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ORIGINAL ARTICLE

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Surgical outcome in pediatric patients with Ebstein's anomaly: A multicenter, long-term study

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Abstract

Objective: Surgical outcomes of pediatric patients with Ebstein's anomaly are often described as part of all-age-inclusive series. Our objective is to focus on patients treated surgically in childhood (0-18 y). We study the intended treatment (biventricular or 1.5 ventricle repair or univentricular palliation), freedom from unplanned reoperation and survival of this specific age group, in a nation-wide study.

Design: Records of all Ebstein's anomaly patients born between 1980 and 2013 were reviewed. Demographic variables, intraoperative procedures and postoperative outcomes were analyzed.

The results of this study have been presented at the 49th Annual Meeting of the Association for European Paediatric and Congenital Cardiology (AEPC), May 20–23 2015, Prague, Czech Republic

The results of this study have been presented at the joint 45th Meeting of the Deutsche Gesellschaft für Thorax-, Herz- und Gefäβchirurgie (DGTHG) and the 48th Meeting of the Deutsche Gesellschaft für Pädiatrische Kardiologie (DGPK), February 13–16 2016, Leipzig, Germany.

Results: Sixty-three patients underwent 109 operations. Median follow-up after diagnosis was 121 months (range 0-216 months). Twenty-nine (46%) patients required surgery in the first year of life, including 21 who required neonatal surgery. The intended treatment was biventricular (n = 37, 59%) and 1.5 ventricle (n = 5, 8%) repair or univentricular (n = 21, 33%) palliation. The one-, five-, and 10-year freedom from unplanned reoperation was 89%, 79%, and 75% respectively. There were nine (14%) in hospital deaths (within 30 d after surgery). Causes of death were low cardiac output syndrome, cardiac failure, hypoxemia, pulmonary hypertension or an unknown cause. There were no late deaths.

Conclusions: Surgery in childhood represents the worse spectrum of disease, biventricular repair is often not applied. Patients often face revision surgery. Mortality is limited to the immediate postsurgical period.

KEYWORDS

congenital, Ebstein's anomaly, neonates, outcome, surgery

1 | INTRODUCTION

Ebstein's anomaly is a complex, congenital disorder characterized by displacement of the effective tricuspid valve (TV) orifice toward the right ventricular apex.[1] Management depends on the severity of clinical symptoms and the hemodynamic consequences of the malformation. The objectives of surgical intervention are to reduce the severity and hemodynamic sequelae of TV regurgitation; to reduce hypoxemia and to tackle intractable tachyarrhythmias.[2] A number of biventricular repair methods including various modifications in technique have been described.[3–6] When TV reconstruction is considered impossible it may be necessary to implant a prosthetic TV to achieve biventricular repair.[5] A bidirectional cavopulmonary connection may be added (1.5 ventricle repair) to unload a malfunctioning right ventricle (RV).[5–7] When biventricular or 1.5 ventricle repair is not possible, a (usually staged) univentricular palliation is performed. The TV may be closed with a patch containing a small perforation to avoid excessive dilatation of the RV, as in a Starnes operation.[8,9] The nteratrial communication is enlarged and a (modified) Blalock-Taussig (BT) shunt is inserted. In some cases, right atrial reduction is performed.

Studies on surgical outcomes in patients with Ebstein's anomaly often include a majority of adults; consequently, long-term outcomes of usually biventricular repair are generally good. Sarris et al. concluded that there was a negative association between mortality and age at surgery, with higher mortality in neonates and infants.[10] Reports on complete cohorts of young patients are, however, scarce. Such studies rarely consider surgical outcomes of an overall study group and inherently may contain a selection bias.

We therefore reviewed outcomes of a complete cohort of Dutch neonates, children and adolescents till 18 years of age with Ebstein's anomaly treated with surgery. We studied cases requiring neonatal surgery as a separate subgroup. We analyzed freedom from unplanned reoperation and survival during childhood.

2 | METHODS

All six Dutch pediatric heart centers included all patients diagnosed with Ebstein's anomaly born between 1980 and 2013. Diagnosis relied on transthoracic echocardiography; the criterion was apical displacement of the TV from the atrioventricular ring >0.8cm/m^2 body surface area.[11] Diagnosis was confirmed during surgery. Some patients were analyzed in our group's earlier study of factors associated with death.[12] The present study included only those patients with an indication for surgery during childhood. Data on demographic variables, intraoperative procedures and postoperative outcomes were collected retrospectively. Disease severity was classified according to the modified Ross Heart Failure Classification for children.[13] We analyzed all available follow-up data before the patient reached 18 years.

The intended surgical strategy could be either a biventricular or 1.5 ventricle repair or univentricular palliation. The decision for the type of repair was based on the size of the RV, the size of the TV annulus and the size of the pulmonary artery in addition to the severity of the tricuspid regurgitation (TR). A biventricular repair was performed in patients with a retained RV function and a favorable anatomy and function of the RV in the absence of severe pulmonary valve stenosis (PS) or atresia (PA). In patients with extensive RV failure, severe TR and/or the presence of other relevant cardiac comorbidity, e.g. PA, a univentricular palliation was the intended surgical strategy. All operations have been analyzed as separate events. Operations which were not part of the original strategy for achieving the intended biventricular or 1.5 ventricle repair or univentricular palliation were categorized as unplanned reoperations. Patients requiring surgery within the neonatal period were analyzed separately as well. The survival curve was obtained using the Kaplan-Meier method.

3 | RESULTS

3.1 | Patients

Figure 1 shows the enrolment of our 63 patients that underwent surgery for their Ebstein's anomaly and/or associated anomalies. The median age at diagnosis was zero months (range: 0–112 months). The median follow-up duration was 121 months (range: 0–216 months). Patient characteristics are presented in Table 1.

3.2 | Cardiac catheterization

Sixteen patients underwent at least one interventional cardiac catheterization. In four patients the atrial septal defect (ASD) was closed with a device; in one of them a balloon valvoplasty of the PV was performed simultaneously. In two cases ASD closure was performed before the first surgery and in two cases it was performed after the last surgery. In a fifth patient a balloon valvoplasty of the PV was performed, but an unplanned surgical commissurotomy of the PV was required the following day due to to hemodynamic instability. A sixth and seventh patients underwent a radiofrequency perforation of an atretic PV before any surgical intervention. Both patients suffered perforation of the right ventricular outflow tract requiring emergency surgery. The remaining nine patients underwent an ablation during the cardiac catheterization. None of the latter patients died.

3.3 | Operations

The median age at first operation was 26 months (range: 0–203 months). Twenty-nine patients (46%) were operated during the first year of life, including 21 requiring surgery within the neonatal period (0–30 d). The surgical strategy was defined by the policy of the local hospital and followed international practice as described in the methods. The intended strategy was biventricular repair in 37 patients (59%), 1.5 ventricle repair in five patients (8%) and univentricular palliation in 21 patients (33%); however, in nine patients (14%) the final repair was a cross-over (Figure 2). Our 63 patients underwent a total of 109 operations; the average number of operations per patients was 1.4 (53 on 37 patients), 1.8 (9/5) and 2.2 (47/21) in the biventricular, 1.5 ventricle and univentricular subgroup, respectively. Twenty-seven patients required a reoperation of whom 18 patients required at least one unplanned reoperation: eight, two and eight in the biventricular, 1.5 ventricle and univentricular group respectively (Table 2). The one-, five- and 10-year freedom from unplanned reoperation was 89%, 79%, and 75%, respectively, but not all patients did already reach the five- or 10-year follow-up at the time the analysis was conducted.

TABLE 1 Demographic data and clinical characteristics at presentation

	Total, n=63 (%)
Male	30 (48)
At presentation	
Median age in months [min; max]	0 [0; 112]
Age < 1month	45 (71)
Heart failure class 4	34 (54)
Cyanosis	33 (52)
Mechanical ventilation	22 (35)
Type of associated anomaly	
ASD/PFO	50 (79)
VSD	14 (22)
Severe PV anomaly[a]	15 (24)
Aortic coarctation	5 (8)
Other[b]	5 (8)
Severity of tricuspid regurgitation	
Mild	13 (21)
Moderate	20 (32)
Severe	21 (33)

Abbreviations: ASD, atrial septal defect; PFO, patent foramen ovale; PV, pulmonary valve; VSD, ventricular septal defect.
[a]Anomaly including severe pulmonary valve stenosis and functional and anatomic pulmonary valve atresia.
[b]Including noncompaction (n = 2), bicuspid aortic valve (n = 1), mitral valve hypoplasia (n = 1), partial anomalous pulmonary venous return (n = 1).
In two patients the severity of TV regurgitation could not be classified at presentation.

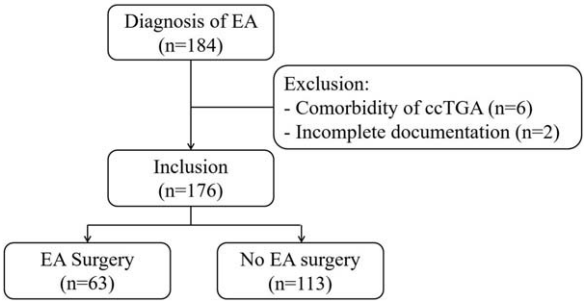

FIGURE 1 Enrolment of 63 pediatric patients diagnosed with Ebstein's anomaly that underwent Ebstein's anomaly related cardiac surgery during childhood or adolescence

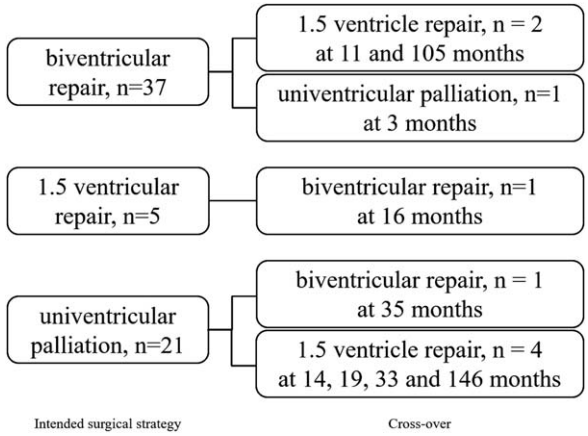

FIGURE 2 Cross-over from the intended surgical strategy in 9/63 pediatric patients with Ebstein's anomaly, including the time period in which the surgical strategy was changed

TABLE 2 Type and number of unplanned reoperations

	Biventricular repair	1.5 ventricle repair	Univentricular palliation
TV (re) reconstruction	4		
TV replacement	2	1	
RV size reduction	1		
RA/RV size reduction and closure multiple VSDs	1		
TV reconstruction and PV homograft	1	2	
Closure PV			1
AP/(M)BT Shunt (re) revision			4
Closure BT shunt, ASD and TV reconstruction			1
PCPC (±TV reconstruction)	3		
TCPC	1		
Fenestration TCPC			1
Conversion to extracardiac TCPC			1
Pacemaker		1	2

Abbreviations: AP, aortopulmonary; ASD, atrial septal defect; (M)BT, modified Blalock Taussig shunt; PCPC, partial cavopulmonary connection; PV, pulmonary valve; RA, right atrium; RV, right ventricle; TCPC, total cavopulmonary connection; TV, tricuspid valve; VSD, ventricular septal defect.

3.4 | Biventricular repair

A biventricular repair was intended in 37 patients. The decision for biventricular repair was based on the severity of the disease, e.g. the retained function of the RV, the anatomy and function of the TV and the absence of severe PV stenosis or atresia (Table 3). The median age at first surgery was 70 months (range: 0–203 months). Six patients underwent neonatal surgery, two of them required additional surgery later in life. The majority received a TV reconstruction (n = 25, 68%). A variety of surgical techniques were used for this reconstruction: 13 patients underwent a Carpentier-Chauvaud repair and in 10 patients a valvuloplasty of the TV was done (in four an Alfieri repair, in five a monocusp repair with annuloplasty, in one patient only an annuloplasty). In two patients the type of surgical reconstruction could not be confirmed retrospectively. In four patients with a TV reconstruction (16%, two patients with a Carpentier-Chauvaud and two other patients with an Alfieri repair) a revision was necessary. Three of these patients received a mechanical valve and one patient got a biological valve.

Due to RV failure a partial cavopulmonary connection (PCPC) was eventually necessary in two patients. The PCPC was performed nine and 105 months after the first surgery. In one additional patient a PCPC was performed three months after a TV reconstruction. Forty-five months later this patient had thrombosis of the mechanical TV which required a switch to univentricular palliation including a total cavopulmonary connection (TCPC).

Five patients had comorbidity of an aortic coarctation. In three of these patients the ASD and/or VSD was closed simultaneously with the aortic arch repair. In the fourth patient with a large VSD the pulmonary artery was banded and ligation of the patent ductus arteriosus (PDA) took place at the same time as the aortic arch reconstruction (as part of an intended biventricular repair).The fifth patient received primarily a coarctation repair. The later was not included in this study as an Ebstein's anomaly associated surgery. However, almost 9 years later the patient required a TV reconstruction as his first Ebstein's anomaly related surgery.

The 37 patients with an intended biventricular repair underwent a total of 53 operations, of which 13 (involving eight patients) were unplanned reoperations (Table 2).

TABLE 3 Clinical and hemodynamic parameter for each intended type of surgery

	All patients (n=63)			Neonates (n=21)	
	bi n=37 (%)	1.5 n=5 (%)	uni n=21 (%)	bi n=6 (%)	uni n=15 (%)
At presentation					
Modified Ross Classification class 4	11 (30)	1 (20)	19 (90)	6 (100)	15 (100)
Severe PS/PA[a]	2 (5)	0 (0)	13 (62)	2 (33)	9 (60)
Mild to moderate TR	25 (68)	3 (60)	12 (57)	5 (83)	8 (53)
Severe TR	11 (30)	2 (40)	8 (38)	1 (17)	6 (40)

Abbreviations: bi, biventricular repair; PA, pulmonary valve atresia; PS, pulmonary valve stenosis; TR, tricuspid valve regurgitation; uni, univentricular repair; 1.5, 1.5 ventricular repair.
[a]Anomaly including severe pulmonary valve stenosis and functional and anatomic pulmonary valve atresia.
In two patients the severity of TV regurgitation could not be classified at presentation.

TABLE 4 Characteristics and causes of death in the study cohort

Age at (last) surgery (months)	Time of death after surgery (d)	Associated anomaly	Intended treatment	No of surgeries	Type of (last) surgery	Cause of death
0	26	ASD, VSD CoA, PDA	bi	1	CoA repair, closure ASD and closure VSD	LCOS, endocarditis
5	1	ASD, VSD, PS	uni	1	Starnes procedure	Cardiac failure
0	1	ASD	uni	1	BT shunt	LCOS
43	4	ASD, PS	uni	2	TV reconstruction, closure ASD and closure BT shunt	Hypoxemia
3	0	VSD	uni	3	PCPC	LCOS/cardiac failure
15	0	PA, PDA	uni	2	MBT	No cause found at autopsy
0	2	PA	uni	1	Valvotomy PV, central AP shunt	LCOS
0	5	PA	uni	2	Enlargement central AP shunt	Reduced venous return after closure sternum? Sepsis?
0	1	Functional PA, ASD, PDA	uni	1	MBT, closure PDA, right atrioplasty	MBT failure due to PPHN and arrhythmias

Abbreviations: ASD, atrial septal defect; AP, aortopulmonary; bi, biventricular repair; BT, Blalock-Taussig shunt; CoA, aortic coarctation; LCOS, low cardiac output syndrome; MBT, modified Blalock-Taussig shunt; PA, pulmonary valve atresia; PCPC, partial cavopulmonary connection; PDA, persistent ductus arteriosus; PPHN, persistent pulmonary hypertension of the newborn; PS, pulmonary valve stenosis; PV, pulmonary valve; TV, tricuspid valve; uni, univentricular repair; VSD, ventricular septal defect.

3.5 | 1.5 Ventricle repair

A 1.5 ventricle repair was intended in five patients. The decision for 1.5 ventricle repair was made at the discretion of the attending surgeon. Clinical and hemodynamic parameters are shown in Table 3. The median age at first surgery was 42 months (range: 17–112 months). None of these patients underwent neonatal surgery. All patients received the PCPC at first surgery. Three patients underwent simultaneously a RV/TV reconstruction (two Carpentier-Chauvaud repairs and one TV valvuloplasty) and ASD closure. In one patient a redirection of the pulmonary veins was simultaneously performed. In one last patient the PV was additionally replaced by a homograft. During a second operation however, the PCPC was taken down, a revision of the TV reconstruction was performed and the PV homograft was replaced by a new homograft. In this patient a biventricular repair was achieved as final repair.

The five patients with an intended 1.5 ventricle repair underwent a total of nine operations, of which four (involving two patients) were unplanned reoperations (Table 2).

3.6 | Univentricular palliation

Univentricular palliation was necessary in 21 patients due to extensive RV failure, the severity of the TV regurgitation or the presence of significant other cardiac abnormalities, e.g. pulmonary atresia (Table 3). The median age at first surgery was less than one month (range: 0–47 months). Fifteen patients (71%) underwent neonatal surgery, 12 of them required additional surgery later in life. At first operation 15 patients received a (modified) BT shunt, four patients received a central aortopulmonary (AP) shunt and two patients a PCPC.

At second operation, three patients underwent a revision or enlargement of the BT shunt or central AP shunt; in one of them the PDA was closed simultaneously. Another patient received a second modified BT shunt to treat a hypoplastic left pulmonary artery. In seven patients a PCPC was performed at second surgery, which took place at an average of 34 months (range: 1–146 months). One patient underwent closure of the PV because of extreme insufficiency before receiving a PCPC during a third operation, at 30 months.

Six patients eventually underwent a TCPC during a second, third or even fourth operation. The mean age at TCPC was 50 months (range: 38–55 months). One patient underwent a Carpentier-Chauvaud repair during the second surgery achieving a final biventricular repair. In four other patients a 1.5 ventricle repair turned out to be adequate. No indications for a TCPC were seen at their most recent outpatient appointment anymore.

The 21 patients with an intended univentricular palliation underwent a total of 47 operations, of which ten (involving eight patients) were unplanned reoperations (Table 2).

3.7 | Neonatal surgery

Twenty-one patients underwent neonatal surgery, 14 of them required further surgery later in life. A biventricular repair was started in six patients (29%), a univentricular palliation was necessary in 15 patients (71%). At first surgery nine patients received a (modified) BT shunt; three received a central AP shunt. Only three patients received a Starnes procedure. Four patients had an aortic

coarctation repaired with simultaneously closure of an ASD and/or VSD, one with contemporarily banding of the pulmonary artery One patient underwent a commissurotomy of the PV and the last patient a valvotomy of the PV.

The 21 patients with neonatal surgery underwent a total of 44 operations, of which nine (involving seven patients) were unplanned reoperations. In two of these patients the unplanned reoperation included a TV reconstruction and revision of TV reconstruction; one of these patients also required a pulmonary homograft. Four patients required a revision or new (M)BT or central AP shunt and in one patient the PV was closed due to severe insufficiency.

In total 7/21 patients died; four died in the neonatal period, the other three died after surgery at one, three and 15 months of age.

3.8 | Surgical complications

Postsurgical complications included arrhythmia (n = 13, 21%), postpericardiotomy syndrome (n = 8, 13%), chylothorax (n = 8, 13%) and infection (n = 7, 11%, no mediastinitis). Further complications included pneumothorax (n = 2), paralysis of the diaphragm (n = 1), vocal cord paralysis (n = 1), hematothorax (n = 1) and a hypoxic ischemic incident resulting in hemiplegia (n = 1).

3.9 | Survival

There were nine (14%) in-hospital deaths (within 30 d after surgery). There were no late deaths. Four patients died in the neonatal period, all but two patients died during the first year of life. Eight of the deaths occurred in the univentricular palliation group. Table 4 shows the characteristics and causes of death for all deceased patients. The one-, five- and 10-year survival rates from time of diagnosis were 89%, 86%, and 86%, respectively. Figure 3 shows the survival of the patients that underwent cardiac surgery for Ebstein's anomaly during childhood (n = 63) vs. the patients that did not undergo surgery for Ebstein's anomaly (n = 113). We included 15 patients with severe PV stenosis or PV atresia. In only two of these patients a biventricular repair was achieved successfully. In the other 13 patients a univentricular palliation was necessary. Five of them died.

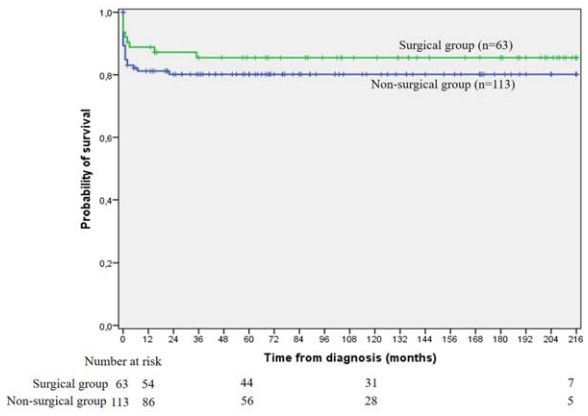

FIGURE 3 Kaplan-Meier estimated survival curve of the patients that underwent Ebstein's anomaly related cardiac surgery during childhood (n = 63) vs. the patients who did not undergo Ebstein's anomaly related cardiac surgery

4 | DISCUSSION

This study reviews the long-term experience in a large all-inclusive cohort of children with Ebstein's anomaly treated surgically, including a high percentage of neonates and infants, to overcome selection bias. Patients were treated with biventricular or 1.5 ventricle repair or univentricular palliation. This study illustrates the extreme variability, complexity and difficulty of this rare cardiac anomaly, especially in neonates. We included all consecutive patients diagnosed with Ebstein's anomaly nationwide, which results in a cohort without bias of patients eligible for certain surgical strategies. Such bias cannot always be excluded in other reports when patients with Ebstein's anomaly are diagnosed elsewhere and are then being transferred to another surgical center for a specific operative correction.

In our cohort a biventricular repair was performed in only half of our patients. These results are difficult to compare to other large studies, because the latter usually include a large number of adults. In one of the largest studies of 539 patients by Brown et al. an AP shunt was created in only 12 patients (6.1% of the patients had received a AP shunt previously in another hospital) and only four patients required a TCPC.[14] However, the mean age of their population at time of their initial surgery in the study group's hospital was 24 years (range: 8 d to 79 y; only one newborn was included). We believe that the surgical outcome of pediatric patients with Ebstein's anomaly should be analyzed separately from adult patients.

In 70% of our patients with neonatal surgery a univentricular palliation was performed. Knott-Craig et al. reported surgical outcomes for a same seize sample (n = 26) of neonatal surgical cases.[3] Twenty-three (88%) neonates received a biventricular repair, one a Starnes palliation and two a BT shunt with or without a pulmonary valvotomy. These results are in line with the study by Boston et al. who reported that 29 of their 32 neonates and young infants had a biventricular repair.[15] In contrast, Reemtsen et al. reported on 16 neonates of which 13 patients underwent right ventricular exclusion and one patient underwent heart transplantation as the initial operation.[16] We cannot rule out that the difference in management strategy among the different studies was influenced by the severity of clinical symptoms and the hemodynamic consequences of the malformation.

When describing freedom from reoperation in pediatric Ebstein's anomaly it is essential to differentiate between operations to achieve a biventricular, 1.5 ventricle repair or univentricular palliation, and all unplanned reoperations. Brown et al. reported a one-, five- and 10-year freedom from any reoperation of 97%, 91%, and 82%, respectively.[14] However, almost all of their patients underwent a biventricular repair and their figures do not differentiate between planned and unplanned reoperations. In our cohort 18/63 patients required one or more unplanned reoperations. The one-, five-, and 10-year freedom

from unplanned reoperations was 89%, 79%, and 75%, respectively. Caution is needed in estimating freedom from unplanned reoperation as not all of our patients have already completed a follow-up period of 10 years after last surgery.

In 2013, Davies et al. described the results of The Society of Thoracic Surgeons Congenital Heart Surgery Database.[17] A total of 595 operations on 498 patients with Ebstein's anomaly were included of which 84% was performed during childhood. They concluded that mortality was highest among neonates. Also in the current study seven of the deceased patients underwent neonatal surgery. Indications for surgery so early in life included severe right-sided heart failure, severe cyanosis and significant associated cardiac anomalies. Therefore, patients requiring neonatal surgery were often in a very unstable clinical condition when undergoing surgery. Patients requiring surgery early in life, especially in the neonatal period, seem to represent the worse spectrum of disease.

4.1 | Study limitations

This study is limited by its retrospective nature. It was not always possible to determine details of the arguments for the intended treatment in each patient retrospectively. Furthermore, over the 33-year period covered by this study there have been changes in treatment strategies and surgical techniques which have influenced surgical outcomes; in particular, advances in technique have improved survival rates for univentricular repair. However, this was not the aim of our study. Carpentier's classification was hardly used in the Netherlands during the 33 years of follow-up and could therefore not be included. Due to the low number of casualties with only nine deaths in the three subgroups, we could not perform reliable analysis to identify risk factors for death. Because of the rarity of the disease a multicenter study was necessary to create this large cohort. Our 63 patients were operated in six university centers. The number of patients treated per center varied between 3 and 15 patients. We cannot rule out that a limited number of operated cases per center and per technique might have influenced the outcome.

4.2 | Conclusions

Patients with Ebstein's anomaly requiring surgery in childhood represent the worse spectrum of disease. Biventricular repair is often not applied. The patients frequently face revision surgery. Mortality is limited to the immediate postsurgical period. To advice on the best type of surgery (biventricular or 1.5 ventricle repair or univentricular palliation) for the individual patient, larger studies are required. However, this is difficult to perform due to the rarity of this disease.

CONFLICT OF INTERESTS

We have no conflict of interest and received no financial support.

AUTHORS CONTRIBUTIONS

Lianne M. Geerdink and Livia Kapusta, Concept/Design, Data Collection, Data analysis/interpretation, Drafting article. Gideon J. du Marchie Sarvaas, Irene M. Kuipers, Willem A. Helbing, Tammo Delhaas, Henriette ter Heide and Lieke Rozendaal, Data Collection, Critical revision of article, Approval of article. Chris L. de Korte, Critical revision of article, Approval of article. Sandeep K. Singh, Tjark Ebels, Mark G. Hazekamp and Felix Haas, Surgery, Critical revision of article, Approval of article. Ad J.J.C. Bogers, Surgery, Drafting article, Critical revision of article, Approval of article.

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