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# Cone Repair in Adult Patients with Ebstein Anomaly

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Ebstein anomaly is a rare congenital heart malformation typically involving the tricuspid valve and the right ventricle that has a wide range of anatomical and pathophysiological presentations. Various surgical repair techniques for Ebstein anomaly have been reported because of its near-infinite anatomical variability. Cone repair for Ebstein anomaly can achieve nearly anatomical reconstruction of the tricuspid valve with promising outcomes. In this article, the surgical techniques for cone repair in adult patients with Ebstein anomaly are described in detail, and clinical experiences and technically challenging cases are presented.

**Keywords:** Ebstein anomaly, Cone repair, Adult patient

## Introduction

Ebstein anomaly is a malformation of the tricuspid valve (TV) and right ventricle (RV) dysplasia with extreme anatomical variability [1,2]. It is a rare disease, accounting for less than 1% of all cases of congenital heart disease [2]. During normal embryological development, the TV leaflets should split and detach from the inner layer of the RV myocardium, a process that is referred to as delamination [1]. Failure of delamination is one of the essential features of the pathological anatomy of Ebstein anomaly, which represents the adherence of primarily the septal and posterior leaflets to the underlying myocardium, leading to downward displacement of the functional annulus and causing the RV to be divided into 2 parts (the functional and atrialized ventricle) (Fig. 1A, B). The atrialized ventricle is usually a thin-walled, noncontractile, and dyskinetic part of the ventricle (Fig. 1A, B). The other features of this condition are redundancy, fenestration, and tethering of the anterior leaflets and dilatation of the true tricuspid annulus (Fig. 2A) [1].

Patients with Ebstein anomaly present with various clinical manifestations depending on their age at presentation, the severity of poor delamination, and the extent of myocardial involvement. Compared to young infants or children, adolescent or adult patients with Ebstein anomaly present with more consistent symptoms and signs, including exercise intolerance, various arrhythmias, and mild cyanosis with a combined atrial septal defect (ASD).

The severity of tricuspid regurgitation (TR) determines whether surgical treatment is indicated. In symptomatic patients with a mild form of Ebstein anomaly and mild TR, non-surgical treatment (e.g., medications, ASD device closure, or arrhythmia intervention) is sufficient. The classic indications of surgical treatment are cardiomegaly, hypoxia, exercise intolerance, new-onset or aggravated arrhythmia, and RV enlargement or dysfunction. However, if patients present with moderate to severe TR, surgical treatment should be considered regardless of the presence of symptoms. The rationale for this approach seems to be that repair is possible in most patients, while TR could advance to right heart failure. Dearani et al. [2] recently suggested from their experiences that repair should be considered for patients with Ebstein anomaly in early childhood.

## Surgical management of Ebstein anomaly

The classic surgical approach to Ebstein anomaly in adults involves so-called "non-cone" procedures, including the Danielson operation [3], the Carpentier operation [4], and the Hetzer operation [5]. The word "cone" derives from a report of da Silva et al. [6], in which they tried to repair the deformed TV based on the principle of Carpen-
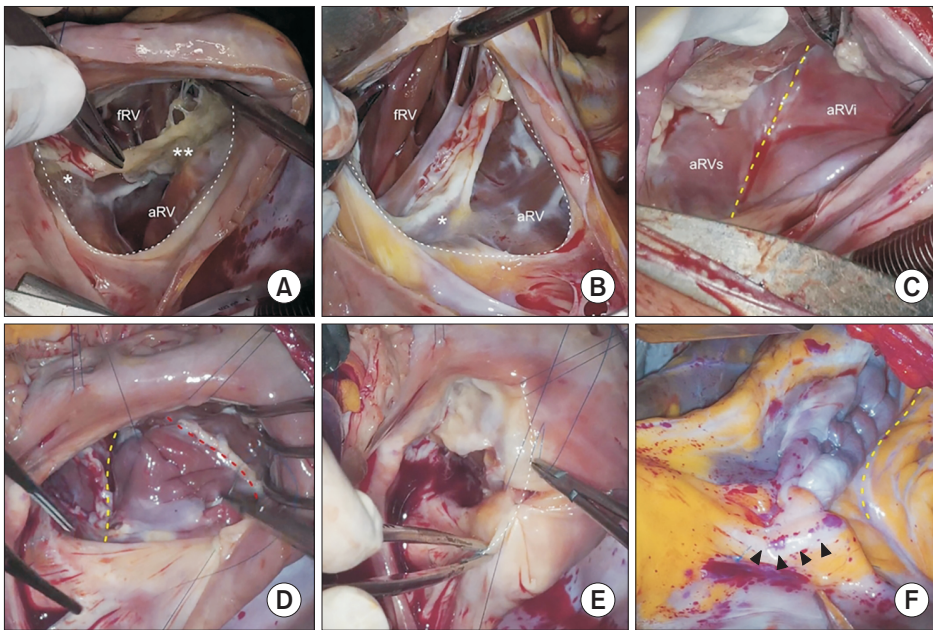
**Fig. 1.** Plication of the atrialized ventricle. (A, B) Displaced septal and posterior leaflets; dotted line: the true TV annulus; * displaced septal leaflet; ** displaced posterior leaflet. (C, D) Internal view of plication of the atrialized ventricle; yellow dotted line: interventricular inferior wall bordered by the posterior descending coronary artery; red dotted line: detached line of the posterior leaflet. (E) Posterior annuloplasty; white dotted line: the true TV annulus. (F) External view of the plicated atrialized ventricle; yellow dotted line: the course of the posterior descending coronary artery, black arrowheads: the right coronary artery. TV, tricuspid valve; aRV, atrialized right ventricle; fRV, functional right ventricle; aRVs, the septal area of the aRV; aRVi, the inferior wall of the aRV.
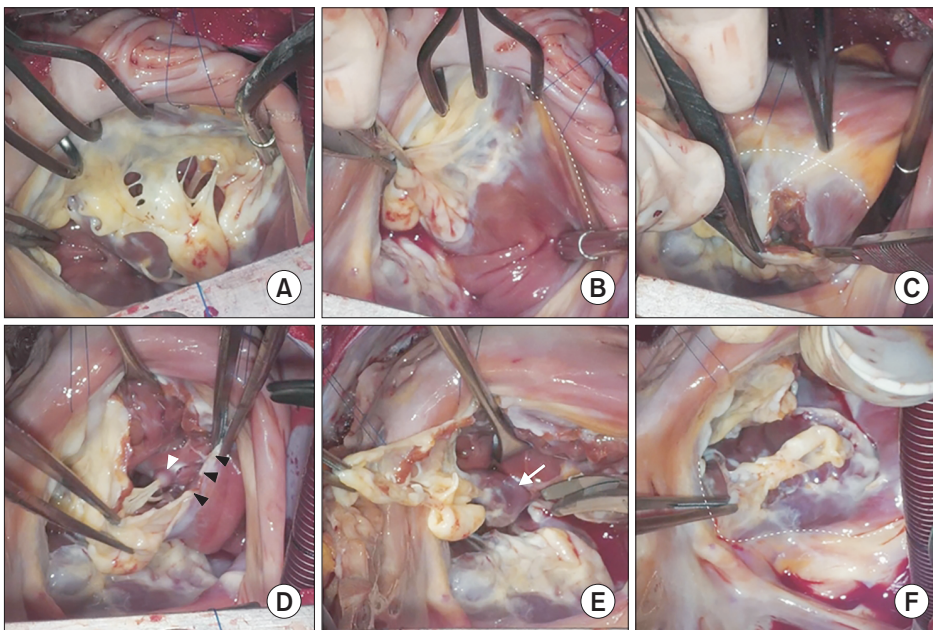


**Fig. 2.** Surgical delamination. (A) The anterior papillary muscle. (B, C) Displacement of the posterior leaflet and starting surgical delamination; dotted line: the true TV annulus. (D) Detachment of the posterior leaflet; white arrowhead: tethering papillary muscle of the anterior leaflet, black arrowheads: detached line of the posterior leaflet. (E) Mobilized anteroposterior leaflet tissue; white arrow: the anterior papillary muscle acting as the cone apex. (F) Mobilizing the displaced septal leaflets; dotted line: the true TV annulus. TV, tricuspid valve.

tier et al. [4], by bringing the TV leaflets to the true tricuspid annulus level and performing longitudinal plication of the atrialized RV as necessary to restore the RV volume and morphology. What is the difference between non-cone and cone repair? In non-cone repair, TV coaptation is accomplished between the anterior leaflets and the ventricular septal wall in a monocuspid manner, thereby generating an off-center diastolic blood flow [6]. These repair techniques may provide excellent short- and mid-term results, but even small residual regurgitation and a slight change in TV coaptation and annulus geometry could result in more valve regurgitation, leading to further changes that eventually culminate in severe regurgitation. However, in cone repair, the cone-shaped valve opens to central blood flow and closes with full coaptation of the leaflets, resulting in central diastolic blood flow [6].

After an initial report of da Silva et al. [6], several authors reported favorable surgical outcomes of cone repair

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**Table 1.** Previous studies analyzing cone repair for Ebstein anomaly

| Authors | Operation year | No. of patients | Mean age at operation (yr) | Early mortality | Mean follow-up (mo) | Late mortality | Reoperation | PPM insertion |
|---------|----------------|-----------------|----------------------------|-----------------|---------------------|----------------|-------------|---------------|
| Da Silva et al. (2012) [7] | 1993–2011 | 100 | 17.5 | 3 (3) | NA | 4 (4) | 4 (4) | 1 (1) |
| Ibraim et al. (2015) [8] | 2009–2013 | 27 | 15.8 | 0 | 32 | 0 | 1 (4) | 3 (11) |
| Lange et al. (2015) [9] | 2010–2013 | 20 | 34.2 | 2 (10) | 11 | 0 | 1 (5) | 0 |
| Belli et al. (2017) [10] | 2007–2014 | 20 | 34.3 | 0 | 34 | 2 (10) | 1 (5) | 0 |
| Holst el al. (2018) [11][a] | 2007–2015 | 101 | 34.1 | 1 (1) | 42 | 0[b] | 7 (7)[b] | 0 |

Values are presented as number or number (%), unless otherwise stated.
PPM, permanent pacemaker; NA, not available.
[a]Among total 235 patients, 101 patients aged 18 years or more were summarized in the table. [b]The exact numbers of late mortality and reoperation were not available in the article.

(Table 1) [7-11]. Most recently, Holst et al. [11] reported the most extensive experiences of cone repair, in which TV leaflet modifications using pericardium were applied to optimize leaflet coaptation in nearly 30% of patients. Close follow-up should be performed to evaluate any changes in the pericardium with these modifications, as well as TV function.

## Anatomical understanding for cone repair

To perform cone repair competently, one must first be familiar with the concept of the cone. A cone is a shape with a flat, round, or oval base and a top that becomes narrower until it forms a point. The base and the top of the cone are integral parts of cone repair. The plane of the base plane is the right-side atrioventricular junction or the true tricuspid annulus, where the TV leaflets should be 360°-attached. The major anterior papillary muscle should be at the top of the cone and keep supporting the leaflets. In some patients, the medial papillary muscle can support part of the septal leaflets that participate in forming the base. Schreiber et al. [12] described Ebstein anomaly as much more than simple downward displacement of the leaflets. The term "more than simple" downward displacement is crucially important for understanding how to make the deformed TV into a new cone. Much more of the septal and posterior leaflets than the anterior leaflets are displaced to the RV apex. The uneven arrangement of each leaflet relative to the true annulus makes the TV apposition plane distorted mainly towards the right ventricular outflow tract (RVOT), and the TV apposition plane is getting more toward the RVOT than the RV apex as poor delamination becomes severe. Therefore, although the amount of leaflet tissue seems small when looking through the TV annulus to the RV apex, even in severe forms of Ebstein anomaly, the total amount of leaflet tissue facing the RVOT is not small and suffices to make a new cone following adequate RV plication, which is a critical point to understand for cone repair.

## Surgical techniques of cone repair

Cone repair consists of 3 main parts: (1) surgical delamination; (2) plication of the atrialized ventricle and reduction and fixation of the dilated true TV annulus; and (3) reattachment of the TV leaflet tissue to the true annulus.

### Surgical delamination

Surgical delamination achieves TV leaflet mobilization by sectioning the abnormal tissues between the TV leaflet and the ventricular wall at the areas of failed embryological delamination. Surgical delamination begins from the point where the anterior leaflets are displaced downward and join the posterior leaflets (Fig. 2B). To obtain as much leaflet tissue from the anterior and posterior leaflets as possible, care should be taken to perform meticulous surgical delamination, such as obtaining extra muscular or fibrotic tissue along with the leaflet and entirely freeing the leaflet tissue from its abnormal attachments to the RV wall and papillary muscles, except for the main anterior papillary muscles at the apex of the cone (Fig. 2C). The role of the septal leaflet, which is usually displaced downward and hypoplastic, in cone repair tends to be underestimated (Fig. 2F). As da Silva et al. [6] emphasized, the septal leaflet is sufficiently developed to take part in the newly constructed TV in more than half of patients. The amount of remaining septal leaflet tissue is different in each patient, and every effort should be made to use it to support the anterosuperior septal area (Fig. 3C). The uppermost part of the septal leaflet remnant could also prevent an injury to the conduction tissue while reattaching the leaflets in this area.
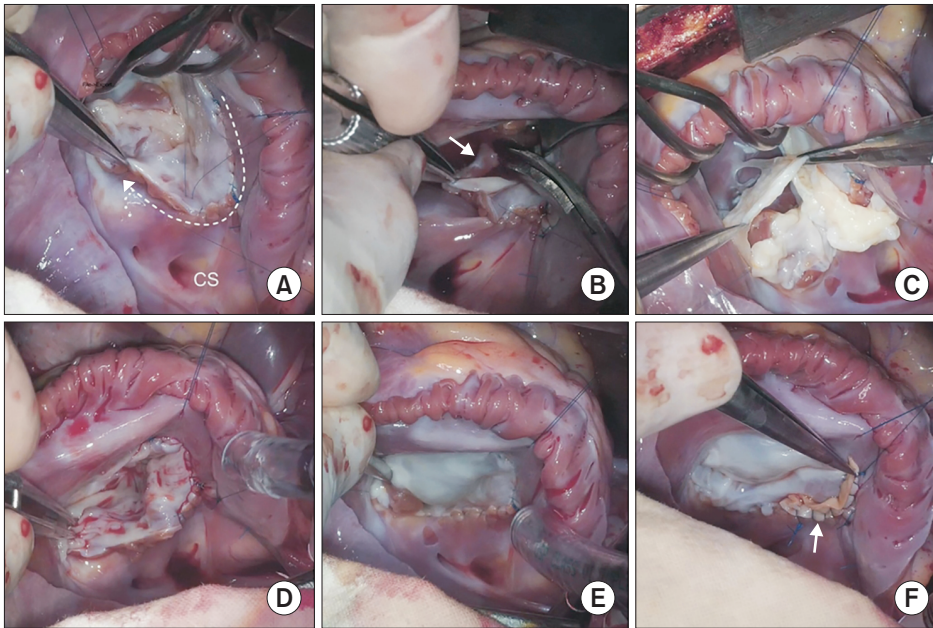
**Fig. 3.** Reattachment of the leaflets. (A) Reattachment of the leaflets clockwise to the true annulus (dotted arrow). (B) Resection of the tethering papillary muscle (arrow). (C) Mobilization of the septal leaflet. (D, E) Saline test using a syringe bulb. (F) Reinforcement of the annulus using a bovine pericardial strip (arrow). CS, coronary sinus.
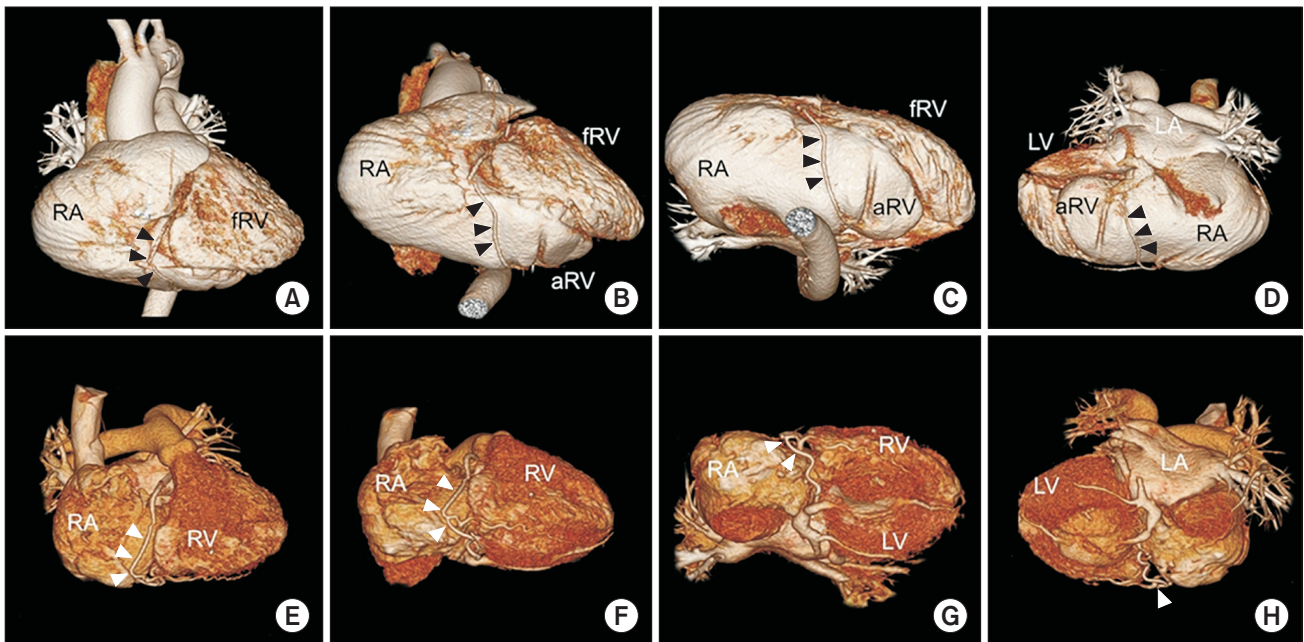


**Fig. 4.** Chest CT angiogram of plication of the atrialized ventricle. The top and bottom panels show preoperative and postoperative CT images, respectively. Column (A, E) anterior-posterior view, (B, F) anterior caudal view, (C, G) view from below, and (D, H) posterior caudal view. CT, computed tomography; RA, right atrium; RV, right ventricle; fRV, functional RV; aRV, atrialized RV; LV, left ventricle; LA, left atrium; white and black arrowheads, the right coronary artery running along the atrioventricular groove.

## Plication of the atrialized ventricle and reduction and fixation of the dilated true tricuspid valve annulus

Whether to perform plication of the atrialized ventricle is still controversial. Recently, Hetzer et al. [13] insisted

that incorporating the atrialized ventricle into the functional RV without its plication has no harmful effects, allows sufficient RV diastolic filling, and could contribute to RV contraction. However, Carpentier et al. [4] emphasized that the shape, compliance, and integrity or impairment of the contractility of the RV may be as important a factor as

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TV dysfunction, and stated that longitudinal plication can preserve the height of the RV and effectively exclude the atrialized chamber (Fig. 4). When patients have Carpentier type B or C anomalies, plication should be actively considered. It is crucial to decide which area should be plicated and how to avoid any kinking or obstruction of the right coronary artery. The atrialized ventricle could be made of 2 parts, such as the septal area and inferior ventricular free wall, divided by the posterior descending coronary artery (Fig. 1C). Therefore, plication should be done only at the inferior ventricular free wall. Its starting point is the most distal part of the atrialized ventricle coming to the atrioventricular junction with a sufficient reduction of the free inferior wall. This longitudinal plication passes through the true annulus to the atrial wall (Fig. 1D, E). In rare cases, a right coronary artery runs across the RV free wall away from the atrioventricular junction, in which case great care should be taken to avoid injuring the coronary artery during plication (Fig. 5).

## Reattachment of the tricuspid valve leaflet tissue to the true annulus

Reattachment of the detached leaflet tissue could be usually performed clockwise to the true TV annulus using a fine running suture (Fig. 3A). Every effort should be made to confirm whether the leaflet tissue is tethered or supported by any other subvalvular structure (Fig. 3B). A saline test using a syringe bulb and putting the leaflet tissue down at the septal area shows valve coaptation and helps reattach the leaflet tissue properly to the true annulus (Fig. 3D, E). Leaflet tissue should be carefully reattached to the atrioventricular conduction area with very superficial sutures or detouring to the ventricular side away from the conduction tissue [6,14]. As mentioned above, the septal leaflet remnant around that area helps avoid any injury to the conduction tissue. Before the procedure concludes, the inferior annulus should be reinforced to prevent the disruption of the repaired annulus using a commercial ring or pericardial strip (Fig. 3F).

## Experiences of cone repair in adult patients with Ebstein anomaly at Sejong General Hospital

### Patient characteristics

Cone repair has been adopted as the procedure of choice for Ebstein anomaly at our institution since 2008. Between
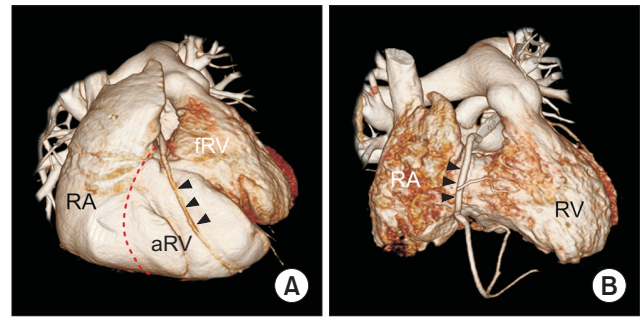


**Fig. 5.** Chest computed tomography angiogram of plication of the atrialized ventricle in a patient with an abnormal right coronary artery course. (A) Preoperative. (B) Postoperative. Arrowheads: the right coronary artery; red dotted line: the atrioventricular groove; RA, right atrium; RV, right ventricle; fRV, functional RV; aRV, atrialized RV.

2008 and 2019, 24 patients with Ebstein anomaly aged 16 years or older (6 male patients and 18 female patients) underwent cone repair. The median age and body weight of the patients at operation were 36 years (range, 17–67 years) and 57 kg (range, 47–89 kg), respectively. Carpentier type B anomalies were the most frequent type (n=17), followed by type A and C anomalies (n=3 each). The other patient had Hetzer repair at another hospital 10 years ago, and it was difficult to confirm the type of the anomaly. The indications for surgery included symptoms of fatigue, cyanosis, shortness of breath, or decreased exercise tolerance. Progressive RV enlargement or dysfunction, atrial tachyarrhythmia, or both are indications for surgery in asymptomatic patients. Preoperative echocardiography showed severe TR in all patients, except for 1 who had moderate TR and hypertrophic cardiomyopathy with severe left ventricular outflow tract obstruction and systolic anterior motion of the mitral leaflets. Nine patients (38%) had preoperative rhythm problems, including atrial fibrillation and flutter (n=5), Wolff-Parkinson-White (WPW) syndrome (n=5), and atrial tachycardia (n=1). Three of the patients with WPW syndrome underwent radiofrequency catheter ablation preoperatively. The associated cardiac lesions were an ASD (n=14) and hypertrophic cardiomyopathy with mitral regurgitation (n=1).

### Surgical details and the postoperative course

Cone repair was performed under mild to moderate hypothermia, bicaval cannulation, and cardioplegia-induced cardiac arrest. The procedures were performed in the same manner as described above. The median cardiopulmonary bypass and aortic cross-clamp time were 192 minutes

(range, 123–311 minutes) and 156 minutes (range, 84–262 minutes), respectively. Longitudinal RV plication was done in 22 patients (92%) and TV annular reinforcement was done in 19 patients (79%) with a commercial annuloplasty ring (n=2) or pericardial strip (n=17). In 8 patients (33%), arrhythmia surgery was performed, including right-side maze (n=4), cavotricuspid isthmus ablation (n=3), and full maze (n=1) procedures. Accessory pathway ablation was performed in 3 patients with WPW syndrome. If present, ASDs were completely closed, and no patients needed the addition of a cavopulmonary connection.

### Surgical results and follow-up

There were no deaths. The median postoperative hospital stay was 10 days (range, 5–77 days). The patient who stayed longest underwent a TV replacement with a tissue valve 2 days after surgery because of posteroinferior annulus disruption, followed by the recurrence of severe TR and a wound problem. Postoperative complications requiring an intervention occurred in 6 patients, including bleeding (n=2), pericardial effusion (n=2), diaphragm palsy (n=1), an operation wound problem (n=1), and complete heart block (n=1). Three patients (13%) experienced complete heart block immediately after surgery, and 2 patients recovered to sinus rhythm on postoperative days 5 and 6, respectively, while 1 patient eventually needed a permanent pacemaker insertion. Twenty-three patients were followed up for a median duration of 37 months (range, 3.2–144 months). One patient from another country underwent cone repair under a charity program, recovered uneventfully, and could not be followed up. One patient (5%) underwent re-repair of the disrupted TV with an annuloplasty ring 17 months after cone repair, and the most recent echocardiography revealed moderate regurgitation and mild stenosis. In this series, 2 patients needed reoperation because of disruption of the posterior annulus, and reinforcement of the new TV annulus was performed using an annuloplasty ring or pericardial strip in both patients. Even though the patients received an anti-arrhythmia intervention preoperatively or concomitantly, 8 patients (35%) experienced various types of arrhythmia during follow-up, including atrial fibrillation/flutter (n=4). Twenty-two patients were followed up using echocardiography for a median duration of 34 months (range, 3.2–144 months), excluding the patient with TV replacement. More than moderate TR at the last follow-up was not found in any patients. However, 3 patients showed moderate TR. Mild tricuspid stenosis was found in 2 patients.

## Conclusion

Cone repair for Ebstein anomaly shows excellent early and mid-term surgical outcomes, providing a well-functioning repaired TV. Furthermore, improved TR and reduced RV size at the last follow-up suggest that cone repair may be advantageous for long-term RV remodeling. Additional long-term research on improving RV function and the durability of TV function is required.

## Conflict of interest

No potential conflict of interest relevant to this article was reported.

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