Reconstructive surgery for Ebstein anomaly: three decades of experience

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ABSTRACT

Objectives

Since 1988, our centre employs vertical plication repair with deattachment and reattachment of the tricuspid valve for Ebstein anomaly. This study describes the characteristics and long-term outcomes of our single-centre cohort.

Methods

Data from all patients operated on between 1988 and 2016 were retrospectively collected. Kaplan–Meier analyses were done for survival data and mixed models were used to analyse longitudinally collected clinical and echocardiography data.

Results

Thirty-six patients (mean age: 25.4 ± 15.9 years, 36% male) were operated on using the Carpentier–Chauvaud 21 (58%) or Cone repair 15 (42%). One patient (3%) died in hospital. Two late deaths were observed, yielding a survival of 97 ± 3% at 25 years. Reoperation was performed in 6 patients after a mean follow-up of 14.1 ± 10.3 years, resulting in a freedom of reoperation of 80 ± 8% at 25 years. During follow-up, predicted probability of being in New York Heart Association III/IV did not exceed 10%. Modelling longitudinal evolution of tricuspid regurgitation showed no major changes over time. Additionally, a rigid ring repair was associated with a higher probability of tricuspid regurgitation, especially after the first years after the operation. A full Cone repair was associated with less progression of tricuspid regurgitation over time.

Conclusions

Repair of Ebstein anomaly is associated with low mortality and morbidity, acceptable reoperation rate and excellent valve function over time, especially in patients with completed Cone repair. Therefore, we conclude that in our centre, repair of Ebstein abnomaly is a durable technique to treat patients.

INTRODUCTION

Ebstein's anomaly is a rare congenital heart disorder first described by Wilhelm Ebstein in 1866 and is characterized by apical displacement of the effective tricuspid valve (TV) orifice, resulting in tricuspid regurgitation (TR) and atrialization of the right ventricle (RV) [1]. The estimated prevalence of Ebstein's anomaly is 0.47 cases per 100 000 people [2]. The severity of Ebstein's anomaly varies from a mild phenotype with limited TV displacement and free moving anterior leaflet (type A) to extensive RV atrialization except for a small infundibular component (type D), as classified according to Carpentier classification [3].

Since 1988, our centre has adopted a repair technique consisting of detachment of the TV leaflets, a vertical plication of the arterialized RV and reattachment of the leaflets in the neo-annulus as described by Carpentier et al. [3] and introduced in our clinic by Chauvaud. In this study, we present our 30-year experience with this repair technique.

METHODS

Patients

All 36 patients operated on at our institution between January 1988 and November 2016 with the Carpentier–Chauvaud–Cone technique for Ebstein's anomaly were analysed retrospectively [3]. In addition, 1 patient had a tricuspid valve replacement at initial surgery, due to limited leaflet tissue, which prohibited adequate repair. Four patients received a univentricular approach directly, due to severe Ebstein and impaired right ventricle function (RVF) and 1 patient only received an atrium septum defect (ASD) closure and bidirectional cavopulmonary connection. Approval of the local Medical Ethics Committee was obtained to conduct this study (MEC-2017-384). If follow-up was done in other hospitals, patients were contacted and consent was obtained to request data of interest in these centres.

Indication for operation

The main indication for operation consisted of complaints of progressive dyspnoea, beyond the stage of mild symptoms and characterized by higher New York Heart Association (NYHA) class. Other indications were progressive exercise intolerance, repeated cerebral vascular events and exercise-induced cyanosis.

Operation technique

Carpentier et al. [3] and Quaegebeur et al. [4] described the operation technique previously in great detail. Median sternotomy was performed with cannulation in the ascending aorta and both caval veins. With mild hypothermia and cardioplegic arrest, a right atriotomy was

performed. The severity of Ebstein's anomaly was classified according to the Carpentier classification [3]. Types B, C and D were found in 11, 23 and 2 patients, respectively.

Thereafter, an incision was made in the enlarged anterior TV leaflet starting at the anteroseptal commissure, extending the incision in a clockwise fashion, if possible also into the posterior leaflet. In most cases, the septal valve leaflet was rudimentary and could not be detached. Delaminating apically, the muscular and trabecular connections between the right ventricular wall and leaflet tissue were dissected, taking care not to perforate the right ventricular free wall. Thereafter, the atrialized part of the RV was longitudinally plicated, as described by Quaegebeur et al. [4]. The detached leaflets were rotated clockwise and sutured back in the neo-annulus. Since 1988, we have tried to create a full 360-degree fit of the TV leaflets on the neo-annulus; however, in most cases, this was not possible with the available leaflet material without inducing stenosis. Hence, a subsequent residual TR was accepted. Following the reported favourable outcomes after Cone reconstruction [5], we systematically intended to create the full 360-degree fit to complete the Cone repair, suturing the posterior valve leaflet and (rudimentary) septal valve leaflet. Nowadays, we see the Cone repair as the preferred end point at Carpentier–Chauvaud repair. A ring was implanted in adult patients when considered to provide additional annular support and when the annular plication and the rotation of the leaflets resulted in a neo-annulus with a configuration that fitted with the available standard rings. The final decision as to whether or not to apply a TV ring was made upon the surgeon's discretion. A bidirectional cavopulmonary connection was performed in case of anticipated impaired right ventricular function in order to attempt to unload the RV. The risk of right ventricular impairment after surgery was more frequently deemed higher in case of a reoperation. The final decision whether or not to perform a bi-directional cavopulmonary connection was made upon the surgeon's discretion. All ASDs were closed, preferably primary.

Postoperative care

Patients received coumarins the first 3 months after operation. When patients were in sinus rhythm with adequate RVF, coumarins were stopped after 3 months. Coumarins were continued when indicated. Standard echocardiography was done at the end of operation, at discharge and at regular outpatient visits.

Follow-up

All patients were followed up in the outpatient clinic by paediatric cardiologists (patients <18 years) and congenital cardiologists (patients >18 years). Relevant events, rhythm status, functional status, echocardiograms and medication were collected longitudinally. Clinical events that were collected were implantable cardioverter defibrillator (ICD)/pacemaker implant, cardioversion, thromboembolism, bleeding, endocarditis, myocardial infarction and admission for heart failure. Vital status was checked in the civil registry. Patients were censored at tricuspid valve replacement, death, loss of follow-up and end of follow-up.

Statistical analysis

Continuous data are presented as mean ± standard deviation, if normally distributed, and as median with interquartile range (IQR), if not normally distributed. Categorical data are presented as percentages. Survival data are presented as Kaplan–Meier estimates with the accompanying standard error or in case of Kaplan–Meier plots, with 95% confidence interval (CI). TV gradient was analysed using mixed models with random intercepts for patients and random slope for time. Dichotomous data were modelled using generalized mixed models with random intercepts for patients. Natural splines for time were added to establish flexibility. All the models only contained time, one other covariate and their interaction term to prevent overfitting. In the generalized mixed model, the marginal probabilities were obtained using a Monte Carlo sampling procedure. For each combination of follow-up time and covariate of interest, 3000 patients were generated with random effect values coming from the normal distribution $N(0, \sigma_b^2)$, where σ_b^2 denotes the estimated variance of the random effects from the model. The mean of the 3000 calculated probabilities was taken as estimate.

Univariable logistic regression was performed to find determinants associated with concomitant ring implant as *post-hoc* analysis. All analyses were done in R (Version 3.3.3) using the 'glm', 'survival' and 'lme4' statistical packages.

RESULTS

Preoperative characteristics

Preoperative characteristics are presented in Table 1. In the 2 patients who underwent prior cardiac surgery, an ASD closure was performed in both patients, one of whom underwent an ASD closure twice. Seven patients had prior episodes of arrhythmias. Supraventricular tachycardia in 5 patients, of whom 1 had an ablation for re-entry tachycardia. The other 4 were treated medically. Another patient was diagnosed with Wolf–Parkinson–White syndrome unsuccessfully treated by an ablation and 1 patient had atrial fibrillation (AF) with sinusbradycardia and subsequent pacemaker implantation. In 20 patients, cor/thorax ratios on X-ray were recorded with a mean of 0.58 ± 0.06.

Procedural characteristics

Median time from diagnosis to operation is 10.3 years (IQR 5.0–12.5). Procedural characteristics are presented in Table 1. In case of a ring implant, a rigid Carpentier-Edwards ring was implanted (subtype of ring: 9 classical and 1 physio). In the 3 patients who underwent concomitant partial cavopulmonary connection, 2 also underwent ring implantation and 1 underwent a Cone reconstruction.

Hospital outcomes

One patient (3%) died 2 days after surgery due to right ventricular failure and ventricular fibrillation unresponsive to therapy. Autopsy revealed an infarction of the RV, which was not caused by the sutures. Rethoracotomy was performed in 2 patients, 1 for bleeding and 1 for suspected cardiac tamponade. Hospital morbidity consisted of episode of ventricular fibrillation in 2 patients (treated by electrical defibrillation), cardiac tamponade in 3 patients (treated by pericardiocentesis in 2) and a pacemaker implant in 1 patient. Median hospital stay is 11 days (IQR 8.5–12.5) and length of intensive care unit stay was reported in 18 patients with a median of 2 days (IQR 2–3).

Table 1: Baseline and operative characteristics

Preoperative characteristics	
n	36
Age[a] (years), mean ± SD	25.4 ± 15.9
Male gender, *n* (%)	13 (36.1)
BMI (kg/m^2), mean ± SD	20.9 ± 4.1
Carpentier classification, *n* (%)	
A	0 (0)
B	11 (31)
C	23 (64)
D	2 (6)
NYHA, *n* (%)	
I	4 (11)
II	12 (33)
III	13 (36)
IV	5 (14)
Unknown	2 (6)
Prior cardiac surgery, *n* (%)	2 (6)
Concomitant ASD,[b] *n* (%)	26 (72)
Concomitant VSD,[b] *n* (%)	2 (6)
Prior arrhythmia, *n* (%)	7 (19)
Prior CVA, *n* (%)	4 (11)
Elevated CVP,[c,d] *n* (%)	6 (17)
Ascites,[c] *n* (%)	0 (0)
Leg oedema,[c] *n* (%)	0 (0)
Hepatomegaly,[c] *n* (%)	5 (14)
Digital clubbing,[c] *n* (%)	5 (14)
Cyanosis,[c] *n* (%)	20 (56)
Diuretics, *n* (%)	1 (3)
Anticoagulation, *n* (%)	4 (11)

Table 1: Baseline and operative characteristics (continued)

Preoperative characteristics	
Digoxin, *n* (%)	2 (6)
Rhythm, *n* (%)	
Sinus	32 (89)
AF	3 (8)
Paced	1 (3)
TR grade, *n* (%)	
None/mild	0 (0)
Moderate	4 (11)
Severe	31 (86)
Unknown	1 (3)
RVF impairment, *n* (%)	
None	26 (72)
Moderate	9 (25)
Severe	1 (3)
Intraoperative characteristics	
CPB time (min), median (IQR)	117 (103–144)
ACC time (min), median (IQR)	88 (73–109)
Concomitant surgery, *n* (%)	
ASD closure	29 (81)
VSD closure	1 (3)
PCPC	3 (8)
Tricuspid ring implant	10 (28)
Cone repair	15 (42)
MAZE	1 (3)

[a]Range (2–58 years).

[b]Observed on echocardiography or during heart catheterization.

[c]Observed at physical examination during the last recorded visit prior to operation.

[d]Diagnosed by visual inspection of external jugular vein.

ACC: aortic cross-clamp; AF: atrial fibrillation; ASD: atrium septum defect; BMI: body mass index; CPB: cardio pulmonary bypass; CVA: cerebral vascular accident; CVP: central venous pressure; IQR: interquartile range; NYHA: New York Heart Association; PCPC: partial cavopulmonary connection; RVF: right ventricle function; SD: standard deviation; TR: tricuspid regurgitation; VSD: ventricular septum defect.

Late clinical outcomes

In total, 500 clinical follow-up moments were recorded during follow-up, with a mean follow-up of 14.1 ± 10.3 years (completeness = 92%). Six patients were partly followed in other centres. Late mortality occurred in 2 patients at 28.6 years and 29.0 years after surgery. Cause of death was non-cardiac in 1 patient and unknown in 1 patient. Overall, survival was 97 ± 3% at 25 years post-surgery. Thromboembolism occurred in 2 patients (1 cerebral and 1 non-cerebral) at 5.2 years and 14.2 years after surgery, yielding a freedom of thromboembolism of 100 ±

0%, 96 ± 4% and 90 ± 7% at 5, 10 and 25 years, respectively. One patient, who was prescribed anticoagulation due to AF, had 2 episodes of epistaxis, which required hospital care at 8.8 years and 9.3 years after surgery. No admissions for heart failure were recorded. Six patients had an episode of NYHA functional class III or IV. Of these 6 patients, 3 were eventually reoperated on. Longitudinal evolution of the probability of being in NYHA class 3 or 4 for an 18-year-old and a 50-year-old patient is presented in Fig. 1.

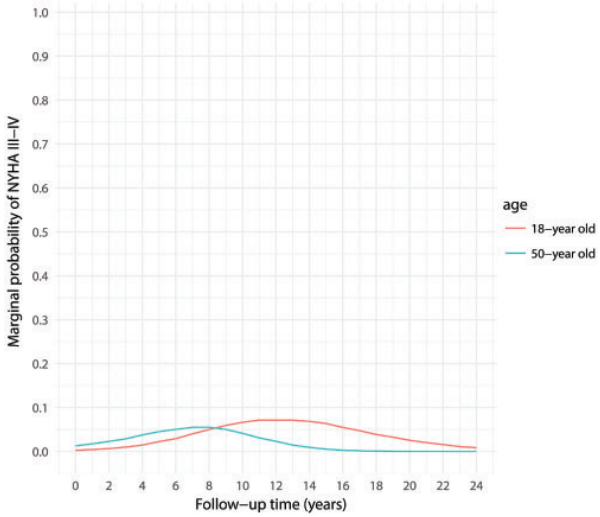


Figure 1: Marginal probability of being in functional class 3 or 4 in an 18-year-old patient and a 50-year-old patient. NYHA: New York Heart Association.

Arrhythmias

In 21 patients, 62 (mean 2.9) episodes of complaints of palpitations were recorded during follow-up, and 5 patients had a subsequent ablation. Ten patients had an electrical cardioversion for supraventricular tachycardia, including AF. In 6 patients, a pacemaker (4) or ICD (2) was implanted during follow-up. Indications for pacemaker implant were atrioventricular block in 3 patients and sinus arrest combined with atrioventricular block in 1 patient. Indications for ICD implant were non-sustained ventricular tachycardia and out-of-hospital cardiac arrest caused by ventricular tachycardia. Freedom of palpitation, cardioversion, ablation and pacemaker/ICD implants are presented in Fig. 2A–D. During follow-up, 13 patients presented with documented arrhythmias, resulting in a freedom of arrhythmia of 73 ± 8%, 64 ± 9% and 53 ± 11% at 5, 10 and 25 years, respectively. The longitudinal evolution of the probability of sinus rhythm versus no sinus rhythm (e.g. AF or paced rhythm) of an 18-year-old patient and a 50-year-old patient is presented in Fig. 3.

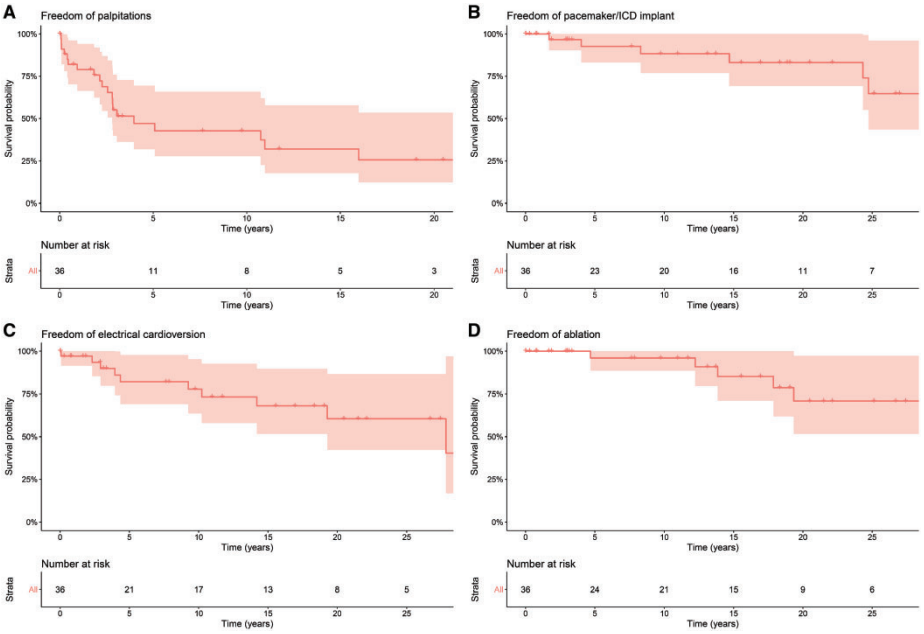


Figure 2: Kaplan–Meier plots of freedom of palpitations (A), pacemaker/ICD implant (B), electrical cardioversion (C) and ablation (D). ICD: implantable cardioverter defibrillator.

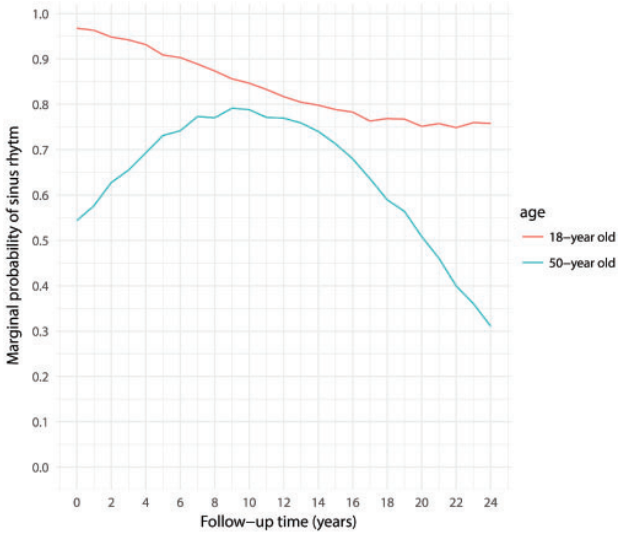


Figure 3: Marginal probability of sinus rhythm in an 18-year-old patient and a 50-year-old patient.

Reoperation

Six patients required 8 reoperations (Table 2). In patients with a tricuspid valve replacement, a St-Jude Medical mechanical valve was implanted. Freedom of reoperation was 88 ± 6%, 80 ± 8% and 80 ± 8% at 5, 10 and 25 years, respectively (Supplementary Material, Fig. S1).

Table 2: Summary of performed reoperations

Patient	First reoperation		Second reoperation	
	Interval (years)	Procedure	Interval (years)	Procedure
1	9.8	Ring implant and PCPC		
2	9.0	TV repair and PCPC		
3	1.6	Re-reconstruction plication, TV repair and ring implant		
4	0.28	TV repair and further detachment of chordal support	0.32	TVR with mechanical prosthesis
5	0.6	TV repair and PCPC	3.0	TVR with mechanical prosthesis and MAZE
6	0.02	TVR with mechanical prostheses		

PCPC: partial cavopulmonary connection; TV: tricuspid valve; TVR: tricuspid valve replacement.

Echocardiography outcomes

During mean follow-up of 14.4 ± 9.8 years, 448 echocardiograms were recorded in 34 patients (mean: 13.2 echocardiograms, range 1–32). Transoesophageal echocardiography directly after the procedure showed a significant reduction (none: 4, trivial/mild: 21, moderate: 5, severe: 3 patients) in TR compared to before surgery ($P < 0.001$). In the 6 patients who required a reoperation, TR grade was severe in 5 patients and moderate in 1 patient on last echocardiogram prior to reoperation. The longitudinal evolution of TV inflow gradient is presented in Fig. 4A, with separate lines for patients receiving a tricuspid ring repair versus repair without a ring. Having a ring repair was not significantly associated with higher overall TV gradient ($P = 0.58$), nor with different changes over time. A higher gradient was not associated with significant TR [odds ratio $(OR)_{per\ 1mmHg}$ 1.03, 95% CI 0.97–1.1; $P = 0.30$].

In Fig. 4B, the evolution of the marginal probability of significant TR is presented. Having a ring as part of the repair was associated with a significantly higher probability of clinically important TR ($P < 0.001$). The determinants associated with ring implantation were older age ($OR_{per\ 1\ year}$ 1.06, 95% CI 1.01–1.03; $P = 0.021$) and a higher systolic blood pressure ($OR_{per\ 1\ mmHg}$ 1.07, 95% CI 1.00–1.14; $P = 0.019$) and preoperative moderate RV impairment (OR 6.89, 95% CI 1.27–37.3; $P = 0.025$).

Carpentier class was not associated with higher probability of moderate-to-severe TR over time ($P = 0.85$) (Supplementary Material, Fig. S2). However, the full Cone repair showed a more

durable result, especially in the long term (Fig. 5). In 6 patients who underwent the full Cone repair, a ring was used.

Ten patients had an episode of severe RV dysfunction on echocardiography, resulting in a freedom of RV dysfunction at 5, 10 and 25 years of 83 ± 7%, 74 ± 9% and 56 ± 12%, respectively. We did not analyse the RV dysfunction longitudinally, because the missing data were highly skewed throughout the years, with more missing data in the early days (Supplementary Material, Fig. S3).

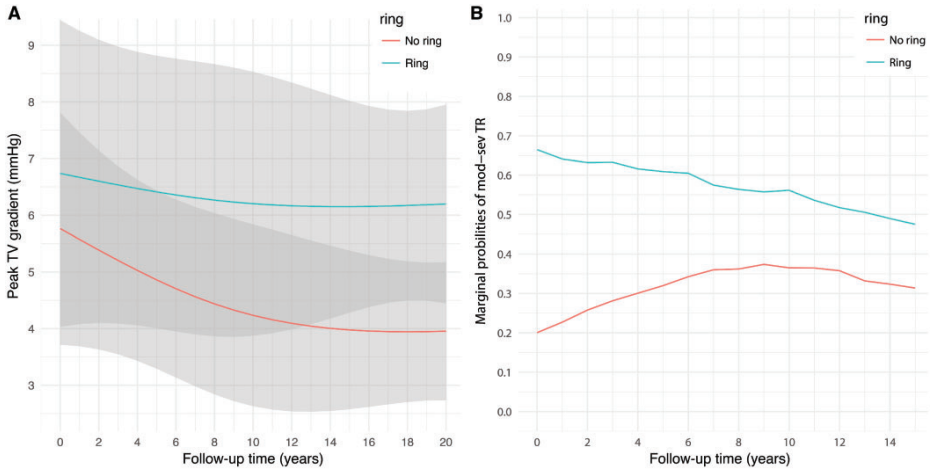


Figure 4: Longitudinal evolution over time of TV peak gradient stratified to a ring implant (A). Marginal probability of moderate-to-severe TR stratified to ring implant (B). TR: tricuspid regurgitation; TV: tricuspid valve.

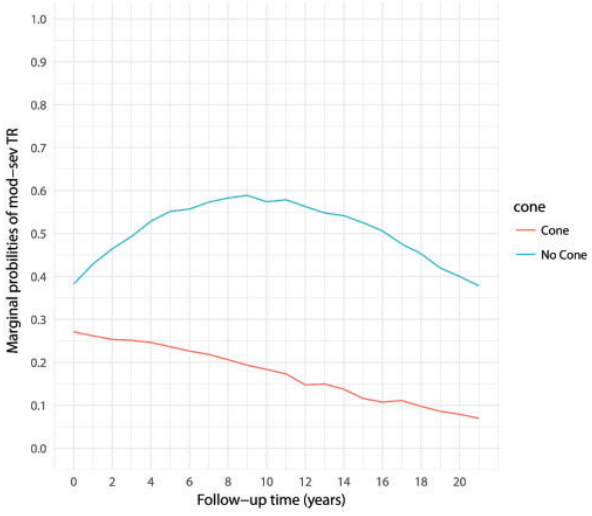


Figure 5: Marginal probability of moderate-to-severe TR stratified to cone repair. TR: tricuspid regurgitation.

Diuretic use

During follow-up, 15 patients were prescribed any form of diuretics. The longitudinal evolution of the probability of prescribed diuretics of an 18-year-old patient and a 50-year-old patient is presented in Supplementary Material, Fig. S4A and B. Seven patients were prescribed furosemide with a mean starting dose of 30 ± 10 mg/24 h (range 20–40 mg). Mean dose at last follow-up was 34.3 ± 9.8 mg/24 h (range 20–40 mg).

DISCUSSION

Ebstein's anomaly has a large variation in TV and right ventricular morphology [6]. Hence, numerous techniques to repair the TV in Ebstein's anomaly have been described [2–4, 7–9]. This study presents a detailed overview of both long-term clinical and echocardiographic outcome after the Carpentier–Chauvaud and Cone repair. As part of continuous evaluation, we have extended our previous reports in terms of the number of patients, the duration of follow-up, and in analysis with advanced statistical methods [10].

Clinical outcomes

Both hospital mortality and morbidity were low in this cohort, which is sustained during follow-up, with an overall survival estimate of 97% at 25 years. These rates are comparable to other series. Brown et al. [11] reported an actuarial survival of 80% at 20 years (337 patients), in which early deaths are excluded and Hetzer et al. [12] reported an actuarial survival of 91% at 20 years (68 patients). Furthermore, these results are very much comparable to other series (Supplementary Material, Table S1).

Functional NYHA III–IV was noted in 6 patients, indicating the overall favourable functional status. Additionally, the predicted probability of being in NYHA class III or IV remains low over time for both 18-year-old and 50-year-old patients, further establishing the low risk of being in NYHA class III/IV during follow-up.

Six patients were reoperated on in this series, yielding a freedom of reoperation of 80% at 25 years. Other studies report actuarial reoperation rates of 73% at 15 years [11]. Da Silva et al. [13] reports 4 reoperations in 52 patients after at least a 10-year follow-up. In this cohort, all patients underwent Cone reconstruction.

Echocardiographic outcome

Assessing the durability of the Carpentier–Chauvaud and Cone repair only by looking at reoperation rates would result in incorrect conclusions, since the reoperation risk may be deemed too high in some patients, and severe residual/recurrent TR or high inflow TV gradient is accepted. Therefore, we modelled the TV gradient and probability of moderate-to-severe TR in order to visualize longitudinal evolution over time.

As expected, TV gradient gradually declined over the years. This may be explained by the fact that the TV annulus dilates throughout the years, resulting in larger TV orifices, and a subsequently lower gradient. A ring implant was associated with a slightly, but not significantly, higher gradient, which was more stable over time.

TR did not increase drastically over the years, indicating a durable repair. However, surprisingly, we found that a ring implant was significantly associated with a higher probability of TR, which is contradictory to the literature [14]. Since only univariable models were used, these findings could be explained by confounding. Therefore, determinants associated with ring implant were explored using univariable logistic regression. An older age, systolic blood pressure and moderate RV impairment were found to be associated with a ring implant. The latter may explain the higher probability of TR in patients with a concomitant ring implant, as RVF impairment is associated with RV dilatation. A heavily dilated RV can result in tethering of the TV, subsequently causing TR. Additionally, confounding by indication may be present, as the choice to implant a ring may be motivated by that fact that the repair is not satisfactory and a ring is implanted to attempt to improve the repair.

When a ring was used, a Carpentier-Edwards rigid ring was applied in all patients. These rings have a predefined shape, designed for a normal TV annulus. We hypothesized that the TV annulus of patients who underwent Carpentier–Chauvaud–Cone repair does not have a normal annulus shape, even if the geometry after the repair resembles the normal annulus shape. In fact, forcing the annulus into the ring-shape might lead to deformation of the created neo-annulus, resulting in malcoaptation of the leaflets and subsequent TR. This also may explain why Dearani et al. [14] found a favourable association with concomitant ring implant, because in this cohort, only flexible rings are used.

Patients with Ebstein's anomaly have a higher risk of arrhythmias compared to the general population [15, 16]. Not surprisingly, the incidence of symptomatic palpitations in this cohort was relatively high. However, a patient may have 1 short episode of AF and undergo cardioversion and live out his remaining life in sinus rhythm [17]. Simply analysing freedom of AF could convey an inaccurate message. Therefore, we collected rhythm status serially and used mixed modelling to analyse these repeated measurements. Using these models, one can visualize the probability of being in sinus rhythm over time with effect plots, as is shown in Fig. 2. Effect plots of the longitudinal evolution of the probability of sinus rhythm of an 18-year-old patient and a 50-year-old patient showed different patterns. In the 18-year-old patient, the risk of having arrhythmia was relatively low after the surgery and this risk gradually increased over time (Fig. 3). In the 50-year-old patient, the risk of arrhythmia was relatively high after the surgery, but this risk decreased during follow-up. When patients become older, the probability of being in sinus rhythm decreases significantly. These different patterns underlie the notion that subject-specific outcome modelling is needed, especially in patients with Ebstein's anomaly, which is known to be very heterogeneous [6].

Strengths and limitations

Strengths of this study include the length of follow-up and the number of follow-up moments. We managed to obtain 500 clinical follow-up moments and 448 echocardiograms. This is mainly due to the fact that patients with congenital heart disease are closely monitored in specialized centres in the Netherlands. The abundant follow-up moments enabled us to use advanced statistical methods to visualize outcomes over time. A limitation is the relatively small single-centre sample size, which limits the use of multivariable modelling, allowing confounding factors to influence results. Other limitations include the possible recall bias in complaints of palpitations and misclassification of NYHA functional class and TR grade. However, we dichotomized these variables to create a more robust measurement, accepting the loss of information paired with dichotomization. Additionally, we did not have magnetic resonance imaging data and echocardiographic assessment of RVF was mostly missing in the early days. In these days, RVF was only reported, if impaired. This prevents longitudinal analyses since the data are not missing at random. Moreover, assessment of right ventricular function is already a semi-quantitative measure, which is expected to change over time. Lastly, the mechanism of the TR (i.e. annular dilation or tethering) was not reported.

CONCLUSIONS

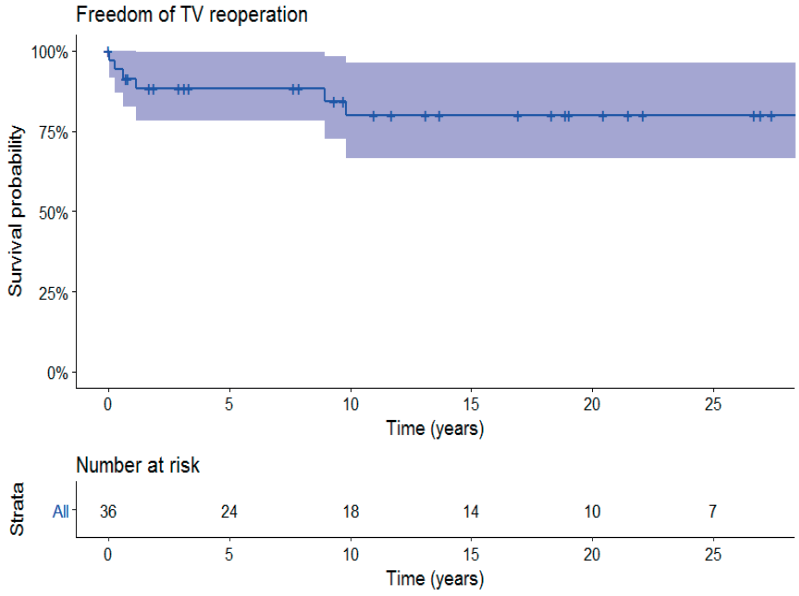
Reconstructive valve surgery for Ebstein is associated with low mortality and morbidity, acceptable reoperation rates and excellent valve function of the TV beyond the first 2 postoperative decades, especially in patients with completed Cone repair. Therefore, we conclude that the Carpentier–Chauvaud (preferably extended into Cone) technique is a durable repair technique for patients with Ebstein's anomaly. When the repair included the use of a rigid ring, more TR was observed. Further studies are needed to evaluate whether this was caused by the rigid ring itself or whether confounding or selection bias was present.

REFERENCES

1. van Son JA, Konstantinov IE, Zimmermann V. Wilhelm Ebstein and Ebstein's malformation. Eur J Cardiothorac Surg 2001;20:1082–5.
2. Boyle B, Garne E, Loane M, Addor MC, Arriola L, Cavero-Carbonell C et al. The changing epidemiology of Ebstein's anomaly and its relationship with maternal mental health conditions: a European registry-based study. Cardiol Young 2017;27:677–85.
3. Carpentier A, Chauvaud S, Mace L, Relland J, Mihaileanu S, Marino JP et al. A new reconstructive operation for Ebstein's anomaly of the tricuspid valve. J Thorac Cardiovasc Surg 1988;96:92–101.
4. Quaegebeur JM, Sreeram N, Fraser AG, Bogers AJ, Stumper OF, Hess J et al. Surgery for Ebstein's anomaly: the clinical and echocardiographic evaluation of a new technique. J Am Coll Cardiol 1991;17:722–8.
5. da Silva JP, Baumgratz JF, da Fonseca L, Franchi SM, Lopes LM, Tavares GM et al. The cone reconstruction of the tricuspid valve in Ebstein's anomaly. The operation: early and midterm results. J Thorac Cardiovasc Surg 2007;133:215–23.
6. Dearani JA, Mora BN, Nelson TJ, Haile DT, O'Leary PW. Ebstein anomaly review: what's now, what's next? Expert Rev Cardiovasc Ther 2015;13: 1101–9.
7. Hardy KL, May IA, Webster CA, Kimball KG. Ebstein's anomaly: a functional concept and successful definitive repair. J Thorac Cardiovasc Surg 1964;48:927–40.
8. Danielson GK, Fuster V. Surgical repair of Ebstein's anomaly. Ann Surg 1982;196:499–504.
9. Schmidt-Habelmann P, Meisner H, Struck E, Sebening F. Results of valvuloplasty for Ebstein's anomaly. Thorac Cardiovasc Surg 1981;29:155–7.
10. Palmen M, de Jong PL, Klieverik LM, Venema AC, Meijboom FJ, Bogers AJ. Long-term follow-up after repair of Ebstein's anomaly. Eur J Cardiothorac Surg 2008;34:48–54.
11. Brown ML, Dearani JA, Danielson GK, Cetta F, Connolly HM, Warnes CA et al. The outcomes of operations for 539 patients with Ebstein anomaly. J Thorac Cardiovasc Surg 2008;135:1120–36, 36.e1–7.
12. Hetzer R, Hacke P, Javier M, Miera O, Schmitt K, Weng Y et al. The longterm impact of various techniques for tricuspid repair in Ebstein's anomaly. J Thorac Cardiovasc Surg 2015;150:1212–19.
13. da Silva JP, da Silva LdaF, Moreira LFP, Lopez LM, Franchi SM, Lianza AC et al. Cone reconstruction in Ebstein's anomaly repair: early and longterm results. Arq Bras Cardiol 2011;97:199–208.
14. Dearani JA, Said SM, O'Leary PW, Burkhart HM, Barnes RD, Cetta F. Anatomic repair of Ebstein's malformation: lessons learned with cone reconstruction. Ann Thorac Surg 2013;95:220–6; discussion 6–8.
15. Walsh EP. Interventional electrophysiology in patients with congenital heart disease. Circulation 2007;115:3224–34.
16. Attenhofer Jost CH, Connolly HM, Edwards WD, Hayes D, Warnes CA, Danielson GK. Ebstein's anomaly—review of a multifaceted congenital cardiac condition. Swiss Med Wkly 2005;135:269–81.
17. Pisters R, Nieuwlaat R, Prins MH, Le Heuzey JY, Maggioni AP, Camm AJ et al. Clinical correlates of immediate success and outcome at 1-year follow-up of real-world cardioversion of atrial fibrillation: the Euro Heart Survey. Europace 2012;14:666–74.

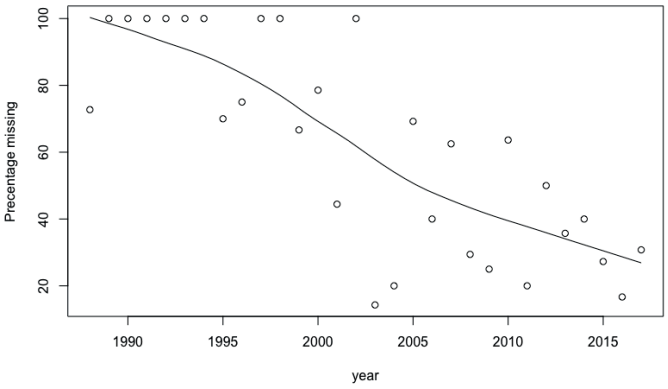
SUPPLEMENTARY MATERIAL

CONTENTS

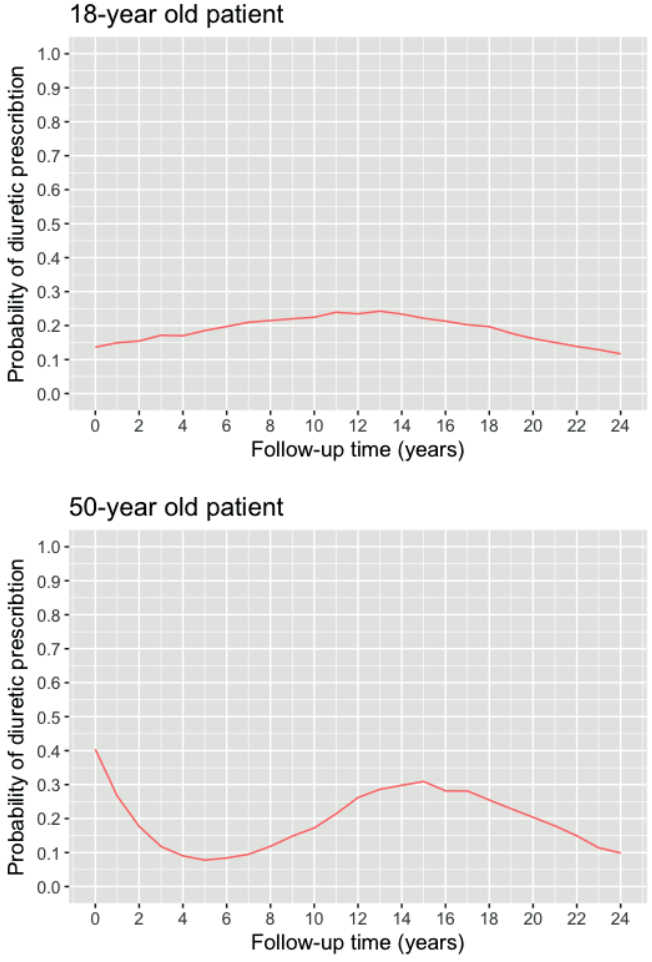


Supplementary Figure 1. Kaplan-Meier plot of freedom from reoperation

Supplementary figure 2. Marginal probability of significant TR stratified to Carpentier class



Supplementary figure 3. A plot of the percentage missing according the year of surgery, with a smoother line



Supplemetary figure 4ab. Marginal probability of diuretic use of a 18-year-old patient (A) and a 50-year-old patient (B)

Supplementary Table 1: Other reported series on reconstructive surgery for Ebstein. KM: Kaplan Meier.

Series	N patients	Mortality	Reoperation
Da Silva, 2011	52	86.2 % at six years (KM estimate)	4 patients with mean follow-up of 57 months
Wu, 2007	78	No hospital mortality	-
Li, 2016	21	None after 9.1 months follow-up	1 after 9.1 months follow-up
Liu, 2011	30	1 (hospital mortality) after 1.9 years follow-up	None after 1.9 years follow-up
Ibragim, 2015	27	None after follow-up 2.7 years	1 (for ASD) after 2.7 years of follow-up
Nguyen, 2014	52	None after follow-up of 42 months	None after follow-up of 42 months