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The modified cone reconstruction in the treatment of Ebstein's anomaly

Case reports

Jun-Sheng Li, MD[a], Gang Chen, MD[b], Jie Ma, MD[c,*], Zi-Xing Yan, MD[c], Dong-Ming Cheng, MD[c], Liang Chang, MD[c], Hai-chun Zhang, MD[c], Jiang-Yan Liu, MD[d]

Abstract

Rationale: To investigate the early and mid-term clinical outcomes of the modified cone reconstruction in the treatment of Ebstein's anomaly (EA) which provide appropriate surgical treatment for clinical and reduce the incidence of re-operation and valve replacement.

Patient concerns: Clinical data of 18 consecutive patients with EA in our hospital between May 2008 and August 2015 were analyzed retrospectively. All patients were diagnosed by echocardiography. Among these patients, according to New York Heart Association functional grade, there were 12 patients with grade II cardiac function and 6 patients with grade III. All patients had severe tricuspid regurgitation grade.

Diagnoses: All patients were diagnosed EA. One case was with acute arterial embolism and amputation of left lower extremity caused by paradoxical embolism of combined secundum atrial septal defect.

Interventions: The modified cone reconstruction in the treatment of EA of the tricuspid valve uses its own tissues to form not only central bloodstream, but also the coaption between 2 leaflets. For those patients whose anterior leaflet developed poor and smaller, the valve leaflet was widened by using autologous pericardial. For all patients, tricuspid annulus were reinforced by autologous pericardial. One case was combined with double-orifice technique due to postoperative poor closure of the tricuspid valve.

Outcomes: There were 2 cases with arrhythmia, and they returned to normal after medication. The rest patients recovered smoothly with no death. Review of echocardiography: 1 patient with moderate regurgitation, the rest of patients' leaflets coapted well and had no tricuspid stenosis. All cases were followed up postoperatively for 9 to 38 months, and there were 14 patients with grade I cardiac function and 4 patients with grade II.

Lessons: The early and mid-term clinical outcomes of the modified cone reconstruction in the treatment of EA were which can make leaflets coapt and had a strong antiregurgitation ability, reducing the incidence of re-operation, valve replacement, and postoperative mortality.

Abbreviations: ASD = atrial septal defect, EA = Ebstein's anomaly, NYHA = New York Heart Association, PDA = patent ductus arteriosus, PFO = patent foramen ovale, TR = tricuspid regurgitation.

Keywords: autologous pericardial, cone reconstruction, double-orifice technique, Ebstein's anomaly

1. Introduction

The significant apical displacement of the part of the tricuspid valve, also called Ebstein's anomaly (EA),[1] is a rare and complex congenital cardiac malformation which was initially founded by Breslaus doctor Wilhelm Ebstein in 1866 when he did the heart autopsy.[2,3] EA accounts for 0.5% to 1% of all congenital cardiac disease[4–6] and only 5% of patients survive beyond the fifth decade.[7,8] From May 2008 and August 2015, 18 consecutive patients with EA were treated by using the modified cone reconstruction in our hospital which achieved satisfactory clinical results. Reports are presented ahead.

2. Methods

2.1. Patients

There were 18 consecutive patients with EA with 8 males and 10 females aged between 5 and 41 years, whose average age was 20.3 ± 12.7 years. According to New York Heart Association functional grade, there were 12 patients with grade II cardiac function and 6 with grade III. All patients presented different

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J-SL and GC equally contributed to the study and should be regarded as co-first authors.

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[a] Department of Cardiothoracic Surgery, Taizhou Central Hospital (Taizhou University Hospital), Taizhou, Zhejiang, [b] The Secondary Ward of Thoracic Surgery, Shanxi Provincial Cancer Hospital, [c] Department of Cardiothoracic Surgery, [d] Department of Ultrasound, Shanxi Medical University Second Hospital, Taiyuan, Shanxi, People's Republic of China.

[*] Correspondence: Jie Ma, Department of Cardiothoracic Surgery, Shanxi Medical University Second Hospital, Taiyuan, Shanxi, People's Republic of China (e-mail: majiexxwk@163.com).

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Table 1

EA Carpentier's classification method with other heart combined deformities.

				Carpentier's classification			
	With ASD	With PDA	With PFO	Type A	Type B	Type C	Type D
Cases	6	2	1	2	10	6	0

ASD = atrial septal defect, EA = Ebstein's anomaly, PDA = patent ductus arteriosus, PFO = patent foramen ovale.

degrees of palpitations, chest stuffiness, and anhelation, among whom 8 were with cyanosis, 4 with edema of lower extremities, and 1 with acute arterial embolism and amputation of left lower extremity caused by paradoxical embolism. Physical examination: a systolic murmur (2–4)/6 was located at the tricuspid valve. Preoperative electrocardiogram: 18 patients presented with right atrial enlargement and 8 patients presented with the right bundle branch block. Preoperative thoracic roentgenoscopy showed different degrees of enlargement in the right atrium and the right ventricle and decrease of pulmonary blood. The cardiothoracic ratio ranged from 0.52 to 0.72 and the average was 0.62 ± 0.06. Preoperative echocardiography: all patients had massive tricuspid regurgitation. There were 6 patients with the atrial septal defect. Among them, 1 had bidirectional shunt and the remaining had left to right shunt; 2 cases were with patent ductus arteriosus and 1 case was with patent foramen ovale. According to Carpentier's classification method, there were 2 patients with type A, 10 with type B, and 6 with type C (Table 1).

2.2. Operative techniques

At first, to find out the development of the tricuspid valve and if there were other combined deformities or not, an oblique incision was made at its proximal attachment to the anterior and moved backwards. The incision terminates when abnormal attachments are completely released from the anterior leaflet, posterior leaflet, and right ventricle. These leaflets are freed as a single piece (Fig. 1A).

Second, cut the papillary muscle abnormally attached at the ventricular wall and leaflets; reserve the junction of the right ventricular apex; provide good exposure to the fused papillary muscle, preserve the space of the subvalvular apparatus, and cut the distal part of the valve leaflet triangularly to form the central bloodstream.

Third, remove the junction between septal and ventricular septum, which can preserve the normal attachment of anterior, tricuspid annulus, and proper subvalvular apparatus. Use all the available tissue to form a cone re-conducted tricuspid valve, stitch up the proximal edge of septal valve and the septal edge of anterior valve, and fix the isolated edge of the posterior valve to the other side of anterior valve. Then a cone-liked reconstructed tricuspid valve are formed (Fig. 1B).

After that, vertically plicate the atrialized right ventricle and stitch it up at endocardium to avoid any coronary injury. Match the plicated true tricuspid annulus with the reconducted cone valve by reconducting the new annulus in the normal anatomical position (Fig. 1C).

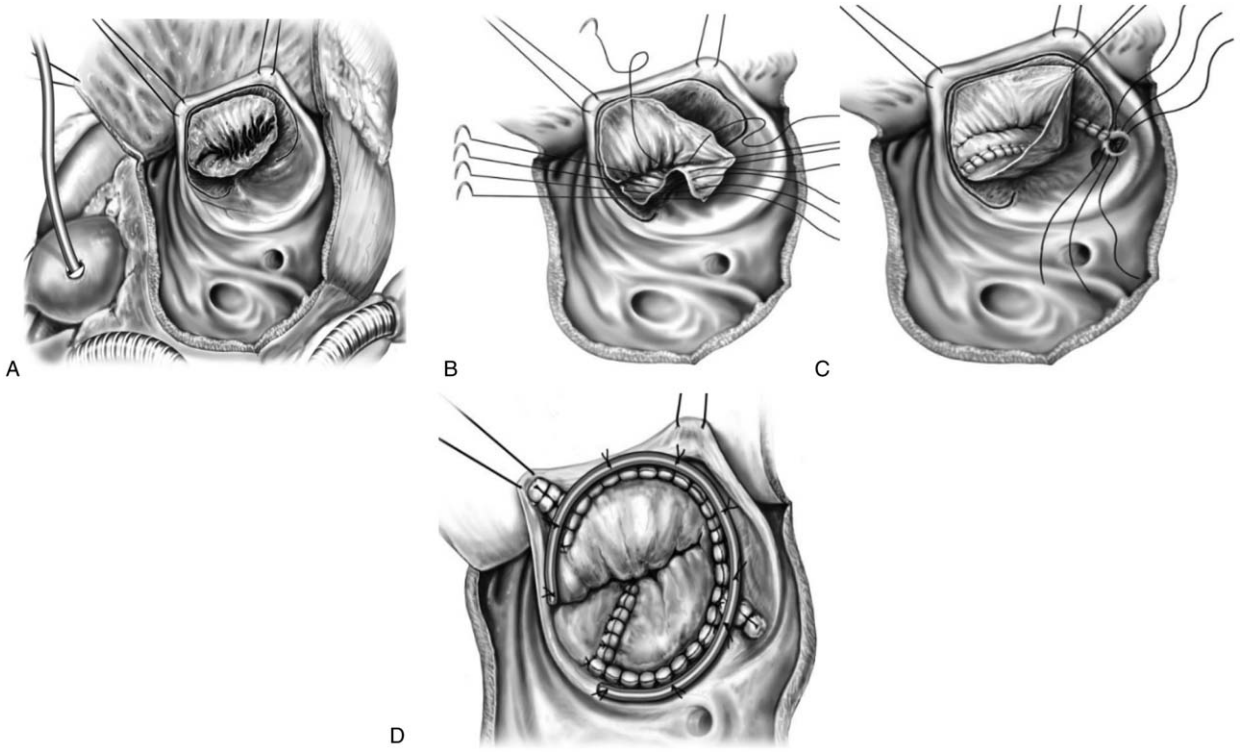

Figure 1. (A) Completely released from the anterior leaflet, posterior leaflet, and right ventricle. These leaflets are freed as a single piece. (B) Use all the available tissue to form a cone re-conducted tricuspid valve. (C) Vertically plicate the atrialized right ventricle and stitch it up at endocardium to avoid any coronary injury. (D) Tricuspid annulus were reinforced by autologous pericardial.

As for the patients whose anterior leaflets have poor development or who have smaller anterior leaflets, autologous pericardium can be used to enlarge leaflets. There were 6 patients who underwent repair of atrial septal defect, 2 had ligation of aorticopulmonary fistula or window, and 1 had acleistocardia suture at the same time. After the operation, right ventricle received routine saline to check the situation of tricuspid valve closure. Because of poor tricuspid valve closure, 1 patient got good effect after combined double-orifice technique (Fig. 1D).

3. Results

For whole group, the time of extracorporeal circulation was 55 to 132 minutes, the average of it was 85.3 minutes; the aortic cross clamp time was 29 to 120 minutes, the average was 63.4 minutes. Arrhythmia happened in 2 patients, both of whom returned to normal after medical treatment. The rest all recovered well without death after operation. Re-examination of echocardiogram showed that 1 had medium tricuspid regurgitation, and all the other 17 patients had a better involution of leaflets without the occurrence of tricuspid stenosis. As for tricuspid regurgitation, 5 had no regurgitation, 4 had insignificant regurgitation, and 8 had little regurgitation. All cases were followed up 9 to 38 months, and data of grade I cardiac function and grade II were 14 and 4, respectively. The thoracic roentgenoscopy for all patients presented that the cardiothoracic ratio had reduced significantly than before (Table 2).

4. Discussion

EA is a rare and complex congenital cardiac malformation in clinical, with poor natural prognosis.[9] It is considered to be caused by the developmental disturbance of primordial valve's connective and muscle tissue in the early stages of embryonic development, which is related to the history of harmful chemical exposure during the early months of pregnancy, abortion, and inheritance. The anatomical features of EA are as below. The septal, posterior leaflet, and anterior leaflet in few cases do not adhere to tricuspid annulus, which have spiral downward displacement to heart ventricle and are divided into atrialized and functional right ventricle.[10,11] EA was divided into 4 types by Carpentier's classification method according to the morphologies and activities of the tricuspid valve and the right ventricle.[12] Type A: atrial ventricle is contractile; septal leaflet and posterior leaflet have the moderate downward displacement; and tricuspid valve has good activities. Type B: atrial ventricular is bigger and has no contractility; septal leaflet and posterior leaflet have the significant downward displacement with dysplasia; and the anterior leaflet has good activities. Type C: the atrial ventricle is large and it cannot contract; septal leaflet and posterior leaflet have the significant downward displacement with severe dysplasia; activities of the anterior leaflet are severely restricted with margo liber tied down. Type D: the large atrial ventricle cannot contract; functional ventricle almost disappears; annulus shows abnormities; valve leaf tissue of the tricuspid valve forms cystic components adhering to the right ventricular wall. Due to the inter-reaction of hypoplasia and dysfunction of the right ventricle and tricuspid incompetence, the right ventricle suffers the overload volume with cardiac enlargement. As a result, patients' life could be threatened by hypoxia, heart failure, and arrhythmia.

According to operation experience in our hospital, surgical treatment should be conducted for patients positively as long as there exist massive tricuspid regurgitation and history of arterial paradoxical embolism. However, based on age of onset, type of abnormal lesion, development of valve, other combined cardiac malformations, and cardiac function, after taking all of these into consideration, when to conduct the operation can be decided. Hunter and Lillehei,[13] Danielson and Fuster,[4] and Carpentier et al[12] all put forward different operational methods in treatment of EA, trying to make the function of the tricuspid valve best. In recent years, surgeons at home conducted many modified Carpentier's classification methods,[14–18] mainly using the bigger anterior leaflet to form the single-leaflet structure based on the anterior leaflet, which got good effects. But as for those whose anterior leaflets are smaller or who have large atrialized ventricle, there is a replacement rate of valve and function of tricuspid valve are poor.

In 1993, da Silva and da Silva Lda[19–21] first put forward the cone reconstruction of the tricuspid valve, which can fully mobilize all the available valve tissues by dissociating and rotating the tricuspid valve clockwise. With this method, the junction of valve and right ventricular apex can be reserved, and not only a cone-reconstructed tricuspid valve can be formed, but also the central bloodstream. As well, the leaflets can coapt well. The right ventricle can get a better anatomic form as well as keep its contractibility. Early follow-up of 52 patients presented that the tricuspid regurgitation had improved significantly, and tricuspid valve still kept a good function in long-term follow-up. In the year of 2012, they summarized and reported the surgical technique and clinical effect of a group of patients who underwent cone reconstruction in details. There are 3 patients dying of low cardiac output syndrome during the stay in hospital; 2 with right ventricular dysfunction and another with bilateral ventricular dysfunction. In the long-term follow-up, 4 patients died of infective endocarditis, heart failure, arrhythmia, and sudden death. Four patients had the second operation because of recurrence of tricuspid regurgitation.[21] After that, Liu et al,[22] Vogel et al,[23] Dearani et al,[6,24] Reddin et al,[25] Sata et al,[26] and Pizarro et al[27] also reported something about reoperation, newborn, and the comparison of traditional operation effect. Patients had a strong ability of antiregurgitation after the cone reconstruction of tricuspid valve, which got good effect.

According to the 18 patients in the group combined with the modified Carpentier's classification method,[17] we summarized some operative experience and improved some. Six key points are as below. First, dissociate all leaflets by excising the secondary

Table 2

Comparison of relative indexes pre/postoperation with 18 EA patients.

	TR (cases)			NYHA (cases)		
	Mild	Moderate	Severe	Grade I	Grade II	Grade III
Preoperation	0	0	18	0	12	6
Postoperation	17	1	0	14	4	0

EA = Ebstein's anomaly, NYHA = New York Heart Association, TR = tricuspid regurgitation.

chordae tendineae and abnormal tissue that limit leaflets movement. Second, reconstruct a cone structure of tricuspid valve by suturing junction. Third, plicate the atrialized right ventricle lengthwise, stitch it up at endocardium to avoid damaging coronary artery, and reduce the tricuspid annulus back to normal anatomical size. Fourth, consolidate the tricuspid annulus by using the autologous pericardium, so does the cone reconstruction, which get good effect. Here are some advantages of it: endocardial cells are easy to elongate and reduce which consolidate the tricuspid annulus; compliance of annulus is not affected; long-term anticoagulation are not needed which avoid the occurrence of thromboembolism; the reoperation can be avoided because of the tricuspid regurgitation caused by tear at the junction of suture lines postoperation. Fifth, the anterior leaflet is widened by using autologous pericardium for those whose anterior leaflets are smaller or have poor development. Sixth, if the tricuspid valve closure is still poor after above steps, double-orifice technique can be applied to avoid the occurrence of valve replacement. Using one to two 4–0 prolene double-ended needle with gasket sews the related leaflets at the middle of new formed orifice, forming a double-orifice tricuspid valve and making the tricuspid regurgitation disappear, which can avoid the occurrence of valve replacement.

There is a patient with EA, severe tricuspid regurgitation and secundum atrial septal defect, and, bidirectional shunt, suffering the acute arterial embolism of left lower extremity, gangrene, and amputation caused by paradoxical embolism in this group. Sudden arterial occlusion is very rare especially in childhood. When thoracic roentgenoscopy, arterial blood gas analysis, coagulation test and echocardiographic of lower extremity deep venous system are all normal, one should consider the possibility of a paradoxical embolism. In the treatment of modified cone reconstruction, ultrasonic cardiograms at postoperative 6 and 9 months showed that there was little tricuspid regurgitation, means patients were recovering well. Patients with massive tricuspid regurgitation combined right to left shunt should perform the surgical treatment actively, avoiding the occurrence of serious complication to provide guarantee for patients' quality of life and to relieve pain of patients and their family.

5. Conclusions

The effects of modified cone reconstruction of tricuspid valve have been well recognized recently and the method is practical, which can use its own tissues to form not only central bloodstream, but also the coaptation between the 2 leaflets. With a strong ability of antiregurgitation, right ventricle can get a better anatomic form as well as keep its contractibility, and both the occurrence of second operation and valve replacement and postoperative mortality can be reduced, which is worth of being generalized.

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