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Results of surgery for Ebstein anomaly: A multicenter study from the European Congenital Heart Surgeons Association

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Additional material is available online.

Objective: Since most centers' experience with Ebstein anomaly is limited, we sought to analyze the collective experience of participating institutions of the European Congenital Heart Surgeons Association with surgery for this rare malformation.

Methods: The records of all 150 patients (median age 6.4 years) who underwent surgery for Ebstein anomaly in the 13 participating Association centers between January 1992 and January 2005 were reviewed retrospectively. Patients with congenitally corrected transposition were excluded.

Results: Most patients (81%) had Ebstein disease type B or C and significant functional impairment (61% in New York Heart Association class III or IV) and 16% had prior operations. Surgical procedures (n = 179) included valve replacement (n = 60, 33.5%), valve repair (n = 49, 27.3%), 1½ ventricle repair (n = 46, 25.6%), palliative shunt (n = 13, 7.26%), and other complex procedures (n = 11, 6.14%). There were 20 hospital deaths (operative mortality 13.3%) after valve replacement in 5 patients, valve repair in 3, 1½ ventricle repair in 7, palliative procedures in 3, and miscellaneous procedures in 2. Younger age and palliative

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procedures were univariate risk factors for operative death, but only age was an independent predictor on multivariable analysis.

Conclusions: Most patients coming to surgery presented in childhood and were significantly symptomatic. More than half underwent valve replacement or repair, but a considerable proportion had severe disease necessitating 1½ ventricle repair or palliative procedures. Operative mortality did not differ significantly among repair, replacement, and 1½ ventricle repair but was associated with palliative procedures for severe disease early in life, young age being the only independent predictor of operative death.

Abbr eviations and Acronyms
ASD = atrial septal defect
ECHSA = European Congenital Heart Surgeons Association
RV = right ventricle/ventricular
TV = tricuspid valve

Ebstein anomaly, named after the German pathologist who first described it[1] in 1866, is a rare complex congenital defect of the tricuspid valve (TV) and the right ventricle (RV).[2,3] It is characterized by a spectrum of several features, the main ones being (1) apical displacement of the functional tricuspid annulus, that is, of the attachments primarily of the septal and posterior leaflets, with consequent reduction of the functional RV size, (2) dilation of the "atrialized" portion of the RV with variable degrees of wall thinning, (3) adherence of the tricuspid leaflets to the underlying myocardium (failure of delamination) with redundancy, fenestrations, and tethering of the anterior leaflet, and (4) dilation of the true tricuspid annulus (atrioventricular junction). These abnormalities cause tricuspid regurgitation resulting in right atrial and RV dilatation, substrates for the development of atrial and ventricular arrhythmias. An atrial septal defect (ASD) is usually present, allowing right-to-left shunting, resulting in systemic arterial desaturation and cyanosis. In approximately 10% to 15% of patients, accessory conduction pathways with Wolff-Parkinson-White syndrome are encountered.[4] Carpentier and associates[5] have proposed a 4-grade classification of the severity of this anomaly: In type A, the volume of the true RV is adequate. In type B, there is a large atrialized component of the RV, but the anterior leaflet moves freely. In type C, the anterior leaflet is severely restricted in its motion and may also cause significant obstruction of the RV outflow tract. Type D is characterized by almost complete atrialization of the RV with the exception of a small infundibular component. A complementary classification system for Ebstein anomaly has been published by Dearani and Danielson[2] as part of the International Congenital Heart Surgery Nomenclature and Database Project, which was adopted by The Society of Thoracic Surgeons, The European Association for Cardio-Thoracic Surgery, and The European Congenital Heart Surgeons Association (ECHSA). This nomenclature system also describes four types of Ebstein anomaly based on the morphology of the anterosuperior leaflet of the TV and then further subclassifies each of these four types based on the morphology of the leading edge of the anterior leaflet.

The wide anatomic spectrum of Ebstein anomaly is reflected in an extremely variable clinical presentation and natural history ranging from very high mortality despite treatment in symptomatic neonates to long-term survival in some adults with and even without treatment.[4] Accordingly, a large variety of surgical approaches for this anomaly have been used, ranging from valve replacement to various types of valve repair, 1½ ventricle "repair," univentricular palliation, and transplantation. Initially, after the first report of prosthetic valve replacement for Ebstein anomaly in 1963,[6] valve replacement was the mainstay of surgical therapy for this condition, but results were not satisfactory.[7] In 1964, Hardy and colleagues[8] introduced a technique of valve repair which, based on the concept of Hunter and Lillehei,[9] emphasized exclusion of the atrialized portion of the RV by transverse plication, transposing the displaced leaflets to the true annulus and reducing the size of the dilated annulus. As the focus of surgery shifted away from valve replacement, numerous types of repair were devised and refined, and indeed excellent outcomes have been reported from various centers. Danielson and colleagues'[10] modification of the Hardy repair (to which posterior TV annuloplasty and reduction right atrioplasty were added) was reproducibly effective and became a very popular, "standard" repair technique. Carpentier,[5] Chauvaud,[11] and their associates introduced a different repair approach involving temporary detachment of the anterior TV leaflet to achieve its complete mobilization, longitudinal plication of the atrialized RV, clockwise advancement of the anterior TV leaflet (which may be augmented with pericardium),[12] and posterior tricuspid annuloplasty (with prosthetic ring placement), resulting in a monocusp valve. Quaegebeur,[13] Chen,[14] and their colleagues also developed a similar technique, without prosthetic ring annuloplasty. Vargas and coworkers[15] introduced the concept of "annuloplasty" at the level of the displaced leaflets, plicating the atrialized RV above the level of the reconstructed valve. Others (Augustin,[16] Schmidt-Habelmann,[17] Hetzer,[18] Friesen,[19] and their associates) have challenged the value of plication of the atrialized RV and have simplified the repair by performing only a posterior plication of

the "annulus," leaving the atrialized chamber untouched above the resulting monocusp valve. More recently, new variations of repair techniques have been reported. These involve mobilization of all TV leaflets and transposition of the septal and posterior leaflets to the true annulus, without plication (the ventricularization method of Ullmann and colleagues[20]) or even with longitudinal excision of the atrialized ventricle (Wu and Huang[21]) in an attempt to reestablish a trileaflet valve.

Despite excellent results reported, most repair series encompass a small number of patients extending over many years, usually excluding the most severe cases of type D disease. In addition, even in the most experienced valve repair centers, there has been increasing realization that results of repair (even for "higher risk" type B and C lesions) can be improved considerably if the RV is unloaded by a concomitant cavopulmonary (Glenn) shunt, that is, by a 1½ ventricle repair.[11,22,23] Furthermore, in many series there are scattered references to a few severe cases (type D) that required a univentricular approach, in addition to the symptomatic neonates who are converted to a univentricular circulation early in life by a Starnes operation[24] and are therefore subsequently tracked toward Fontan palliation.[24-28] Finally, valve replacement (without atrialized RV plication) continues to appear in various reports as a good option for many nonpediatric patients and also seems to be associated with good long-term results.[29,30]

Given the preference, supported by excellent reported results, of some well-known centers toward one or another of the many available types of surgical procedure for Ebstein anomaly, and also that most centers' experience with this condition is limited to a small number of patients accumulated over decades, we sought to analyze the collective experience of participating institutions of the ECHSA with the full spectrum of types and results of surgery for Ebstein anomaly in Europe in the current era.

Methods

The records of all 150 patients with the diagnosis of Ebstein anomaly who underwent surgical therapy in 1 of the 13 participating ECHSA centers between January 1992 and January 2005 were reviewed retrospectively. Patients with congenitally corrected transposition and "Ebsteinoid" malformation of the systemic atrioventricular valve were excluded. Demographic and functional parameters were evaluated, including age, gender, weight, functional class, presence and degree of cyanosis, cardiothoracic ratio, presence of preoperative arrhythmias, prior operations, echocardiographic and/or angiocardiographic data, and associated anomalies. The various types of surgical procedures performed were categorized as valve replacement, valve repair, 1½ ventricle repair (if valve repair or replacement with closure of atrial communication was supplemented by a Glenn shunt), palliative (systemic or cavopulmonary [Glenn] shunt or Fontan operation), or other complex or miscellaneous procedures. Associated procedures and operative variables (cardiopulmonary bypass and aortic crossclamp times) were also noted. Surgical outcome parameters (including mortality, causes of death, complications, and functional outcome, when available) were evaluated.

Statistical Analysis

Hospital mortality was defined as death during surgery or within 30 days of surgery, according to the definition used in the ECHSA database (now the European Association of Cardiothoracic Surgery database). Ratios are given with 95% confidence limits. A nonparametric test for trend was used to assess the univariate association between mortality and age and between mortality and number of operations, while the association between mortality and type of procedure was assessed by a χ^2 test. Moreover, a nonparametric Mann-Whitney test was used to assess the association between age as a continuous variable and mortality, as well as number of operations as a continuous variable and mortality. A univariate logistic regression model was used to estimate the odds for death (along with their 95% confidence intervals) by age, type of operations and number of operations. Multivariable logistic regression was carried out to assess the independent effect of age, type of operation, and number of operations on mortality. This process was repeated for age and number of operations as continuous rather than categorical variables. All statisitcal analyses were carried out in SPSS version 11.0 (SPSS, Inc, Chicago, Ill).

Results

Patient ages ranged from 1 day to 48.3 years (9.5 ± 10.2 years [mean ± standard deviation], median 6 years). Age distribution is shown in Figure E1. Patient weights ranged from 2 to 87 kg (28.4 ± 21.9 kg, median 20 kg). Eighty patients (53.4%) were female. Participating centers and corresponding number and age of patients are shown (listed in random order) in Table E1. Most patients were symptomatic, with 3% in New York Heart Association class I, 36% in class II, 52% in class III, and 9% in class IV preoperatively. Cyanosis was common, with median oxygen saturation 87% (mean 83.5% ± 15.8%). Cardiomegaly was a prominent feature, with a median cardiothoracic ratio of 66%. Most patients (80%) were in sinus rhythm (in 52% right bundle branch block was noted), 8% had Wolff-Parkinson-White syndrome, 9% had atrioventricular block (8.7% first degree and 0.66% second degree), and 3% had supraventricular arrhythmias. Prior surgical procedures had been performed in 24 patients (16%) and included TV replacement in 5, repair in 9, 1½ ventricle repair in 1, and various palliative operations in 8 patients. In 67% of patients the diagnosis was established by echocardiography alone, in 31% by both echocardiography and cardiac catheterization study, and in 2% only catheterization had been performed. Of the patients who had an explicit severity grade assigned to them (n = 120), 8.3% were considered to have type A anatomy, 50% type B, 30.8% type C, and 10.8% type D (Figure E2). TV annular displacement was severe in 29.8% and moderate in 51.7% of patients. RV function was assessed by echocardiography and graded as

TABLE 1. Clinical characteristics, associated lesions and preoperative risk factors (as percentage of the total number of patients, n = 150)

NYHA (%)	
Class I	3
Class II	36
Class III	52
Class IV	9
Grade of Ebstein anomaly (%)	
Grade A	8.3
Grade B	50
Grade C	30.8
Grade D	10.8
TV annular displacement (%)	
Mild	8.7
Moderate	51.7
Severe	34
RV function (%)	
Normal	8.8
Mild	26
Moderate	58
Severe	19
ASD (%)	70
VSD (%)	7.33
Pulmonary atresia (%)	6.66
WPW syndrome (%)	8
DiGeorge syndrome (%)	0.66
Guilber syndrome (%)	2
Other chromosomal anomalies (%)	2.66
Acidosis (%)	1.33
Pulmonary hypertension (%)	0.66
Mechanical ventilatory support (%)	4.66
Endocarditis (%)	4
Septicemia (%)	3.33
Neurologic deficits (%)	6.66
Seizures (%)	0.66
Renal anomaly (%)	2
Other (%)	7.33

NYHA, New York Heart Association; *TV,* tricuspid valve; *ASD,* atrial septal defect; *VSD,* ventricular septal defect; *WPW,* Wolff-Parkinson-White syndrome.

normal in 8.8% of patients and mildly impaired in 23%, whereas 51.3% had moderate and 16.8% severe RV dysfunction. In 106 cases (70%) the presence of an ASD was explicitly recorded. Anatomic and functional features, associated lesions, and preoperative risk factors are summarized in Table 1.

The surgical procedures performed (n = 179) included valve replacement (n = 60, 33.5%), valve repair (n = 49, 27.3%), 1½ ventricle repair (n = 46, 26.2%), palliative shunt (n = 13, 6.7%), and other complex or miscellaneous procedures (n = 11, 8.5%). The distribution of these procedures is depicted in Figure 1, whereas details of these and associated procedures are summarized in Table E2.

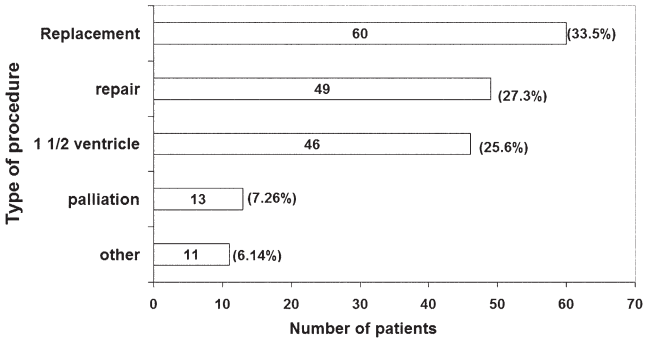

Figure 1. Distribution of the various types of surgical procedures performed for Ebstein anomaly.

Of the surgical procedures performed, 150 (83.7%) were first-time operations and included (1) TV replacement in 51, (2) repair in 47 (17 Carpentier, 8 Danielson, 1 Vargas, 1 Quaegebeur, 2 De Vega, 1 Knott-Craig plasty, and 17 other types of valve repair), (3) 1½ ventricle repair in 36 (16 had TV repair and Glenn shunt, 13 TV replacement and Glenn shunt with or without concomitant VSD and/or ASD closure, and 7 had various other procedures), (4) palliative procedures (eg, systemic-pulmonary shunt) in 12 cases, and (5) miscellaneous other procedures in 4 patients. In 24 patients, 30 reoperations were performed, including 9 TV replacements, 2 TV repairs, 11 1½ ventricle operations (in 10 patients), a palliative procedure (in 1 patient), and 7 other procedures (in 4 patients). Median cardiopulmonary bypass time was 98 minutes (mean 105 ± 38 minutes), and median crossclamp time 54 minutes (mean 57 ± 25 minutes).

There were 20 hospital deaths (operative mortality 13.3%, 95% confidence interval 8.3%-19.8%). Causes of death include 12 (60%) cardiac (low cardiac output, cardiac arrest, pulmonary hypertension, Glenn thrombosis) and 8 (40%) noncardiac (eg, pneumonia, sepsis, respiratory, renal, or multiple organ failure). Differences in operative mortality between institutions did not reach statistical significance. Operative mortality according to type of surgical procedure was 5 of 54 (9.2%) in valve replacement, 3 of 42 (7.1%) in valve repair, 7 of 42 (16.6%) in 1½ ventricle repair, 3 of 4 (75%) in palliative procedures, and 2 of 8 (25%) in miscellaneous procedures. This distribution is shown graphically in Figure 2 and summarized in Table E3, along with results of univariate analysis of the association of mortality with type of procedure. The large and clinically very important differences in mortality among the five types of operative procedures are statistically significant overall ($P = .01$). The highest mortality was noticed for palliative procedures, followed by other types of procedures, 1½ ventricle repair, valve replacement, and valve repair. Compared with palliative procedures, the odds for death were significantly lower

CHD

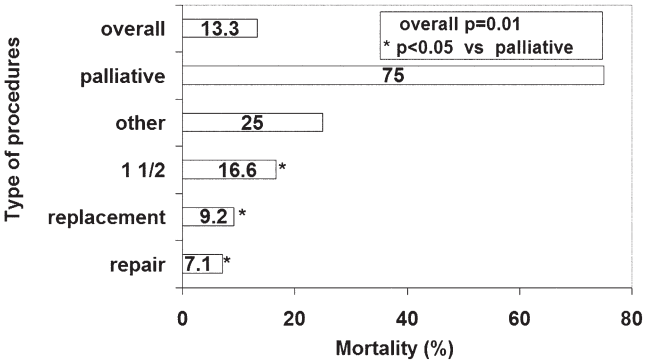

Figure 2. Distribution of operative mortality according to the type of surgical procedure.

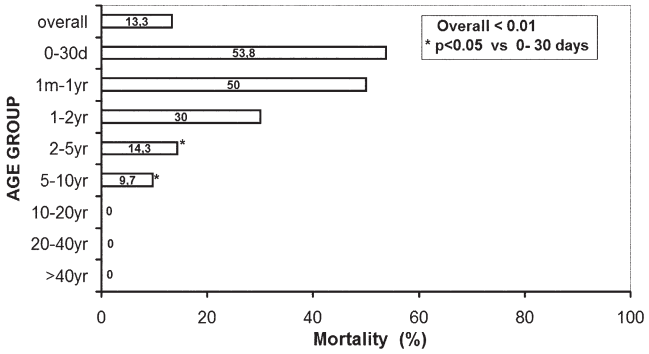

Figure 3. Distribution of operative mortality by age group.

CHD

for 1½ ventricle repair, valve repair, and valve replacement procedures. Pairwise comparisons of repair versus replacement (odds ratio 1.32, $P = .71$), repair versus 1½ ventricle repair (odds ratio 2.6, $P = .17$), and replacement versus 1½ ventricle repair (odds ratio 1.96, $P = .27$) showed that the observed and potentially clinically important differences did not reach statistical significance.

The relationship between age and mortality is shown graphically in Figure 3 and summarized in Table E3, along with results of univariate analysis, which demonstrated that as age decreases, mortality increases, being significantly higher in the infant and neonate age groups. This clinically important finding of statistically significant association of higher mortality with younger age is also confirmed when age is treated as a continuous variable (odds ratio 0.67, confidence interval 0.54-0.83, $P < .001$).

Notably, although reoperation(s) were associated with a trend toward higher mortality, this did not achieve statistical significance ($P = .58$).

Analysis of the multivariable association between mortality and age, type of operation, and number of operations, depicted graphically in Figure E3 and summarized in Table E3, showed that only younger age was independently associated with mortality ($P = .02$).

Important postoperative parameters included mechanical ventilation time (median 1 day), intensive care unit stay (2 days), and hospital stay (16.5 days). Postoperative complications occurred in 114 patients (76%), included arrhythmias, delayed sternal closure, and low cardiac output, and are summarized in Table E4.

Discussion

Reflecting the great anatomic heterogeneity of Ebstein anomaly, the clinical presentation of patients with this condition requiring surgical therapy also varies widely, but some generalizations can be made from this study. Most patients with Ebstein anomaly coming to surgery in the participating centers presented in childhood (median age 6 years), were at least moderately symptomatic (60% New York Heart Association class II-IV), and had mild-to-moderate cyanosis (median oxygen saturation 87%) and increased cardiothoracic ratio (median 66%), whereas a significant percentage (16%) had one or more prior operations. Echocardiography alone seems to be the most useful diagnostic tool. Although most patients were judged to have type B or C disease suitable for biventricular repair (valve replacement or some type of repair), many patients had severe enough disease that necessitated either a 1½ ventricle repair (addition of a Glenn shunt) or even various univentricular palliative operations.

Many centers of excellence focusing on one particular type of surgical approach for Ebstein anomaly have reported outstanding results with their preferred procedure (be it some variation of repair or replacement) for most of their patients (albeit frequently these series do not include patients at the extremes of age or disease). However, our results indicate that, across many European centers, the patient population with Ebstein anomaly is served by a wide variety of surgical procedures. Almost two thirds of operations (n = 109, 61%) were biventricular repairs, that is, valve replacement or repair, with more replacements (33.5%) than repairs (27.5%). It is noteworthy that 25.6% of procedures (n = 46) were 1½ ventricle repairs, whereas 13.4% (n = 24) involved univentricular palliation or other complex procedures for severe disease, presumably owing to serious impairment of RV function. Because of the retrospective and multiinstitutional nature of the study, it was not possible to delineate precise criteria used by participating centers for choosing different procedures. However, there was an obvious association of various palliative and complex procedures and very young age and a tendency toward valve replacement rather than repair in older children, as well as toward 1½ ventricle repair in older patients.

Importantly, although operative mortality was not low (13.3%), it was associated with palliative surgery and very

young age. Although only younger age emerged as an independent predictor of death on multivariable analysis, clinically, palliative surgery was clearly associated with very young age (especially neonates and infants), whereas operative mortality was significantly lower in older children and zero beyond the first decade of life. Cause of death was primarily cardiac failure, while sepsis, respiratory failure, and organ failure may have been facilitated by low cardiac output in several cases. These findings underscore the fact that Ebstein anomaly is a disease not only of the TV but also of the RV and support the principle of management of this malformation depending on age at manifestation, clinical condition, anatomic severity, and associated malformations, employing an eclectic approach rather than attempting to always use the same procedure.

It is widely accepted and confirmed by our data that symptomatic neonates and infants with Ebstein anomaly tend to have complex anatomy, worse pathophysiology, and a generally unfavorable prognosis. This is the case despite various conservative or aggressive approaches, including temporizing with prostaglandin E_1 infusion while awaiting reduction in pulmonary vascular resistance[28] or converting the heart into single ventricle physiology (Starnes operation[24]), although early comprehensive biventricular repair advocated by Knott-Craig and associates[27] has been associated with improved outcome in some patients.

The higher observed mortality of 1½ ventricle repair in our data (compared with reports in the literature[11,22,23]) may reflect more severe underlying disease in this subgroup in our series. Conversely, it is possible that some patients in our replacement or repair subgroups may have possibly benefited from the unloading effect of a supplementary Glenn shunt with consequent reduced mortality.

With regard to possible further analysis of outcomes according to specific type of repair and underlying anatomic parameters, the heterogeneity of types of surgical procedures performed, as detailed in Table E2, and of various selection criteria used in participating institutions did not permit any definitive conclusions.

Although 8% of patients had Wolff-Parkinson-White syndrome, only one surgical ablation procedure (surgery for WPW [n = 1]) was reported in our series (listed in Table E2). It is likely that, in many institutions, interventional catheter techniques may have been used at various time intervals either preoperatively or postoperatively, but such information on the cardiologic management of these patients was not collected in our study.

Our study design and limitations did not permit evaluation of long-term functional outcome in these patients. Such an analysis is clearly of paramount importance when considering the relative merits of the great variety of surgical procedures used but very difficult to achieve given the rarity of the condition and the length of follow-up necessary, which ideally ought to be measured in decades.

Limitations

This study is limited by its nature as a retrospective study carried out in different institutions in 13 different countries spanning a 13-year period. As a consequence, the data are not homogeneous, with two thirds being contributed by just two centers. Accordingly, the small number of patients treated at most centers does not permit a meaningful assessment of the effect of center volume on outcome. However, the data represent *the total and unselected surgical experience of the participating ECHSA institutions during the study period.* Furthermore, although all patients who underwent surgery for Ebstein anomaly at the participating institutions were recorded in the home surgical databases and analyzed, it was not possible to gather information on all patients with this diagnosis who did not have surgery but were being followed up medically during the study period. It was not possible to collect detailed echocardiographic and/or cardiac catheterization information that would permit classification of *all* patients into one of the established (Carpentier scheme) severity grades of the disease. Likewise, because of variations in patient populations served by participating centers and variability in organization of medical care in different countries, complete follow-up information could not be obtained for most patients. Accordingly, it seems desirable to design a long-term prospective study that will ensure the use of uniform criteria to (1) characterize the severity of disease and (2) standardize assessment of the anatomic, physiologic, and overall clinical outcome (including objective assessment of exercise tolerance) of the different types of surgical procedures used for the management of Ebstein anomaly.

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Discussion

Dr Gordon K. Danielson *(Rochester, Minn).* Dr Sarris, I would like to congratulate you and the members of the ECHSA for this large review from 13 centers over 13 years.

Controversy still exists regarding Ebstein anomaly. Can you draw conclusions from your review of these medical centers regarding 3 questions I'd like to ask?

First, we all agree that a competent TV repair is the ideal. However, not all valves can be repaired and it is known that the RV is myopathic and does not tolerate tricuspid regurgitation well. It is also known that all tricuspid bioprostheses will ultimately need to be replaced. Now, the question is this: is it better for the patient to leave the operating room with a competent TV even though it means bioprosthetic valve replacement, or is it acceptable to reduce the tricuspid regurgitation with a repair by only 1 or 2 echocardiographic grades as some medical centers have suggested?

Dr Sarris. Dr Danielson, first let me thank you for your kind comments. It is certainly an honor to have our paper discussed by one of the world's authorities on this disease, the Dean of Ebstein Surgery, as Dr Williams mentioned.

Your question is obviously an important one. We cannot truly answer it directly based on our data, especially since, as I mentioned in the limitations section of this study, there was no uniform approach necessarily in all participating centers in how Ebstein disease is handled. However, the data do suggest that, probably in most of the participating centers, there is a preference toward leaving the operating room with as good a valve as possible, since more valve replacements have been performed in this series than valve repairs. Clearly, this distribution of procedure type is different from what some centers have reported, for example, Carpentier's group, with almost universal application of repair. Thus, I believe our data show that the practice in the participating centers is more along what you are suggesting and what you have preached.

Dr Danielson. The second question is in regard to the addition of a bidirectional cavopulmonary shunt at the time of operation. There are some downsides to that shunt that aren't usually mentioned. These include prolonging the operative time, limiting access to the heart for catheter procedures at a later time (such as for diagnosis and treatment of arrhythmias), and the late systolic pulsations that develop in the head and neck of some patients.

Can you draw conclusions from this study about what you think the prevailing criteria are for performing this shunt at the time of operation? Also, if the decision is made to perform this shunt, is

that decision made preoperatively, intraoperatively, or at the time of weaning from cardiopulmonary bypass?

Dr Sarris. Again, our data do not allow us to answer this question precisely because each center has handled patients according to its own protocol. This is a retrospective study, the surgical approach was not uniform across centers, and consequently it is not possible for us to discern whether the decision to add a cavopulmonary shunt was made preoperatively or intraoperatively.

However, we can answer this question indirectly by noting that there is a substantial proportion of patients with advanced disease, as judged by either echocardiography or degree of annular displacement or by the Carpentier grade, and many of our patients did receive supplementary cavopulmonary shunts in conjunction with either valve repair or replacement. I can only presume that these patients were the ones with more severe disease, but it is not possible to know whether the decision was made preoperatively or postoperatively.

Dr Danielson. The last question relates to atrial arrhythmias. If there is a history of arrhythmias preoperatively, are they treated surgically at the time of operation, or are they left to be treated later, if possible, by catheterization techniques, as some centers recommend?

Dr Sarris. Indeed, our data showed that a significant fraction of patients had arrhythmias. Preoperatively 8% had Wolff-Parkinson-White syndrome. Postoperatively, the most common complication, as mentioned in the manuscript, was arrhythmias. However, in this retrospective multi-institutional study, we did not have data regarding whether arrhythmias, particularly Wolff-Parkinson-White syndrome, were treated by catheter-based intervention or intraoperatively.

Dr Christian Kreutzer *(Buenos Aires, Argentina).* I am quite surprised by the bad results that you show us with a 1½ ventricle repair. Tchervenkov, 5 years ago, presented at the meeting of this Association a wonderful series of valvuloplasties in which the bidirectional Glenn shunt was a protective factor for early survival (unpublished data; 2000).

I wonder if the reason for the low results is the association of a bidirectional Glenn with a valve replacement. I think when you do a bidirectional Glenn, you unload the right ventricle, and the flow that you have going through the prosthetic valve is so low that the valve gets clotted or invaded with pannus.

We have done that in Argentina a few times, and we have abandoned that idea because the function of the valve gets disastrous in a few months. I think it is a real contraindication to do the bidirectional Glenn with a valve replacement.

Do you notice any difference in between mortality in 1½ ventricle repairs between valve replacements and valve plasties?

Dr Sarris. There was no difference in mortality between these two subgroups. Approximately half of the 1½ ventricle repair patients in our study had valve repairs and the other half had valve replacement, both supplemented with bidirectional cavopulmonary shunts.

We are aware of the reports in the literature of much lower mortality in 1½ ventricle repairs, but we believe that the difference between those reports and our findings must pertain to the patient population that was subjected to this operation. Clearly, if the criteria for using 1½ ventricle repair are relaxed, then the overall results of this approach will be better. It is possible that if more 1½ ventricle repairs were performed in this particular cohort of patients, that is, with more liberal criteria, then this particular subgroup would have lower mortality.

Dr Carlo F. Marcelletti *(Palermo, Italy).* I believe this group of patients that you have reported is quite inhomogeneous. They have only the name of the basic disease in common. However, if you try to look at different groups by age, you find that the neonatal age is the age when the Ebstein anomaly is basically not treatable, because the disease not only involves the heart but mostly involves the lungs. Severe Ebstein anomaly at that age involves undeveloped lungs. I think most of us who have tried to do any type of palliation between 0 and 30 days of age have always failed.

In our center, we like to consider neonates with Ebstein anomaly to be medical patients. The administration of prostaglandin and nitrous oxide is probably the best palliation that we can use.

Dr Sarris. Your question?

Dr Marcelletti. Why did you put the neonates in this group of patients? They are absolutely a different subset of patients. I think they have nothing to do with patients who undergo valve replacement, 1½ ventricle repair, because they are not treatable.

Dr Sarris. We are well aware that the neonates and infants are a special subgroup with much more severe presentation and higher mortality no matter what you do, whether you apply a more conservative approach of watching and using prostaglandin, or a more radical approach such as conversion to a single ventricle physiology, the Vaughn Starnes approach, or using what may be perhaps an even more aggressive approach, the Knott-Craig type of repair. There are reports in the literature of reasonable results with all of these approaches.

Once again, this is a retrospective study documenting the practice of Ebstein surgery in 13 participating ECHSA centers, and there is no uniform approach across these centers. The results do confirm what you have just said, that surgical mortality is much higher in the neonates and infants. It is 50%, but there *are* survivors. Our study documents the practice of Ebstein surgery for all age groups in the participating institutions.

Dr Hillel Laks *(Los Angeles, Calif).* In older children, meaning not neonates or infants, we have found that there is a higher risk and more difficulty postoperatively in those who have ASDs with right-to-left shunts. In that group, rather than using a bidirectional Glenn, we use an adjustable ASD, which can then subsequently be closed as the RV recovers. We found this extremely useful in reducing the postoperative difficulty. Was this used in any of these patients as opposed to the bidirectional Glenn?

Dr Sarris. I don't believe so, although this is an approach that you have introduced and popularized for a number of reasons. I don't believe that this was used in any of these patients.

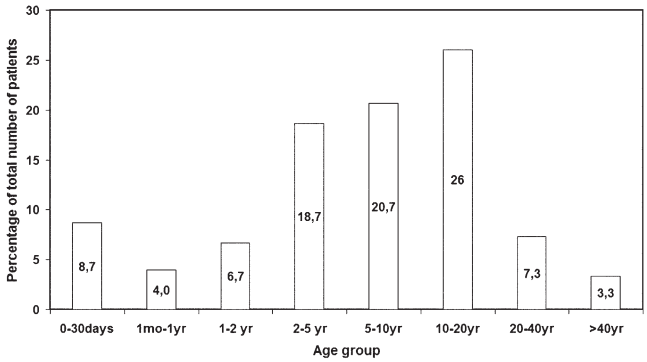

Figure E1. Age distribution as percentage of total number of patients (n = 150) by age subgroup.

TABLE E1. Participating centers with corresponding number and ages of patients*

Center	No. of patients	Age (median, y)	Age range
1	21	3.5	4 d-44.8 y
2	4	13.4	8.5-35.4 y
3	3	11.8	1 d-15.9 y
4	3	47.4	46.6-48.3 y
5	6	1.6	3 d-21.6 y
6	6	2.8	3 d-31.8 y
7	3	31	3-33 y
8	78	6.8	25 d-47.4 y
9	10	7.2	1 d-21 y
10	5	18.4	6.2-45.2 y
11	8	5.6	1.1-11.1 y
12	1	15.9	15.9 y
13	2	8.9	5.5-12.4 y
Total	150	6.4	1 d-48.3 y

*In random order.

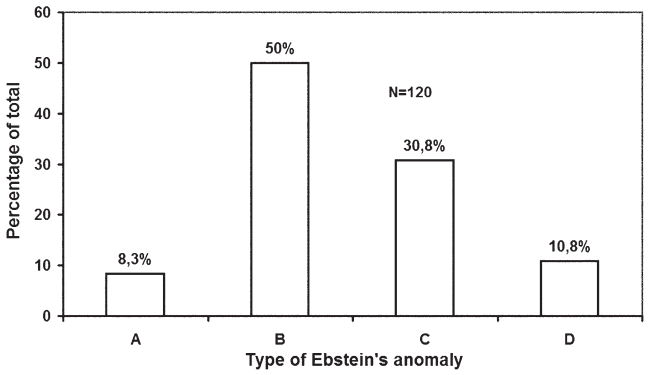

Figure E2. Patient distribution into the types of Ebstein anomaly according to Carpentier's classification.

TABLE E2. Types of procedures performed (n = 179)

Replacement (n = 60)	Replacement TV (± ASD repair) (n = 58)	Replacement TV + ASD + VSD repair (n = 2)				
Repair (n = 49)	Carpentier (n = 17)	Danielson (n = 8)	Other valve repair (n = 20)	Vargas (n = 1)	DeVega (n = 2)	Knott-Craig (n = 1)
1½ ventricle repair (n = 46)	Valve repair + Glenn (n = 16)	Valve replacement + Glenn (n = 13)	Glenn + ASD closure (n = 10)	TV replacement + Glenn + VSD (n = 2)	Glenn + RVOT resection (n = 1)	Glenn + PV replacement (n = 2)
	Hemi-Fontan (n = 2)					
Palliation (n = 13)	mBT shunt + PDA closure (n = 5)	Central shunt (n = 2)	Central shunt + ASD enlargement + TV closure (n = 1)	mBT shunt + TV closure (n = 1)	mBT Shunt + PVotomy (n = 2)	mBT shunt + TV closure + atrial septation (n = 1)
		Fontan (n = 1)				
Other (n = 11)	Pacemaker (n = 2)	PDA closure (n = 2)	Ablation (n = 1)	Other (n = 6)		

TV, Tricuspid valve; *ASD*, atrial septal defect; *VSD*, ventricular septal defect; *RVOT*, right ventricular outflow tract; *PV*, pulmonary valve; *mBT* shunt, modified Blalock-Taussig shunt; *PDA*, patent ductus arteriosus.

TABLE E3. Analysis of operative mortality

Mortality by type of procedure	Total	Dead	Mortality (%) (95% CI)	Univariate OR for death (95% CI)	*P* value
Type of operation					
Palliative	4	3	75 (19.4-99.4)	1.0	
1½	42	7	16.6 (6.9-31.3)	0.06 (0.006-0.74)	.03
Repair (TV plasty)	42	3	7.1 (1.5-19.5)	0.025 (0.002-0.33)	.005
Replacement	54	5	9.2 (3.0-20.3)	0.034 (0.003-0.39)	.007
Other	8	2	25.0 (3.2-65.1)	0.11 (0.007-1.77)	.12
Overall	150	20	13.3 (8.3-19.8)		.01

Mortality by age group	Total	Dead	Mortality (%) (95% CI)	Univariate OR for death (95% CI)	*P* value
Age					
0-30 d	13	7	53.8 (25.1-80.7)	1.0	
1 mo-1 y	6	3	50 (11.8-88.2)	0.86 (0.12-5.94)	.87
1-2 y	10	3	30 (6.7-65.2)	0.37 (0.06-2.08)	.26
2-5 y	28	4	14.3 (4-32.6)	0.14 (0.03-0.65)	.012
5-10 y	31	3	9.7 (2-25.7)	0.09 (0.02-0.46)	.004
10-20 y	39	0	0	—	—
20-40 y	11	0	0	—	—
>40 y	5	0	0	—	—
Overall*	123	20	13.98 (8.7-20.7)		<.001

Multivariable association between age, type of procedure, number of operations, and mortality

Risk factors	Multivariate odds for dying (95% CI)	*P* value
Age	0.69 (0.55-0.87)	.02
Type of procedure		
Palliative	1.0	.62
1½	0.30 (0.02-3.85)	.36
Repair (TV plasty)	0.15 (0.01-2.13)	.16
Replacement	0.35 (0.02-5.0)	.44
Other	0.54 (0.02-13.5)	.71
No. of operations	0.87 (0.26-2.84)	.82

CI, Confidence interval; *OR*, odds ratio; *TV*, tricuspid valve. *Information is missing for the ages of 7 surviving patients.

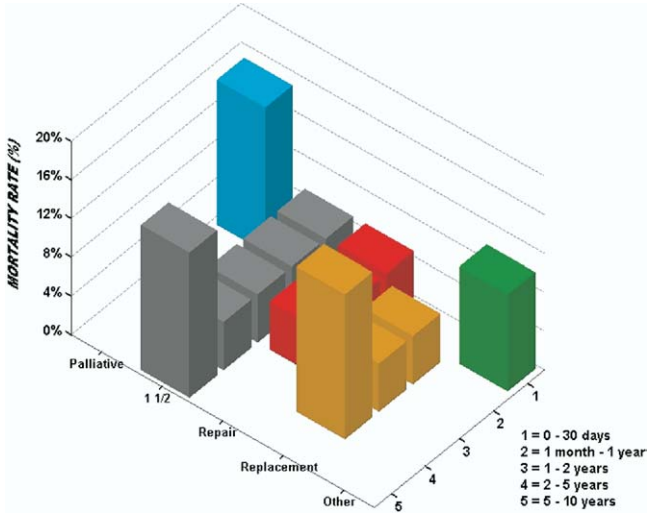

Figure E3. Graphic depiction of the relationship of age group, type of procedure, and mortality.

TABLE E4. Postoperative complications noted in 114 patients

Complications	No. of patients	%
Arrhythmia	39	34.3
Sternum left opened	18	15.8
Temporary complete AV block requiring pacemaker	17	14.9
Low cardiac output	12	10.5
Pneumonia	8	7
Pericardial effusion requiring drainage	7	6.1
Pleural effusion requiring drainage	7	6.1
Cardiac arrest	7	6.1
Systemic vein obstruction	6	5.3
Acute renal failure requiring temporary dialysis	4	3.5
Respiratory insufficiency requiring mechanical ventilatory support $>$ 7 d	4	3.5
Pulmonary hypertension crises (PA pressure $>$ systemic pressure)	4	3.5
Permanent complete AV block requiring pacemaker	4	3.5
Wound infection	4	3.5
Septicemia	4	3.5
Neurologic deficit persisting at discharge	4	3.5
Tracheostomy	2	1.8
Bleeding requiring reoperation	2	1.8
Mediastinitis	1	0.9
Chylothorax	1	0.9
Wound dehiscence	1	0.9
Other	1	0.9

AV, Atrioventricular; *PA*, pulmonary artery.

Results of surgery for Ebstein anomaly: A multicenter study from the European Congenital Heart Surgeons Association

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