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Case Report

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# Two Ventricle Early Repair in Very Critical Neonatal Ebstein's Anomaly Using Cone Procedure after Starnes

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## 1. Abstract

Ebstein's anomaly (EA) is a rare congenital cardiac anomaly. Presentation varies from a severe symptomatic form during the neonatal period to an incidental detection later in life due to the wide morphological variation of the condition. Neonates not eligible or failing medical management should be surgically managed either with a single ventricle palliative approach or a more desirable two ventricle repair with a neonatal TV valvuloplasty. In this case series, we present 3 (three) neonates with severe EA in cardiogenic shock, dependent on prostaglandin and mechanical ventilation after birth who underwent Starnes procedure in the neonatal period as palliative solution prior to an early two ventricle repair using Cone procedure.

## 2. Introduction

Symptomatic Ebstein's Anomaly (EA) in the neonatal period remains one of the most challenging diseases in pediatric cardiology despite the improvement in surgical techniques developed in recent years [1]. Early surgical intervention in neonates with EA and hemodynamic instability consists of primary repair or palliative treatment (univentricular procedure). The decision regarding biventricular or univentricular treatment depends on the function of the right ventricle (RV), the amount of tricuspid valve tissue and the presence of anatomical or functional pulmonary atresia. Univentricular treatment has shown good early survival and encouraging medium and long-term results, however patients remain subject to univentricular physiology complications [2]. Primary two ventricle repair in the neonatal period or in early childhood has shown variable results, with high mortality, especially in neonates with severe EA and pulmonary atresia [3]. We propose the use of Starnes procedure as palliative solution during neonatal period for those extremally seek patients intended to provide an early two ventricle circulation with the cone procedure during the first six months of life.

## 3. Case Report

This is a case series of our first 3 (three) patients with extremely severe EA with hemodynamic instability in the neonatal period. All of them prostaglandin-dependent, using two or more vasoactive drugs and on mechanical ventilation. In one patient, functional atresia of the pulmonary valve was observed, while other two was anatomical atresia of the same valve. The Celemarjer score was 3 (three) in two patients and 4 (four) in one. Demographic data is present in Table 1. All patients had a greatly enlarged cardiac area with varying degrees of pulmonary hypoplasia (Figure 1). The prenatal diagnosis of Ebstein's anomaly was confirmed through transthoracic echocardiography after birth (Figure 2). In all patients, a palliative procedure was performed using the Starnes procedure, modified for two ventricle correction in the future, which consisted of right ventricle exclusion through a polytetrafluoroethylene (PTFE: 0,5mm) fenestrated (5mm) patch sutured above the tricuspid ring, associated with atrioseptectomy, right atrial reduction and a systemic-to-pulmonary shunt (we performed central shunts with PTFE grafts (3,5mm) from the ascending aorta to the pulmonary trunk, in a horseshoe shape [5]. In all patients, chest was kept open after the surgical procedure and closure was performed after clinical stabilization. The mean time between surgery and chest closure was 72 hours (ranging from 48 to 96 hours). The hemodynamic

improvement after the Starnes procedure was accompanied by a significant decrease in the cardiac area in all patients (Figure 3). This tree patients were discharged from hospital and followed up weekly on an outpatient basis until the second surgery. Two ventricle correction, using the cone technique described by Da Silva, was planned to be performed during the first six months of life [4]. The decision to go directly to two ventricle repair is made for all patients, and during the weaning on CPB the hemodynamic status and the arterial saturation must be adequate. If there is some kind of hemodynamic impairment a bidirectional cavo-pulmonary anastomosis should be done promptly. In one patient, correction was performed at the age of 3 months and 6,2kg, with the Cone technique and enlargement of the right ventricular outflow tract using bovine pericardium monocusp patch. In the second patient, at 4 months of life and 6,8kg, tricuspid valve repair was performed with expansion of the septal leaflet using autologous pericardium patch, treated in glutaraldehyde, and enlargement of the right ventricular outflow tract with PTFE patch (0,5mm thick). A third patient was operated with 6 months and 8,2kg, the Cone procedure was done with enlargement of the septal leaflet with autologous pericardium (Figure 4), associated with pulmonary valve commissurotomy. Postoperative echocardiographic control in patients 1,2 and 3 showed no significant gradient across the tricuspid valve, with mild regurgitation and good anterograde flow through the pulmonary valve (Figure 5), patients 1 and 2 shows moderate to important pulmonary regurgitation as expected. In all cases, the interatrial septum was reconstructed with a fenestrated (4-5 mm) PTFE patch (0.5 mm thick).

The echocardiographic assessment 3 and 6 months after the Cone procedure showed good biventricular function, absence of gradient across the tricuspid valve and mild reflux in all patients. The last follow up is presented in Table 2. The current oxygen saturation is above 90% on all patients, and functional class I (NYHA), with normal growth and development.

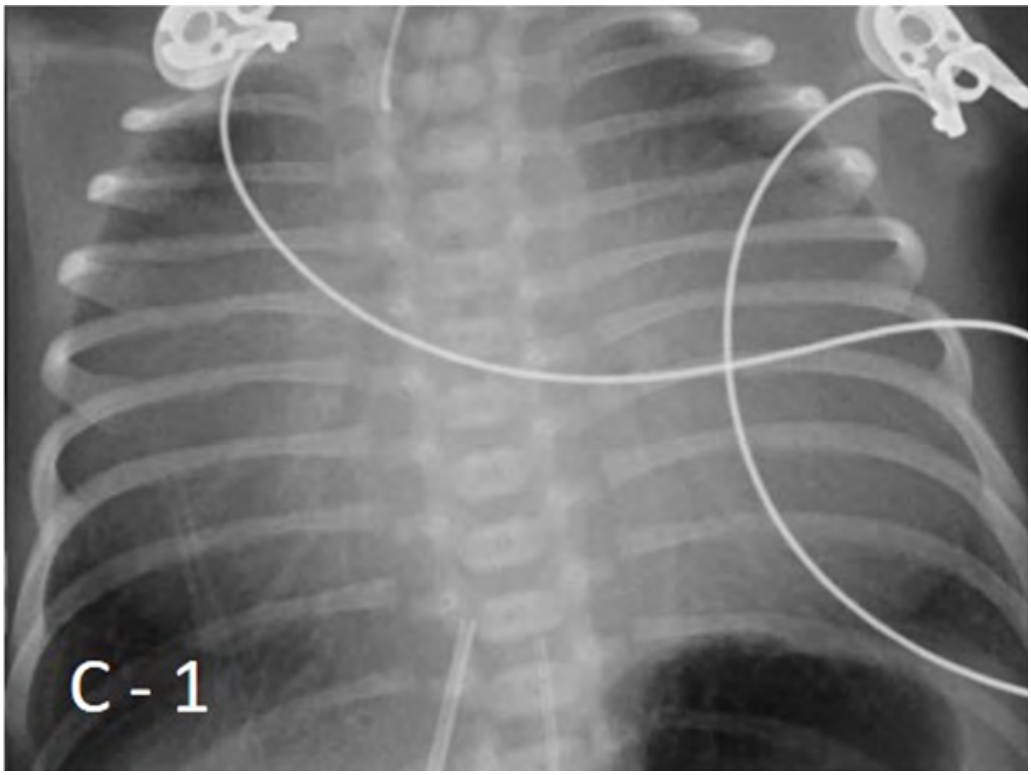


**Figure 1:** Preoperative x-ray showing the cardiac area.

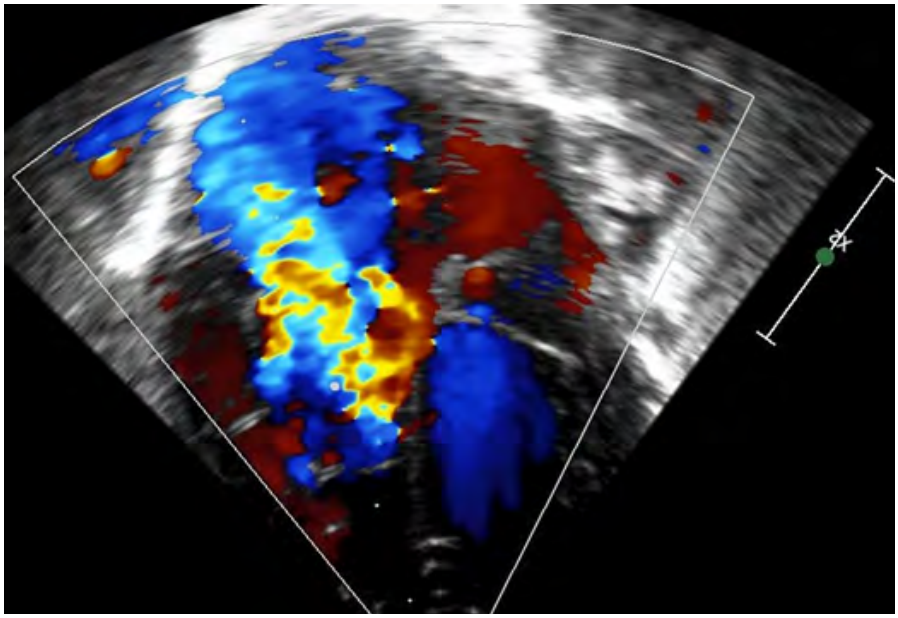

**Figure 2:** Preoperative echocardiography shows the amount of tricuspid regurgitation.

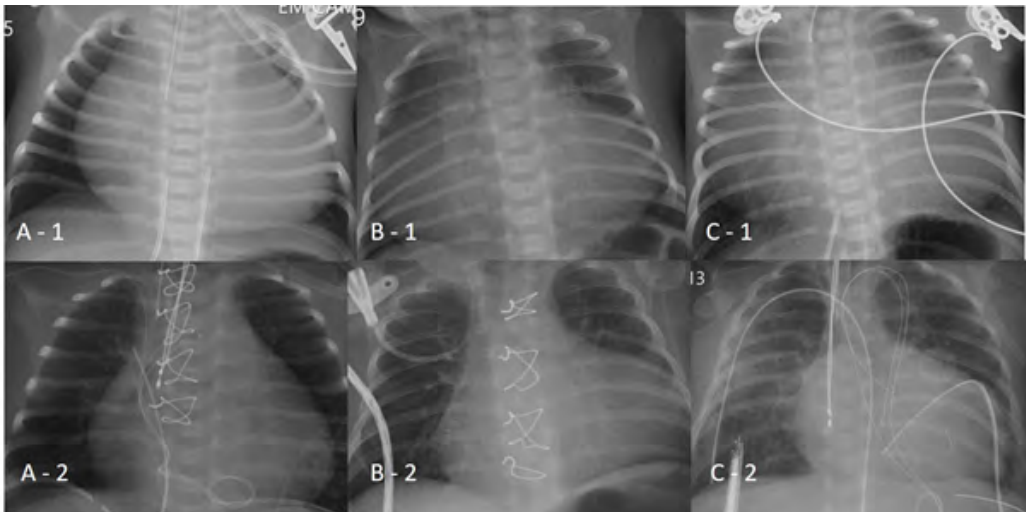


**Figure 3:** Post-operative xRay shows the reduction of the cardiac area after Starnes procedure in 3 patients.

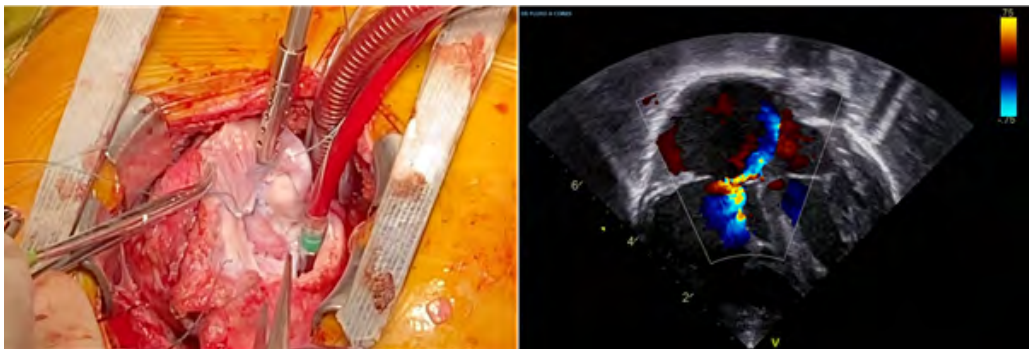

**Figure 4:** Rigth: Intraoperative aspect of the da Silva cone procedure with enlargement of septal leaflet with autologous pericardium. Left: Echocardiographic Doppler of the tricuspid valve after surgery.

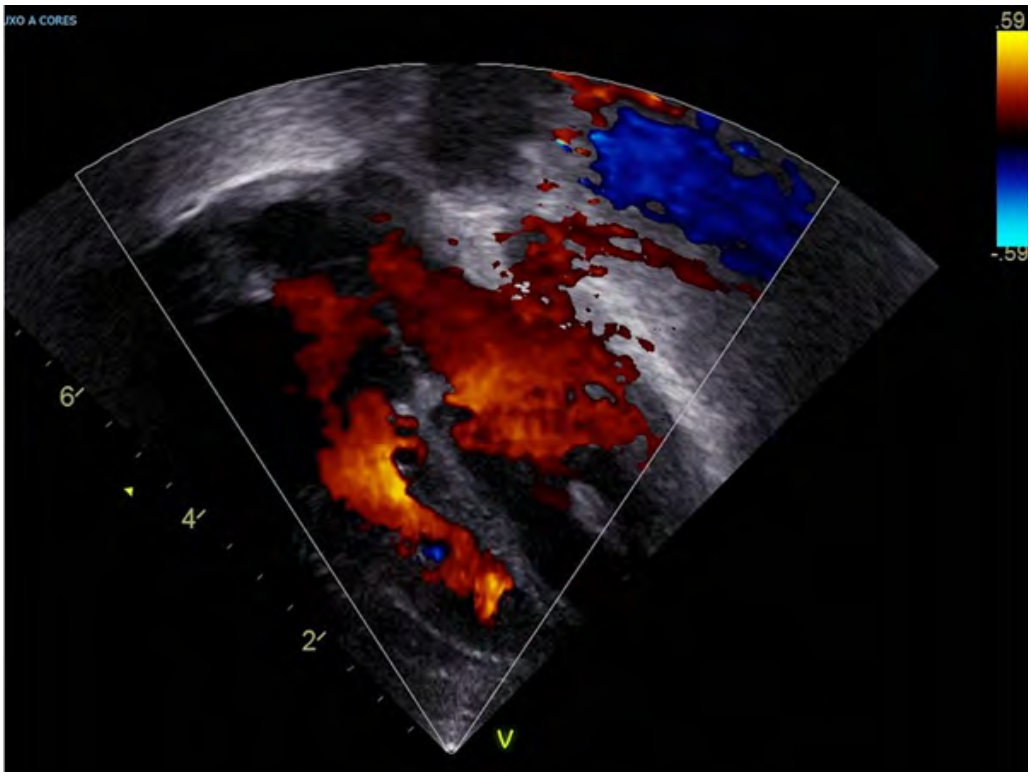


**Figure 5:** Post-operative echocardiography

## 4. Discussion

Ebstein's anomaly has a wide spectrum of anatomical and clinical presentation, from very mild cases to cases in which symptoms start in the intrauterine period and may even progress to fetal death. In cases with severe impairment at birth, extreme cardiomegaly with pulmonary hypoplasia, pulmonary blood flow may be compromised in the neonatal period. Many patients have anatomical or functional pulmonary atresia, requiring prostaglandins to maintain ductus arteriosus patency. In this scenario, early surgical treatment is essential, however, is accompanied by high morbidity and mortality when reconstruction of the valve is tried [6]. Currently, the standard treatment for Ebstein's anomaly with neonatal clinical repercussions is the technique developed by Starnes. However, most of the times this technique is used as a bridge for univentricular repair, with patients who survive the postoperative period undergoing the Glenn and Fontan procedure later. The Cone technique after palliation with the Starnes procedure was described by Da Silva and seems to be a great strategy for this group of patients [7]. The idea of an early two ventricle repair is that the Cone procedure is performed around 4 - 6 months of age, which allows for a technically more feasible and lasting valve repair. This early repair without a Glenn anastomosis is more physiologic and as far as we know was not tried. Another possible advantage of this technique would be the improvement in right ventricular function during the first months after Starnes procedure, which has been demonstrated in the literature and can be observed in our series [8]. There is a concern about the size and the function of the right ventricle in this patient to go further with two ventricle correction without a Glenn anastomosis. Nonetheless we show that even in these very complex cases of neonatal Ebstein was possible to go through two ventricle circulation. After a mean follow up of 9 months the right ventricle is presenting with good function and adequate size in all three patients. It's necessary to increase the number of patients and a long term follow up to ensure that these patients will continues doing well, but the possibility to treat these very seek patients with two ventricle circulation already drive us to a direction that is bright them univentricular outcomes.

## 5. Conclusion

Two ventricle repair is possible even in very symptomatic patients with neonatal Ebstein's anomaly. The initial correction with the Starnes procedure for clinical stabilization, maintenance of pulmonary flow and reduction of the cardiac area must be followed by correction with the Cone technique. In this series all three patients were treated with preservation of two ventricle circulation, what is a huge progress regarding the concerns that are involved in univentricular and 1 and ½ circulation.

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