CASE REPORT

Surgical treatment option in a patient with neonatal Ebstein's disease. Case report

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Ebstein's anomaly is a rare congenital heart disease characterized by displacement of the tricuspid valve insertion and impairment of right ventricular function. Its presentation in the neonatal stage may require surgical intervention, as it is a severe form of presentation. The surgical procedure is based on the analysis of the anatomical characteristics of each patient. A neonatal case is reported herein in which the Starnes procedure was successfully performed.

Key words: Ebstein's anomaly; Neonate; Starnes surgery.

La anomalía de Ebstein es una cardiopatía congénita poco frecuente caracterizada por el desplazamiento de la inserción de la válvula tricúspide y afectación del funcionamiento del ventrículo derecho. Su presentación en la etapa neonatal llega a requerir intervención quirúrgica, pues es una forma de presentación grave. El procedimiento quirúrgico es basado en un análisis de las características anatómicas de cada paciente. Se reporta un caso neonatal que se realizó procedimiento de Starnes con éxito.

Palabras clave*: *Anomalía de Ebsten; Neonatos; Cirugía de Starnes.

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Ebstein's anomaly accounts for 1% of all congenital heart diseases. It is a right ventricular myopathy with tricuspid valve implantation failure and highly variable tricuspid valve morphology usually resulting in severe failure. It is the only congenital heart lesion that has a range of clinical presentations, from the severely symptomatic newborn to an asymptomatic adult.

It is known to be severe depending on the age of presentation especially in the neonatal period, with mortality of approximately 50% of cases without treatment. Surgical treatment options are the Starnes procedure, Knott-Craig repair, Da Silva and Carpentier cone surgery. Overall survival estimated for all surgical intervention patients was 66.7% at 1 year, 62.2% at 5 and 10 years, and 51.9% at 15 years [1]. The main causes of death were heart failure, perioperative death and sudden death. The main predictors of death were degree of severity assessed by echocardiogram, fetal presentation and right ventricular outflow tract obstruction [2].

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CLINICAL CASE

This is a male patient with a maternal history of gestational diabetes and hypothyroidism at 24 gestation weeks, and medical treatment was started. At 26.3 gestation weeks, a fetal echocardiogram was performed and congenital heart disease was found. The echocardiographic report showed a heart in situs solitus, CTI of 0.72, significant dilatation of the right atrium and normal ventricles. Loss of valvular offset at the expense of the tricuspid valve with septal and posterior leaflets inserted 12.5 mm apical to the mitral valve, maximum regurgitant jet velocity at the tricuspid valve of 260 cm/sec, so a diagnosis of Ebstein's anomaly, patent foramen ovale and ventricular arterial concordance, pulmonary valve with antegrade flow without significant insufficiency.

The baby was presented at the medical-surgical session and birth was planned at our institution for diagnosis and immediate comprehensive management. A single live product was obtained, male at term, 37.1 gestation weeks, abdominal route, cried and breathed at birth, APGAR 8/9, SA 1, capurro 37.1 gestation weeks, weight 3100 gr, size 51cm. Basic resuscitation maneuvers were required.

On physical examination she presented with vital signs: HR 140 bpm, RR 63 per min, BP 70/43 (55) mmHg, SaO 85%. At birth, prostaglandins were started at 0.03mcg/kg /

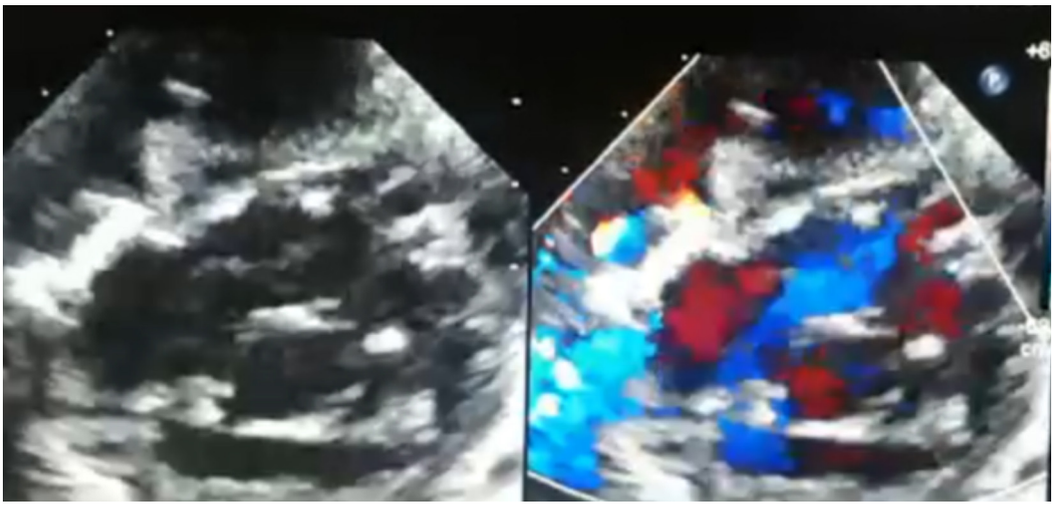

Figure 1. Preoperative echocardiogram showing tricuspid valve with 33% attachment of the septal leaflet to the interventricular septum.

min and sildenafil 1mg/kg. With saturations between 75-88%, however, he presented desaturation events of up to 60%; thus, it was decided to start conventional mechanical ventilation. Electrocardiogram study showed HR 150, sinus rhythm, P axis +60º, QRS axis +120º, PR 100 ms, QRS 80 ms, QT 280 ms, QTc 443 ms, right atrial enlargement, Crochetage sign suggestive of atrial septal defect. Chest X-ray with situs solitus, mesocardia, levoapex, CTI 0.88, right atrial and ventricular enlargement, decreased pulmonary flow. Echocardiogram at birth reported VA connection, single outflow tract due to pulmonary valvular atresia, interatrial septum with 9mm bidirectional CC fenestrated ASD , TAPSE 15mm, ARVD 29%, tricuspid valve with 33% attachment percentage of septal leaflet to ventricular septum, moderate tricuspid insufficiency with gradient 45mmHg, ventricular septum intact, LVEF 61%, pulmonary artery valvular plane 5.5mm (Z score - 2.03) Truncus 6.6mm (z score -1.2) pulmonary branches: Left branch 3.6mm (Z score - 1.07) Right branch 2.9mm (Z score - 2.83) (**Fig 1**).

He was admitted to the operating theatre at five days of life undergoing RV exclusion with placement of a 4 mm fenestrated Gore-tex 0.6 membrane at the level of the tricuspid annulus (**Fig. 2**) and a central aorto-pulmonary end-to-side fistula due to hypoplasia of the pulmonary arteries (**Fig. 3**), in addition to right atrial reduction plasty.

During his evolution, the patient presented arrhythmias compatible with atrial flutter, for which he was started on amiodarone, which he remains on until now. He also presented data on sepsis associated with health care, which responded to broad spectrum antimicrobial management. Patient is currently being treated with inodilators, anticoagulants and diuretics at conventional doses.

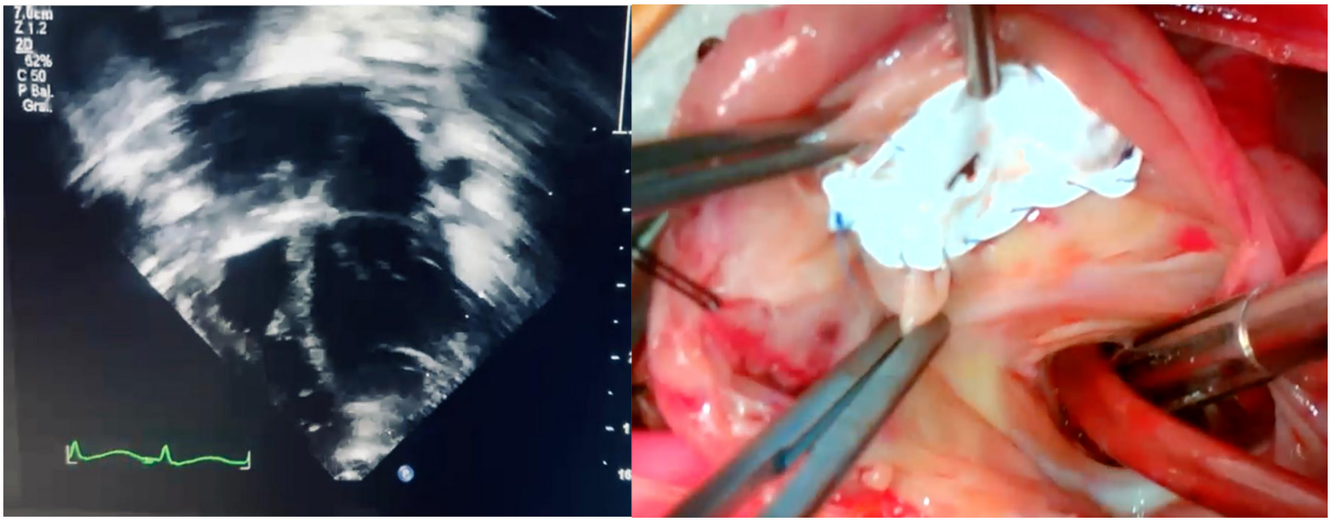

Figure 2. Starnes Surgery: Gore-tex membrane is placed at the anatomical level of the tricuspid valve annulus with a 4-mm fenestration.

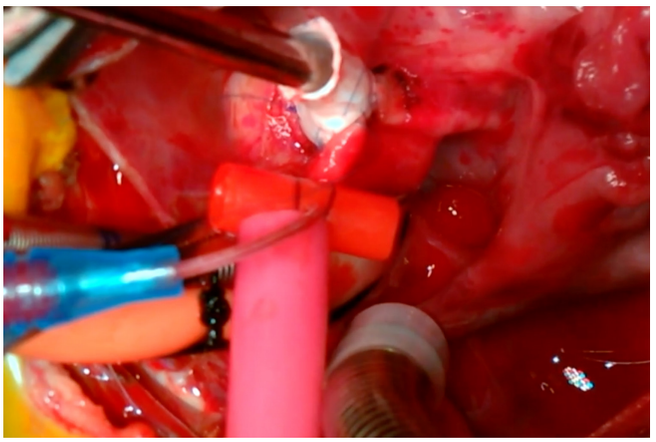

Figure 3. Starnes Surgery: Central aortic-pulmonary end-to-side fistula due to hypoplasia of the pulmonary arteries

COMMENT

Ebstein's anomaly is a heart disease that requires timely diagnosis in order to be able to treat patients appropriately and avoid complications related to early mortality caused by pulmonary hypertension and severe heart failure. Prenatal diagnosis currently makes a difference, as in the case presented here.

Treatment is initiated at diagnosis, and according to the clinical picture of each patient; it can range from being asymptomatic, requiring medical treatment as a complex heart disease, to management in intensive care and surgery in the first days of life [1].

Neonatal Ebstein's disease is an entity that requires a different treatment approach. In severe patients, the goal of treatment is to counteract cardiomegaly and decrease pulmonary vascular resistances as well as increase pulmonary flow. Routine echocardiographic evaluation and dynamic assessment for prostaglandin weaning, ventilation and tolerance to full enteral feeding should be performed [3,4].

Surgical intervention in the neonatal period should be considered as the first option in patients with severe Ebstein's anomaly. However, standardized surgical options and perioperative management protocols have not been established, particularly in this age group. Different surgical options and techniques have been described, including the one described by Knott-Craig et al. [3], which consists of creating a competent monocuspid valve from the anterior leaflet, closure of fenestrated atrial septal defect, reduction atrioplasty, with a survival rate of 80%. However, Muzino et al. describe the performance of cone surgery of first intention; however, the sample in their study consisted of only 10 patients with a mortality rate of 50%, so we do not consider it as an option [5]. The surgical procedure of choice for our patient was to perform Starnes surgery, since he had a diagnosis of anatomical pulmonary atresia associated with severe tricuspid insufficiency. The aim is to exclude and decompress the affected right ventricle; ensure a wide atrial septal defect; perform a reduction atrioplasty; and provide adequate pulmonary flow through a systemic pulmonary shunt [6].

Our proposal management in accordance with the Sushee 2020 and Knott-Craig 2021 algorithms will be to perform an early cavo-pulmonary superior shunt, this does not contraindicate assessing before the third stage (Fontan) whether the patient may be a candidate for correction one and a half, removing the fenestrated patch and, as reported by José Pedro Da silva, performing cone surgery, with pulmonary valve plasty to bring it to physiology with two functional ventricles. [7].

In summary, in the previous decade, survival for the neonatal patient diagnosed with severe Ebstein's anomaly was low. Lack of understanding of this pathology (morphology and pathophysiology) influenced this outcome, as did unclear surgical treatment options. The anatomy of the tricuspid valve has a high range of variation ranging from minimal changes to being attached to an atrialized right ventricular chamber.

In the neonatal stage, management remains complex. Neonates may have a rapid progressive deterioration with severe heart failure, cyanosis and acidosis. Without surgical treatment most die. The surgical goal should be palliation for optimal survival, which can be achieved with valve repair, RV exclusion or cardiac transplantation.

The Starnes procedure is mostly used in cases with severe spectrum of Ebstein's anomaly, where there is a small RV, a very tightly attached valve with difficult delamination and an obstructed RVOT. This results in significant RV dilatation and its impact on the LV. Therefore, the goal is to decompress the RV and allow the LV to function more effectively [6].

The surgical procedure aims to increase survival, the intervention options are based on the manifestations, characteristics of each patient and the experience of the cardiovascular group. The different surgical techniques written by the authors have been developing, giving favorable results. However, more complex techniques such as Starnes [6] and cone surgery allow a more physiological evolution. Da Saliva and co-workers show encouraging results in cone reconstruction surgery after Starnes and it may become the intervention of choice [7].

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