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Abstract

Introduction: Congenital heart disease (CHD) is evident at birth and can compromise the structure and function of an infant's heart. CHDs are a diverse group in terms of complexity, severity and type. The purpose of this study was to determine the outcomes of different heart surgery outcomes including palliative, closed, open, and complete repair surgery, of patients diagnosed with CHD between January 2017 and September 2021.

Method: This was a cross-sectional retrospective study at the Al Jalila Children's Specialty Hospital, Dubai, United Arab Emirates involving 353 pediatric patients.

Results: Of the 353 patients, 182 (51.6%) were male while 171 (48.4%) were female. A total of 295 infants had not undergone any palliative care (83.6%), and 58 patients had gone through a redo either twice, thrice, or four times after undergoing open, closed, complete repair, or palliative surgery. The patients who underwent complete repair had a higher survival rate beyond eight weeks after surgery (96.4%) with a p-value of 0.001 (p < 0.05).

Conclusion: This study highlighted the outcomes of various types of surgery often used in surgical interventions in the management of CHD.

Keywords: Congenital Heart Disease (CHD); Surgical Interventions; Heart Defect

Introduction

Congenital heart disease (CHD) is a heart defect that can be present from birth and can be either asymptomatic or life-threatening. Some factors that predispose individuals to CHD have been identified. However, for the vast majority of CHD patients, no specific causes have been found. This is because the development of CHD is influenced by both genetic and environmental factors [1,2].

It is the most frequent birth defect, affecting one neonate out of every 120-166 live births, and globally, there are approximately 1,310,000 infants born every year with CHD [3]. As adult survival rates have climbed to 90 - 95% in developed countries from fewer than 20% in the presurgical era, the prevalence of CHD in the population has dramatically grown. In 2017, North Africa, Oceania, the Caribbean,

the Middle East, Southeast Asia, and central sub-Saharan Africa had the highest rates of infant mortality because of congenital cardiac disease [4]. The incidence of severe CHD in the Al-Qassim area of the Kingdom of Saudi Arabia is roughly 5.4 per 1000 live births, which is nearly twice as high as the incidence in the USA, which is only three per 1000 [5]. Additionally, research indicates that some CHD subtypes may predominate in kids with consanguineous parents [6].

The incidence of adult CHD is anticipated to surpass that of neonatal/infant CHD by the 2020s [7]. Approximately 15% of CHD patients who have had cardiac surgery in the past will need additional procedures (for example, conduit exchange, valve repairs, or replacement) [7,8].

Surgery is the most common intervention in the management of CHD. By the intended effects of treatment, surgeries for congenital cardiac disease can be classified as palliative or reparative. When a child is very young for surgical repair, a palliative procedure is typically needed to improve a defective cardiac function and minimize the condition [9]. The objective is to reduce cyanosis, manage heart failure, or set up the circulation for eventual repair when the infant reaches an age and weight appropriate for the existing treatments. Open heart surgery involves the use of cardiopulmonary bypass which aids in maintaining the lung heart-lung function [10]. It maintains the perfusion of the tissues during the process of surgery. Closed heart surgery involves a sternotomy or thoracotomy. The chest is entered from the front and/or the side. Closed heart surgery is commonly palliative and is often done in children with CHD who require staged heart operations [11]. It includes procedures such as Ballock-Taussig shunts, ligation of patent ductus arteriosus, and pulmonary artery repair [12].

Purpose of the Study

The purpose of this study was to find out the outcomes of different heart surgeries including palliative and complete repair surgery [13].

Materials and Methods

This was a cross-sectional retrospective study conducted at Al Jalila Children's Specialty Hospital in Dubai, United Arab Emirates (UAE). The data extracted was from January 2017 up to September 2021. The process of data collection commenced in June and ended in September. Dubai Scientific Research Ethics Committee (DSREC) Dubai Health Authority approved the study (approval number: DSREC-SR-07/2022_13). Data was extracted from the Cerner database (Oracle Cerner, Kansas City, Missouri, United States), which is the primary database used at the Children's Specialty Hospital.

Participants

The selected population of 400 patients were all children diagnosed with CHD who underwent cardiac surgery at Children's Specialty Hospital between January 2017 to September 2021. Patients with missing data and those who had contraindications were excluded. Additionally, patients with co-morbidities such as diabetes, obesity, infections, and other diseases that might affect the outcome of the surgery were also excluded from the study.

Variables

The independent variable was surgery, while the dependent variable was the prognosis related to various types of surgeries. The exposure was the children diagnosed with CHD and the outcome was the patient's successful recovery or passing away. If the patient passed away eight weeks after the surgery, it was considered most likely due to another reason and not due to the cardiac surgery. Confounding factors were addressed through the process of stratified sampling and the exclusion criteria as it was highly representative of the target population.

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Bias

The method of selection was through secondary data, using the Cerner database at the hospital which made sure patients would remain anonymous as only the medical record number alongside information like gender, nationality, and prognosis were retrieved. This addressed information bias, which is one of the limitations of a cross-sectional study. Potential selection bias also existed as the patients at the hospital were not representative of all the patients with CHDs across the UAE.

Statistical analysis

The data collected was organized based on the age, gender, diagnosis, kind of surgery performed, number of surgery stages, prognosis, and outcomes of each patient, using Microsoft Excel (Microsoft Corporation, Redmond, Washington, United States).

The Statistical Package for Social Sciences (SPSS) software was used to run the chi-square test to obtain the p-value to prove the significance between the type of surgery and its success. For all the continuous variables, standard deviation (SD) and mean were determined. Following double entry, data was checked before analysis and all patients were filtered using the exclusion criteria.

Results

A total of 400 infant patients were extracted from the Cerner databases and 353 met the inclusion criteria. Of these, 182 (51.6%) were male while 171 (48.4%) were female. The mean height of the patient was 68.7 ± 28.4 cm, the mean weight was 8.20 ± 9.82 kg, and the mean age in months was 19.50 ± 3.31 months (Table 1).

Characteristics	Mean	SD
Height (cm)	68.7	28.4
Weight (Kg)	8.20	9.67
Body surface area	37	28
Age (months)	19.50	37.31

Table 1: Patient characteristics (N = 353).

Out of the 353 patients, 295 (83.6%) infants did not require any redo surgeries, and 58 patients had gone through a redo either twice or thrice after undergoing open, closed, total repair, or palliative surgery. Forty-four (12.5%) patients underwent surgery once after the surgery, 10 (2.8%) patients went through surgical palliation twice after having the heart surgery, and four (1.1%) patients underwent surgical palliation three times after the heart surgery. In addition, out of the total patients, 62 underwent closed heart surgery (17.6%), 291 patients underwent open heart surgery (82.4%), 277 had complete repair (78.5%), and 76 patients had palliative repair (21.5%). Most of the patients who had surgery at the Children's Speciality Hospital went through open heart surgery (82.4%), followed by complete repair surgery (78.5%).

Surgery details		Frequency	Percentage	
Number of Redo Surgeries	0	295	83.6	
	1	44	12.5	
	2	10	2.8	
	3	4	1.1	
Operation Type 1	Closed	62	17.6	
	Open	291	82.4	

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Operation Type 2	Complete repair	277	78.5	
	Palliative	76	21.5	
Outcome	Alive	333	94.3	
	Dead	20	5.7	

Table 2: Distribution of mortality according to type of surgery and number of redo surgeries.

Closed heart surgery had a higher mortality rate compared to open heart surgery, even though general open heart surgery is more complex, with patients with closed surgery 1.5 times more likely not to survive compared to open heart surgery (p value = 0.368). Palliative surgery patients were four times more likely not to survive compared to complete repair surgery (p value = 0.001).

Types of Surgeries	Mortality Outcomes				n volue	
	Survived		Died			p-value
	Frequency	Percentage	Frequency	Percentage		
Operation Type 1	Complete repair	267	96.4	10	3.6	0.001
	Palliative	66	86.8	10	13.2	
Operation Type 2	Closed	57	91.9	5	8.1	0.368
	Open	276	94.8	15	5.2	

Table 3: Association of the different types of operation with mortality.

P < 0.05 is considered significant. Chi-square test was used to derive p values.

The difference between complete and palliative surgery is statistically significant. This is perhaps because, in complete repair, the surgical outcomes are almost similar to a normal heart whereas in palliative repair surgery, only connections were made between specific arteries to improve blood flow in an infant's heart. In contrast, the difference between open or closed surgery is not statistically significant and this could be due to the reason that at Al Jalila Children's Speciality Hospital, closed heart surgery is reserved for complex diseases like pulmonary atresia, and open heart surgery is opted for the complete repair of simple CHDs like tetralogy of Fallot, atrial septal defect, and ventricular septal defect. However, in certain cases like patent ductus arteriosus ligation, which is a simple heart defect, a closed procedure had to be done, and in cases of CHDs like hypoplastic left heart syndrome, an open heart procedure had to be done in three stages: Norwood procedure, bidirectional Glenn (BDG) shunt procedure, and Fontan procedure.

Discussion

A total of 353 patients with a mean age of 19.50 months took part in the present study. A high number of cases were observed in Asia at 76.2% (United Arab Emirates at 31.2%), followed by Africa at 18% with Australia and South America having the least cases at 0.3% and 0.9% respectively.

The advancement of medicine has made it possible to surgically treat or cure a growing number of congenital cardiac abnormalities while achieving a high general survival rate. After a successful procedure, most patients, particularly those with straightforward problems, have a good life expectancy [14]. This study showed that complete repair heart surgery is associated with a higher survival rate as compared to palliative and closed heart surgery. This is similar to other studies. In a cohort study involving 3283 patients, the survival after one year, five years, 10 years, 20 years, and 25 years were 98%, 97.8%, 97.1%, 95, and 94.5%, respectively in patients having tetralogy of Fallot [15]. Some of the deaths observed in complete repair surgery could be due to sudden deaths, congestive heart failure, and cardiac arrhythmias.

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In another retrospective study by Kwak., *et al.* involving patients with trisomy 18 who had total cardiac repair and palliative care, those who had total cardiac repair surgery had better survival outcomes [16]. Out of the total 11 patients (36.4%), four survived. No patients who had palliative care only survived. In a study by Elassal., *et al.* the outcome of corrective or complete repair heart surgery was reported to be encouraging [17]. Furthermore, in a retrospective study done in Shahid Modarres Hospital, Tehran, Iran, involving 202 patients, out of the 143 patients that had open heart surgery, only 21 (15%) patients didn't survive one year after the operation, indicating better survival rates statistically [18].

Minotti., *et al.* in a prospective study involving 218 neonates, reported that complete repair surgery in the management of aortic coarctation is associated with good late and early outcomes [19]. Most neonates (81.6%) underwent end-to-end anastomosis repair procedures associated with very low mortality (2%). These findings correlate with other studies elsewhere. However, patients presenting with coarctation of the aorta often have an increased risk of developing hypertension [20,21].

In a retrospective study to repair atrial septal defects involving 1326 children, no mortalities were reported, correlating well with the findings of the present study [22,23]. Atrial septal defect closure, despite its low mortality rate, is associated with cardiopulmonary bypass-related morbidity and sternotomy. Palliations are surgical treatments used to treat severe hemodynamic abnormalities caused by CHDs, usually in advance of phased repair. Symptoms are treated, but the condition cannot be stopped from getting worse. A total of 66 pediatrics underwent palliative procedures. Fifty-six of them survived while 10 of them passed away representing a 13.2% mortality rate.

Delgado-Corcoran., *et al.* in a retrospective study involving 1389 patients of whom 112 received palliative care, reported that patients receiving palliative care had high mortality rates [24]. These findings are consistent and correlate with the findings of this study which found that the mortality rates of pediatrics who underwent palliative surgery were higher than those of pediatrics who had complete repair surgery. According to Trowbridge., *et al.* pediatrics who receive palliative care have a lower likelihood of dying during resuscitation than those who do not receive palliative care [25]. Furthermore, Bertaud., *et al.* report that pediatric palliative surgery is vital in improving symptom control and quality of life, especially in pediatrics who cannot survive the disease [26]. Palliative care plays an important role in improving symptom control with a less positive impact on mortality rates, thus resonating well with the findings of our study.

In another multinational, multicenter prospective study on palliative surgery outcomes, 1004 infants were studied, and the study reported a high infant mortality rate, especially in infants who received systemic to pulmonary hunt palliative interventions. Of these, 33% died within one year after surgery.

Our study findings are contrary to what was observed by Bockeria., *et al.* in a retrospective study involving 29 patients, of whom 26 had tetralogy of Fallot and three had a double outlet right ventricle [27]. They reported that the mortality of patients who had palliative surgery improved. In this group of patients, palliative surgical procedures continue to be an effective strategy that improves patients' conditions, raises artery oxygen saturation, develops the pulmonary vascular bed and the left ventricle, and serves as a staging area for the complete repair of CHDs in the future.

Strengths and Limitations of the Study

The strengths of the study were that we were able to obtain a good sample size to differentiate between the types of surgery and analyze the mortality rates. Some of the biases of a cross-sectional study were addressed and resolved, such as information bias. However, there were problems with generalizability as this was a single-center study. A retrospective, cross-sectional analysis was used in this study, and the patient numbers for the different surgery types weren't equalized. In addition, the study did not highlight the distribution of mortality outcomes or survival based on gender to provide a better understanding and clear picture of how different surgeries and their outcomes vary with gender.

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Areas for future research

This study can be further developed by expanding the scope of the data collection by incorporating other hospitals in the research for it to be later generalized to the UAE population. Another aspect that could be explored is the relationship of age, gender, and ethnicity with the different surgery groups, with an equal number of patients in each.

Conclusion

This study highlighted the outcomes of various types of surgery often used in surgical interventions in the management of CHDs such as atrial and ventricular septal defects, coarctation of the aorta, or teratology of Fallot. The study found that complete repair heart surgery, a form of open-heart surgery, is associated with a better prognosis as compared to palliative procedures.

These findings would help in determining the estimated effectiveness of various cardiac surgical treatments in CHD patients in the United Arab Emirates. Aiding patients in choosing the type of surgery with a better prognosis would be more accurate and precise.

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