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Cone reconstruction after carpentier repair in ebstein anomaly: yes we can!

Alessandro Giamberti[1*], Federica Torchio[1], Massimo Chessa[2,4], Stefan Veljkovic[3], Ana Perunicic[3] and Aleksandra Nikolic[3]

Abstract
The management of recurrent tricuspid regurgitation (TR) after initial tricuspid valve (TV) repair in patients with Ebstein's anomaly (EA) represents a complex challenge. Traditionally, tricuspid valve replacement has been the first-line option for these patients. The use of Cone Repair for recurrent TR remains under-reported. Herein, we report a successful cone repair for recurrent TR, twelve years after a Carpentier repair in a 39-year-old male patient.

Keywords Adult with congenital heart disease, Ebstein's anomaly, Cone repair, Cardiac surgery

Introduction

Ebstein's Anomaly (EA) is a rare congenital heart disorder occurring in approximately 1 per 200. 000 live births and accounting for less than 1% of all cases of congenital heart disease [1]. EA is a congenital valvular and ventricular dysplasia of the right-sided heart presenting a wide spectrum of severity of the anatomical abnormality [2].

Different surgical techniques have been proposed in the attempt to repair the EA since the first surgical case report published in 1958 [3]. For many years the monocusp reconstruction techniques (Danielson or Carpentier) [4, 5] were the most widely used. There is now a consensus in preferring a more anatomical repair technique introduced by Jose da Silva at the end of 80s and published in 2007 [6], the Cone Repair. Compared to previous repair techniques, the Cone Repair showed better results and several advantages [7, 8]. In few selected cases the Cone Repair has been reported after tricuspid valve replacement or Starnes Procedure or other repairs [9–11]. We report here the case of an EA adult patient who successfully underwent the cone repair twelve years after a Carpentier repair.

Case report

A 39-year-old male patient presented to our department with severe tricuspid regurgitation. Twelve years prior, he underwent a Carpentier repair for EA. The patient had a childhood diagnosis of Wolff-Parkinson-White syndrome requiring multiple transcatheter radiofrequency ablations before and after the previous surgery.

The patient arrived in NYHA functional class II-III in sinus rhythm. Auscultation revealed a grade 4/6 precordial systolic murmur with palpable tremor over the xiphoid process. Chest X-ray showed an increased cardiothoracic index (70%). Trans-thoracic echocardiography depicted a severely enlarged right atrium, severe tricuspid regurgitation with the apical displacement of the septal leaflet (20 mm/m2) and severe annular dilation (54 mm) (Fig. 1). The right ventricle was dilated with moderate systolic dysfunction. The interatrial septum

*Correspondence:
Alessandro Giamberti
alessandro.giamberti@grupposandonato.it
[1]Department of Congenital Cardiac Surgery, IRCCS Policlinico San Donato, Via Morandi 30, San Donato M.se (Mi) 20097, Italy
[2]ACHD Unit - Pediatric and Adult Congenital Heart Centre, IRCCS Policlinico San Donato, San Donato M.se, (Mi), Italy
[3]Dedinje Institute for Cardiovascula Disease, ACHD Unit, Belgrade, Serbia
[4]Faculty of Medicine and Surgery, Vita - San Raffaele University, Milano, Italy

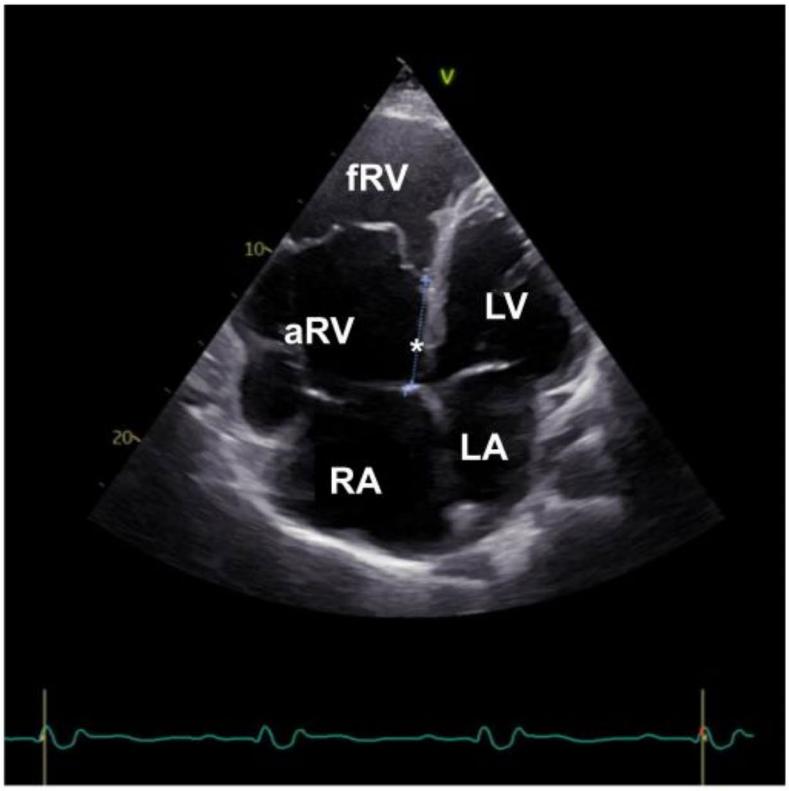

Fig. 1 Pre-operative apical 4-chamber echocardiography showing typical features of EA with apical displacement (46 mm) of the septal leaflet. RA, right atrium; aRV, atrialised right ventricle; fRV, functional right ventricle; LA, left atrium; LV, left ventricle; * septal leaflet

appeared intact and left ventricular systolic function was within normal limits.

A cardiac magnetic resonance (CMR) revealed a similarly enlarged right atrium and severe tricuspid valve regurgitation. We were also able to appreciate his native TV leaflets (Fig. 2).

Given these findings, the patient was scheduled for a possible re-repair procedure employing the Cone Repair.

Surgical technique

After secondary sternotomy and lysis of adhesions, a cardio-pulmonary by-pass (CPB) with standard aortic and bi-caval cannulation and left vent was started. After aortic cross-clamping and Del Nido cardioplegic cardiac arrest, a transverse incision of the right atrium was performed in the direction of the cavo-tricuspid isthmus. The tricuspid annulus appeared dilated with a diameter greater than 50 mm. A rigid ring had not been used in the previous operation. The posterior and the septal leaflets were displaced to the ventricular apex, and they had not been touched.

The anterior leaflet appeared wide with residual focal failing of delaminations to the free wall of the ventricle. The clockwise rotation of the anterior flap (typical of the Carpentier repair technique) was reduced probably due to the progressive dilation of the right ventricle (RV) and the tricuspid annulus.

We performed a CR including complete mobilization and detachment of tricuspid leaflets including the septal leaflet. Once the leaflets were detached and completely delaminated from the right ventricular wall, they were sutured together creating a cone the vertex of which remains fixed at the RV apex. The true tricuspid annulus was plicated to match the proximal circumference of the already-created cone-shaped valve. The zone between the septal and the posterior leaflet of the cone was then enlarged using a piece of heterologous pericardial patch (Fig. 3). At this point the new valve, generally slightly rotated clockwise, was sutured to the new annulus paying extreme attention to shift the suture line re-attachment for the septal leaflet slightly lower in the ventricular septum avoiding injury of the conduction tissue.

The valve was tested with saline injection in the RV, and finally a flexible annuloplasty ring (Medtronic Contour 3D, 34 mm) was placed to stabilize a competent repair.

The weaning from CPB was easy and an intra-operative transesophageal echocardiography showed a very good result of the repair with trivial residual TR and no gradient across the Cone.

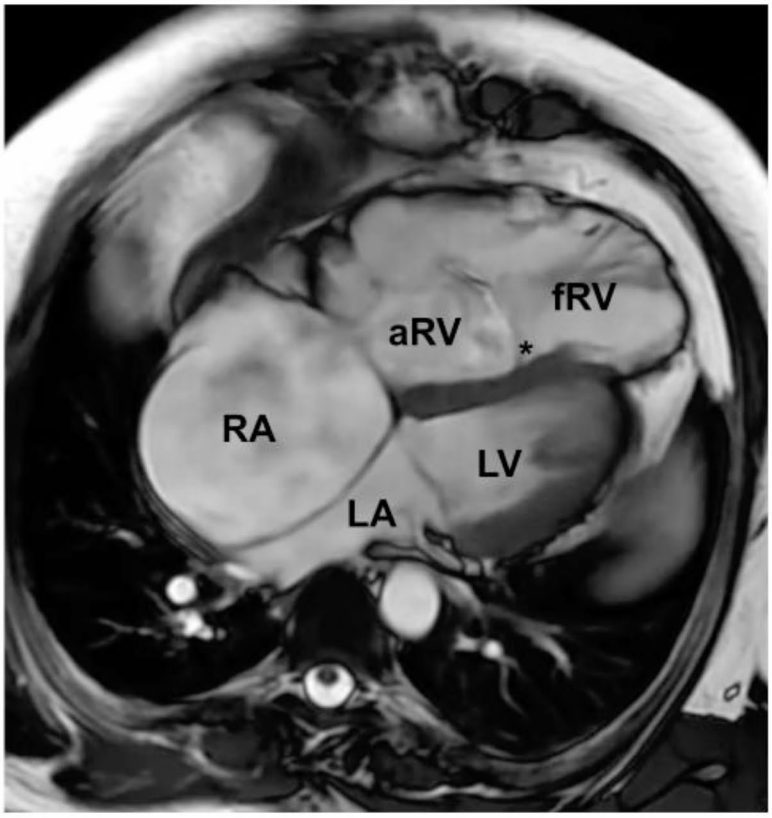

Fig. 2 Magnetic resonance cine-SSFP images in long axis. Special focus on the tricuspid valve and apical displacement of the septal leaflet. RA, right atrium; aRV, atrialised right ventricle; fRV, functional right ventricle; LA, left atrium; LV, left ventricle; * septal leaflet

The postoperative course was uneventful. The patient was extubated the following day and discharged home on postoperative day 10.

At the 6-month follow-up, the patient is NYHA Class I. We observed trivial regurgitation, with a right atrial area of about 25 cm^2, an end-diastolic right ventricular diameter of about 44 mm and conserved left ventricular ejection fraction. Pre-operative medications were gradually tapered and then discontinued.

According to our protocol, the patient will undergo a CMR 12 months after surgery.

Discussion

There is a general consensus considering the Cone Repair superior when compared to previous repair technique in EA in term of residual TR, freedom from re-operation, and life expectancy [7, 8]. Advantages of the Cone Repair include growth potential and durability of the native tissue, no need for long-term anticoagulation therapy, and a more physiological TV and RV geometry [2, 7, 8].

In all previous repairs the displaced septal leaflet was ignored with the consequence of very often moderate or more TV regurgitation from the septal or antero-septal commissure with a tendency to increase in the follow-up [9]. Our patient is an example with a reparative technique that highlighted a residual TR that increased over the years until it became severe.

In our patient the decision to re-repair the TV instead of a prosthetic replacement was based on feasibility because the septal leaflet had not been touched, the anterior and posterior leaflets were still usable, and the annulus was dilated without the presence of a previous rigid ring. The use of a pericardial patch also contributed to stabilizing the cone, as previously demonstrated by Belli [12].

In patients with EA in natural history or after repair procedures with severe TR older than age 50 to 55 years, the surgical choice is often to replace the valve with bioprosthetic rather than mechanical valve related to their acceptable durability, no need for anticoagulants, and the possibility of subsequent transcatheter "valve-in-valve" procedure.

TV replacement when compared to CR was associated with a significant higher risk of TV reoperation and TV stenosis, and worse RV function [8].

We believe that regardless of the patient's age or surgical history, every effort must be made to perform a repair technique with a good result. The literature of

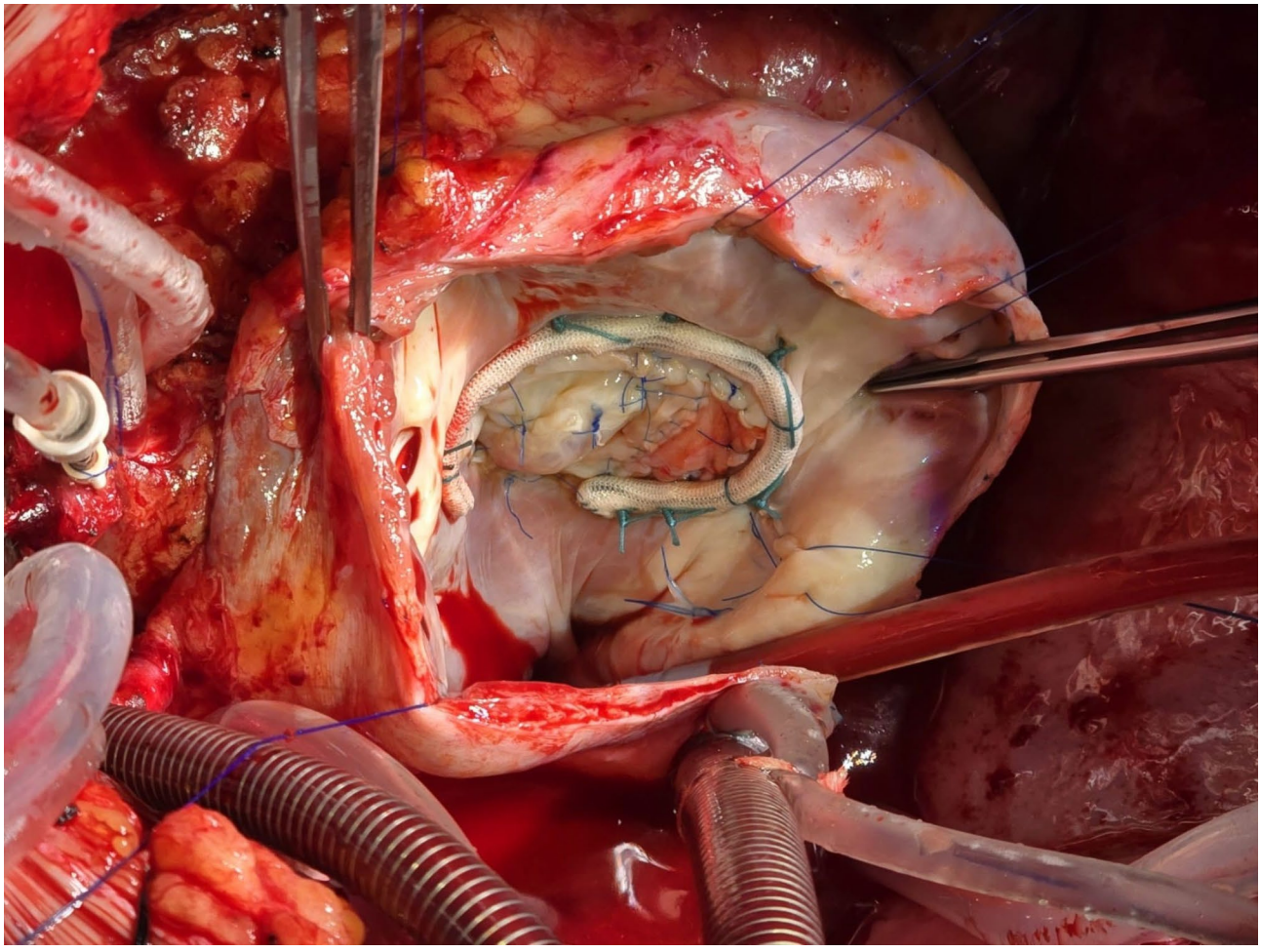

Fig. 3 Intra-operative view showing the reattachment of the cone and the leaflet augmentation with heterologous pericardial patch

recent years has demonstrated that CR is the operation of choice, and our case demonstrates how this technique is a feasible approach in reoperations for EA.

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Author contributions
AG: chief surgeon and conceived the manuscript. FT, MC: have made substantial contributions to conception and design of the manuscript. SV, AP, AN: helped to draft the manuscript. AG, FT, AN: reviewed the manuscript. AG: gave final approval of the version to be published. All authors read and approved the final version of the manuscript.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Conflict of interest
The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Ethics approval and consent to participate
Not applicable.

Competing interests
The authors declare no competing interests.

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and accompanying images. All the authors agree to the publication of the article.

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