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Early Surgical Morbidity and Mortality in Adults with Congenital Heart Disease: The University of Michigan Experience

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ABSTRACT

Objectives. To review early surgical outcomes in a contemporary series of adults with congenital heart disease (CHD) undergoing cardiac operations at the University of Michigan, and to investigate possible preoperative and intraoperative risk factors for morbidity and mortality.

Methods. A retrospective medical record review was performed for all patients ≥18 years of age who underwent open heart operations by a pediatric cardiothoracic surgeon at the University of Michigan Congenital Heart Center between January 1, 1998 and December 31, 2004. Records from a cohort of pediatric patients ages 1–17 years were matched to a subset of the adult patients by surgical procedure and date of operation.

Results. In total, 243 cardiac surgical operations were performed in 234 adult patients with CHD. Overall mortality was 4.7% (11/234). The incidence of major postoperative complications was 10% (23/234) with a 19% (45/23) minor complication rate. The most common postoperative complication was atrial arrhythmias in 10.8% (25/234). The presence of preoperative lung or liver disease, prolonged cardiopulmonary bypass and aortic cross clamp times, and postoperative elevated inotropic score and serum lactates were significant predictors of mortality in adults. There was no difference between the adult and pediatric cohorts in terms of mortality and morbidity.

Conclusions. The postoperative course in adults following surgery for CHD is generally uncomplicated and early survival should be expected. Certain risk factors for increased mortality in this patient population may include preoperative presence of chronic lung or liver dysfunction, prolonged cardiopulmonary bypass and aortic cross-clamp times, and postoperative elevated inotropic score and serum lactate levels.

Key Words. Adults; Congenital Heart Disease; Morbidity; Mortality; Risk Factors

Introduction

There are an increasing number of children with congenital heart disease (CHD) surviving to adulthood because of significant advances in operative techniques and medical care. Fifty years ago, only 25% of infants born with CHD survived beyond the first year of life,[1] but today 80–85% can expect to reach adulthood[2] with an estimated 800 000 adults living with CHD in the United States alone.[1] As this patient population has aged, new challenges have surfaced including the potential for additional operations to prevent further progression of residual or recurrent heart disease in patients with complex defects originally repaired during childhood. These cardiac operations in adults are becoming more commonplace in many congenital heart centers across the country. Despite the increasing frequency of adults with CHD undergoing cardiac operations, there are limited published data on early outcomes. A contemporary review of 438 adult patients with CHD over a 13-year period showed an overall mortality of approximately 6%, with increasing trends in the percentage of repeat operations and in the complexity of diagnoses and operative procedures in the most recent era.[2] Other publications in adults with CHD suggest that early operative mortality is increased in cyanotic patients and in patients undergoing repeat operation.[3–5]

The objectives of this study were: (1) to describe the early postoperative course in adults with CHD undergoing cardiac surgery at a single institution; (2) to compare early morbidity and mortality data in adult CHD patients with a cohort of pediatric CHD patients undergoing similar surgical procedures during the same era; and (3) to investigate possible preoperative and intraoperative risk factors for significant morbidity and mortality in adults with CHD.

Methods

A retrospective chart review was conducted of the medical records for all patients ≥18 years of age undergoing cardiac operations by any of the 3 pediatric cardiothoracic surgeons at C.S. Mott Children's Hospital (University of Michigan) for a 7-year period between January 1, 1998 and December 31, 2004. Patients operated at University Hospital were excluded from analysis. The research protocol was approved by the Institutional Review Board for Human Subject Research at the University of Michigan Medical School and informed consent was waived.

Preoperative data collected included the primary diagnosis, type of procedure, age at operation, gender, previous cardiac surgeries (number and type), pulse oximetry on room air, and comorbidities. The preoperative comorbidities of interest included a history of arrhythmias (atrial and/or ventricular), presence of a genetic syndrome (e.g., Marfan's, Down, DiGeorge, etc.), history of neurological abnormalities (seizures, encephalopathy, brain abscess), history of heart block and/or pacemaker dependence, presence of liver dysfunction and/or hepatitis (B or C), significant ventricular dysfunction, presence of chronic lung disease, and current alcohol or cigarette use. Liver dysfunction was defined by liver function tests ≥twice the upper limits of normal or by cirrhosis noted on liver biopsy. Significant ventricular dysfunction was present if it was classified as "severe" or "cardiomyopathic" on a preoperative echocardiogram report or hospital admission note. Chronic lung disease was defined by abnormal pulmonary function tests (consistent with an obstructive or restrictive pattern) and a requirement for chronic medical therapy.

Postoperative data collected included mortality, cardiopulmonary bypass and aortic cross clamp times, inotropic score (dopamine [μg/kg/min] + dobutamine [μg/kg/min] + 100 × epinephrine [μg/kg/min] + 10 × milrinone [μg/kg/min]), blood lactate, significant postoperative complications, length of mechanical ventilation, and intensive care unit (ICU) and hospital lengths of stay. Significant postoperative complications included arrhythmias requiring antiarrhythmic treatment, renal insufficiency (creatinine ≥ 2 × preoperative values), seizures or encephalopathy, reoperation during the same hospitalization, mediastinal bleeding (requiring re-exploration), ventricular dysfunction, liver dysfunction (elevated liver enzymes ≥ 2 × preoperative values), gastrointestinal bleeding, and atrioventricular block requiring pacemaker placement.

In order to compare adult patients to pediatric patients, a temporally matched (operation within 1 month of adult patient) cohort of pediatric patients (aged 1–17 years) who had undergone Ross procedure were compared with the adult patients who had undergone Ross operation. Similarly, a group of pediatric patients who underwent right ventricle to pulmonary artery (RV to PA) conduit replacement were used for comparison to the adult patients with RV to PA conduit replacement.

For all analyses, the Student's *t*-test was used to analyze normally distributed continuous variables. Wilcoxon rank sum scores were used to analyze continuous variables which were not normally distributed. The chi-squared test was used to determine significance for categorical variables. Multivariable logistic models were composed to evaluate the independent impact of potential risk factors on mortality.

Results

In total, 243 cardiac surgical operations were performed in 234 adult patients with CHD during this 7-year period. These operations represented approximately 5% of all cardiac operations performed by our pediatric cardiothoracic surgeons during that same era (epicardial pacemaker lead placement and minor surgical procedures excluded). A total of 233 patients underwent surgery requiring cardiopulmonary bypass and 1 patient with a vascular ring underwent repair without bypass. All patients were recovered in the pediatric cardiothoracic intensive care unit (PCTU). The mean age at time of surgery was 30 ± 11 years (range: 18–67 years), with an approximately equal male to female ratio (49% male [n = 115], 51% female [n = 119]). Sixty-one percent of the patients had a history of previous cardiac surgery, and 10% of the patients had cyano-

Table 1. Congenital Heart Disease Primary Diagnostic Categories

Diagnoses	n = 234
Left ventricular outflow tract lesions	69
Aortic stenosis	23
Aortic regurgitation	22
Aortic root disease	12
Coarctation of the aorta ± VSD	7
Subaortic stenosis	4
Vascular ring	1
Right ventricular outflow tract lesions	64
Tetralogy of Fallot	34
Tetralogy of Fallot with pulmonary atresia	9
Double chamber right ventricle ± VSD	8
Double outlet right ventricle ± VSD	5
Pulmonary stenosis	4
Absent pulmonary valve syndrome	3
Pulmonary atresia with intact ventricular septum (biventricular repair)	1
Left-to-right shunts	47
ASD ± PAPVR	27
VSD	4
AVSD	10
PAPVR with no ASD	6
Single ventricle	12
Double inlet left ventricle	5
Tricuspid atresia	3
Double outlet right ventricle, VSD, hypoplastic left ventricle	2
Unbalanced AVSD	1
Tetralogy of Fallot with VSD	1
Acquired heart disease	7
Cardiomyopathy	2
Trauma	2
Endocarditis	1
Rheumatic disease	1
Transplant coronary artery vasculopathy	1
Other	35
D-TGA ± VSD	12
Ebstein's/tricuspid valve regurgitation or stenosis	9
L-TGA ± VSD	4
Mitral valve disease	4
Truncus arteriosus	3
Cor triatriatum	2
Coronary artery abnormalities	1

VSD, ventricular septal defect; ASD, atrial septal defect; PAPVR, partial anomalous pulmonary venous return; AVSD, atrioventricular septal defect; D-TGA, dextro-transposition of the great arteries; L-TGA, levo-transposition of the great arteries.

sis at rest (defined as an arterial O_2 saturation <94%). The primary cardiac diagnoses were categorized as: left ventricular outflow tract abnormalities (29%; nearly two-thirds congenital aortic stenosis or regurgitation), right ventricular outflow tract abnormalities (27%; primarily tetralogy of Fallot and variants), left-to-right shunts (20%), or functional single ventricle lesions (5%). Seven patients had acquired heart disease, and 35 patients had a variety of other complex cardiac lesions (Table 1).

Preoperative Comorbidities

Several of the adult patients with CHD had a history of preoperative arrhythmias [42 (18%) atrial and 19 (8%) ventricular]. There were 24 patients (10%) diagnosed with a genetic syndrome [Marfan's syndrome (n = 7) Down syndrome (n = 8) DiGeorge syndrome (n = 3) and other (n = 6)]. Twenty adults (8%) had a history of a neurological abnormality (seizures, encephalopathy, or brain abscess), and 13 patients (6%) had a history of liver dysfunction and/or hepatitis. There were 9 patients (4%) with significant ventricular dysfunction, and 13 patients (6%) had a history of heart block and/or pacemaker dependence. Three patients carried the diagnosis of chronic lung disease, and 49 patients (22%) were current cigarette smokers or had a significant past history of cigarette use. There were 11 patients (5%) with a history of heavy alcohol use (defined as ≥6 drinks per day).

Table 2. Cardiac Surgical Procedures

Surgery	n = 243
Aortic valve replacement ± root replacement	50
Pulmonary valve replacement	35
ASD ± PAPVR repair	35
Right ventricle to pulmonary artery conduit	24
Tricuspid valve repair/replacement	13
Primary repair of right ventricular outflow tract	10
Valve sparing aortic root replacement	12
Mitral valve repair/replacement	9
Fontan/Fontan revision	8
Orthotropic heart transplant	8
Coarcation repair	6
Aortic aneurysm repair	6
Primum ASD ± mitral valve repair	6
Aortic valve repair	5
VSD closure	4
Mustard revision	4
Cor triatriatum repair	2
Vascular ring	1
Pulmonary artery unifocalization	1
Coronary unroofing	1
Left ventricle to aorta conduit	1
Right ventricular repair after trauma	1

ASD, atrial septal defect; PAPVR, partial anomalous pulmonary venous return; VSD, ventricular septal defect.

Intraoperative and Postoperative Results for Entire Cohort of Adult Patients

The most common cardiac surgical procedures performed in these adult CHD patients were aortic valve ± aortic root replacement (n = 50), pulmonary valve replacement (n = 35), atrial septal defect closure ± repair of partial anomalous pulmonary veins (n = 35), and RV to PA conduit replacement (n = 24) (Table 2). The median cardiopulmonary bypass time for all procedures was 101 minutes (range: 0–584 minutes) with a median aortic cross clamp time of 46 minutes (range: 0–294 minutes). The median days intubated was 0.5 days (range: 0.5–30 days), and the reintubation

Table 3. Postoperative Mortality

Patient	Age (y)	Diagnosis	Previous Operations	Current Operation	Cause of Death
1	47	TOF/PI	TOF repair and RV to PA conduit	Pulmonary valve replacement	Ventricular failure
2	20	DORV, TGA, hypoplastic LV	Classic Fontan	Extracardiac Fontan	Sepsis and multisystem organ failure
3	30	Ebstein's anomaly	None	Tricuspid valvuloplasty, PM/ICD placement	Sepsis and ventricular failure
4	24	TOF/PA	none	Pulmonary artery unifocalization	Cardiac tamponade and multisystem organ failure
5	30	D-TGA/severe RV dysfunction	Mustard operation	Orthotropic heart transplant	Sudden cardiac arrest
6	30	TOF	Waterston shunt, TOF repair with conduit, and conduit replacement	Conduit replacement, RPA stent	Massive stroke (presumed air embolus)
7	60	PAPVR/coronary artery disease	none	PAPVR repair and coronary bypass	Mediastinitis, sepsis and multisystem organ failure
8	20	Marfan's, aortic aneurysm, mitral regurgitation	Aortic root resection and aortic valve replacement	Mitral valve replacement and Aortic arch graft	Diffuse brain edema, withdrew support
9	61	TOF	Classic BT shunt, Potts shunt, repair of TOF, and pacemaker	Pulmonary valve replacement, tricuspid valve repair	Ventricular failure
10*	41	TOF	TOF repair, RV to PA conduit	Pulmonary valve replacement	Late sternal wound infection, sepsis; died during surgical debridement
11*	22	IAVSD and cardiomyopathy	none	Closure of primum ASD, repair of tricuspid valve	Arrhythmias and ventricular failure

*Late death.
TOF, tetralogy of Fallot; PI, pulmonary insufficiency; RV, right ventricle; PA, pulmonary atresia; DORV, double-outlet right ventricle; D-TGA, dextro-transposition of the great arteries; PM, pacemaker; ICD, implantable cardiac defibrillator; RPA, right pulmonary artery; PAPVR, partial anomalous pulmonary venous return; BT, Blalock-Taussig; IAVSD, incomplete atrioventricular septal defect; ASD, atrial septal defect.

rate was 3% (n = 7). The median inotropic score at PCTU admission was 3 (range: 0–150), and at 24 hours was 3.5 (range: 0–170). The median lactate level at PCTU admission was 1.8 mmol/L (range: 0.5–9.4 mmol/L), and at 24 hours was 1.2 mmol/L (range: 0.3–34 mmol/L). The median ICU length of stay was 1 day (range: 1–89 days), and the median hospital length of stay was 5 days (range: 3–103 days). Overall mortality was 4.7% (11/234) with 9 early deaths (<30 days after surgery) and 2 late deaths (≥30 days) (Table 3).

Table 4. Postoperative Complications

Complication	Percentage	n
Arrhythmias	14	33
Atrial	10.8	25
Ventricular	3.4	8
Renal insufficiency	4.3	10
Seizures/encephalopathy	3.9	9
Reoperation	2.6	6
Bleeding requiring re-exploration	2.6	6
Ventricular dysfunction	1.7	4
Liver dysfunction (failure, varices)	0.4	1

Postoperative Complications

The overall incidence of significant postoperative complications was 29% (68/234). Complications were further subdivided into major and minor complications. A major complication was defined as cardiac arrest, reoperation, acute lung injury, ventricular failure requiring mechanical support, renal failure requiring renal replacement therapy, hepatic failure, acute neurological injury with sequelae, and pacemaker placement. Minor complications were defined as arrhythmias requiring therapy, renal insufficiency (not requiring dialysis), seizures or encephalopathy with no long-term sequelae, deep venous thrombosis, pulmonary emboli, and surgical bleeding not requiring reintervention. There were 23 (9.8%) major complications and 45 (19%) minor complications. The most common postoperative complication was atrial arrhythmias in 25 patients (10.8%) (Table 4).

Predictors of Mortality and Morbidity

Bivariate analysis demonstrated that preoperative chronic lung disease ($P < .001$) or preoperative hepatic dysfunction ($P = .017$) was associated with an increased rate of mortality. However, there were only 3 patients in this study with chronic lung disease (with 2 deaths), and 3 patients with

Table 5. Functional Single Ventricle Surgical Procedures

Surgery	n
Fontan revision	5
Extracardiac Fontan	3
Heart transplant	2
Bidirectional Glenn	1
Left ventricle to aorta conduit	1

liver dysfunction or cirrhosis (with 2 deaths). Again, on bivariate analysis, prolonged cardiopulmonary bypass ($P = .0017$) and aortic cross-clamp ($P = .05$) times were significant intraoperative risk factors for death. Postoperatively, an elevated initial lactate ($P = .03$), lactate at 24 hours ($P = .0037$), initial inotropic score ($P = .003$) or inotropic score at 24 hours ($P < .0001$) was each associated with increased rate of mortality. Multivariable logistic models demonstrated that the impact of chronic lung disease ($P = .0005$) and prolonged cardiopulmonary bypass time ($P = .0084$) were independent predictors of mortality.

Longer ICU and hospital length of stay was found to be associated with preoperative liver dysfunction ($P = .005$ and $P = .001$) and with cyanosis at rest ($P = .001$ and $P = .0001$). Patients with liver dysfunction ($P = .004$), chronic lung disease ($P = .04$), or cyanosis ($P = .0003$) also remained on the ventilator longer. Adult patients with a history of preoperative atrial arrhythmias were significantly more likely to have postoperative atrial arrhythmias ($P < .001$). In contrast, preoperative ventricular arrhythmias did not predict the presence of postoperative ventricular arrhythmias. A preoperative history of seizures or encephalopathy did not correlate with early postoperative neurological outcomes.

Functional Single Ventricle Patients

Although the numbers were small, patients with single ventricle physiology appeared to be at a particularly high risk for postoperative complications. There were 12 adult patients with functional single ventricle physiology who had undergone previous palliation during childhood with an aortopulmonary shunt, a hemi-Fontan/bidirectional Glenn procedure, a Fontan procedure, or some combination of all 3 procedures. Most of the adult patients in this study underwent Fontan revision (Table 5). One patient (8.3%) died secondary to sepsis and multiorgan system failure, and 2 patients (17%) required epicardial pacemaker placement. Furthermore, the incidence of minor complications was 83% (10/12) and the most common postoperative complication was atrial arrhythmias.

Comparison to a Similar Pediatric Cohort

Overall survival following open heart surgery in adult patients in this study was 95.3%, which compares favorably with the survival rate observed in children less than 18 years old at our institution. To better compare morbidities and mortality of adult patients to those of pediatric patients, 2 subgroups of pediatric patients were compared with adult patients: (1) patients with aortic valve disease who underwent a Ross operation; and (2) patients with tetralogy of Fallot or variant who underwent placement of an RV to PA conduit. These particular surgical procedures were selected because they are commonly performed in both adult and pediatric patients with CHD.

There were 20 adults who underwent a Ross operation who were compared with 20 pediatric patients (1–17 years of age) who underwent a Ross operation. The adult and pediatric cohorts were similar with regard to preoperative comorbidities. The rate of reoperation was 60% in the adult patients, and 65% in the pediatric patients. The overall complication rate in the Ross procedure group was 30% in both adult and pediatric cohorts. There was no difference between pediatric and adult Ross groups in cardiopulmonary bypass time or inotropic score. There was a tendency for aortic cross-clamp time to be longer for the adult patients. There was no difference in hospital length of stay, ICU length of stay or intubation time (Table 6).

Similarly, comparison of 14 adult patients who underwent RV to PA conduit replacement to 14 pediatric patients undergoing the same procedure showed no differences in mortality ($P > .99$) or in the postoperative course (Table 7). In addition, there was no difference in the complication rates between the 2 groups (adult cohort: 28%, pediatric cohort: 21%; $P = $ NS).

Discussion

In this study, we analyzed the preoperative characteristics, postoperative course, and early surgical outcomes following operative repair of CHD in adults. In our institution, adult patients with CHD constitute approximately 5% of all open heart operations performed by our pediatric cardiothoracic surgeons. More than 60% of our adult patients had significant preoperative comorbidities

Table 6. Pediatric Patients Compared with Adults Following Ross Operation

	Pediatric Patients (n = 20)	Adult Patients (n = 20)	*P* Value
Hospital LOS (d): median (range)	4.0 (2–10)	5.5 (3–12)	.15
ICU LOS (d): median (range)	1.0 (1–6)	1.0 (0–2)	>.99
Intubation time (h): median (range)	0.5 (0.5–3.0)	0.5 (0.5–2.0)	>.99
Cardiopulmonary bypass time (min): mean (SD)	160 (33)	168 (34)	.47
Aortic cross clamp time (min): mean (SD)	106 (19)	121 (27)	.06
Initial inotropic score: median (range)	0 (0–5)	0 (0–5)	.19
24-h inotropic score: median (range)	0 (0–11)	0 (0–0)	.16

Ross mortality: 1/20 pediatric patients compared with 0/20 adults—$P > .99$ (Fischers exact test).
LOS, length of stay; ICU, intensive care unit.

Table 7. Pediatric Patients Compared with Adults Following RV to PA Conduit Replacement

	Pediatric Patients (n = 14)	Adult Patients (n = 14)	*P* Value
Hospital LOS (d): median (range)	5 (3–20)	5.5 (1–14)	.95
ICU LOS (d): median (range)	1.0 (1–13)	1.5 (1–11)	>.99
Intubation time (d): median (range)	0.5 (0.5–14)	0.5 (0.5–5)	.96
Cardiopulmonary bypass time (min): mean (SD)	88 (55)	110 (42)	.26
Aortic cross clamp time (min): median (range)	0 (0–54)	0 (0–3)	.99
Initial inotropic score: median (range)	0 (0–7)	3 (0–15)	.24
24-h inotropic score: median (range)	0 (0–27)	0 (0–15)	.57

RV to PA conduit mortality: 1/14 pediatric patients compared with 1/14 adults—$P > .99$ (Fischers exact test).
RV to PA conduit arrhythmias: preoperative—0/14 pediatric patients compared with 2/14 adults—$P = .48$; postoperative—1/14 pediatric patients compared with 0/14 adults—$P \geq .99$.
RV, right ventricle; PA, pulmonary atresia; LOS, length of stay.

that placed them at increased surgical risk. Despite these risks, a relatively high rate of complex lesions and difficult operative procedures (including reoperations), the overall mortality was only 4.7% which compares favorably with our surgical mortality in children <18 years old with CHD, and to published mortality rates from other centers specializing in adults with CHD.[1–5] Major complications were observed in 10% of patients and minor complications in 19%. The most common complication was arrhythmia which occurred in 14% of patients, and over 75% of these arrhythmias were atrial in origin.

A recent study by Sirnathan et al.[2] demonstrated an increase in the percentage of reoperations and surgical complexity in adults with CHD over the last decade compared with 2 earlier surgical eras. However, mortality (6.3%), complication rates, ICU and hospital lengths of stay remained constant throughout their entire surgical experience. These authors hypothesize that these somewhat surprising findings may reflect the relatively young age and paucity of major comorbidities in their study patients. Although the average age of our patients was also relatively young (approximately 30 years old), the high incidence of significant comorbidities in our adult population suggests that good outcomes may be expected regardless of the presence or absence of many preoperative risk factors. In agreement with our data, other studies have found a similarly low early postoperative mortality rate (6–7.6%) in adults with CHD independent of preoperative risk factors.[3–5]

Dore et al.,[3] in a review of 295 adult patients with CHD, found an increased mortality in patients with cyanosis and advancing age. They also demonstrated a higher mortality in patients who underwent reoperations compared with those undergoing their first operation. A similarly designed study by Mott et al.[4] in 102 adult patients demonstrated a longer hospital stay but no higher mortality risk in those patients with lower oxygen saturations. Additionally, this study did not find a significant increase in mortality in patients undergoing reoperation. In our adult subjects, we observed a significant increase in ICU and hospital length of stay in those patients with room air pulse oximetry <94%, but unlike the study from Dore et al.,[3] cyanosis did not predict an increase in mortality. Similar to Mott et al.,[4] we found no effect of reoperation on mortality, ICU or hospital length of stay, despite a 60% incidence of reoperation in our patient population.

Arrhythmias are the most common consequence of CHD in the adult population with atrial

flutter being the most frequent arrhythmia.[6] Atrial flutter has a particularly high prevalence in patients with certain anatomic substrates including univentricular heart, transposition of the great arteries, and tetralogy of Fallot.[7] As expected, postoperative arrhythmias are also not uncommon in adult patients with CHD.[4,6,8] In some studies, the rate of postoperative arrhythmias has been found to be as high as 30%.[5] In our patients, postoperative arrhythmias occurred in 14% of patients and preoperative atrial arrhythmias were predictive of postoperative recurrence.

After arrhythmias, renal insufficiency was the second most common complication and occurred in approximated 4% of our adult patients. However, only 3 patients required renal replacement therapy (with continuous venovenous hemofiltration) and no patient required chronic dialysis.

In this retrospective review, we were able to identify some additional independent perioperative predictors of mortality not previously described. The preoperative presence of chronic lung disease or liver dysfunction, and an elevated inotropic score and lactate level postoperatively were associated with increased mortality. In addition, longer cardiopulmonary bypass and aortic cross-clamp times were also significantly associated with mortality; however, total bypass time has consistently been found to be associated with increased mortality in pediatric patients with CHD.[9] Lastly, we were unable to prove any difference in morbidity or mortality between a subset of adults with CHD and a matched cohort of pediatric patients between 1 and 17 years undergoing similar operative procedures during this same time period.

Several risk stratification models have been published in adult patients undergoing open heart surgery for indications other than CHD (usually coronary artery bypass grafting).[10–13] Two of the more publicized risk stratification models include the EuroSCORE[10] and the clinical severity score from the Cleveland Clinic.[11] The EuroSCORE[10] stratifies patients as low, moderate or high risk for mortality based on preoperative factors such as age, serum creatinine, history of chronic pulmonary disease, previous cardiac surgery, angina, left ventricular dysfunction, and recent myocardial infarction. During development of the Cleveland Clinic severity score,[11] factors such as age, hematocrit, serum creatinine, history of chronic pulmonary disease, previous cardiac surgery, and severe ventricular function were found to be associated with increased mortality. In contrast to these patients, adults with CHD represent a relatively new and small population and thus there are limited data identifying pre- and postoperative risk factors for mortality. Berat et al.[5] reviewed their postoperative experience in a cohort (n = 66) of adult CHD patients and identified several perioperative risk factors including cyanosis, poor ventricular function, and an underlying diagnosis of transposition of the great arteries or pulmonary atresia. However, as these authors indicated, this study was limited by the small sample size and heterogeneous population.

Recently, a consensus-based risk adjustment (RACHS-1)[14] was developed by an 11-member panel of pediatric cardiologists and cardiac surgeons to stratify in-hospital mortality risk in patients with CHD by age, diagnosis, and surgical procedure. These authors identified patients with single ventricle anatomy undergoing the Norwood procedure or the Damus-Kaye-Stansel procedure as those with the highest surgical mortality (category 6). The RACHS-1 study also identified that younger age, prematurity, and the presence of major noncardiac structural anomalies increased mortality risk. Similar to these findings, we observed a substantially increased risk for morbidity and mortality in our subgroup of adult patients undergoing cardiac surgery with a diagnosis of single ventricle anatomy. Recognizing the relatively small numbers of adult patients with CHD at any single center, it will require a multi-institutional approach in order to develop and validate a risk-adjustment score for this ever-increasing patient population.

Conclusions

The postoperative course in adults following congenital heart surgery is generally uncomplicated and survival should be anticipated. We have identified several perioperative risk factors for increased morbidity and mortality in this patient population, including the presence of chronic lung disease and liver dysfunction, increased cardiopulmonary bypass and aortic cross-clamp times, and elevated inotropic scores and lactate levels in the immediate postoperative period. However, further research is required in order to develop a risk stratification model for the emerging population of adults with CHD undergoing cardiac surgery.

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