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Biventricular Repair in Very Critical Neonatal Ebstein's Anomaly using Da Silva's Cone Procedure after Modified Starnes Palliation

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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 biventricular repair with a neonatal TV valvuloplasty. In this case series, we present 5 (five) 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 biventricular 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 biventricular 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]. Biventricular correction 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].

3. Case Report

This is a case series of 5 (five) patients with extremely severe EA with hemodynamic repercussions in the neonatal period. All of them prostaglandin-dependent, using two or more vasoactive drugs and mechanical ventilation. In two patients, functional atresia of the pulmonary valve was observed, while in the others there was anatomical atresia of the same valve. Mean birth weight was 2,101g, ranging from 1,760g to 2,850g, with initial postnatal oxygen saturation between 40% and 75% under mechanical ventilation. 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 biventricular correction in the future, which consisted of right ventricle exclusion through a polytetrafluoroethylene (PTFE) 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 from the ascending aorta to the pulmonary trunk, in a horseshoe shape) [5]. The pulmonary artery was preserved in all five patients, but one patient developed circular shunt after palliation and the pulmonary trunk was ligated at

this moment. 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 average age at the time of surgery was 14 days of life; in 1 patient whose birth weight was 1,760g, we chose to wait until a weight of 2,500g was reached and surgery was performed on the 44th day of life. The average weight at surgery was 2,625g, ranging between 2,540g and 3,120g. In 1 patient, there was complete atrioventricular block, requiring permanent pacemaker implantation that was accomplished on the 7th postoperative day.

The hemodynamic improvement after the Starnes procedure was accompanied by a significant decrease in the cardiac area in all patients (Figure 3). There was two hospital death, which occurred in the first patient with 48 days after the Starnes procedure. This death was due to a septic condition triggered by broncho aspiration. Another patient died 24 days after surgery due septic shock and multiple organ disfunction.

The three patients discharged from hospital were followed up weekly on an outpatient basis until the second stage. Biventricular correction, using the cone technique described by Da Silva, was planned to be performed in the first six months of life [4]. So far, three patients who were discharged have already undergone biventricular repair.

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 (pulmonary atresia). In the second patient, at 4 months of life and 6,8kg, tricuspid valve repair (Cone technique) 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. 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 of both patients showed no significant gradient across the tricuspid valve, with mild regurgitation and good anterograde flow through the pulmonary valve (Figure 5). In both cases, the interatrial septum was reconstructed with a fenestrated (4-5 mm) PTFE patch (0.5 mm thick). The current oxygen saturation of patients who have already undergone biventricular correction is currently 86-94%, and these patients are in functional class I, with normal growth and development.

The echocardiographic assessment 3, 6 months and one year after the Cone procedure showed good biventricular function, absence of gradient across the tricuspid valve and mild reflux in patients (Table 1 and 2).

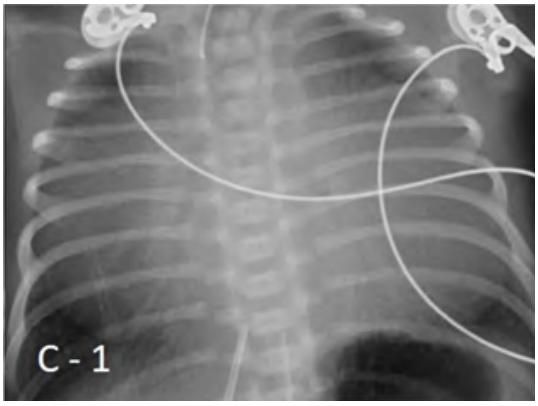

Figure 1: Preoperative xRay showing the cardiac area

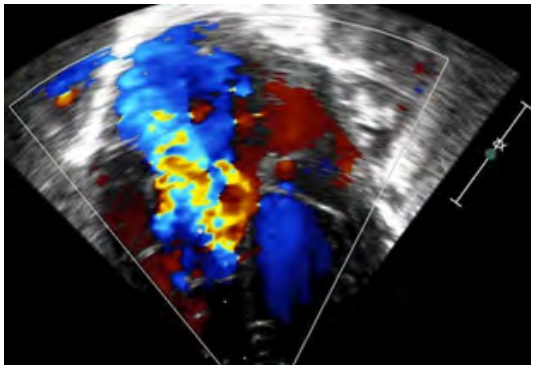

Figure 2: Preoperative echocardiography shows the amount of tricuspid regurgitation

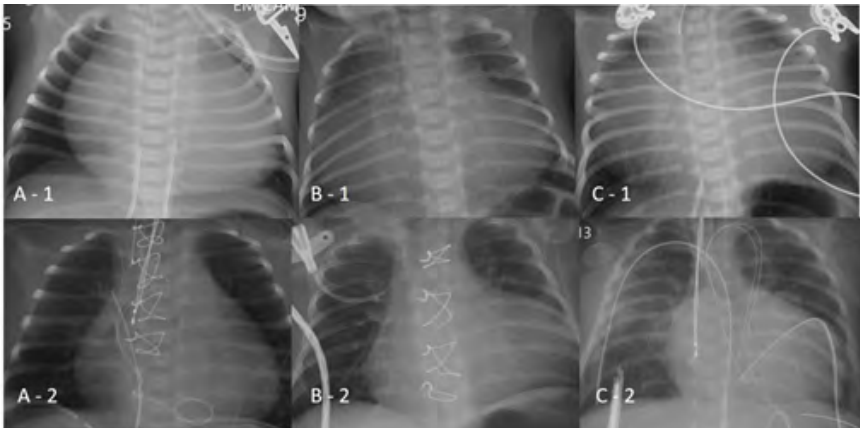

Figure 3: Post-operative xRay shows the reduction of the cardiac area

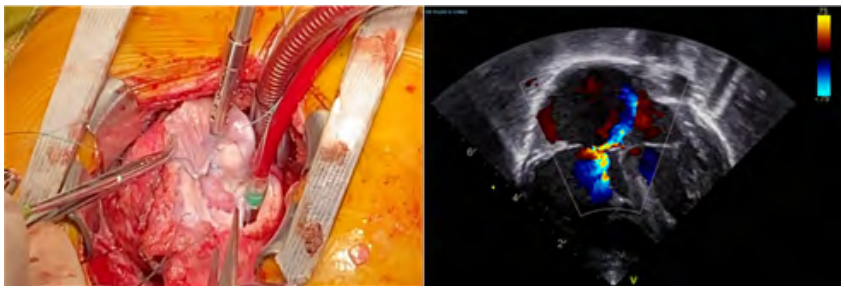

Figure 4: Intraoperative aspect of the da Silva cone procedure with enlargment of septal leaflet with autologous pericardium

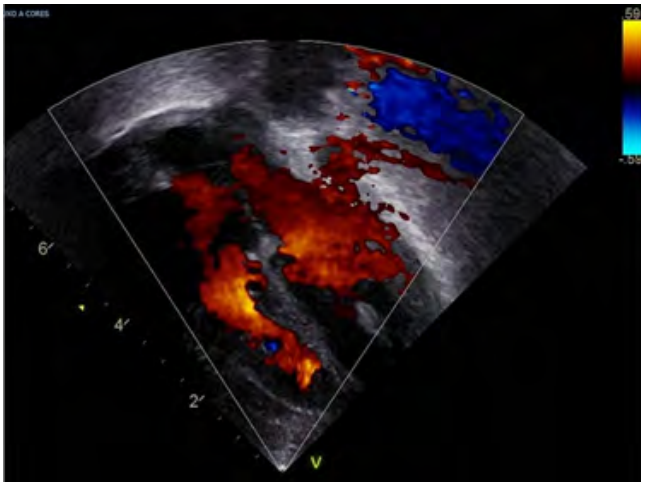

Figure 5: Post-operative echocardiography during diastole shows no gradient through tricuspid valve

Table 1:

Patient	Birth date	Weigth Birth(g)	Gestacional Age (weeks)	Cardio-Toracic Index	Celemanjer Grade	Starnes Age(days)	Starnes Weigth	Saturation after Starnes	Cone procedure Age (months)	Cone Weigth
1	6/4/2018	1.76	36	0.85	4	44	2.54	79%	-	-
2	01/22/2018	2.55	37	0.82	3	5	2.509	82%	-	-
3	10/18/2020	2.02	35	0.78	3	8	2.237	80%	6	4.78
4	6/3/2021	2.64	38	0.86	4	6	2.43	84%	3	5.37
5	06/22/2023	3.63	39	0.81	3	5	3.12	83%	4	4.59

Table 2:

Patient	Birth date	Last Follow up	Weigth (g)	NYHA	Tricuspid Valve Regurgitation	Function RV	RV dilatation	Pulmonary valve Regurgitation	Saturation O2(%)
1	6/4/2018	-	-	-	-	-	-	-	-
2	01/22/2018	-	-	-	-	-	-	-	-
3	10/18/2020	1/3/2024	13.83	I	Mild	Normal	None	Important	94%
4	6/3/2021	02/20/2024	11.72	I	Mild/moderate	Normal	Mild	Important	92%
5	06/22/2023	7/3/2024	8.5	I	Mild	Normal	Mild	Trivial	96%

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 [6].

Currently, the standard treatment for Ebstein's anomaly with neo-

natal 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 is that the Cone procedure is performed around 4 - 6 months of age, which allows for a technically more feasible and lasting valve repair. 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].

5. Conclusion

Biventricular 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 that survived until the cone procedure were treated with preservation of biventricular circulation, what is a huge progress regarding the concerns that are involved in univentricular and 1 and ½ circulation.

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