



Outcome of cardiac surgery in adults with congenital heart disease: A single center experience

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Objectives: Adult survivors with congenital heart diseases represent a large and growing population, yet the published data does not represent the magnitude of their needs specifically in the Middle East. We aimed to review our experience at King Faisal Heart Center, Riyadh, Saudi Arabia for the outcome of adult patients with congenital heart disease who underwent either primary or redo surgery.

Methods: A retrospective study at a tertiary care hospital. All patients who underwent surgery either as the first surgery or as a reoperation for congenital heart disease aged >16 years old at the time of cardiac surgery in the period between January 1, 2008 and January 1, 2013. We looked for incidence of postoperative bleeding, arrhythmias, acute kidney injury, neurological complications, duration of mechanical ventilation, hospital and intensive care unit (ICU) stay. Additionally, we assessed the mortality and 1- and 5-year survival.

Results: Ninety-eight patients were included in our study. Fifty-two (53%) were females and 46 (47%) were males, with a mean age of 26 ± 8.4 years and a mean weight of 62 ± 22.8 kg. Forty-nine patients (50%) required redo surgery. Ten patients (10%) suffered from postoperative bleeding. Eight patients (8%) had postoperative arrhythmias, of which two patients required permanent pacemaker insertion. Three patients (3%) had postoperative acute kidney injury and seven patients (7%) suffered from neurological complications. The mean duration of ventilation was 1.3 ± 2 days, with a mean ICU and hospital stay of 3.7 ± 3 days, and 10 ± 7 days, respectively. The overall mortality rate in our series was 4% with a 1–5-year survival of 96%.

Conclusion: Adult patients with congenital heart disease are prone to immediate postoperative multisystem complications, yet the majority of them are reversible. Their 1- and 5-year survival rate is excellent. Further follow up studies are required.

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1. Introduction

Adult survivors with congenital heart diseases (CHD) are a truly growing and demanding population. Gilboa et al. [1] estimated adults living with CHD in the United States only to be 1.4 million. The surgical treatment of CHD in adults has recently showed significant growth and this is possibly due to multifactorial reasons related to advances in diagnosis and treatment as well as patient awareness [2,3]. Some patients with CHD undergo primary surgery for defects that did not require surgery during childhood, or for lesions that were not recognized early in life. Other patients undergo reoperation after corrective repair or further palliation after a first palliative surgery [4].

Extended survival and increased frequency of procedures leads to complex anatomy and diverse clinical conditions which may consequently lead to complicated surgical intervention(s). Reduced myocardial function, decreased vascular compliance, increased arrhythmias, dysfunction of other organ systems, and age-associated comorbidities are other factors that affect outcomes [5].

Published studies report diverse array of risk factors for morbidity and mortality after adult congenital heart disease (ACHD) surgery. Some studies showed that severe postoperative complications were common, whereas others reported low mortality rates in this patient population. Although reoperations in ACHD have become very frequent today, our knowledge regarding this growing patient population is still limited [6].

As the outcome data as well as evidence-based data to guide management for this patient population are still limited [7], and even more limited for the Middle East population, we decided to review our experience at Heart Center in King Faisal Specialist Hospital and Research Center (KFSHRC) looking for the outcome of adult patients with congenital heart disease who underwent either primary or redo surgery at our center.

2. Materials and methods

Institutional review board approval and waiver of consent were obtained. A nonrandomized retrospective chart review was performed for all patients who underwent surgery either as the first surgery or as a reoperation for congenital heart disease aged ≥ 16 years old at the time of cardiac surgery over a 5-year period, between January 1, 2008 and January 1, 2013.

We predefined the patients' main complications to look for as: incidence of postoperative bleeding,

Abbreviations

ACHD	adult congenital heart disease
ACHS	adult congenital heart surgery
AKI	acute kidney injury
ASD	atrial septal defect
AS	aortic stenosis
AV	aortic valve
AVSD	atrioventricular septal defect
CHB	complete heart block
CHD	congenital heart disease
CRRT	continuous renal replacement therapy
CT	computed tomography
DILV	double inlet left ventricle
ECMO	extracorporeal membrane oxygenation support
H	hour
ICU	intensive care unit
IVS	intact ventricular septum
KFSHRC	King Faisal Specialist Hospital & Research Center
KG	kilogram
LTGA/PA	L-transposition of great arteries and pulmonary atresia
LV-PA	left ventricle to pulmonary artery
MV	mitral valve
ML	milliliter
PA	pulmonary atresia
PV	pulmonary valve
RACHS-1	risk adjustment in congenital heart surgery-scores
RV-PA	right ventricle to pulmonary artery
SAM	Subaortic membrane
SD	standard deviation
TAPVD	total anomalous pulmonary venous drainage
TOF	Tetralogy of Fallot
TV	tricuspid valve
VSD	ventricular septal defect
Y	years

arrhythmias, acute kidney injury (AKI), and neurological complications. Postoperative bleeding was defined as blood loss >1.5 mL/kg/h for 6 consecutive hours within the first 24 hours [8]. We have followed the RIFLE (Risk, Injury, Failure, Loss of kidney function, End stage renal disease) criteria for diagnosis and staging of AKI [1]. Neurological complications were defined in our study as either seizures or radiological evidence of any pathology for patients who needed to have brain computed tomography (CT) done.

We also predefined the primary outcomes as: duration of mechanical ventilation, intensive care unit (ICU) and hospital stay. Secondary outcomes were: operative mortality, and 1- and 5-year survival rates. Mortality was defined as per the Society of Thoracic Surgeons operative mortality definition: (1) all deaths, regardless of cause, occurring during the hospitalization in which the operation was performed, even if after 30 days (including patients transferred to other acute care facilities); and (2) all deaths, regardless of cause,

occurring after discharge from the hospital, but before the end of the 30th postoperative day [9].

We have used the adult congenital heart surgery (ACHS) mortality score [10] for risk stratification in our patients. The procedure with the lowest model-based estimate of mortality accompanying ACHS mortality score was atrial septal defect repair (0.1), and the highest was Fontan revision (3.0).

The outcome parameters were further compared between primary congenital heart surgery and redo surgery for the same group of patients.

Information on patients' demographics, diagnosis, cardiac surgical procedure(s), and all outcome parameters were gathered from patients' medical records and the hospital electronic database.

The statistical analysis was performed using the software package SAS version 9.4 (SAS Institute Inc., Cary, NC, USA). Descriptive statistics for continuous variables are reported as mean and standard deviation and categorical variables are summarized as *n* (%). Continuous variables are compared by the independent *t*-test, whereas categorical variables were compared and relative risks and confidence intervals calculated by the Chi-square or Fisher exact test. The level of statistical significance was set at *p* < 0.05.

3. Results

A total of 98 patients underwent surgery either as the first surgery or as a reoperation for congenital heart disease during the study period. Forty six (47%) were males and 52 (43%) were females with a mean age and weight of 26 years (Y) and 62 kilograms (kg) respectively (Table 1). The patients had a different range of congenital heart diseases (Table 2). The cardiac surgical procedures that were either conducted separately or in conjunction with one another and their ACHS risk stratification are shown in Table 3.

Half of the patients included in our cohort had their surgery as primary intervention, whereas the other half had a redo surgery for a primarily corrected congenital heart disease during their childhood (Fig. 1). Twenty-two (45%) patients from the redo group underwent right ventricle to pulmonary artery (RV-PA) conduit placement or replacement.

The overall incidence of complications was 18% with some of the patients suffered from more than one complication. Ten patients (10%) suffered from postoperative bleeding. Three patients (3%) had postoperative AKI, of which only one patient postFontan procedure needed continuous renal

Table 1. Demographic data for all 98 patients included in the study.

Variables	N (%)
<i>Sex</i>	
Male	46 (47)
Female	52 (53)
<i>Age (y)</i>	
Mean ± SD	26 ± 8.4
Range	17–47
<i>Weight (kg)</i>	
Mean ± SD	62 ± 22.8
Range	30–107

SD = standard deviation.

Table 2. The primary congenital heart disease for all 98 patients.

Congenital heart disease	N
Tetralogy of Fallot (TOF)	17
Atrial septal defect (ASD)	16
Ventricular septal defect (VSD)	14
Subaortic membrane (SAM)	7
Truncus arteriosus	5
Others e.g., AVSD, DILV, DTGA, LTGA, PS, PA/IVS, PA/VSD, AS	40

AS = aortic stenosis; AVSD = atrioventricular septal defect; DILV = double inlet left ventricle; DTGA = D-transposition of great arteries; LTGA = L-transposition of great arteries; PA/VSD = pulmonary atresia/ventricular septal defect; PS = PA/IVS pulmonary stenosis/intact ventricular septum.

Table 3. Surgical procedures and ACHS mortality scores for all 98 patients.

Cardiac surgery	N	ACHS mortality score
ASD closure	19	0.1
VSD closure	16	0.5
AVSD repair	3	0.5
TV repair	10	0.6
TV replacement	1	0.9
MV repair/MV replacement	5	0.4/1.5
AV repair/AV replacement	7	0.3/0.6
Ross procedure	11	0.7
PV valvuloplasty	5	0.4
PV replacement	1	0.2
TOF repair	2	0.6
LVOTO relief	6	0.2
RV-PA conduit	22	0.7
Fontan	5	1.8
Arch repair	1	0.5
Others (TAPVD, PA augmentation)	5	–

ACHS = adult congenital heart surgery; ASD = atrial septal defect; AV = aortic valve; MV = mitral valve; PA = pulmonary artery; PV = pulmonary valve; RV-PA = right ventricle to pulmonary artery; TAPVD = total anomalous pulmonary venous drainage; TOF = Tetralogy of Fallot; TV = tricuspid valve; VSD = ventricular septal defect; LVOTO = left ventricular outflow tract obstruction.

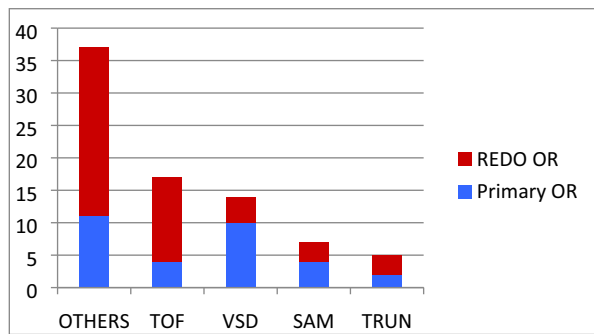


Figure 1. Number of patients who needed redo surgery and their diagnosis. SAM = subaortic membrane; TOF = tetralogy of Fallot; VSD = ventricular septal defect; OR = operation.

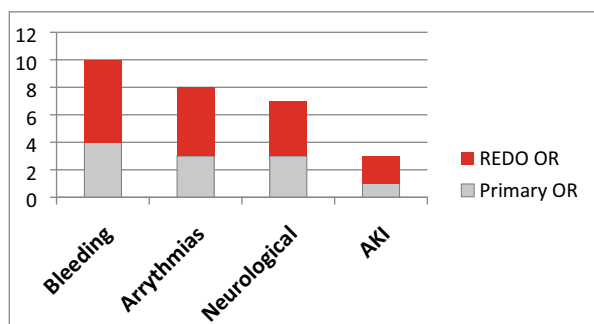


Figure 2. Postoperative complications for all patients. AKI = acute kidney injury; OR = operation.

replacement therapy (CRRT). Eight patients (8%) had postoperative arrhythmias. Three patients had complete heart block (CHB, 2 of them needed permanent pace maker insertion), four had ventricular tachycardia, and one patient had flutter. Seven (7%) patients suffered from neurological complications in the form of six postoperative seizures, one with brain edema and one with ischemia. All the complications were more predominant in the redo surgery group (Fig. 2).

The mean ventilation days were 1.3 days with a mean ICU and hospital stay of 3.7 days and 10 days, respectively. The mortality in this group of patients was 4% (Table 4). The four patients who died included three patients from the redo group: two patients postFontan procedure (22 years old and 17 years old), one L-transposition of great arteries and pulmonary atresia (LTGA/PA) post left ventricle to pulmonary artery (LV-PA) homograft who suffered from severely depressed systemic ventricular function and suffered from multiple organ failure. The deceased patient from the primary group was post atrial septal defect (ASD) repair who developed postoperative persistent tachyarrhythmia that led to cardiac arrest and needed rescue extra-

Table 4. Primary and secondary outcome for all 98 patients.

Variable	N (%)
Ventilation (d)	1.3 ± 2
• Mean ± SD	
ICU Stay (d)	3.7 ± 3
• Mean ± SD	
Hospital Stay (d)	10 ± 7
• Mean ± SD	
Mortality	4 (4)
Survival (1-y)	94 (96)
Survival (5-y)	94 (96)

ICU = intensive care unit; SD = standard deviation.

corporeal membrane oxygenation support (ECMO) support, but never recovered. Ninety-four patients (96%) survived for 1 year and 5 years following their surgical procedures.

4. Discussion

Advanced diagnostic tools and improved medical management allow the majority of newborns with congenital heart diseases to survive to adulthood. Only patients with relatively simple lesions, such as isolated ventricular septal defects or patent ductus arteriosus without pulmonary hypertension or ASD are finally cured with primary cardiac surgery [11]. The short- and long-term outcomes of adults with CHD became a timely topic of increasing clinical interest.

Patients' presentation in the current era is significantly variable, especially in the Middle East. Some patients undergo primary surgery for defects that did not require surgery during childhood, or for defects that went unrecognized or for those who were diagnosed but missed follow up. Other patients undergo redo surgery for subsequent consequences of prior palliative operations.

During the 5-year period of our study, 50% of the patients underwent redo surgery for their CHD. Our incidence is comparable with the 58% incidence of reoperation in the series published by Dore et al. [12]. The most common cause for reoperation in our series was for RV-PA conduit insertion or change. It has been reported by other groups that pulmonary valve and right ventricular outflow tract reconstruction procedures are the most common reoperations among ACHD [11,13]. Tetralogy of Fallot (TOF) was one of the largest subgroups in our series, and this group is known for their need of redo surgery because of frequent problems with pulmonary regurgitation, right ventricular outflow tract obstruction, and conduit failure. This may be due to the regional differences in CHD prevalence. A report has previously shown

that there is relatively more right-sided and less left-sided lesions in Asia [14]. In the series from Toronto [15], 46% of TOF patients required reoperations.

One would expect that this group of patients would have a very high risk of postoperative complications due to the complexity of their cardiac lesions, risk of reopening the sternum in redo cases, and other possible comorbidities faced during adulthood. The overall complication incidence in our series was 18% and that was similar to a couple of previous reports [6,16], and <28% rate of postoperative complications reported by Mascio et al. [11].

Our results for postoperative complications have both similarities and differences with other reports. Our patients had less incidence of postoperative arrhythmias and AKI than what was reported by Giamberti et al. [6]. As expected, the rate of complications was higher in the redo surgery group, but at the same time we reported less incidence of individual complications such as arrhythmias and AKI which we think is related to having better patient follow up and early planning for redo surgeries. None of the patients included in the study needed chest reexploration for bleeding. We attributed this to the fact that all our patients were operated upon by pediatric cardiac surgeons. Studies have shown that redo sternotomy carries a negligible risk of injury and postoperative morbidity when it is done in large centers by pediatric cardiac surgeons [6,17].

We reported 7% neurological complications in the study group. Neurological injuries are a known complication after intervention for congenital cardiac disease [18]. It can be related to individual patient factors, including genetic predisposition, sex, race and socioeconomic issues, or factors related to operative variables including circulatory arrest, cardiopulmonary bypass, and low cardiac output state.

The average length of stay and ventilation days were relatively short in our study and well comparable with other studies [11,19]. We believe that caring for these patients postoperatively in a specialized cardiac intensive care unit by a trained team in CHD is one of the contributing factors for better patients' outcomes.

Surgical risk stratification for adult patients with CHD undergoing primary or redo surgery remains challenging. Previous studies compared the use of Aristotle and risk adjustment in congenital heart surgery (RACHS-1) scores for adults undergoing congenital heart surgeries but the results were mixed with variable degrees of discrimination

[20,21]. The ACHS scoring is so far the only scoring system available for the adult age group and its use proved to have accurate estimation of adjusted mortality risk for the case mix of the majority of CHD surgeries (52 procedural groups) [10].

Several authors demonstrate low mortality rates after adult CHD surgery [7,11,22,23]. Mortality was 4% in our series. All the patients who died in our redo surgery group suffered from complex cyanotic congenital heart defects. It is well known from studies that this type of patient has high early operative mortality rates [6,7]. We had two mortality cases postFontan procedure who developed multiple organ failure and our results were comparable with those reported by Giamberti et al. [6]. The in-hospital mortality postFontan reported by Mascio et al. [11] was 11% versus 2.1% overall. This is probably due to the consequences of the chronic Fontan circulation that may lead to comorbidities, in addition to the progressive deterioration of cardiac function.

The overall 1- and 5-year survival in our patient cohort was 94% which matches well with the 97.6% survival rate at 1 year and 95.2% survival rate at 5 years reported by Kogon et al. [21]. It has even been reported that in the current era, survival for patients with CHD intimates that for the healthy adult population [24].

To our knowledge this is the first study conducted in the Middle East looking at outcome of adult patients with CHD. We reported favorably comparable results of morbidity, mortality, and survival with different centers worldwide. In view of this growing heterogeneous patient population, further efforts from research scientists and clinical teams are needed to cope with patients' needs and improve their quality of life.

4.1. Limitations

This study is limited by being a single center observational one and the inherent limitations of a retrospective data review. The number of patients is relatively small and very heterogeneous in nature which made it difficult for patients grouping. Therefore, patients had to be grouped according to their primary cardiac defect, and different surgical procedures they underwent. We have used the ACHS score for risk stratification being the most recent and applicable to adult patients with CHD; even though it does exclusively stratify all the procedures but it did cover the majority, and we are aware that this is still an area of challenge for this group of patients worldwide. Also, the late follow-up for the

patients was limited to survival although looking for quality of life and longer-term complications in this group of patients will be important, but it was beyond the scope of this study.

5. Conclusion

Adult patients with congenital heart disease are prone to immediate postoperative multisystemic complications, yet the majority of them are reversible. Their 1- and 5-year survival rate is excellent. Further follow up studies are required.

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Conflicts of interest

The authors declare that there is no conflict of interest.

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