

Perioperative Risk Factors for Mortality in Patients Undergoing Surgery for Ebstein's Anomaly at a Single Institution: A Retrospective Analysis

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ABSTRACT

Background: Mortality after cardiac surgery for Ebstein's anomaly ranges from 2.5% to 31%. Independent predictors for mortality and morbidity remain poorly defined because of the low incidence of this congenital anomaly. To identify potentially modifiable factors, this retrospective study investigates the prognostic value of perioperative variables for mortality and morbidity.

Methods: We reviewed the charts of 171 patients with Ebstein's anomaly who were at one point in follow-up at our center. Only patients who underwent cardiac surgery for this anomaly were included. The primary endpoint was a composite of mortality or rehospitalization for cardiac reasons within 1 year of surgery. Logistic regression and Cox regression models were used to study the predictive value of various variables.

Results: We identified 32 patients (median age 12 years; range 7 days to 70 years) who underwent a total of 49 surgical procedures for Ebstein's anomaly at our institution between November 1987 and March 2015. The following variables were significantly associated with the primary outcome: increased severity of tricuspid valve stenosis (odds ratio 2.089; 95% confidence interval 1.175 to 3.713) and right ventricular dysfunction (1.826; 1.109 to 3.006), partial corrective surgery (versus corrective surgery) (6.709; 1.436 to 31.344), occurrence of major postoperative complications (5.460; 1.419 to 21.008), and increased length of stay in the intensive care unit (1.051; 1.010 to 1.093). A better outcome was observed with the use of intraoperative cardioplegic arrest (0.185; 0.063 to 0.550), atrial septal defect closure during surgery, and longer duration of surgery (0.991; 0.984 to 0.998).

Conclusion: Several patient-specific characteristics and perioperative characteristics were associated with a poorer outcome after cardiac surgery for Ebstein's anomaly. The outcome seems to be primarily determined by the severity of the valve dysfunction and right ventricular performance, with only a minor role for perioperative surgical or anesthetic technical determinants.

INTRODUCTION

Ebstein's anomaly [Ebstein 1866] is a rare congenital heart defect characterized by a broad spectrum of tricuspid valve (TV) and right ventricular (RV) dysplasia [Kouchonkos 2003; Attenhofer 2006]. Indications for surgery vary widely and depend on clinical presentation. Patients with severe forms of this anomaly require surgery, with a major distinction being made at the neonatal age. When severe cyanosis persists in neonates after the decrease of pulmonary vascular resistance, partial corrective surgery is considered. Cyanosis results from inadequate pulmonary flow due to low RV stroke volumes, caused by tricuspid valve regurgitation (TR) or stenosis (TS), pulmonary valvular stenosis (PS), RV dysfunction, or right-left atrial shunting. At the neonatal age, the options most frequently considered for palliation include stenting of

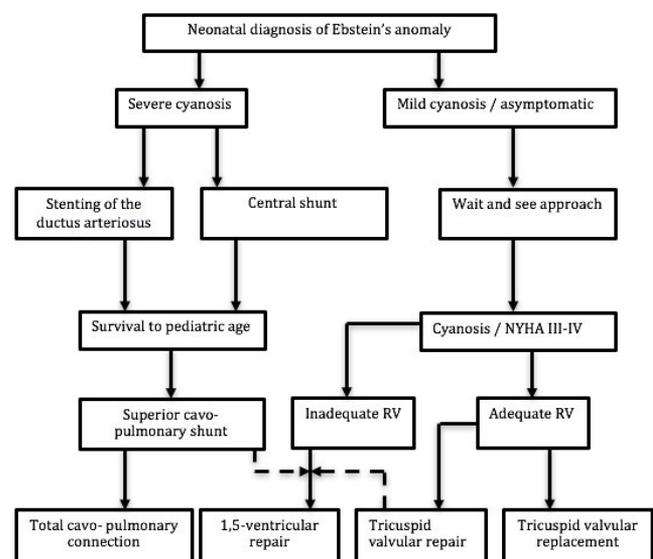


Figure 1. Strategy for surgery for Ebstein's malformation at the University Hospitals of Leuven.

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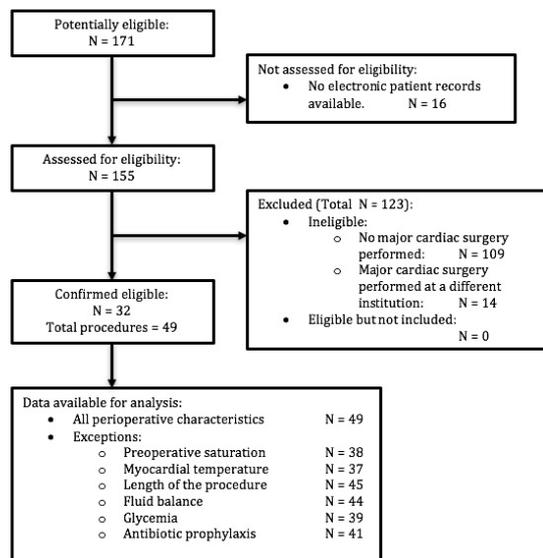


Figure 2. Study flow, in accordance with STROBE guidelines [Von Elm 2014].

the ductus arteriosus or placement of a systemic to pulmonary artery shunt. When patients survive the critical neonatal period, a single ventricle pathway is considered, with first the placement of a superior cavopulmonary shunt (bidirectional Glenn shunt) and later a total cavopulmonary connection (Fontan procedure) [Starnes 1991; Fontan 1971]. In patients who tolerate the Ebstein malformation without severe decompensation or cyanosis, a conservative strategy is appropriate.

After infancy, surgery is considered in case of functional limitation [New York Heart Association (NYHA) class III or IV] caused by progressive exercise intolerance with cyanosis or right-sided heart failure caused by marked TR (grade 3 to 4 or 4) or low RV stroke volume [Knott-Craig 2007]. In these patients, a corrective procedure is first considered if feasible. Tricuspid valve repair is preferred over valve replacement, because valve repair has the potential to be more durable and avoids the potential complications of valve prostheses [Brown 2008].

Different types of valve repair have been described. Corrective surgery is not always feasible, however, in particular when the functional portion of the right ventricle is too small or there is an associated right ventricular outflow tract (RVOT) obstruction. In these cases, a palliative derivation of the blood flow must be achieved, which is known to carry a higher risk of mortality and morbidity after surgery [Brown 2008]. The aim of a partial corrective approach is to decompress the malformed right ventricle, improve the function of the systemic ventricle, and provide adequate pulmonary blood flow. This can be achieved with the placement of a bidirectional Glenn shunt to achieve a 1.5-ventricular repair [Quinonez 2007]. The strategy for surgery in our center for Ebstein's malformation is summarized in Figure 1.

In general, mortality after surgery for Ebstein's anomaly is reported to vary widely, between 2.5% and 31%. Perioperative morbidity and mortality have been demonstrated to be related to severity of TR or TS; the size, thickness, and function of the

Table 1. Baseline Characteristics of All 32 Patients Who Underwent Cardiac Surgery for Ebstein's Anomaly at the University Hospitals of Leuven*

Characteristic	Value
Sex, M/F (%)	56/44
Age (y)	12 (0.02 to 70)
Cyanosis, Y/N (%)	48/52
Functional limitation (NYHA classification)	2 (0 to 4)
Severity of deformity [Carpentier 1988]	2 (1 to 4)
Partial corrective surgery	22 (44)
BT shunt	5 (10)
Bidirectional Glenn	7 (14)
Fontan circulation	7 (14)
LV assistive device placement	1 (2)
Ventricular septal defect closure	2 (4)
Corrective surgery	27 (56)
Tricuspid valve repair	10 (21)
Tricuspid valve replacement	13 (27)
Half ventricle repair	2 (4)
Heart transplantation	2 (4)

*Data are median (95% CI) or n (%) unless noted otherwise.

RV; left ventricular (LV) systolic function; the presence of an atrial septal defect (ASD); and the type of surgery [Kouchonkos 2003; Attenhofer 2006].

Data on further characteristics associated with mortality or morbidity after surgery remain scarce because of the low incidence of this anomaly. In particular, little is known about the influence of variables related to the immediate perioperative period that may be amenable to intervention by the surgeon or the cardiac anesthesiologist/intensivist. Therefore, we aimed to assess the prognostic value of certain perioperative characteristics for the outcome of patients who underwent surgery for Ebstein's anomaly in our institution.

METHODS

Patient Selection

This retrospective study was approved by the Committee on Medical Ethics of the KU Leuven (March 2, 2016; registration number mp11152). We searched the database of the University Hospitals of Leuven for all patients who were at any point in follow-up for Ebstein's anomaly at our institution from January 1987 to July 2015. Patients in continuous follow-up and patients referred for surgery were evaluated for inclusion.

Included were patients who had undergone cardiac surgery for Ebstein's anomaly and for whom electronic anesthesia and intensive care unit (ICU) records were available.

Table 2. Analysis of Preoperative Characteristics for Association with the Primary Endpoint (a Composite of the Occurrence of Mortality or Rehospitalization for Cardiac Reasons, within 1 Year of Surgery)*

Characteristic	Group 1 (n = 22)	Group 2 (n = 27)	P Value, Group 1 versus 2, Mann-Whitney U	Odds Ratio (95% CI)	P Value, Logistic Regression
Sex, F/M (%)	58/42	67/33	.35	1.29 (0.46 to 4.87)	.50
Age (y)	5 (0.02 to 46)	20 (0.04 to 70)	<.05	0.98 (0.94 to 1.00)	.09
Cyanosis, Y/N (%)	62/38	37/63	.51	1.34 (0.76 to 2.35)	.32
Functional limitation (NYHA class)	2 (0 to 4)	2 (0 to 4)	.13	1.20 (0.70 to 2.04)	.52
Severity of deformity [Carpentier 1988]	4 (1 to 4)	2 (1 to 4)	.07	1.69 (0.96 to 2.97)	.07
Severity of TR (grade)	0.5 (0 to 4)	1.5 (0 to 4)	.14	0.70 (0.48 to 1.03)	.07
Severity of TS (grade)	1 (0 to 3)	0 (0 to 3)	<.05	2.09 (1.18 to 3.71)	<.05†
Severity of RV dysfunction (grade)	2 (1 to 4)	1 (1 to 4)	<.05	1.83 (1.11 to 3.01)	<.05†
Severity of RV dilatation (grade)	4 (0 to 4)	2 (0 to 4)	.01	1.56 (0.86 to 2.83)	.15
Severity of LV dysfunction (grade)	0 (0 to 4)	0 (0 to 4)	.40	1.44 (0.87 to 2.40)	.16
Prior cardiovascular procedures (n)	1 (0 to 4)	1 (0 to 3)	.08	1.35 (0.76 to 2.41)	.31
History of ASD closure Y/N (%)	19/81	18/82	.96	1.87 (0.42 to 8.38)	.41
History of stenting ductus of Botalli Y/N (%)	19/81	0/100	<.05	3.59 (0.74 to 17.46)	.11
Endocarditis prior to surgery Y/N (%)	5/95	7/93	.71	4.50 (0.41 to 61.11)	.22
History of dysrhythmias Y/N (%)	24/76	41/59	.22	N.A.	.54
RACHS-1 score [Jenkins 2004]	3.0 (2.0 to 4.0)	3.0 (2.0 to 4.0)	.79	0.79 (0.18 to 3.57)	.76
Aristotle Basis score [Lacour-Gayet 2004]	7.0 (6.0 to 9.3)	7.5 (6.0 to 9.0)	.44	1.11 (0.62 to 1.96)	.73
STAT mortality score [O'Brien 2009]	0.7 (0.2 to 2.1)	0.7 (0.2 to 3.4)	.48	0.27 (0.03 to 2.14)	.21
Year of operation	2006 (1997 to 2015)	2005 (1987 to 2014)	.57	1.04 (0.99 to 1.01)	.08

*Data are median (95% CI) unless noted otherwise.

†Significant difference in regression analysis of endpoints.

Patients with associated defects were included if cardiac surgery was aimed to treat Ebstein's anomaly. We included all cardiac procedures under extracorporeal circulation (ECC) that were performed with the intention to treat the Ebstein malformation. These surgical procedures were separated into 2 subgroups: corrective procedures (repair or replacement of the deformed tricuspid valve; or heart transplantation) and partial corrective procedures (all procedures with the aim to improve hypoxemia and/or the hemodynamic consequences of the deformity, when a corrective approach was not yet possible; including but not limited to the construction of a Glenn shunt construction, Fontan circulation, or the placement of a Blalock–Taussig shunt). Data collection took place between August 2015 and February 2016.

Study Endpoint

The primary endpoint of this study was a composite consisting of early mortality (all-cause mortality, <30 days), late mortality (all-cause mortality, >30 days), or rehospitalization for a cardiac cause within 1 year after surgery. This endpoint was analyzed in accordance with the guidelines for reporting mortality and morbidity after valve surgery as proclaimed by

the Ad Hoc Liaison Committee for Standardizing Definitions of Prosthetic Heart Valve Morbidity [Akins 2008].

Predictive Value of the Perioperative Factors

Several preoperative, perioperative, and postoperative characteristics were selected to identify predictors of outcome. These characteristics were evaluated for each single procedure.

Preoperative characteristics included patient sex, age at the time of the procedure, year of the procedure, preoperative oxygen saturation, history of cardiovascular procedures, history of arrhythmias, endocarditis at the time of operation, severity of the malformation, severity of TR, severity of TS, severity of RV dysfunction, and severity of LV dysfunction.

The severity of the anomaly was defined according to the definition of Carpentier [1988], proposing 4 grades of Ebstein's anomaly: Type A (1), the volume of the true right ventricle is adequate; Type B (2), there is a large atrialized component of the right ventricle but the anterior leaflet moves freely; Type C (3), the anterior leaflet is severely restricted in its movement and may cause significant obstruction of the right ventricular outflow tract; or Type D (4),

Table 3. Analysis of Intraoperative Characteristics for Association with the Primary Endpoint (a Composite of the Occurrence of Mortality or Rehospitalization for Cardiac Reasons, within 1 Year of Surgery)*

Characteristic	Group 1 (n = 22)	Group 2 (n = 27)	P Value, Group 1 versus 2, Mann-Whitney U		P Value, Logistic Regression
			Whitney U	Odds Ratio (95% CI)	
Type of operation, partial corrective/corrective (%)	82/18	30/70	<.05	6.71 (1.44 to 31.34)	< .05†
Concomitant procedure Y/N (%)	50/50	67/33	.26	0.45 (0.10 to 2.05)	.30
ASD closure	0/100	30/70	<.05	NA	< .05†
Duration of the procedure (min)	252 (65 to 365)	245 (130 to 450)	<.05	0.99 (0.98 to 0.99)	< .05†
Duration of ECC (min)	90 (0 to 188)	98 (0 to 230)	.33	0.99 (0.98 to 1.00)	.09
PC administered (n)	1 (0 to 8)	1 (0 to 3)	.19	1.12 (0.73 to 1.72)	.59
FFP administered (n)	1 (0 to 8)	1 (0 to 6)	.90	1.00 (0.71 to 1.41)	.99
Fluid balance (mL)	125 (-2760 to 1810)	22.5 (-2300 to 2710)	.62	0.63 (0.29 to 1.36)	.24
Glucose levels at the onset of surgery (mg/dL)	123.5 (65 to 221)	106 (71 to 207)	.10	0.97 (0.81 to 1.16)	.73
Glucose levels at the end of surgery (mg/dL)	136 (86 to 265)	125.5 (89 to 204)	.26	1.09 (0.99 to 1.19)	.08

*Data are median (95% CI) unless noted otherwise. NA indicates not applicable.

†Significant difference in regression analysis of endpoints.

almost complete atrialization of the right ventricle with the exception of a small infundibular component.

Severity of regurgitation, stenosis, and ventricle dysfunction was assessed with transthoracic echocardiography and evaluated with standardized scales. These scales defined the deformity as light, moderate, severe, or very severe.

The Risk Adjustment for Congenital Heart Surgery (RACHS-1) score [Jenkins 2004], the Aristotle Basis Score [Lacour-Gayet 2004], and the Society of Thoracic Surgeons-European Association for Cardio-Thoracic Surgery (STAT) mortality score [O'Brien 2009]—risk scoring systems that offer prognostic value in patients undergoing congenital heart surgery—were calculated for each patient, including children.

Intraoperative characteristics included the type of operation (corrective as opposed to partial corrective approach), type of concomitant procedure (eg, ASD closure, ablation of Kent bundles), duration of operation (in hours), length of ECC (in hours), numbers of transfused blood products: packed cells (PC) and fresh frozen plasma (FFP), fluid balance during ECC, and blood glucose levels at the beginning and end of the procedure.

Postoperative characteristics included length of stay in ICU; total hospital length of stay; duration of mechanical ventilation (defined by day of extubation); occurrence of major complications in the ICU, medium-care unit, or normal ward (including acute hemodynamic collapse with ventricular failure, septic shock, massive pericardial or pleural effusion, chylothorax, low output syndrome, third-degree atrioventricular block, phrenic nerve paralysis, or lactate acidosis); postoperative severity of TR (derived from the records of the postoperative transthoracic echocardiography); and the number of cardiovascular procedures performed within 1 year postoperatively.

Statistical Analysis

Analyses were performed using SAS software (version 9.4 for Windows; SAS institute, Raleigh, NC) and SPSS software (version 23.0 for Mac OSX; IBM, Armonk, NY). Summary statistics are presented as medians and ranges for continuous variables and as frequencies and percentages for categorical variables.

Categorical variables were compared using χ^2 test. The Mann-Whitney U test was used to compare continuous variables for nonpaired samples. Logistic regression models were applied to study the prognostic effect of variables on the primary endpoint. Generalized estimating equations (GEEs) were used to account for clustering of surgeries within patients. Results are presented as odds ratios (ORs) with 95% confidence intervals (CIs) [Lin 1989]. All tests are 2-sided. A probability value of ≤ 0.05 is considered statistically significant for all tests. Statistical tests were performed without correction for multiple testing.

RESULTS

We identified 171 patients who had been at any point in follow-up for Ebstein's anomaly at the University Hospitals of Leuven from November 1987 to May 2015 (Figure 2).

One hundred thirty-nine patients who did not meet the inclusion criteria were excluded from this retrospective analysis. Thirty-two patients who underwent a total of 49 procedures that were performed at our institution between November 12, 1987, and March 25, 2015, were included. Baseline data of the included patients are summarized in Table 1.

Of the 32 patients included, 12 (38%), undergoing a total of 22 procedures (45%), reached the primary endpoint (Group 1) and were compared with 20 patients (62%), undergoing a

Table 4. Analysis of Postoperative Characteristics for Association with the Primary Endpoint (a Composite of the Occurrence of Mortality or Rehospitalization for Cardiac Reasons, within 1 Year of Surgery)*

Characteristic	Group 1 (n = 22)	Group 2 (n = 27)	P Value, Group 1 versus 2, Mann–Whitney U	Odds Ratio (95% CI)	P Value, Logistic Regression
ICU length of stay (d)	6 (2 to 54)	3 (2 to 20)	<.05	1.05 (1.01 to 1.09)	< .05†
Total hospital length of stay (d)	16 (8 to 54)	12 (8 to 47)	.08	1.03 (0.99 to 1.07)	.0600
Major postoperative complications Y/N (%)	91/9	37/61	<.05	5.46 (1.42 to 21.01)	< .05†
Postoperative TR (grade)	0.5 (0 to 4)	1 (0 to 4)	.24	0.85 (0.32 to 2.25)	.7451

*Data are median (95% CI) unless noted otherwise.

†Significant difference in regression analysis of endpoints.

total of 27 procedures (55%), who did not meet the primary endpoint (Group 2).

Patients in Group 1 had significantly higher severity of TS, RV dysfunction, and RV dilatation; had significantly more frequent stenting of the ductus of Botalli; were significantly younger (Table 2); more frequently underwent partial corrective surgery; underwent no ASD closure during the procedure (Table 3); had longer length of stay at the ICU; and suffered more frequently from major postoperative complications (Table 4).

In a logistic regression analysis, we identified the following factors to be significantly associated with the occurrence of the primary endpoint: increased severity of TS and RV dysfunction; partial corrective surgery (versus corrective surgery); increased length of stay in the ICU; and the occurrence of major postoperative complications.

Conversely, the following factors were significantly associated with a lower risk of occurrence of the primary endpoint: ASD closure during surgery and a longer duration of surgery.

DISCUSSION

In the present retrospective analysis, we identified several perioperative variables that were associated with morbidity and mortality after cardiac surgery for Ebstein's anomaly in our institution. Most of these predictors are patient related or operator related and therefore not amenable for modification in perioperative management (eg, severity of structural heart disease). No predictive factors observed in the intraoperative period are modifiable or could become targets for future preventive strategies.

Data on perioperative and postoperative factors that affect mortality and morbidity after cardiac surgery for Ebstein's anomaly remain scarce. Several reasons contribute to this lack of data: the relatively low prevalence of the disease (Ebstein's anomaly is a rare congenital heart deformity with an incidence of 1 to 5 in 200,000 [Starnes 1991]); the wide disease spectrum and hence a significant heterogeneity of anatomic deformities, hemodynamic profile, and clinical presentation; the lack of randomized controlled trials and evidence to support management strategies in international guidelines; and,

consequently, the need for individualized surgical treatment [Baumgartner 2010; Warnes 2008].

Patient-Related Characteristics

Our data show that the severity of valvular dysfunction is one of the most important predictors for outcome. We identified increased severity of the RV dysfunction and tricuspid valvular stenosis as strongly associated with poorer outcome after cardiac surgery for Ebstein's anomaly. Our findings are consistent with the results of previous studies: Celermajer [1994] found the severity of Ebstein's anomaly, right ventricular outflow tract (RVOT) obstruction, and presentation as a fetus to be associated with poorer survival. Likewise, Brown [2008] identified RV or LV systolic dysfunction, RVOT obstruction, and hypoplastic pulmonary arteries as risk factors for poorer outcome in a retrospective study of 539 patients undergoing tricuspid valve repair or replacement for Ebstein's anomaly. In accordance with other studies, we could not identify age to be associated with a worse outcome, suggesting that not age but rather the severity of the deformation is the determining factor. In fact, patients who underwent cardiac surgery in early infancy were far more likely to have a more severe deformity than patients in whom cardiac surgery could be postponed to adulthood.

Risk Scoring Systems

The RACHS-1 score [Jenkins 2004], the Aristotle Basis Score [Lacour-Gayet 2004], and the STAT mortality score [O'Brien 2009] are risk scoring systems that offer prognostic value in patients undergoing congenital heart surgery. We calculated these risk scores for both pediatric and adult patients. Interestingly, we found in this retrospective study no significant association between the risk scores and outcome after surgery. This finding is most probably because, despite their use in the adult population [Kogon 2014], the systems still lack validation for adult congenital heart surgery and therefore may not be appropriate for prediction of operative risk in adult patients undergoing cardiac surgery for Ebstein's anomaly.

Perioperative Characteristics

In our study, a partial corrective procedure had a significantly and dramatically higher risk for poorer outcome

compared with a corrective procedure. It is obvious that the impossibility to perform a corrective procedure is a reflector of disease severity. Moreover, of all 22 partial corrective procedures performed in our institution, 10 were done on neonatal patients. Management of a neonatal symptomatic Ebstein's anomaly remains extremely challenging, even in highly experienced institutions [Sano 2014]. Consequently, mortality in neonates is inevitably higher [Yu 2013].

The same considerations might also hold true for our finding that the use of cardioplegic arrest and increased duration of surgery were associated with a better outcome. In fact, many partial corrective approaches (eg, the placement of a Blalock–Taussig shunt) are shorter than corrective approaches and are frequently performed without cardiopulmonary bypass (CPB) or on the beating/fibrillating heart, obviating the need for cardioplegic arrest.

We found that ASD closure during cardiac surgery for Ebstein's anomaly was associated with a lower risk for mortality or rehospitalization. Again, this most probably reflects the severity of the underlying malformation. In 80% to 94% of all patients with Ebstein's anomaly, an interatrial communication is present [Attenhofer 2006]. This communication is usually associated with a right-to-left-shunt, although left-to-right shunting may be present in young patients with mild forms of the anomaly [Starnes 1971]. In neonates with a severe deformity of the tricuspid valve and right ventricular deterioration, the interatrial right-to-left-shunt must remain open to provide adequate cardiac output. Hence, ASD closure is considered during corrective approaches only when RV stroke volume is sufficient to provide an adequate forward cardiac output.

A poorer outcome was also found if the length of stay at the ICU increased, which is certainly a reflection of the severity of the underlying condition and a marker of a complicated postoperative course. As a matter of fact, the occurrence of cardiac postoperative complications was associated with poor outcome.

It is interesting to note that perioperative factors that are well known from the literature to worsen outcomes could not be identified as risk factors in our study (eg, the need for and amount of intraoperative transfusion [Bennett-Guerrero 2010; Guzzetta 2011] or fluid balance [Lex 2016]). However, the 2 groups of patients did not differ in these important determinants of postoperative outcome, most likely reflecting a uniform and standardized treatment strategy for all patients in our center. This holds also true for intraoperative glycemic control, the impact of which is under debate in the literature [Vlasselaers 2009; Agus 2012].

Limitations

We acknowledge that our study suffers from several limitations. It is based on retrospective data from a single institution spanning a wide range of time and patient ages. It can be argued that the study has a significant lack of power due to the low number of events and small number of analyzed patients. Power is also decreased owing to the heterogeneity of included patients and procedures. This is inherent to the rarity of Ebstein's anomaly and the setup of this study.

Consequently, a nonsignificant result for a factor should not be interpreted as proof that this variable is unrelated to the endpoint; it indicates only that the data are not robust enough to show a significant association.

Another limitation of this retrospective study is the lack of data on postoperative RV function. We were unable to include this characteristic in our analysis for several reasons: postoperative transthoracic echocardiography (after the patient was discharged from the ward) was frequently performed at non-standardized time points, and in many cases the follow-up was performed at another center. Although we acknowledge that postoperative RV function is an important outcome parameter, with decreased RV function being predictive for rehospitalization [Davlouros 2006], the lack of inclusion of this parameter does not invalidate our belief that immediate perioperative factors appear not to be related to long-term outcome.

Conclusion

In a retrospective analysis, we assessed the outcome of 32 patients with Ebstein's anomaly who underwent cardiac surgery in the University Hospitals of Leuven, and we identified several characteristics associated with 1-year postoperative mortality and morbidity. The outcome seems to be primarily determined by severity of the anatomic deformity and functional decompensation, with only a minor role or no role for perioperative surgical or anesthetic technical determinants.

REFERENCES

- Agus MS, Steil GM, Wypij D, et al. Tight glycemic control versus standard care after pediatric cardiac surgery. *N Engl J Med* 2012;27;367:1208-1219.
- Akins CW, Miller DC, Turina MI, et al. Guidelines for reporting mortality and morbidity after cardiac valve interventions. *Ann Thorac Surg* 2008;8:1490-1495.
- Attenhofer Jost CH, Connolly HM, Dearani JA, Edwards WD, Danielson GK. Ebstein's anomaly. *Circulation* 2006;115:277-285.
- Baumgartner H, Bonhoeffer P, De Groot NM, et al. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). *Eur Heart J* 2010;31:2915-2957.
- Bennett-Guerrero E, Zhao Y, O'Brien SM, et al. Variation in use of blood transfusion in coronary artery bypass graft surgery. *JAMA* 2010;304:1568-1575.
- Brown ML, Dearani JA, Danielson GK, et al. The outcomes of operations for 539 patients with Ebstein anomaly. *J Thorac Cardiovasc Surg* 2008;135:1120-1136.
- Carpentier A, Chauvaud S, Mace L, et al. A new reconstructive operation for Ebstein's anomaly of the tricuspid valve. *J Thorac Cardiovasc Surg* 1988;96:92-101.
- Celermajer DS, Bull C, Till JA, et al. Ebstein's anomaly: Presentation and outcome from fetus to adult. *J Am Coll Cardiol* 1994;23:170-176.
- Davlouros PA, Niwa K, Webb G, Gatzoulis MA. The right ventricle in congenital heart disease. *Heart* 2006;92(Suppl 1):i27-i38.
- Ebstein W. On a very rare case of insufficiency of the tricuspid valve caused by a congenital severe deformity. *Arch Anat Physiol Wiss Med*

1866;7:238-254.

Fontan F, Baudet E. Surgical repair of tricuspid atresia. *Thorax* 1971;26:240-248.

Guzzetta NA. Benefits and risks of red blood cell transfusion in pediatric patients undergoing cardiac surgery. *Paediatr Anaesth* 2011;21:504-511.

Jenkins KJ. Risk adjustment for congenital heart surgery: The RACHS-1 method. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann* 2004;7:180-184.

Knott-Craig CJ, Goldberg SP, Overbolt ED, Colvin EV, Kirklin JK. Repair of neonates and young infants with Ebstein's anomaly and related disorders. *Ann Thorac Surg* 2007;84:587-592.

Kogon B, Oster M. Assessing surgical risk for adults with congenital heart disease: Are pediatric scoring systems appropriate? *J Thorac Cardiovasc Surg* 2014;147:666-671.

Kouchonkos NT, Blackstone EH, Doty DB, Hanley FL, Karp RB. Ebstein's anomaly. In: Kirklin JW, Barratt-Boyes BG, eds. *Cardiac Surgery*. 3rd ed. New York: Churchill Livingstone; 2003:1177-1199.

Lacour-Gayet F, Clarke D, Jacobs J, et al. The Aristotle score: A complexity-adjusted method to evaluate surgical results. *Eur J Cardiothorac Surg* 2004;25:911-924.

Lex DJ, Tóth R, Czobor NR, et al. Fluid overload is associated with higher mortality and morbidity in pediatric patients undergoing cardiac surgery. *Pediatr Crit Care Med* 2016;17:307-314.

Lin DY, Wei LJ. The robust interference for the Cox proportional hazard

model. *J Am Stat Assoc* 1989;84:1074-1078.

O'Brien S, Clarke D, Jacobs J, et al. An empirically based tool for analyzing mortality associated with congenital heart surgery. *J Thorac Cardiovasc Surg* 2009;138:1139-1153.

Quinonez LG, Dearani JA, Puga FJ, et al. Results of the 1.5 ventricle repair for Ebstein's anomaly and the failing right ventricle. *J Thorac Cardiovasc Surg* 2007;133:1303-1310.

Sano S, Fujii Y, Kasahara S, et al. Repair of Ebstein's anomaly in neonates and small infants: Impact of right ventricular exclusion and its indications. *Eur J Cardiothorac Surg* 2014;45:549-555.

Starnes VA, Pitlick PT, Bernstein D, Griffin ML, Choy M, Shumway NE. Ebstein's anomaly appearing in the neonate. A new surgical approach. *J Thorac Cardiovasc Surg* 1991;101:1082-1087.

Vlasselaers D, Milants I, Desmet L, et al. Intensive insulin therapy for patients in paediatric intensive care: A prospective, randomised controlled study. *Lancet* 2009;14;373:547-556.

Von Elm E, Altman D, Egger P, et al. The Strengthening, the Reporting of Observational Studies in Epidemiology (STROBE) statement: Guidelines for reporting observational studies. *Int J Surg* 2014;12:1495-1499.

Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA guidelines for the management of adults with congenital heart disease: A report of the American College of Cardiology/ American Heart Association Task Force on Practice Guidelines. *J Am Coll Cardiol* 2008;52:e143-e263.

Yu JJ, Yun TJ, Won HS, et al. Outcome of neonates with Ebstein's anomaly in the current era. *Pediatr Cardiol* 2013;34:1590-1596.