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Surgical Outcome for Congenital Heart Malformations in the Adult Age: A Multicentric European Study

G. Stellin, V.L. Vida, M.A. Padalino, and G. Rizzoli

Congenital heart diseases are currently treated in the pediatric age. However, a conspicuous number of patients requires treatment in the adult age. This study has been undertaken by members of the European Congenital Heart Surgeons Association with the aim of evaluating the impact of cardiac surgery in this particular age group. We have collected data from 1,247 patients who underwent 1,287 operations during a 5-year period between January,1 1997 and December 31, 2001. Patients were divided into three groups: (1) palliative procedures (4.4%), operation performed to improve patients' clinical status without restoring normal anatomy or physiology; (2) repair (79.3%), operation performed to achieve an anatomic or physiologic repair 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); (3) reoperation (16.3%), all the reoperations performed after repair (either anatomic or physiologic). Hospital mortality (within 30 days) was 2.4% (range, 0% to 15.3% in different centers). Kaplan-Meier estimates shows a 94% survival at 4 years, which is higher for repair (95%) as compared with reoperations (92%) or palliations (88%). Surgery for congenital heart disease in the adult age is a safe, beneficial, and low-risk treatment that modifies patients' natural history by improving their clinical status.

Key words: Congenital malformations; adult; multicentric European study.

Congenital heart diseases are currently treated in the pediatric age. However, a conspicuous number of patients require treatment in the adult age.[1] To evaluate the impact of cardiac surgery in this particular group, we have embarked on a multicentric European study involving 19 centers from 13 different European countries.

Materials and Methods

Included in this multicentric study are 1,247 adult patients with congenital heart disease, 634 men (51%) and 613 women (49%), ranging in age between 18 and 78 years (mean, 33.6 years). There were 1,287 procedures performed between January 1, 1997 and December 31, 2001, in 19 European centers (median, 45 procedures/center; range, 11 to 202 procedures). Procedures were divided into three groups:

Group I. Palliative procedures (4.4%): There are 57 palliative procedures performed in 57 patients. In this subset we have included any operations performed to improve the patient's clinical status, without restoring normal anatomy or physiology. Among palliation, 35% were bidirectional cavopulmonary anastomosis, 24% systemic-to-pulmonary artery shunts, 9% pulmonary artery banding, etc.

Group II. Repair (79.3%): Included are 1,021 operations to achieve an anatomic or physiologic repair 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. Among repairs, 31% were atrial septal defect (ASD) closure, 5% ventricular septal defect closure, 5% aortic valve replacement, etc.

Group III. Reoperation (16.3%): All the reoperations performed after repair (either anatomic or physiologic). There were 209 reoperations

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Table 1. Diagnoses and Diagnoses Leading to Surgery Classified According to Pediatric European Database Classification

Categories	Diagnosis No. (%)	Diagnosis Leading to Surgery No. (%)
Septal defects	695 (39.9)	646 (40.9)
Pulmonary venous anomalies	68 (3.9)	68 (4.3)
Systemic venous anomalies	7 (0.4)	6 (0.4)
Right heart lesions	362 (20.8)	316 (20.0)
Left heart lesions	312 (17.9)	291 (18.4)
Single ventricle	54 (3.1)	44 (2.8)
TGA	51 (2.9)	19 (1.2)
DORV	13 (0.7)	6 (0.3)
Thoracic arteries/veins	99 (5.7)	84 (5.3)
Lung disease	9 (0.5)	6 (0.3)
Electrophysiologic	23 (1.3)	24 (1.5)
Other	50 (2.9)	69 (4.6)
Total	1,743 (100)	1,579 (100)

Abbreviations: TGA, transposition of the great arteries; DORV, double outlet right ventricle.

performed in 193 patients. Among reoperation, 30% were conduit replacement, 8% aortic valve replacement, 6% ventricular septal defect closure, etc.

Diagnoses, procedures, extracardiac anomalies, preoperative risk factors, and postoperative complications were classified according to Pediatric European Database classification. We have 1,743 anatomic diagnoses in 1,287 patients. Listed in Table 1 are diagnoses of "basic" heart malformations and diagnoses "leading to surgery" before each operation.

Preoperative Data

A clinical evaluation was performed in all patients before surgery (98% of data available). Noninvasive preoperative diagnosis by means of 2-dimensional echocardiography Doppler (± other noninvasive techniques) was possible for 491 operations (39%). Cardiac catheterization was used in adjunct in the remaining 770 patients (61%).

According to a preoperative clinical evaluation and instrumental evaluation, 408 patients (34.4%) were classified in the New York Heart Association (NYHA) class I, 524 in NYHA class II (42.1%), 241 in NYHA class III (19.3%), and 51 in NYHA class IV (4.2%).

A preoperative electrocardiogram was performed in all patients. Eighty-eight percent of patients were in sinus rhythm. Major arrhythmias included atrial fibrillation in 4%, complete atrioventricular block in 2%, and atrial flutter in 1%.

Associated extracardiac anomalies were present in 24 patients and included Down's syndrome in 17 (1%), Turner's syndrome in 4 (0.2%), and Williams Beuren syndrome in 3 (0.2%). Other nonsignificant chromosomal anomalies were present in 13 patients (0.8%). Other noncardiac nonchromosomal anomalies were found in 35 patients (2.2%).

Preoperative risk factors were present in 305 patients and included cyanosis in 187 patients (14.5%), preoperative arrhythmias in 31 (2.4%), preoperative neurological deficit in 19 (1.4%), preoperative endocarditis in 16 (1.2%), and other risk factors in 52 patients (4%).

Statistical Analysis

Survival estimate curves for all procedures for different operative categories (palliation, repair, and reoperation), according to the main pathologies (ASD, ventricular septal defect, aortic valve disease, single ventricle, tetralogy of Fallot, and pulmonary atresia), for ASD in comparison with other pathologies and according to preoperative cyanosis were analyzed according to the Kaplan-Meier method. Hazard ratio was estimated by means of Cox analysis of the influence of operative categories of correction, pathologies, and preoperative risk factors.

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Table 2. Procedures Classified According to Pediatric European Database Classification

Categories	Procedure No. (%)
Septal defects	676 (40.6)
Pulmonary venous anomalies	65 (3.9)
Systemic venous anomalies	6 (0.3)
Right heart lesions	321 (19.3)
Left heart lesions	304 (18.2)
Single ventricle	28 (1.6)
TGA	8 (0.4)
DORV	2 (0.1)
Thoracic arteries and veins	85 (5.1)
Lung disease	8 (0.4)
Electrophysiologic	37 (2.2)
Palliative procedures	51 (3.0)
Miscellaneous	81 (4.9)
Total	1672 (100)

Abbreviations: TGA, transposition of the great arteries; DORV, double outlet right ventricle.

Operative Results

There were a total of 1,672 surgical procedures performed in 1,287 patients (100% of data available). In Table 2, we have summarized the procedures according to the European database short list categories. T2

Thirty-one patients died in hospital (within 30 days) with an overall mortality of 2.4% which ranged from 0% to 15.3% in different centers. The most common causes reported were low output syndrome (6 patients, 0.7%), sepsis (7 patients, 0.5%), and perioperative bleeding (2 patients, 0.1%). Five patients died from other less-frequent causes (0.1%). No data were available in 11 cases.

Mean intensive care unit stay was 2.9 days (range, 1 to 173 days); 4.4 days in group I, 2.7 days in group II, and 4.4 days in group III.

Major postoperative complications were reported in 286 patients (23.1%). The most common complications included: postoperative arrhythmias (81 events), bleeding requiring reoperation (47 events), pericardial effusion requiring drainage (21 events), postoperative mechanical ventilation for more than 7 days (20 events), acute renal failure requiring temporary dialysis (18 events), and low output syndrome (17 events). Other less common complications were reported for 105 operations (8.5%).

Cardiac rhythm at the moment of discharge from the hospital showed 90% of patients with sinus rhythm by electrocardiogram, while 6% showed major arrhythmias that included atrial fibrillation in 3.6%, complete atrioventricular block (ill degree) in 2.2%, and atrial flutter in 0.6%, etc.

Follow-Up

Follow-up period ranged from 1 month to 60.5 months (mean, 17.7 months; median, 11.6 months) and data were available for 810 patients (63% of data available).

There were 82 adverse events reported in 75 patients (10%). Late death occurred in 11 patients (1.5%). In five patients, death was noncardiac related. Reoperation was necessary in 19 patients (2.5%) and interventional cardiology procedure in 3 (0.4%). Other less common adverse events were reported in 49 patients.

At latest follow-up, 77.6% of the patients are in NYHA class I, 20.2% in HYHA class II and 2.2% in HYHA class III. This compares favorably with preoperative data. Considering the Ability Index at last follow-up, it was I degree in 82.7%, II degree in 14.8%, and III degree in 2.5%.

Statistical Analysis

Survival curves according to Kaplan-Meier showed:

1. Survival at 5 years for all procedures is 94%, with 95% confidence interval between 92% and 96%.
2. Survival estimate is 88% at 4 years for palliative procedures, 95% at 5 years for repairs and of 92% reoperations (Fig 1). F1
3. Survival estimates is 98% at 5 years for ASD and 91% at 5 years for all other pathologies (Fig 2). F2
4. Survival estimates according to main pathologies is 99% at 5 years for ASD, 95% at 5 years for aortic valve disease, 92% at 4 years for ventricular septal defect, 86% at 3 years for single ventricles, and 61% at 3 years for tetralogy of Fallot and pulmonary atresia (Fig 3). F3
5. Survival estimate is 96% at 5 years for preoperative acyanotic patients and 83% at 4 years for preoperative cyanotic patients (Fig 4). F4

Hazard ratio by means of univariate Cox analysis is 3.1 for palliation ($P = .03$), 0.41 for repair ($P = .008$), and 1.87 for reoperation ($P = .07$).

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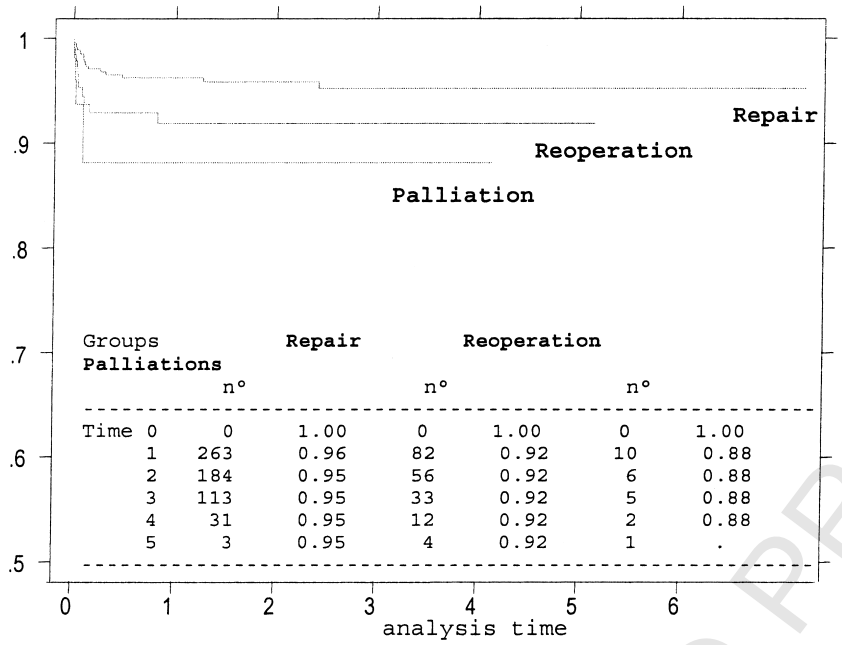

Figure 1. Survival estimates according to Kaplan-Meier, by operative categories.

Hazard ratio by means of univariate Cox analysis is 6.5 for tetralogy of Fallot ($P < .0001$), 3.4 for single ventricle ($P = .04$), and 0.11 for ASDs ($P < .0001$). Hazard ratio by means of univariate Cox analysis is 4.9 for Down syndrome ($P = .008$) and 5.2 for preoperative cyanosis ($P < .0001$) (Table 4).

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Discussion

The number of adult patients with congenital heart defects has been growing steadily since the 1960s. In 2000, the number of adult patients is roughly equal to those under pediatric age. Twenty years from now, the number of adult

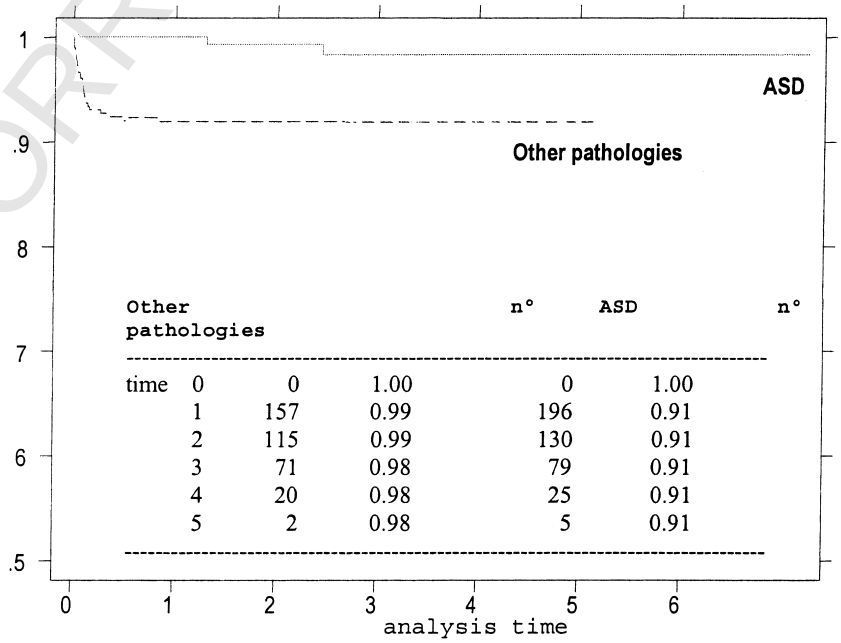

Figure 2. Survival estimates according to Kaplan-Meier, by ASD versus other pathologies. ASD, atrial septal defect.

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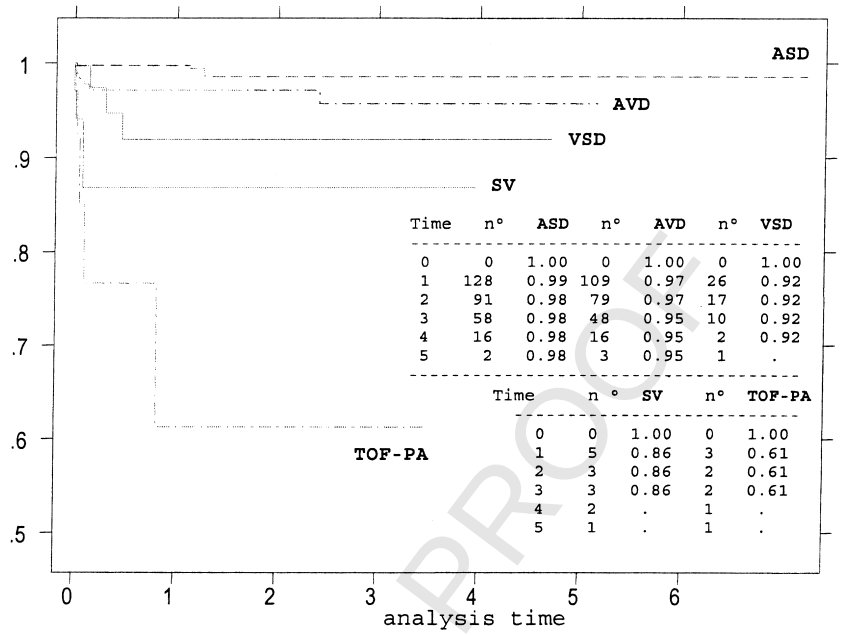

Figure 3. Survival estimates according to Kaplan-Meier, by pathologies. ASD, atrial septal defect; AVD, aortic valve disease; VSD, ventricular septal defect; SV, single ventricle; TOF-PA, tetralogy of Fallot-pulmonary atresia.

patients will exceed those in pediatric care by a considerable margin.[1]

Surgery for congenital heart disease has also changed considerably in the last two decades. Primary repair has steadily decreased beyond the pediatric age because of the improved results with correction in neonates and infants and better screening of asymptomatic patients. However, the success in managing infants and children with CHID have created an emerging population of adult patients who may require reoperation after an initial repair of their heart malformation.[2-4] AQ: 2

There are no data available in the literature analyzing the impact of cardiac surgery, referring

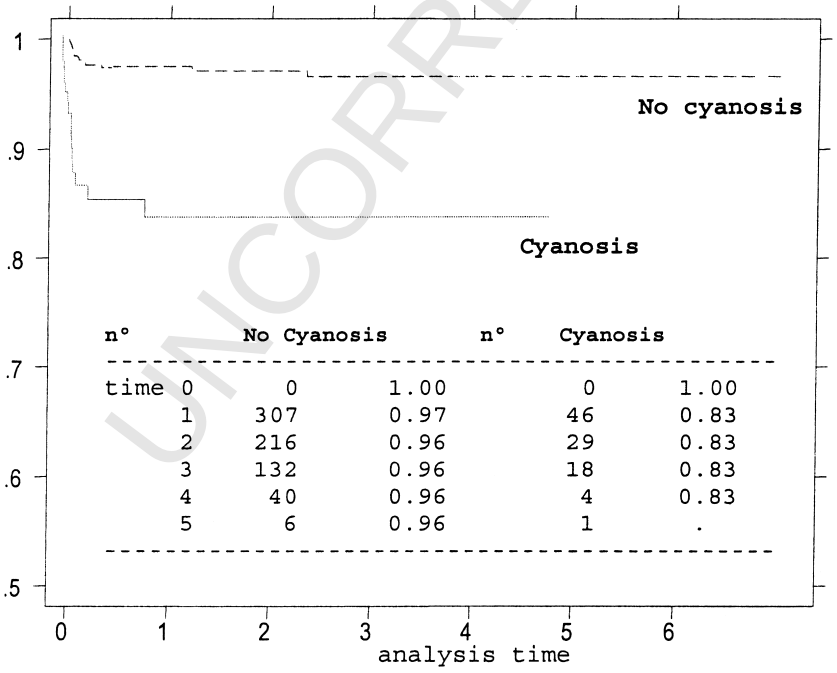

Figure 4. Survival estimates according to Kaplan-Meier, by preoperative cyanosis.

to the most recent years. For this reason, our study has been undertaken with the aim of assessing the results of cardiac surgery in this particular age group and analyzing data of a large population referred in a short period of time.

Surgery for CHID in adult is based on the need to improve the patient's natural history. While natural history in children is well known, natural history of the adult population is less certain or unknown.[3] Adult patients are generally referred for surgery because of increasing symptoms not controlled by medical therapy.[4-8]

In our analysis of a multicentric experience, the most striking result is that the risk for cardiac surgery in the entire population is indeed very low (2.4%) and comparable to that in the pediatric population.[5]

Our statistical analysis shows an estimated cumulative survival of 94% at 4 years, which is higher for complete repair (95%) as compared with reoperations (92%) and palliations (88%). This showed that when repair of congenital heart defects is not performed at a "proper time," this can still be carried out with a low mortality risk, improving patients' quality of life. This is also true for patients who underwent a physiologic correction by means of a Fontan repair and one and a half ventricle repair.[8,9]

Reoperation after repair also shows a good survival estimate. This will include patients with a residual lesion after repair (ie, residual ventricular septal defect, residual right or left ventricular outlet obstruction, residual valve dysfunction, etc) and/or patients who developed further heart disease after their initial repair (ie, right ventricular failure for pulmonary regurgitation after Fallot' tetralogy repair, conduits obstruction/overgrowth, valve diseases, coronary arteries disease, right atrium enlargement/thrombosis after Fontan procedures, etc) In this group of patients, the intrinsic risk for intraoperative hemorrhage caused by repeated sternotomies also needs to be taken into account.

A palliative procedure performed in the adult is undertaken with the aim of temporarily improving clinical status in patients with no chance for correction. Surgery in this group still shows a positive impact by improving quality of life with an acceptable mortality. However, in this group, results should be compared with those for heart or heart/lung transplantation in CHID.

Repair of simple lesions such as ASDs, ventricular septal defects, or atrioventricular septal defects shows a better survival estimate when compared with lesions characterized by a functionally single ventricle. However, survival estimate is superior for functionally single ventricle lesions when compared with a complex biventricular lesion as tetralogy of Fallot and pulmonary atresia.

Approximately one third of our population consisted of repair of ASDs. It is impressive to see that still, in the adult age, so many ASDs are closed surgically rather than with a transcatheter device. In our Kaplan-Meier survival estimates (Fig 9), we compared survival at 5 years of ASD alone with other pathologies. Surgery for lesions other than ASD still shows a good 5-year survival (91%).

It is also noteworthy that[4,5] patients presenting with a preoperative cyanosis have a higher surgical risk and lower survival estimates at 4 years when compared with noncyanotic patients.

Conclusion

This study has been undertaken by pediatric cardiac surgeons with the aim of evaluating the impact of cardiac surgery in this particular age group. We were able to collect follow-up information on 63% of patients.

Surgery for CHID in the adult age is a safe, beneficial, and low-risk treatment that modifies the patient's natural history by improving their clinical status. Kaplan-Meier survival curves show a cumulative survival of 94% at 4 years, which is higher for complete repair (95%) as compared with reoperations (92%) or palliations (88%). Correction of CHID in the adult age is often a special problem requiring expertise and experience to ensure good outcome.[3] Most congenital heart surgery should be performed by congenital heart surgeons. However, most congenital heart surgeons are based in pediatric units and practice primarily in children. Mixed units should be developed where pediatric cardiologists cooperate with adult cardiologists and where the pediatric cardiac surgeons can find proper facilities to treat adults with congenital hearts outside pediatric units.[4-6]

Acknowledgment

On behalf of the Members of the European Congenital Heart Surgeons Association: W. Daenen,

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