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Original article

Clinical Profile of Adult Patients Referred to Congenital Heart Surgery: A Single Center Experience in Egypt

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Abstract

Background: A major barrier for adult congenital heart disease (ACHD) services in developing countries is the insufficient data on the disease burden and specific needs of this population. Our goal was to the clinical profile and challenges of ACHD patients in Egypt who require further surgical correction. **Methods:** The study comprised ACHD patients (above 14 years of age) scheduled for further surgical correction following a heart team discussion at a tertiary cardiac center in Egypt. Data on age, gender, presentation, functional capacity, resting oxygen saturation, diagnosis, previous intervention and its timing, and the type of planned surgery were collected. **Results:** Throughout one year, 103 cases (49.5% males, mean age 23 years) were referred to surgery out of 134 cases discussed by the heart team. One-third (34%) were cyanotic. The majority presented in NYHA class II (44.7%) and III (35.95%) and had a previous surgical or trans-catheter intervention (56.3%) at a median age of four years. The most frequent diagnoses were Tetralogy of Fallot (ToF) (23.3%), atrial septal defect (ASD) (18.4%), double outlet right ventricle (11.6%), and transposition of great arteries (7.8%). The most common planned surgical procedures were Fontan (20.3%), ASD closure (18.4%), pulmonary valve replacement (14.5%), total ToF repair (9.7%), subaortic membrane resection (6.7%) and tricuspid valve surgery (5.8%). **Conclusion**

ACHD patients in our database who require additional surgical correction are heterogeneous, with highly variable presentations and surgical complexity. An efficient ACHD program mandates a multidisciplinary team that is familiar with this patient population's unique demands and difficulties.

Keywords: Adult; Congenital; Heart; Surgery; Egypt

Introduction

Over the past decades, the population of adult patients with congenital heart disease (CHD) has remarkably grown worldwide. This is mainly due to the advances seen in congenital heart surgery and pediatric cardiac care. Some reports postulate that the adult congenital heart disease (ACHD) population is currently outnumbering the pediatric CHD group in Europe and the United States [1,2]. Data from the developing world are scarce, but some reports show similar trends [3,4].

Despite significant advances in the field of congenital heart surgery in recent years, significant challenges remain, particularly for surgeons operating on adult patients with congenital defects. Most cardiac surgical repair procedures during childhood are not curative [1], and many of the remaining cardiac lesions may progress or begin to affect cardiac structure and function over time. Tissue valves and conduits frequently require repeated interventions when they degenerate. Redo surgeries entail a higher risk sternotomy, which may injure previous conduits or dilated aortas adherent to the sternum[5].

In developing countries, where facilities for congenital cardiac surgery are not widely available, patients with CHD who survive until adulthood have a different profile than those in developed countries [4]. Most ACHD patients have either mild lesions (left or overlooked in childhood) [3] or severe CHD that was operated on during infancy and childhood. Only a small fraction survives despite complex unrepaired defects or because of palliative or incomplete repair.

One of the major challenges facing ACHD service in low- and middle-income countries is the lack of adequate data on the disease burden and specific needs of this peculiar group [4,6]. Clearly, data on the various presentations and types of required surgeries are insufficient.

Materials and methods

Our retrospective descriptive study included all CHD patients above 14 years of age, who sought medical advice at Al Nas Hospital (an Egyptian tertiary cardiac center) and required surgical intervention between March 2022 and February 2023; a total of 220 patients. At the outpatient clinic, all referred patients were evaluated by one of the ACHD team cardiologists. After all non-invasive testing and hemodynamic studies were completed, candidates for surgical intervention were presented in a heart team meeting that included a congenital cardiac surgeon, a congenital structural interventionist, a radiologist, and a cardiologist. Accepted cases by the committee were included in the study. Age, gender, presentation, functional capacity, resting oxygen saturation, diagnosis, previous intervention and its timing, surgery type, need for cardiac magnetic resonance imaging (CMR) or computed tomography (CT), and need for pre-surgery catheter hemodynamic assessment were all collected. An informed consent was obtained from all individual participants, or their parents (or legal guardians) included in the study. The study was approved by Al Nas hospital ethical committee on February 2024 (code

no. RHDIRB-NA-240823-01UC-NPO-N0323-0001).

Statistical analysis

Descriptive statistics were tested for normality. Continuous variables were presented as median and interquartile range (IQR) as the data were skewed, while categorical data were presented as number (%).

Results

Out of 220 patients who attended the ACHD clinic; the heart team committee discussed 134 patients and decided on surgery for 103 cases, while the remaining 86 patients were either given medical treatment/follow-up decisions or scheduled for trans-catheter intervention.

Half of the patients were males (49.5%), the median age was 21 years (IQR: 17-31 years), 34% were cyanotic, and the majority presented in NYHA functional class II (44.7%) and III (35.95%). Fifty-eight patients (56.3%) had a form of previous surgical or trans-catheter intervention at a median age of 4 years (IQR 2.1-11.8 years) at the time of the first procedure: 15.5% of them had two procedures, while 7% had three or more separate surgical or catheter interventions. The types of these surgeries are summarized in (Table 1).

Tetralogy of Fallot (ToF) was the most frequent diagnosis among our cohort (24 patients, 23.3%): 8 did not have any form of previous intervention, 2 had Blalock Taussig (BT) shunts during childhood, and 14 had a previous total repair. One of the uncorrected patients presented with a large ascending aortic aneurysm and a ruptured sinus of Valsalva. Nineteen patients (18.4%) were referred for surgical atrial septal defect (ASD) closure: 11 had large secundum ASDs not amenable for trans-catheter closure, 7 had sinus venosus ASDs (6 of them had associated partial anomalous pulmonary venous drainage), and one patient had primum ASD [partial atrioventricular (AV) canal]. Twelve patients (11.6%) had a double outlet right ventricle; 9 with a previous palliative shunt with/without pulmonary artery banding; and one patient had total repair (Rastelli's operation). Eight patients (7.8%) had transposition of great arteries (TGA); two of them had congenitally corrected TGA with severe incompetence of the tricuspid (systemic) valve. The other 6 patients had D-TGA with protected pulmonary circulation in addition to either ASD or ventricular septal defect (VSD); 3 of them had previous Glenn procedures, and the remaining 3 had previous BT shunts. Five patients (4.8%) had a common AV canal; four with previous pulmonary artery banding (one of them had severe pulmonary stenosis), and 2 had previous total repair. Five patients (4.8%) had subaortic membranes with significant left ventricular outflow tract obstruction. Six patients (5.8%) had Ebstein's anomaly; one had a previous repair, and another had a tricuspid valve replacement. Four patients (3.8%) had VSDs; two were residual defects post-surgical closure associated with significant aortic regurgitation due to cusp prolapse, one was a restrictive defect with left ventricular dilatation; the last one had previous surgical closure with iatrogenic severe tricuspid incompetence. Three patients (2.9%) had tricuspid atresia; all had

previous Glenn procedures. Three patients (2.9%) had severe pulmonary regurgitation after previous surgical or catheter pulmonary valvotomy. Two patients (1.9%) had a double-chambered right ventricle. Two patients (1.9%) had partial anomalous pulmonary venous drainage. Two patients had unrepaired Shone complex diseases (1.9%). Two patients (1.9%) had a double inlet left ventricle. Single patients had the following diagnoses: supravalvular aortic stenosis, branch pulmonary stenosis, double outlet left ventricle, aortic paravalvular leak, pulmonary stenosis, and right atrial appendage aneurysm. The recommended surgeries by the heart team are listed in (Table 2).

Other than echocardiography, 65% of the patients required further non-invasive testing: 39 (37.8%) had CMR, 23 (22.3%) had a CT scan, and 5 (4.8%) had both tests. Pre-operative cardiac catheterization was performed in 26 patients (25.2%).

Table 1 Types of previous surgeries/ catheter interventions.

Type of previous surgeries/interventions	Number of patients
Total Fallot repair	14
BT (Blalock- Taussig) shunts	15
Glenn	14
PA banding	7
ASD closure	2
VSD closure	4
Surgical aortic coarctation repair	2
Balloon pulmonary valvuloplasty	2
Subaortic membrane resection	2
Rastelli	1
Kawashima	1
Common AV canal repair	2
Surgical pulmonary valvotomy	1
LPA stent	1
TV repair	2
TVR	1
AVR	2 (one with Kono procedure)

BT shunt: Blalock Taussig shunt, **PA**: pulmonary artery, **ASD**: atrial septal defect, **VSD**: ventricular septal defect, **common AV canal**: common atrioventricular canal, **LPA**: left pulmonary artery, **TV**: tricuspid valve, **TVR**: tricuspid valve replacement, **AVR**: aortic valve replacement.

Table 2 Types of required surgeries for ACHD patients.

Type of surgery	Number of patients (percentage)
Total cavo-pulmonary anastomosis (Fontan surgery)	21 patients (20.3%); Completion Fontan surgery (after a previous Glenn shunt) in 16 patients (15.3%) and without previous surgery in 5 patients (4.8%)
Surgical ASD closure	19 patients (18.4%) (with PAPVD repair in 6 patients and pulmonary valvotomy in 1 patient)
Surgical pulmonary valve replacement	15 (14.5%) patients (with RVOT reconstruction in 1 patient, VSD closure in 1 patient, TV repair in 2 patients, and repair of LPA origin stenosis in one patient)
Total ToF repair	10 patients (9.7%) (with MAPCAs surgical closure in 2 patients and after RVOT stenting in 2 patients)
Tricuspid valve surgery	6 patients (5.8%). - TV repair in 5 patients (4.8%) in addition to ASD closure in one patient and Glenn surgery in another patient. - TV replacement in one patient who had a common AV canal, PA banding in infancy, and two previous attempts for repair, and presented with massive TR
Subaortic membrane resection	7 patients (6.7%); catheter coiling of a saccular aortic aneurysm was required in one patient, additional mitral valve repair in 2 patients (Shone complex patients), aortic valve replacement in one patient
Surgical VSD closure	3 patients (2.9%); with aortic valve repair in 2 patients.
- **Total repair of DORV** - **Tricuspid (Systemic) valve replacement** - **Double-chambered RV repair** - **Repair of PAPVD** - **Repair of bilateral branch pulmonary artery stenosis**	2 patients (1.9%) each
- **Aortic valve repair** - **Hepatic incorporation surgery** - **Mitral valve replacement** - **Partial AV canal repair** - **RAA aneurysm excision** - **Redo AVR with relief of RVOT obstruction** - **Redo degenerated Tricuspid valve replacement in addition to mitral valve replacement** - **Repair of aortic aneurysm and ruptured sinus of Valsalva in a post-TOF repair patient** - **Replacement of RV-PA conduit** - **Surgical correction of CAVC and PS,** - **Surgical correction of RVOT obstruction in a previously repaired ToF patient,** - **Surgical correction of supravalvular AS**	One patient (0.9%) each

ASD: atrial septal defect, **AVR**: aortic valve replacement**, AS**: aortic stenosis, **CAVC**: common atrioventricular canal, **DORV**: double outlet right ventricle, **LPA**: left pulmonary artery stenosis, **MAPCAs**: Major aortopulmonary collaterals, **PAPVD**: partial anomalous pulmonary venous drainage, **RVOT**: right ventricular outflow tract, TV: tricuspid valve, **RV**: right ventricle, **RV-PA**: right ventricle to pulmonary artery, **RAA**: right atrial appendage aneurysm, **ToF**: Tetralogy of Fallot.

Discussion

The care of ACHD patients in developing countries constitutes a major challenge. In a previous report, the number of ACHD centers in Africa was 0.1 per 10 million inhabitants in contrast to a substantially higher number in Europe (3.6) and North America (1.7) [7]. The African research production in the field of ACHD was also extremely lower than in Europe and the United States. Consequently, ACHD patients in those developing countries are ill-defined, making future planning more challenging [7].

In this study, we describe the surgical needs of a cohort of ACHD patients in Egypt. Determining the clinical profile, different presentations, and types of required surgical interventions should help initiate a service for this underserved and poorly defined group.

More than half of the patients had a previous surgery (palliative or total repair). Patients who never had any previous intervention were either patients with simple defects [including ASDs, VSDs, and subaortic membrane (25% of all cases)] or with more complex but neglected diseases (e.g., single ventricle physiology and unrepaired ToF). On the other hand, smaller numbers of ACHD patients in developing countries had received a form of correction previously (35% in Ghana and only 19% in Egypt) [3,4,8]. In contrast, over 90% of ACHD in developed countries had a previous intervention in infancy or childhood [3]. Our patient cohort represents a challenge for the surgical team who face the complexity of both unrepaired naturally surviving adults, and those with previous repair presenting with residual defects or delayed second-stage single ventricle repair.

Out of all patients requiring surgery, 13.6% had a previous Glenn shunt during childhood and presented with progressive effort intolerance and desaturation. There is insufficient evidence in Egypt about the frequency of single ventricle palliative surgeries, the age at which these surgeries take place, and the percentage of patients who undergo Fontan procedures after the first stage of superior cavo-pulmonary anastomosis. Many ACHD patients in Egypt require Fontan surgery for a variety of reasons, including inadequate resources, a lack of supplies and proper conduits, and insufficient expertise at some hospitals to deal with patients requiring univentricular repair in childhood. Furthermore, the debate about the optimal timing for Fontan completion renders them a lesser priority in many pediatric cardiac facilities.

Delaying the Fontan procedure has been proposed by believers in the “ticking clock theory”: any Fontan circulation will eventually fail, and should thus be postponed to the last feasible stage [9]. However, it should be emphasized that contemporary Fontan surgical procedures have improved long-term prognosis [9], and several reports have shown that earlier Fontan surgery is associated with better outcomes [10,11]. Fontan completion in adulthood should therefore be considered the exception and can pose a challenge in treating ACHD patients in developing countries. Nonetheless, few reports have

shown acceptable short- and mid-term outcomes [12,13].

Patients with ToF, whether native or previously repaired, represented almost one-fourth of all ACHD patients in this study. Many previous reports have shown that patients with repaired ToF have good mid-term survival, approaching 90% at 20 years post-repair [14,15]. This survival has led to increasing numbers of adult patients with repaired ToF presenting to ACHD centers worldwide. These patients frequently present with significant morbidities, including severe pulmonary regurgitation and right ventricular dilatation requiring re-intervention, arrhythmias, residual shunts, and progressive effort intolerance [16,17].

In developing countries, naturally surviving adults with ToF are among those seeking medical help. In our cohort, unrepaired ToF, with or without previous palliative shunts, represented more than one-third of all ToF patients. Few previous studies investigated the outcome of late repair of ToF in adulthood, with controversial results [18–20]. Management of unrepaired adult patients with ToF is usually considered a real challenge as these patients are subjected to chronic hypoxemia, increasing the risk of myocardial dysfunction, arrhythmias, and other complications like cerebral insults caused by right to left shunting [19].

For our patients with repaired ToF, surgical pulmonary valve replacement was frequently indicated for severe pulmonary incompetence and/or stenosis. In general, it is considered safe, with low short- and mid-term mortality [21]. However, an increased risk of mortality was reported in redo surgeries [22], older patients, and those with concomitant surgeries [23]. In recent years, percutaneous pulmonary valve replacement has become a less invasive and effective alternative to address native pulmonary valve regurgitation/stenosis, and dysfunctional conduits. The learning curve and the procedure's cost are its main obstacles in developing countries, making it largely unavailable [24,25].

Multimodality cardiovascular imaging is becoming an integral part of the evaluation of ACHD patients [26–28]. In the present study, the final decision about the indication and type of surgical procedure required either a CMR or a CT scan in two-thirds of the patients and cardiac catheterization with hemodynamic assessment and/or angiography in one-fourth of the patients.

The choice of the imaging modality was usually based on the clinical question, in addition to the strengths and limitations of each modality. Performing and interpreting these tests requires trained personnel with sufficient knowledge and understanding of congenital heart anatomy, pathophysiology, and surgical/transcatheter interventions. Certainly, building an efficient ACHD service requires a well-organized partnership between ACHD-specialized cardiologists, congenital cardiac surgeons, and cardiac imagers with this kind of training and expertise.

Study limitations

The present study has some limitations. It described only ACHD patients who needed further surgical correction, in a single center. Thus, this cohort does not represent the whole population of ACHD patients in Egypt. The outcome of surgeries was not described, as it was outside the scope of this

study. However, we believe it will be highly beneficial to report the results, complications, and challenges faced by the surgical team operating on this group of patients.

Conclusion

Our ACHD patients who require further surgical correction is a heterogeneous cohort, with various presentations and needs. Most of them had already undergone at least one cardiac intervention. The required surgeries are highly variable in complexity. Creating an efficient ACHD program necessitates the collaboration of dedicated cardiologists, radiologists, and cardiothoracic surgeons who are familiar with the unique demands and difficulties of this group of patients.

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