart disease, which rarely requires surgical intervention in the neonatal period. Therefore, the case involving surgical treatment for neonatal Ebstein’s anomaly with circular shunting by Yohsuke Yanasa et al. is very interesting [1].
The clinical symptoms vary in severity depending on the anatomical malformation. Severe tricuspid regurgitation and right ventricular hypoplasia may produce a critical state in the neonate. Concomitant pulmonary regurgitation is rare, with the reversal of blood flow to the right ventricle, resulting in a ‘circular shunt’ and necessitating urgent surgical treatment for severe neonatal Ebstein’s anomaly.

Between 2006 and 2010, 107 patients (including one neonate and one infant) underwent surgery for Ebstein’s anomaly at Bakoulev Scientific Center for Cardiovascular Surgery at the Russian Academy of Medical Sciences. In our practice, due to the decrease in pulmonary vascular resistance, minimal antegrade blood flow via pulmonary valve was restored but it was not sufficient to ensure adequate oxygen saturation. We performed a modified Blalock-Taussig shunt with a 4-mm graft. Postoperative oxygen saturation was 80% and the baby was discharged. Six months later, a successful one and a half surgical repair was performed.
Surgery in the neonatal period has previously focused on palliation and conversion to single-ventricle physiology. Successful two-ventricle repair with good clinical results can safely be performed in the neonatal period but requires an individual approach in each case.

References