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CLINICAL RESEARCH

Ebstein's anomaly in adults: Modified cone reconstruction of the tricuspid valve is associated with promising outcomes

Anomalie d'Ebstein chez l'adulte : la chirurgie de cône de reconstruction valvulaire tricuspide est prometteuse

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KEYWORDS
Congenital heart disease;
Surgery;
Tricuspid valve;
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Summary
Background. — Ebstein's anomaly is a complex malformation. Justification of a procedure in mildly symptomatic adults is debatable: repair techniques are demanding and valve replacement is associated with poorer outcome.
Objectives. — We report our initial experience with the cone procedure versus medical follow-up.
Methods. — Patients aged ≥ 15 years with Ebstein's anomaly were enrolled during 2007—2014. The cone procedure was performed in consecutive patients with severe tricuspid regurgitation (TR); those with less severe disease did not undergo surgery, although some underwent percutaneous catheter atrial septal defect closure.
Results. — The cone procedure was performed in 20 patients (mean age 34.3 ± 14.4 years; TR grade 3.3 ± 0.7) because of impaired functional capacity: six New York Heart Association (NYHA) class II, 14 class III. No surgical patient died during a mean (range) follow-up of 2.8 (0.5—5.0) years. One patient required subsequent repair for suture dehiscence 6 months postoperatively.

Abbreviations: ASD, atrial septal defect; NYHA, New York Heart Association; SD, standard deviation; TR, tricuspid regurgitation.

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All patients presented with mild or less TR at last echocardiographic follow-up. NYHA functional class was significantly improved at follow-up ($P < 0.0001$): 16 NYHA class I and four class II. Of 24 non-surgical patients (mean age 37.3 ± 16.9 years; TR grade 2.2 ± 0.8), seven underwent percutaneous ASD closure. During a mean (range) follow-up of 4.8 (3.6—5.0) years, two patients died: one sudden death and one stroke. **Conclusion** The cone repair of the tricuspid valve in adults with Ebstein's anomaly provided excellent mid-term results and significantly improved functional status. This procedure might be considered even in mildly symptomatic patients in the presence of severe valve regurgitation.

MOTS CLÉS
Cardiopathie congénitale ;
Chirurgie ;
Valve tricuspide ;
Ventricule droit

Résumé L'anomalie d'Ebstein est une malformation complexe. L'indication opératoire chez un patient peu symptomatique est débattue, la réparation de la valve est requise car le remplacement valvulaire est associé à de mauvais résultats.

Objectifs. — Nous rapportons notre expérience initiale de chirurgie de cône ainsi que le suivi médical.

Méthodes. — Les patients de plus de 15 ans furent inclus entre 2007 et 2014. La chirurgie de cône fut indiquée chez les patients avec fuite tricuspide sévère, ceux ayant une forme moindre de la maladie ne furent pas opérés, certains eurent une fermeture percutanée de la communication interauriculaire.

Résultats. — La chirurgie de cône fut réalisée chez 20 patients (âge moyen de $34,3 \pm 14,4$ ans ; fuite tricuspide de grade $3,3 \pm 0,7$) en raison de leur symptomatologie fonctionnelle : 6 étaient en NYHA classe II, 14 en classe III. Il n'y a eu aucun décès de patients opérés durant in suivi de 2,8 (0,5—5,0) ans. Un patient a dû être réopéré 6 mois après pour lâchage de suture. La fuite tricuspide était de faible grade chez tous les patients opérés. La classe fonctionnelle NYHA a été améliorée de façon significative ($p < 0,0001$) : 16 étaient en NYHA classe II, 4 en classe III. Parmi les 24 patients non opérés (âge médian de $37,3 \pm 16,9$ ans ; fuite tricuspide de grade 2,2 0,8), sept patients eurent une fermeture percutanée de la communication interauriculaire. Durant un suivi moyen de 4,8 (3,6—5,0) ans, deux patients décédèrent : 1 mort subite et 1 accident vasculaire cérébral.

Conclusion. — La chirurgie de cône de reconstruction valvulaire tricuspide chez des patients adultes avec anomalie d'Ebstein donne d'excellent résultats à moyen terme et améliore significativement le statut fonctionnel. Cette chirurgie peut être discutée chez des patients peu symptomatiques avec des fuites tricuspides sévères.

Background

Ebstein's anomaly was described for the first time by Wilhelm Ebstein in 1866 [1] and accounts for less than 1% of congenital heart disease (approximately 1:200,000 births) [2]. This anomaly has always been difficult to classify, because of a specific pathophysiology involving tricuspid valve regurgitation and a primary abnormality in the development of the right ventricle, frequently associated with a right-to-left shunt at atrial level [3]. Another characteristic of this disease is the wide variety of its types and ages of detection, related to a wide spectrum of severity of the anatomical abnormality, ranging from minimal apposition of the septal leaflet of the tricuspid valve to a quasi-absence of functional right ventricle.

The therapeutic approach to Ebstein's anomaly in paediatric cardiology is still debated. Surgery has had a central place — since the 1970s — for most congenital heart defects and allows — for a significant proportion of patients — a full recovery with good long-term prognosis. However, the management of Ebstein's anomaly, particularly in adults, is still the source of much debate on the appropriateness of an invasive procedure for many reasons. First, the surgical repair to directly correct the valvular abnormality — the tricuspid plasty — is technically challenging. Second, Ebstein's disease can be very well tolerated despite severe anatomical lesions. Third, effective tools to stratify the patient's prognosis, and predict the occurrence of right heart failure are still lacking.

Several surgical techniques have been developed. The first ones consisted of valve replacement by mechanical prosthesis then bioprothesis [4,5]. The first valve repair was simply forming a 'mono-cusp' valve by associating a transverse plication of the atrialized ventricle to a posterior annuloplasty [6]. Carpentier et al. published, in 1988, an innovative technique based on the complete detachment of

the proximal insertion of the anterior leaflet associated with the most effective 'surgical delamination' possible and the re-implantation at the anatomic tricuspid annulus level [7]. A longitudinal plication of atrialized ventricle, a prosthetic ring and, eventually, a superior cavopulmonary anastomosis were associated for most of the patients. This surgery was guided by preoperative ultrasound assessment of the state of the anterior leaflet in a specific detailed classification [7]. Until 2007, the Mayo Clinic team continued to use this technique. Also, after stopping the original technique, they preferred a functional reconstruction technique using anterior leaflet with annuloplasty, moving the anterior pillar and making a 'mono-cusp' valve. The proportion of tricuspid valvuloplasties performed in this group remained below 30% [8]. An elective resection of atrialized ventricle was preferred to the systematic plication, with a conservative attitude when the atrialized ventricle showed residual contractility.

The difficulty in Ebstein's anomaly is the lack of valvular tissue and the importance of defects of the sub-valvular apparatus, making anatomic repair in three leaflets illusory. Since the late 1990s, introduced by da Silva et al. [9], there has been a new surgical repair technique called 'cone reconstruction' that brings several important innovations. The first is to replace the tricuspid annulus in its anatomical position; the second is to repair the valve with two sheets, opening in the right ventricle in an axis more 'physiologically' directed to the apex [9]. Since the publication of the paper by da Silva et al. [9] in 2008, we opted for this new technique for all types of Ebstein's patients of any age. In our current practice, the original 'cone' technique was modified, particularly in the adult population, including the use of prosthetic annulus and/or leaflet patch augmentation.

The aims of this study were to evaluate the technique of modified 'cone' reconstruction of the tricuspid valve in patients aged 15 years or older with Ebstein's anomaly and to compare their outcomes to those of patients who had received only medical follow-up, including some who benefited from percutaneous closure of their atrial septal defect (ASD) by cardiac catheterization.

Methods

During 2007—2014, consecutive patients with Ebstein's disease underwent 'cone' reconstruction. Surgical patients were compared with those who were not proposed for surgical procedure, some of whom underwent percutaneous ASD closure.

Medical history prior to inclusion was collected through analysis of medical records and interviews of the patients. The following symptoms were exhaustively sought: dyspnoea, cyanosis, weakness, palpitations, angina pectoris and episodes of faintness or syncope. Dyspnoea was graded according to the New York Heart Association (NYHA) classification into 4 grades. Saturation at rest was systematically recorded at inclusion. Ongoing medical treatments were collected.

A 12-lead electrocardiogram was recorded in all patients at inclusion and at every follow-up visit, searching for: sinus rhythm or not, right bundle branch block, pre-excitation and first-degree atrioventricular block. Chest radiography was available for most patients (except for five patients in the medical group) and was used to measure cardio-thoracic index. Echocardiography was available in all patients. The degree of tricuspid regurgitation (TR) was graded into four classes [10], and the existence of an atrial shunt and its type (ASD or patent foramen ovale) were sought, as well as any association with other congenital heart defects. Accurate and reproducible data for assessing right ventricular function were only available for a limited number of patients so were not used for this study. All surgical patients also underwent a transoesophageal echocardiography before leaving the operating room and a transthoracic echocardiography prior to hospital discharge, to evaluate surgical results and track complications.

The modified 'cone' reconstruction tricuspid valve repair technique

The repair consisted, firstly, of a basal detachment of anterior and posterior leaflets. The access thus created allowed optimal surgical 'delamination' of both leaflets, of the septal leaflet when it existed, and also allowed the release of the sub-valvular apparatus, especially working on the anteromedial papillary muscle. It is essential to keep the distal attachments of the leaflets to the ventricle, but it is necessary to reduce their number and to extend them (Fig. 1A).

After this initial preparation phase, the 'atrialized' intermediate chamber was plicated by means of continuous monofilament sutures. This plication, as well as the annular plication, was performed in a limited manner by the insertion of two or three polyester sutures at the level of the caudal segment of the tricuspid annulus. Then, the free end of the posterior leaflet was rotated in a clockwise direction to be sutured to the free end of the anterior leaflet, forming a cone. The modification essentially aimed to preserve the septal commissure, not to result in an atrioventricular valve without any commissure and, on the other hand, to avoid extensive plication of the atrialized segment, particularly the annulus. Meticulous attention was paid to avoid coronary artery distortion. The posterior leaflet was slid and fixed to the septum to form a septal leaflet. The septal leaflet, when it had retained a certain mobility, was used to expand the cone, using a suture zone between the septal and posterior leaflets. The latter was then enlarged by means of an autologous pericardial patch in 14 at the beginning of the present experience. This patch formed part of the cone (Fig. 1B). The prosthetic ring (Uniring®, Peters Surgical, Bobigny, France) was used in 16 patients in the current study. The prosthetic ring was used mainly to support leaflet repair sutures. The size of the annulus was adjusted to reduce the posterior annulus without any particular size target allowed by the specificity of the device, with the number of stitches varying between 10 and 14. Only four patients underwent repair without an additional patch or ring. In three patients, leaflet coaptation was optimized by the addition of Sebening stitches [11]. Finally, the atrial septal defect was closed.

The percutaneous ASD closure technique

In the medical group, some patient benefited from an ASD catheter closure, which involved the implantation of a

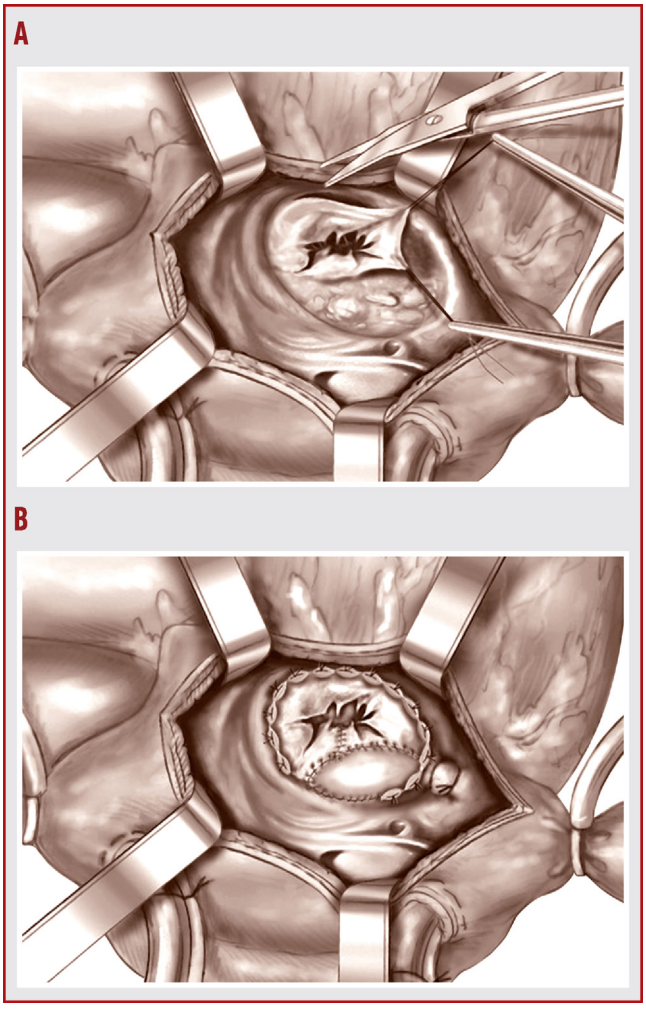

Figure 1. Cone reconstruction' tricuspid valve repair. A. Subtotal detachment of leaflet tissue. B. Repair completed using ring and pericardial augmentation patch.

self-expandable prosthesis of variable size, usually of the Amplatzer type, by femoral venous access, on the edges of the ASD [12]. The principal indication for ASD closure was cyanosis due to right-to-left shunt, particularly during exercise, which limited functional capacity.

Postoperative data collection and follow-up

All events during hospitalization were collected exhaustively. The types of prosthetic equipment used were noted. In the postoperative period, the following data were collected: duration of stay in the intensive care unit, time to extubation, duration of catecholamine therapy, occurrence of infectious events related to surgery or resuscitation and occurrence of other complications. Indications for immediate surgical reintervention were the occurrence of severe bleeding, the presence of a residual TR greater than grade II and the occurrence of surgical site infection. The occurrence of arrhythmias or conduction disorders after surgery was investigated. The need for peri- or postoperative transfusion was noted.

We paid particular attention to the data permitting comparison with previous studies, i.e. mortality and morbidity, supraventricular arrhythmias, heart failure and the increase in functional status after the procedure. Event rates associated with the disease were assessed using a composite criteria consisting of recurrence of supraventricular tachycardia or occurrence ≥ 2 months after procedure, cardiac insufficiency, endocarditis, stroke or sudden death.

Forty of 44 patients completed a telephone survey (two patients with mental retardation were helped with for this). Patients were asked about the same types of symptoms as those sought during the first interview. In case of death, questioning of relatives or doctors allowed us to ascertain its causes and circumstances.

Patients were asked about the need for hospitalization for whatever reason, focusing on heart failure, arrhythmias and the use of rhythmic procedures. Information on the need for surgery (first time or reoperation) and its indication was collected.

Two questions derived from a standardized and validated test of quality of life seeking a possible global subjective functional benefit were asked: 'Since the surgery, do you feel: much better, a little better, unchanged, at bit worse or much worse than before?' and 'If you were to give your opinion on the support around the intervention, would you be: very satisfied, quite satisfied, not really satisfied or not satisfied at all?'.

Statistical analysis

Data are expressed as counts and percentages for categorical variables and means for continuous variables. StatView® 4.0 and Excel 5.0 softwares were used. Differences between groups were analysed by the Chi^2 test, the paired *t*-test, the Fisher least significant difference test and analysis of variance. P values < 0.05 were considered statistically significant.

Results

Patients characteristics

The mean $\pm$ SD (range) ages at inclusion were 34.3 ± 14.4 (17—60) years in the surgical group and 37.3 ± 16.9 (15—75) years in the medical group ($P = 0.55$) and, overall, 28 (64%) were women (Table 1). The surgical patients had significantly worse NYHA functional status than patients in the medical group ($P = 0.0023$).

At echocardiography, atrial shunt was found in 68% of patients, with no significant difference between the two groups (Table 1). TR grade was significantly more serious in the surgical group ($P < 0.0001$). Echocardiography allowed the diagnosis of associated congenital heart defects in the surgical group (three right partial anomalous pulmonary venous return, one ventricular septal defect and one moderate hypoplasia of the left pulmonary artery) and in the medical group (one partial anomalous pulmonary venous connection, one unique coronary ostium and one dextrocardia).

Table 1 Patient characteristics.

	Surgical group ($n=20$)	Medical group ($n=24$)	*P*	Catheter subgroup ($n=7$)
Age (years)	34.3 ± 14.4	37.3 ± 16.9	0.55	48.2 ± 16.2
Women	13 (65)	15 (63)	NS	4 (57)
NYHA class	2.4 ± 0.6	1.7 ± 0.7	0.0023	2.0 ± 0.8
I	0	9 (38)	—	2 (29)
II	6 (30)	13 (54)	—	3 (43)
III	14 (70)	2 (8)	—	2 (29)
Other symptoms				
Cyanosis	7 (35)	6 (25)	NS	3 (43)
Asthenia	6 (30)	4 (17)	NS	1 (14)
Palpitations	12 (60)	10 (42)	NS	1 (14)
Angina pectoris	0	1 (4)	NS	0
Faintness/syncope	2 (10)	1 (4)	NS	1 (14)
Saturation at rest (%), mean	95.8	94	NS	90.5
Medical history of SVT				
Wolff Parkinson White	3 (15)	3 (13)	NS	1 (14)
Atrial	7 (35)	6 (25)	NS	4 (57)
Medical history of rhythmic procedure	2 (10)	6 (25)	NS	3 (43)
Electrical cardioversion	1 (5)	3 (13)		2 (3)
Radiofrequency	1 (5)	4 (17)[a]		1 (1)
Medical history of cardiac insufficiency	1 (5)	1 (4)	NS	0
Medical history of stroke	2 (10)	2 (8)	NS	1 (14)
Electrocardiogram				
Sinus rhythm	19 (95)	23 (96)	NS	6 (86)
Right bundle branch block	18 (90)	19 (79)	NS	6 (86)
First-degree atrioventricular block	3 (15)	5 (21)	NS	3 (43)
Pre-excitation	3 (15)	1 (4)	NS	0
Cardio-thoracic index, mean	0.58	0.59	NS	0.6
Haematocrit (%), mean	45.6	50.7	0.02	53.7
Tricuspid regurgitation grade	3.5 ± 0.7	2.3 ± 0.8	< 0.0001	2.6 ± 0.6
I	0	3 (13)	—	0
II	2 (10)	13 (54)	—	3 (43)
III	7 (35)	7 (29)	—	4 (57)
IV	11 (55)	1 (4)	—	0
Atrial shunt	13 (65)	17 (71)	NS	7 (100)
Other cardiac congenital anomaly	5 (25)	3 (13)	NS	0
Medical treatment	14 (70)	12 (50)	NS	5 (71)
Diuretics	4 (20)	2 (8)	NS	0
Anticoagulants (VKA or similar)	7 (35)	8 (33)	NS	5 (71)
Antiarrhythmic drugs	6 (30)	7 (29)	NS	4 (57)
Beta-blocker, ACEI	6 (30)	6 (25)	NS	3 (43)

Data are expressed as mean ± standard deviation or number (%) unless otherwise indicated. ACEI: angiotensin-converting enzyme inhibitor; NYHA: New York Heart Association; SVT: supraventricular tachycardia; VKA: vitamin K antagonist.
[a] Two Wolff Parkinson White, two flutter.

Early outcomes

Modified 'cone' repair was performed in all patients in the surgical group. Two patients required a second bypass for a significant residual TR at transoesophageal echocardiography (Tables 2 and 3), leading to a good functional result. Fifteen of 20 patients (80%) had trivial postoperative TR, while five patients had a moderate residual TR (Fig. 2A). The mean gradient after repair was 2.8 mmHg. Two patients (6.2—8.3%) had a gradient > 5 mmHg in the immediate postoperative period.

In terms of early complications related to the surgical procedure, two patients had postoperative heart failure that required mechanical ventilation, catecholamine support and nitric oxide. It was not necessary to use mechanical circulatory support in these patients. No severe surgical site

Table 2 Early complications after surgery or percutaneous catheter closure (within 30 days).

	Surgical group ($n=20$)	Catheter subgroup ($n=7$)
Failure of procedure	0	1 (14)
New cardiopulmonary bypass	2 (10)[a]	0
Extracardiac infection	1 (5)	0
Mediastinitis/endocarditis	0	0
Cardiac insufficiency	1 (5)	0
Supraventricular tachycardia	3 (15)	1 (14)
Atrioventricular block requiring pacemaker implantation	1 (5)	0
Stroke	0	1 (14)
Pericardial effusion requiring drainage	1 (5)	0
Blood cells transfusion	7 (35)	0

Data are expressed as number (%).
[a] Both immediately after the procedure.

Table 3 Events during follow-up.

	Surgical group ($n=20$)	Medical group ($n=22$)	*P*	Catheter subgroup ($n=6$)
Death	0	2 (9)	NS	1 (17)
Reintervention	2 (10)	3 (14)	NS	2 (33)
Mediastinitis	0	0	—	0
Endocarditis	0	1 (5)	NS	1 (17)
Supraventricular tachycardia	4 (20)	7 (32)	NS	5 (83)
Early first episode (< 2 months)	3 (15)	3 (14)		1 (17)
Late first episode (> 2 months)	1 (5)	0		0
Recurrence	2 (10)	4 (18)		4 (67)
Atrioventricular block requiring pacemaker	0	0	—	0
Stroke	0	1 (5)[a]	NS	1 (17)[a]
Parietal complications	6 (30)	—	—	—
Superficial infection	0	—	—	—
Scar disjunction	3 (15)	—	—	—
Keloid scar	2 (10)	—	—	—
Pain	2 (10)	—	—	—
Cardiac insufficiency	1 (5)	2 (9)	NS	1 (17)
Requiring hospitalization	0	2 (9)		1 (17)
Disease evolution[b]	3 (15)	8 (36)	NS	4 (67)
Hospitalization (all-cause)	3 (15)	6 (27)	NS	3 (50)

Data are expressed as number (%).
[a] Within 2 months.
[b] Recurrence of supraventricular tachycardia or occurrence ≥ 2 months after procedure, cardiac insufficiency, endocarditis, stroke or sudden death.

infection was observed in the surgical group. One patient had pericardial effusion and required surgical drainage.

At repair, no concomitant arrhythmia procedure was performed, as per the decision of the referring cardiologist. Two patients had postoperative rhythm disorders and one had a complete atrioventricular block requiring pacemaker implantation (Table 2).

Seven patients in the medical group underwent successful closure of an atrial shunt by Amplatzer prosthesis. In two of them, a moderate residual shunt was identified by transthoracic echocardiography at the end of examination. Another patient underwent percutaneous closure, but the procedure failed due to insufficient support for the prosthesis (Table 2). No in-hospital procedural complications were reported in the catheterization subgroup.

Late outcomes

The median (range) durations of follow-up were 2.8 (0.5—5.0) years in the surgical group and 4.8 (3.6—5.0) years in the medical group. Two patients of the medical group were lost to follow-up (one of whom was in the catheterization subgroup), but none in the surgical group. The overall complication rates associated with the disease were 15% in

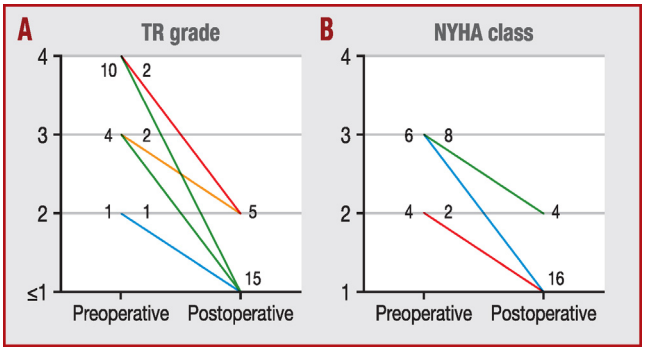

Figure 2. Comparisons of (A) New York Heart Association (NYHA) class and (B) tricuspid regurgitation (TR) grade before and at last visit in the surgical group ($n = 20$).

the surgical group and 36% in the medical group. After discharge, no patients died in the surgical group, whereas two patients (5.7%) in the medical group died during follow-up: one sudden death 10 days after the ASD closure and one stroke (1 month after the cardiac catheterization process) (Table 3).

In the surgical group, at last visit, 18 patients (90%) had improved NHYA functional class (the two patients preoperatively in class II remained in class II) (Fig. 2B). No surgical patients displayed any postoperative cyanosis. Two of three patients suffering from asthenia were improved and eight of 12 patients reporting palpitations were not suffering these after the operation.

In the surgical group, three patients (15%) developed de novo atrial tachycardia (flutter) within the first 2 months (Table 3). Two of them underwent percutaneous radiofrequency ablation and one underwent electrical cardioversion.

In the catheterization subgroup, residual shunts described evolved to complete closure within 6—12 months. Two patients in the catheterization subgroup required reintervention (Table 3): one right heart failure occurred 5 years after the procedure and required surgery for tricuspid valve replacement in another institution; and one patient developed a late confirmed infectious endocarditis on the pacemaker leads, 3 years after ASD closure, and needed a surgical procedure. The existence of a residual shunt (due to a dehiscence of the edges of the prosthesis) meant that the device had to be extracted and the ASD closed by a pericardial patch. In this procedure, which was carried out at our centre, no valve repair was performed.

By subjective functional overall assessment, 18 surgical patients (90%) 'felt better' after surgery and 19 patients (95%) were 'satisfied' with the global surgical management (Table 4).

Table 4 Questionnaire evaluation of functional status after surgery.

	Surgical group ($n = 20$)
Amelioration	
Much better	12 (60)
A little better	6 (30)
Unchanged	1 (5)
A bit worse	1 (5)
Much worse	0
Satisfaction	
Very satisfied	8 (40)
Quite satisfied	11 (55)
Not really satisfied	1 (5)
Not satisfied at all	0

Data are expressed as number (%).

Discussion

The present study analysed 20 consecutive adults who underwent surgery for Ebstein's disease. All surgical patients benefited from native valve preservation. Although two patients (10%) required a second bypass run before leaving the operating room for residual TR, the immediate success rate was very high, regardless of disease severity.

Tricuspid reconstruction in an adult population using the 'cone' concept seems to be a surgical procedure with encouraging early and medium outcomes in terms of functional improvement, morbidity and mortality. In our series, tricuspid 'cone' reconstruction was associated with no mortality, a moderate percentage of specific complications and an excellent success rate. It resulted in significant improvement in functional status compared with medical treatment, with a significant degree of patient satisfaction.

Several studies corroborate the improvements obtained with the surgical management of Ebstein's patients. In the largest series of patients operated on for Ebstein's anomaly, published by the Mayo Clinic, 539 patients (mean age 24.1 ± 18.1 years) underwent tricuspid valve surgery [8,13]. Mortality at 30 days was 4.9% after tricuspid valvuloplasty and 6.8% after valve replacement. Carpentier's group reported on a series of 191 patients (mean age 24.4 ± 15 years) [14]. They reported significant immediate postoperative mortality due to right heart failure, but this decreased from 12% to 7% by systematically adding a bidirectional cavopulmonary derivation. da Silva et al. presented a series of 52 patients who underwent 'cone' reconstruction [15], with mortality of the procedure reduced to 3.8%. In the latest study published by the Mayo Clinic, mortality related to cone reconstruction was reduced to 1.0% [16].

Regarding the complications before hospital discharge (within 30 days), arrhythmias are the most frequent in all series. However, postoperative heart failure still remains a real challenge, causing 100% of the immediate deaths in the last Mayo Clinic series [16], 75% in da Silva's series [15] and 50% in Carpentier's group [14]. Mechanical circulatory support must be added to drug support and nitric oxide when these are insufficient. In our series, two patients (10%) developed an episode of heart failure (mainly right) requiring mechanical ventilation, catecholamine and nitric oxide support, but without need of mechanical circulation support or new surgical procedures.

There are two possible explanations for the low incidence of heart failure in our study. Firstly, the modified 'cone' reconstruction technique with the principle of closure of the ASD produced good functional results. Secondly, we must take into account the characteristics of our cohort, which

included adults with no previous history of severe heart failure episodes (despite a severe mean TR grade) and only two patients with a cardio-thoracic index ≥ 0.65. To predict heart failure, reliable tools for estimating right ventricular function preoperatively need to be found, covering systolic function and, more importantly, ventricular compliance. Magnetic resonance imaging and three-dimensional echocardiography are now used for this objective [17,18].

Finally, regarding the complications of atrialized ventricle plication, several authors have reported a risk of postoperative ischaemia (0.4% postoperative myocardial infarction in a series from the Mayo Clinic) by 'kinking' of the right coronary artery [19]. Neither myocardial infarction nor ventricular tachycardia was reported in our series, taking care to make this plication after analysis of coronary anatomy and with superficial endocardial points.

In adults, a significant discrepancy between the annular size and the surface of the available leaflet tissue is not uncommon. On the other hand, despite this inconvenient, restrictive leaflet opening, and also direct anterior leaflet insertion into the right ventricle can be observed. In this population, insistence on conventional 'cone' repair can require excessive annular plication and can also result in restrictive leaflet opening. The use of a prosthetic ring and leaflet augmentation (Fig. 1B) can be helpful to obtain good functional results despite these unfavourable anatomical factors.

The advantages of the 'cone' technique are numerous. The new tricuspid valve is reconstructed in the anatomical plane which, combined with longitudinal plication, brings the right ventricle to a sub-normal morphology. Then, valvular coaptation is done, more likely with two leaflets, reducing the risk of residual regurgitation. The use of a prosthetic ring also has the advantage of annular reinforcement, which reinforces repair and, eventually, prevents dehiscence. Finally, we did not observe severe tricuspid stenosis at hospital discharge. Nevertheless, we must remember that every patient was at least 15 years old.

ASD closure was always performed. However, one patient received a concomitant bidirectional cavopulmonary shunt for a postoperative left heart preload failure suspected during the intervention.

Functional improvement

In our study, 80% of patients were in NYHA class I and 20% in NYHA class II after surgery, significantly better than before surgery ($P < 0.0001$), which showed a good correlation between cardiac and functional status. The results of our study also showed an improvement in overall subjective functional assessment in 18 of 20 patients (90%).

Unfortunately, these excellent results could not be confirmed by objective tests, e.g. peak exercise oxygen consumption, as these were only available for a low percentage of patients. Patients were referred to our centre for surgery from numerous different hospitals and it is very difficult to develop standardized and comparable preoperative and follow-up examinations.

Place of interventional catheterization

The indications for catheter closure of ASD are limited. It should be remembered that the existence of a right-to-left shunt reflects low right ventricular compliance. There is, therefore, a risk of worsening or onset of subsequent congestive heart failure. In addition, although patients initially feel a symptomatic improvement because of the correction of hypoxaemia, the procedure does not correct TR and cannot improve long-term prognosis. It seems that percutaneous ASD closure to relieve cyanosis in the Ebstein's setting is not recommended. The remaining indications for percutaneous closure are probably the rare cases of documented paradoxical embolism with reduced Ebstein's anomaly without functional impact on the right ventricle.

Study limitations

The present study included a cohort of adult patients presenting with Ebstein's anomaly analysed in two non-randomized groups: medical and surgical. There are fundamental differences in the baseline characteristics of these two groups. The 20 severe patients underwent surgical repair during the study period. The remaining patients were not considered for surgical procedure because of their functional status, degree of TR and the referring cardiologist's decision. This study design, along with the small size of the cohort, meant that statistical comparisons between the two groups were not appropriate.

The cone reconstruction technique is the first technique to approach the restoration of a normal right heart anatomy. The annulus returned to its anatomical position and the cone restored a sub-normal filling flow. We could suppose that these new morphologic conditions can lead to a better right heart remodelling, restoring an almost normal right ventricular volume, and may also permit an improvement in right ventricular compliance. The decrease of the incidence of complications, preventing the occurrence of arrhythmias and right heart failure, may be suspected. Unfortunately, our present results, including the comparison with the medical group, do not provide firm conclusions. The number of surgical patients was small and the follow-up period does not exceed the mid-term. Adding to the good immediate results, the benefits of this anatomical reconstruction technique would probably be more visible during a longer follow-up and in larger studies. However, long-term follow-up data are missing, so while we would like to propose this low-risk surgery in asymptomatic adult Ebstein's patients presenting with TR, longer follow-up data are required.

Conclusion

Given the very low morbi-mortality and the high success rate of repair (regardless of anatomical form) afforded by experienced teams, we believe that a modified 'cone' reconstruction should be proposed for every symptomatic Ebstein's patient. Although the benefit of surgery on the long-term evolution of Ebstein's anomaly in adults has not yet been precisely determined, this short series leads us to propose an intervention on the tricuspid valve in all patients

presenting more than moderate regurgitation before they progress to the stage of decompensated heart failure.

Tricuspid 'cone' reconstruction seems to be the surgical procedure that has given the best results in terms of functional improvement and morbidity and mortality in the short and medium term. In our series, modified 'cone' repair was associated with low mortality, a low percentage of specific complications and an excellent success rate. It allowed a significant improvement in functional status in comparison to medical treatment, with a significant degree of patient satisfaction.

We would suggest surgery for all patients with moderate or severe TR and, particularly, we observed that percutaneous ASD closure, despite reducing cyanosis, should be considered with extreme caution.

Developing tools for accurate and reproducible prognostic evaluation and assessing the potential benefit in terms of reduction in the incidence of arrhythmias and heart failure warrant a larger series and a long-term follow-up.

After the study period, eight additional adults have undergone the cone procedure for Ebstein's repair, one of whom had already undergone a failed tricuspid valve repair attempt. No mortality occurred and, at discharge, all patients had mild or less residual regurgitation. With this increasing experience, the need for valvular patch augmentation — as well as prosthetic annuloplasty — were not required.

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Disclosure of interest

The authors declare that they have no competing interest.

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