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Risk of Surgery for Congenital Heart Disease in the Adult: A Multicentered European Study

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Background. Surgery for congenital heart disease (CHD) has changed considerably during the last three decades. The results of primary repair have steadily improved, to allow treating almost all patients within the pediatric age; nonetheless an increasing population of adult patients requires surgical treatment. The objective of this study is to present the early surgical results of patients who require surgery for CHD in the adult population within a multicentered European study population.

Methods. Data relative to the hospital course of 2,012 adult patients (age ≥ 18 years) who required surgical treatment for CHD from January 1, 1997 through December 31, 2004 were reviewed. Nineteen cardiothoracic centers from 13 European countries contributed to the data collection.

Results. Mean age at surgery was 34.4 ± 14.53 years. Most of the operations were corrective procedures (1,509 patients, 75%), followed by reoperations (464 patients,

23.1%) and palliative procedures (39 patients, 1.9%). Six hundred forty-nine patients (32.2%) required surgical closure of an isolated ostium secundum atrial septal defect. Overall hospital mortality was 2%. Preoperative cyanosis, arrhythmias, and NYHA class III-IV, proved significant risk factors for hospital mortality. Follow-up data were available in 1,342 of 1,972 patients (68%) who were discharged home. Late deaths occurred in 6 patients (0.5%). Overall survival probability was 97% at 60 months, which is higher for corrective procedures (98.2%) if compared with reoperations (94.1%) and palliations (86.1%).

Conclusions. Surgical treatment of CHD in adult patients, in specialized cardiac units, proved quite safe, beneficial, and low-risk.

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Surgery for congenital heart disease (CHD) has changed considerably during the last three decades. The results of primary repair of CHD have steadily improved and allowed to treat almost all patients within the pediatric age group. This was in part due to a more widespread screening of asymptomatic patients and to the optimization of the results of early correction in neonates and infants. Nonetheless, an increasing population of adult patients requires surgical treatment for CHD. This selected

group of patients includes the following: (1) patients with recognized CHD who have survived into adult age without any surgical treatment and without developing an irreversible damage to their heart and lungs; (2) patients who are candidates for correction after previous palliative procedures; (3) adult patients who either require reoperations because of late complications or residual defects; (4) patients who require additional palliative procedures or even heart transplantation [1–4], and (5) patients with previously undetected CHD who remained asymptomatic during childhood. Ostium secundum atrial septal defects (ASD II) provide the best example of CHD within this last group of patients [4–6]. The majority of the latter patients usually maintain a good or only mildly decreased functional status

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Table 1. Main "Basic Diagnoses" Total Number of Patients = 2,012)

Diagnosis Category	n	Diagnosis Type	n		
Septal defects	895	ASD (ostium secundum and sino venosus types)	649		
		Partial AVSD	119		
		VSD	108		
		Complete AVSD	12		
		Truncus arteriosus	7		
Pulmonary venous anomalies	108	PAPVC	93		
		TAPVC	7		
		Pulmonary venous stenosis	4		
		Cor triatriatum	4		
Systemic venous anomalies	5	Anomalous connections/obstruction	5		
Right heart lesions	420	TOF	219		
		TOF-PA	70		
		RVOT obstruction	33		
		Pulmonary valve stenosis	27		
		Ebstein's tricuspid valve	27		
		TV stenosis/regurgitation non-Ebstein related	20		
		Pulmonary valve regurgitation	18		
		PA-IVS	6		
		Left heart lesions	311	Congenital aortic valve stenosis	88
				Congenital aortic valve stenosis + regurgitation	63
Subaortic fibrous stenosis	56				
Congenital aortic valve regurgitation	50				
Congenital mitral valve stenosis/regurgitation	37				
Other	17				
Single ventricle	70	Single right ventricle	58		
		Single left ventricle	12		
Transposition of the great arteries	73	Congenitally corrected TGA	35		
		Complete TGA-VSD	22		
		Complete TGA-IVS	16		
Double outlet right ventricle	16	TOF type	10		
		Uncommitted VSD type	5		
		TGA type	1		
Thoracic arteries and veins	101	Isthmic aortic coarctation	69		
		Coronary artery anomalies	12		
		Patent ductus arteriosus	10		
		Vascular ring	5		
Electrophysiology	5	Congenital III degree AV block	5		
Miscellaneous	8	Cardiac tumor	3		
		Other	5		

ASD II = ostium secundum atrial septal defect; AV = atrioventricular; AVSD = atrioventricular septal defects; IVS = interventricular septum; PA = pulmonary atresia; PAPVC = partial anomalous pulmonary venous connections; RVOT = right ventricular outflow tract; TAPVC = total anomalous pulmonary venous connections; TGA = transposition of the great arteries; TOF = tetralogy of Fallot; VSD = ventricular septal defect.

for several years and generally are referred for surgical closure of the ASD II because of increasing symptoms not controlled by medical therapy [7-9]. The objective of this study is to present the early surgical results of patients who require surgery for CHD in the adult population within a multicentered European study population.

Material and Methods

Data were reviewed retrospectively relative to the hospital course of adult patients (≥ 18 years) who required

surgical treatment of CHD from January 1, 1997 through December 31, 2004. Nineteen cardiothoracic centres from 13 European Countries contributed initially to the data collection.

Individual consent was not obtained by patients enrolled in this study. However, this is a retrospective study, patients are not identified, and the chairperson of each Ethics Committee approved to send their data with the aim of publication. This study was undertaken under the aegis of the European Congenital Heart Surgeons Association. Previous multicentered studies have already

Table 2. Main Diagnoses Leading to Surgery Listed According to the Operative Surgical Category

Main diagnoses leading to palliative procedure (n = 39)	
Single ventricle	17 (53.1%)
TOF-PA	10 (28.1%)
Corrected TGA	6 (9.5%)
Ebstein's tricuspid valve	2 (6.2%)
Other	4 (3.1%)
Main diagnoses leading to corrective procedures (n = 1509)	
Isolated ASD II	649 (43.0%)
Congenital AoV stenosis/regurgitation	162 (10.7%)
Partial AVSD	98 (6.5%)
VSD	96 (6.3%)
PAPVC	93 (6.1%)
Coarctation of the aorta	54 (3.6%)
RVOTO	52 (3.4%)
Subaortic stenosis	48 (3.2%)
Single ventricle	38 (2.6%)
Congenital MV insufficiency/stenosis	37 (2.5%)
Tetralogy of Fallot	37 (2.5%)
Ebstein anomaly	19 (1.3%)
PDA	10 (0.7%)
Supravalvar aortic stenosis	6 (0.3%)
Other diagnoses leading to correction ^a	94 (6.4%)
Main diagnoses leading to reoperation (n = 464)	
Pulmonary valve insufficiency	119 (25.7%)
Conduit failure	79 (17.0%)
RVOTO	56 (12.1%)
AoV insufficiency	25 (5.4%)
Mitral valve insufficiency/stenosis	21 (4.5%)
Residual VSD	19 (4.1%)
Subaortic stenosis	18 (3.9%)
Postoperative third degree AV block	16 (3.5%)
AoV stenosis	14 (3.1%)
Residual coarctation of the aorta	11 (2.3%)
Prosthetic valve failure	11 (2.3%)
Cardiomyopathy	11 (2.3%)
Atriopulmonary connection failure	11 (2.3%)
Tricuspid valve regurgitation	8 (1.7%)
Obstructive pulmonary vascular disease	2 (0.4%)
Other diagnoses leading to reoperation	43 (9.4%)

^a Other diagnoses not listed in the table included diagnostic of single or few patients.

AoV = aortic valve; ASD II = ostium secundum atrial septal defect; AVSD = atrioventricular septal defect; DORV = double outlet right ventricle; MAPCAs = main pulmonary artery collaterals from aorta; MV = mitral valve; PA = pulmonary atresia; PAPVC = partial anomalous pulmonary venous connections; PDA = patent ductus arteriosus; PV = pulmonary valve; RVOTO = right ventricle outflow tract obstruction; TGA = transposition of the great arteries; TOF = tetralogy of Fallot; VSD = ventricular septal defect.

been published under the aegis of the European Congenital Heart Surgeons Associations (ECHSA) [10, 11], which also founded the "European Congenital Database" [12–14].

Cardiac rhythm, preoperative risk factors, extra-cardiac anomalies, diagnoses leading to surgery, surgical procedures, and postoperative complications were identified

according to the Pediatric European Database Classification [12–14].

For the analysis, surgical procedures were divided into three categories, including the following: (1) palliative procedures; (2) corrective procedures; and (3) reoperations. Palliative [15] procedures include all operations performed to improve the patient's clinical status, without separation between the systemic and the pulmonary circulations. Corrective procedures [15] are operations employed to achieve an anatomical or physiologic correction with separation of the pulmonary from the systemic circulation. Included in this group are also "Fontan-type" repair and "one and a half ventricle repair." Finally, reoperations refer to procedures performed after an initial correction.

Because patients requiring surgical closure of an isolated ASD II represent a large group within our adult population with CHDs, and because this operation carries a low mortality and morbidity rate, additional analysis was performed excluding the ASD II group, with the aim to better evaluate the impact of cardiac surgery on this age group of patients.

Results are presented as mean and standard deviation (median and range were used if data were not normally distributed). To assess if there was a statistical difference between the groups, data were analyzed with analysis of variance. A multiple comparison test (Tukey test) was further employed to assess statistical significance between groups. A Wilcoxon signed rank test was used to assess the change in NYHA status at follow-up of patients who were discharged from the hospital in comparison with their preoperative clinical status [16]. Level of significance was set at a *p* value of 0.05 or less.

Overall survival probability, as well as survival probability for different surgical categories (palliative procedures, corrective procedures, and reoperation), and for patients requiring isolated ASD II closure in comparison with patients requiring surgical treatment for all other patients were analyzed according to the Kaplan-Meier method. Patients only who were discharged from the hospital and were subsequently clinically followed were entered in the probability of survival analysis.

Results

Two thousand twelve patients (≥ 18 years of age) requiring surgical treatment for CHD were included in this study. Of the 2,012 patients, 1,005 were males (50%) and 1,007 females (50%). Mean age at surgery was 34.4 ± 14.53 years. Preoperative diagnosis was achieved by echocardiography in 1,225 of the 2,012 patients (60.9%). Magnetic resonance and computer tomography of the thorax were done in addition to echocardiography as an adjunct in 647 patients (32.1%) and cardiac catheterization 399 patients (19.8%).

According to the preoperative clinical evaluation, 647 patients (32.1%) belonged to New York Heart Association (NYHA) class I, 908 in NYHA class II (45.1%), 384 in NYHA class III (19.1%), and 73 in NYHA class IV (3.7%).

A preoperative electrocardiogram revealed a sinus rhythm in 1,813 of 2,012 patients (90.2%). Preoperative

Table 3. Main Surgical Procedures, Listed Within Their Respective Surgical Categories

	n (%)	Hospital mortality - n (%)
Palliative procedures (n = 39):		
Systemic-to-pulmonary shunt	18 (46.9%)	2 (11.1%)
BCPS	15 (37.4%)	1 (6.6%)
MAPCAs unifocalization	3 (9.5%)	-
Pulmonary artery banding	3 (6.2%)	-
Main corrective procedures (n = 1509):		
Isolated ASD II closure	649 (43.0%)	1 (0.2%)
AoV replacement	111 (7.4%)	2 (1.8%)
Partial AVSD correction	98 (6.5%)	-
VSD closure	96 (6.3%)	1 (1%)
PAPVC correction	93 (6.1%)	1 (1%)
Coarctation of the aorta repair	54 (3.6%)	-
RVOTO correction	52 (3.4%)	-
Ross operation	51 (3.4%)	-
Subaortic stenosis resection	48 (3.2%)	1 (2%)
Tetralogy of Fallot correction	37 (2.5%)	2 (5.4%)
Mitral valve replacement/plasty	37 (2.5%)	-
Total cavopulmonary connection	36 (2.4%)	3 (8%)
Ebstein's tricuspid valve plasty	19 (1.3%)	-
PDA closure	10 (0.7%)	-
Supravalvar aortic stenosis correction	6 (0.3%)	-
Other corrections ^a	94 (6.4%)	7 (7.4%)
Main reoperations (n = 464):		
Conduit placement	123 (26.5%)	4 (3.1%)
Conduit reoperation	67 (14.4%)	-
Pulmonary valve placement	45 (9.8%)	1 (2.2%)
Aortic valve replacement	35 (7.6%)	-
Residual VSD closure	19 (4.1%)	1 (5.2%)
RVOTO reoperation	19 (4.1%)	-
Subaortic stenosis resection	18 (3.9%)	-
Mitral valve replacement	16 (3.5%)	-
Pacemaker implantation	16 (3.2%)	-
Heart transplantation	15 (3.2%)	7 (46%)
Ross operation	11 (2.3%)	-
Isthmic aortic coarctation redo	11 (2.3%)	-
Mitral valvuloplasty	9 (2.0%)	1 (11%)
Total cavopulmonary connection	9 (2.0%)	1 (11%)
Tricuspid valve placement	8 (1.7%)	-
Other reoperations	43 (9.4%)	3 (6.9%)

^a Other diagnoses or surgical treatments not listed in the table included diagnostic of surgical groups of single or few patients.

AoV = aortic valve; ASD II = ostium secundum atrial septal defect; AVSD = atrioventricular septal defect; BCPS = bidirectional cavopulmonary shunt; DORV = double outlet right ventricle; MAPCAs = main pulmonary artery collaterals from aorta; MV = mitral valve; PA = pulmonary atresia; PAPVC = partial anomalous pulmonary venous connections; PDA = patent ductus arteriosus; PV = pulmonary valve; RV-MPA = right ventricle-to-main pulmonary artery; RVOTO = right ventricle outflow tract obstruction; TGA = transposition of the great arteries; VSD = ventricular septal defect.

arrhythmias were present in the remaining 199 patients (9.9%) and included the following: (1) atrial fibrillation in 106 patients (53.3%); (2) III degree AV block in 44 patients (22.1%); (3) supraventricular tachycardia in 21 patients (10.6%); (4) atrial flutter in 15 patients (7.5%); and (5) other arrhythmias in 13 patients (6.5%). Twenty-two patients had a previous pacemaker implantation.

Preoperative variables which are commonly associated to an increased hospital morbidity and mortality were

identified in 397 patients (19.7%), being multiple in 150 patients (37.7%). Preoperative cyanosis (247 patients, 12.3%) ($p < 0.0001$), preoperative arrhythmias (199 patients, 9.9%) ($p < 0.03$), and preoperative NYHA class III and IV (< 0.0001) were the only causes linked to an increased hospital mortality. The principal basic diagnoses and diagnoses leading to surgery according to surgical category (ie, palliative procedures, corrective procedures, or reoperations), are presented in Tables 1 and 2. Listed in

Table 4. Postoperative Complications

Complication	n
Arrhythmias	154
Bleeding	63
Low output syndrome	42
Pericardial effusion requiring drainage	35
Pleural effusion	35
Pneumothorax	32
Reoperation during same admission	29
Mechanical ventilation ≥7 days	25
Third degree atrioventricular block requiring permanent pace maker	22
Wound infection	22
Pneumonia	19
Postoperative septicemia	19
Neurological deficit persisting at discharge	19
Renal failure	17
Tracheostomy	12
Laryngeal nerve injury	8
Cardiac arrest	6
Phrenic nerve injury	6
Other postoperative complications	19

Table 3 are the main surgical procedures within their respective surgical categories. One-fourth of the patients (528 of 2,012, 26.2%) required additional surgical procedures: 504 for intracardiac anomalies (95.4 %) and 24 for extracardiac anomalies (4.6%). Intraoperative atrial ablation was performed concomitantly in 42 patients (2%), while 3 patients required a positioning of an internal cardiac defibrillator. A pacemaker implantation was necessary in 38 patients (1.9%).

Major postoperative complications occurred in 444 patients (22%). One hundred eighteen patients (5.9%) presented multiple complications (Table 4). There were 40 hospital deaths (2%). The most common causes were the following: (1) sepsis (13 patients, 32.5%); (2) low cardiac output syndrome (12 patients, 30%); followed by (3) arrhythmia (7 patients, 17.5%); (4) perioperative

bleeding (5 patients, 12.5%); and (5) neurological events (3 patients, 7.5%). Mortality was lower for corrective procedures (1.2%) when compared with reoperations (4.4%) and with palliative procedures (7.7%)

At the time of hospital discharge, 92% of the survivors were in sinus rhythm (1,813 of 1,972 patients). Postoperative arrhythmias were present in the remaining 159 patients (8%), of which 109 were untreated arrhythmias and 55 new-onset arrhythmias. They included atrial fibrillation in 76 patients (47.8%), third degree AV block in 64 patients (40.2%), atrial flutter in 7 patients (4.4%), and other arrhythmias in 12 patients (7.6%).

Regarding surgical categories, most of the procedures were corrective procedures (1,509 patients, 75%), followed by reoperations (464 patients, 23.1%), and palliative procedures (39 patients, 1.9%). Patients who underwent corrective procedures presented with a significantly lower mortality rate ($p = 0.001$) requiring also less time in the intensive care unit ($p = 0.002$) and a shorter total hospital stay ($p = 0.0001$) (Table 5).

Six hundred forty-nine patients underwent isolated ASD II closure (32.2%), while 1,363 patients (67.8%) required surgical treatment for all other CHD. Patients who underwent isolated ASD II closure had a less complicated postoperative course, requiring less time in the intensive care unit ($p < 0.0001$) and a shorter total hospital stay ($p < 0.0001$) with a hospital mortality of 0.2%. Nonetheless, even after eliminating patients who underwent isolated ASD II closure, the overall mortality of the remaining 1,324 patients was still low (2.9%).

Follow-up data were available in 1,342 of 1,972 patients (68%) who were discharged from the hospital. Fourteen European centers provided us with follow-up data, which was 91% completed. Five European centers could not provide follow-up data but only with operative data. In these centers, patients were referred for surgical treatment and were later followed by the referring physician of peripheral cardiologic units.

Late deaths occurred in 6 patients (0.5%), 2 of which were not cardiac-related. Reoperation was necessary in 12 of 1,342 patients (0.9%), within a mean time of 29.5 ±

Table 5. Surgical Categories

Variables Analyzed	Palliative Procedures	Corrective Procedures	Reoperations	p Value
Number of procedures	39 (1.9%)	1509 (75%)	464 (23.1%)	
Sex M/F	20/19	778/731	207/257	0.03
Mean age in years (SD)	29.48 (11.6) ^{a,c}	36.21 (15.1) ^b	28.94 (10.7) ^c	<0.0001
Preoperative sinus rhythm	37 (94.9%) ^b	1383 (91.7%) ^b	393 (84.7%) ^c	<0.0001
Preoperative cyanosis	35 (89.7%) ^d	143 (9.5%) ^b	69 (14.9%) ^c	<0.0001
Postoperative complications	20 (51.6%) ^d	312 (20.7%) ^b	111 (23.9%) ^c	<0.0001
Sinus rhythm at discharge	34 (94.4%)	1377 (92.4%)	402 (90.3%)	0.33
Mean ICU stay/days (SD)	3.71 (4.4) ^c	2.31 (5.4) ^b	3.25 (5.6) ^b	0.002
Mean hospital stay/days (SD)	14.17 (8.0) ^b	10.54 (9.0) ^b	12.53 (9.7) ^c	0.0001
Hospital mortality (%)	3 (7.7%) ^c	18 (1.2%) ^b	19 (4.1%) ^c	0.0001

^a In each category in which the p value is significant, those values with different letters (b, c, and d) are statistically significantly different in the multiple comparison (Tukey) test ($p < 0.05$).

ICU = intensive care unit.

after their initial correction, and including mainly conduit placement-change (190 of 464 patients, 41%), pulmonary valve change (45 of 464 patients, 10%), and aortic valve replacement (35 of 464 patients, 7.5%). The relative low percentage of reoperations (23.1%) among all surgical procedures could be justified by the increasing number of interventions in the catheterization laboratory (such as for atrial septal defect closure, radiofrequency ablation, pacemaker implantation, etc). As a consequence the number of patients with a residual isolated ASD II could also be underestimated. In fact, many patients could probably have been referred for percutaneous transcatheter treatment after surgery and therefore not entered in our surgical database.

A follow-up analysis shows an overall survival probability of 97% at 60 months, which is higher for corrective procedures (98.2%) if compared with reoperations (94.1%) and palliations (86.1%). Survival probability of patients requiring closure of an isolated ASD II is 99.1% at 60 months. After excluding the ASD II patients, survival rate at 60 months for all other patients was still 95.5%.

The majority of these patients survived into adult age (≥ 18 years) in good functional status, considering that 77.2% of them was in preoperative NYHA class I or II. Our data suggest that an adult congenital patient is treated early in his clinical presentation (NYHA class I-II), the hospital outcome can be better when compared to that of patients who presented with worst functional status (NYHA class III-IV) with a significant increase in functional status at midterm survival: 1,306 of 1,342 patients (97.4%) who were discharged from the hospital were in NYHA class I-II at the 18.5 ± 15.1 months last follow-up control ($p < 0.001$).

Obviously, long-term postoperative functional results of these patients are extremely important to know, unfortunately we were able to collect follow-up information only in 68% of the patients and limited to 60 months after the operations (mean of 18.5 ± 15.1 months). The main reason for this limitation of this study was that most of the patients were referred to the tertiary centers for surgical treatment and were later followed by the referring physician of peripheral cardiologic units. According to that, believing that tertiary units have the duty to find out what happens to the patients referred to them for surgery, we are now developing an educational program with the aim to involve peripheral unit in our data collection.

According to our data, surgical treatment of CHD in adults when performed in specialized European cardiac facilities, proved to be quite safe, beneficial, and low risk. We believe that a 2% hospital mortality in our study population compared well with the 5% mortality rate from the European Congenital Database [19–25]. It is obvious that in the adult population we do not encounter very complex CHD; ie, hypoplastic left heart syndrome, complex functional single ventricle malformations, heterotaxia syndrome, etc.

The low mortality for adult patients who require surgery for CHD, when compared with the pediatric population, may be attributable to the high level of specializa-

tion coming from the pediatric cardiac units participating in the study. In fact, the care of adult patients with corrected or uncorrected CHD is growing and already is a challenging new field in cardiology. It requires knowledge of the following: (1) pathology of congenital cardiac malformations; (2) treatment options (ie, surgery versus interventional cardiology); (3) postoperative cardiac and noncardiac complications and sequelae; (4) acquired cardiac and noncardiac diseases that appear with age; and eventually, according to the high incidence of preoperative and postoperative arrhythmias (5) the need for expertise in electrophysiology [3, 17, 18]. To address the many different needs of this complex and growing patient population, and in order to have a good outcome, this complex group of patients with CHD should be addressed to these units, and also new specialized units are needed around the world [26–27].

Furthermore, while a risk-adjusted method for surgery of acquired heart diseases and surgical treatment of pediatric patients with congenital heart disease have been well established and refined over the last 10 years, [28–31], a risk-adjusted model to predict in-hospital mortality for adult patients with CHD is still lacking. This will be of fundamental importance to allow a meaningful comparison and to evaluate surgical risks of this emerging group of patients.

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