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Adult Congenital Heart Disease Registry at Cairo University: A Report of the First 100 Patients

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Abstract

Aims: To establish a clinical registry for adult patients with congenital heart disease (CHD) managed in Cairo University Hospitals, aiming at description of the pattern and clinical profile of such patients. **Methods:** Patients were recruited from both Cardiovascular Medicine Department Outpatient Clinic and inpatient wards of Cairo University Hospitals. Clinical data were collected from hospital records and directly from patients by treating cardiologists. Collected data were then registered in a dedicated database system and subsequently analyzed. **Results:** Patients (49% males) ranged in age from 16 to 63 years, with a median of 25 years. Fifty-one patients were in the age-group from 20 to 30 years, with only 9% aged 50 years or older. Seventy-eight patients had acyanotic lesions, with atrial septal defect being the most common primary diagnosis (20% of total lesions). The remaining 22 patients had cyanotic heart disease, with tetralogy of Fallot being the predominant diagnosis (45% of cyanotic lesions). Six patients presented with infective endocarditis in the setting of CHD. Four women (8% of females) presented during pregnancy. Forty-six patients were sent for surgical correction/repair, while percutaneous intervention was planned in 20 patients. **Conclusions:** A new registry of adult patients with CHD managed in Cairo University Hospitals provides useful information, including the extent to which congenital heart defects are underdiagnosed and undertreated during infancy and childhood. In addition, those who were previously treated early in life require long-term follow-up in specialized centers. Establishment of a multidisciplinary team with expert physicians (cardiologists, dentists, obstetricians, and psychiatrists), cardiac surgeons, and nurses may be facilitated by development of a dedicated database system. Continuous financial support is a major challenge.

Keywords

adult congenital heart disease, clinical registry, challenges, Cairo University Hospitals, Egypt

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Introduction

Congenital heart disease (CHD) is the most common category of congenital defects, with a prevalence of 8 to 10 per 1,000 live births.[1] Improved surgical care, together with the advances in medical management, has led to a remarkable increase in survival of patients with CHD. Accordingly, in some developed countries, the number of adults now exceeds the number of children with CHD, and, as these patients mature into adulthood, many develop late cardiac complications.[2-4] Despite these facts, in some parts of the world there is little information about this group of patients and their needs. Specifically, in Egypt, data on the clinical profile of adult patients with CHD is lacking. We aimed to establish a registry to better understand the clinical characteristics of these patients with an ultimate goal of providing optimal care. A secondary hope is that this registry facilitates creation of a research program to further improve outcomes.

Patients and Methods

For this report, the population consists of the first enrolled 100 patients older than 16 years of age who had any form of CHD, whether repaired or not, and managed in our department.

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Abbreviations and Acronyms

ACHD	adult congenital heart disease
ASD	atrial septal defect
BAV	bicuspid aortic valve
CHD	congenital heart disease
CONCOR	CONgenital CORvitia
IE	infective endocarditis
PDA	patent ductus arteriosus
PS	pulmonary valve stenosis
TEE	transesophageal echocardiography
TOF	tetralogy of Fallot
VSD	ventricular septal defect

Table 1. Causes of Referral of Patients to Our Center.

Causes of Referral	Number of Patients
Possible percutaneous/surgical intervention	57
Regular follow-up visits	15
Heart failure	6
Assessment prior to noncardiac surgery	6
Suspicion of infective endocarditis	6
Pregnancy	4
Arrhythmias	4
Resistant hypertension	1
Stroke	1

The registry was approved by the hospital ethical committee, and all patients gave informed consent before participation. Patients with Marfan syndrome as well as those with mitral valve prolapse were not included in the registry. We recruited outpatients as well as hospitalized adults. Many of the patients were referred to us from general cardiologists for possible percutaneous/surgical intervention. Pregnant women were referred for assessment before delivery, and patients scheduled to undergo noncardiac surgery were referred from different surgical wards.

Data were collected by cardiologists either directly from the patients seen in the outpatient clinic or from the hospital records of hospitalized patients. All data were recorded in a dedicated electronic database system and analyzed after enrollment of the 100th patient. Data collected included demographic features (date of birth, gender, and contact information), clinical characteristics, diagnostic studies, and interventions performed. An electrocardiogram and transthoracic echocardiography were performed for all patients. Other investigations including transesophageal echocardiography (TEE), Holter monitoring, cardiac catheterization, and cardiac magnetic resonance imaging were performed as indicated for each individual patient. The complexity of the CHD was defined using the 32nd Bethesda Conference–Task Force 1 recommendations.[5]

Statistical Analysis

Statistical analysis was performed using SPSS version 17.0 for data management. Statistical methods used were only descriptive. Categorical variables were presented as numbers and percentages. Continuous variables were presented either as mean (with standard deviation) for normally distributed variables or as median (with interquartile range) for nonnormally distributed data.

Results

Baseline Characteristics

One hundred patients (age > 16 years) were enrolled in the registry. Sixty-three patients were from Cairo and the Nile Delta region, 29 from Upper Egypt and El Fayoum, with the remaining patients from Hurghada, Suez, and Alexendria. Fifty-seven patients were referred to our center for possible percutaneous or surgical intervention (Table 1). Age of patients ranged between 16 and 63 years, with a median of 25 years. About half (51%) of the patients were in the age-group from 20 to 30 years, with only 9% of the patients aged 50 years or older. The age range at the time of diagnosis ranged from birth to 58 years, with a median of 19 years. Forty-five patients were diagnosed during adulthood, with atrial septal defect (ASD) being the most commonly encountered lesion. Thirty-seven patients were diagnosed early in life (since birth and during infancy), with ventricular septal defect (VSD) and tetralogy of Fallot (TOF) being the most prevalent lesions. Clinically, none of the patients had manifestations suggestive of a syndromic CHD. Genetic testing was not performed.

Fifty-one patients were females. Atrial septal defect, pulmonary valve stenosis (PS), and aortic coarctation were more common in females, while VSD and TOF were more common in male patients. Four women (mean age 27 years and 8% of females) were pregnant and referred for assessment of their cardiac condition prior to delivery. One of the four patients referred during pregnancy (27-year-old) had undergone a bidirectional Glenn procedure before conception. This patient was kept in the intensive care unit prior to delivery for proper monitoring of her oxygen saturation and hemodynamics as well as the status of the fetus. Oxygen saturation was in the mid-80% range. The remaining three pregnant women had unrepaired lesions. One patient had a single moderate-sized muscular VSD associated with a small- to moderate-sized patent ductus arteriosus (PDA), resulting in left ventricular dilation and mild pulmonary hypertension. During pregnancy, this 39-year-old woman developed symptoms of congestive heart failure (New York Heart Association functional class 3) and received anticongestive therapy in the form of oral diuretics. The second patient had a large secundum ASD and right ventricular dilation. The last pregnant patient had a large hemodynamically significant secundum ASD associated with severe PS (with peak instantaneous Doppler gradient = 70 mm Hg) and right ventricular enlargement. This patient developed infective endocarditis (IE) 2 months after a vaginal delivery. She presented with fever, loss of appetite, and shortness of breath. Vegetations were detected on both the mitral and pulmonary valves, with severe mitral valve regurgitation that necessitated surgical intervention. Only one pregnancy was delivered by cesarean section, while the other three had successful uncomplicated vaginal deliveries. All patients received intravenous

Table 2. Anatomical Types of Atrial Septal Defect (ASD) and Ventricular Septal Defect (VSD) Detected in the Registry.

Type of the Lesion	Number of Patients
ASD	
Secundum ASD	19
Isolated secundum ASD	15
Associated with persistent left superior vena cava	1
Associated with polysplenia syndrome	1
Associated with valvular pulmonary stenosis	1
Associated with rheumatic mitral stenosis	1
Sinus venosus ASD	1
VSD	
Surgically repaired, anatomy unknown	5
Perimembranous VSD	9
Isolated VSD	2
Associated with other acyanotic lesions	5
Associated with prolapsing right coronary cusp	1
Complicated by pulmonary hypertension	1
Muscular VSD	2

prophylactic antibiotics prior to delivery. Four healthy babies were delivered (mean gestational age, 36 $\pm$ 1 weeks). Birth weights were 2,700 to 3,100 g (median 2,800). Congenital heart disease was excluded clinically in all of them.

Of the 100 adult patients with CHD, 7 had significant pulmonary artery hypertension. Five of them (median age 35 years) were female (two patients had ASD, one had VSD, one had PDA, and one had atrioventricular canal defect), while the remaining two patients were male (median age 51 years), both of whom had ASDs.

Type of Cardiac Lesions

Seventy-eight patients had acyanotic lesions, with ASD being the most common (20% of total lesions and 25% of the acyanotic population). Four patients with ASD had pulmonary hypertension, one had concomitant valvular PS, and another had associated rheumatic mitral stenosis (Lutembacher syndrome). In all, 17 patients had VSD (Table 2), 12 patients had valvular PS, and 10 patients had aortic coarctation. Less frequently encountered acyanotic lesions were fibromembranous subaortic stenosis (seven patients), atrioventricular canal defect (five patients), and PDA (three patients).

Twenty-two patients had cyanotic heart disease, with TOF being the predominant defect (45% of cyanotic lesions). Other cyanotic lesions included tricuspid atresia (three patients), other functionally univentricular heart (two patients), corrected transposition of the great arteries (two patients), double outlet right ventricle (two patients), truncus arteriosus (one patient), one patient with transposition of the great arteries with previous Senning procedure, and one patient with double-chambered right ventricle with VSD.

Overall, only 19 patients had undergone therapeutic interventions before enrollment in the registry, including surgery (17 patients) or a catheter-based intervention (2 patients).

The most commonly affected valve in our registry was the pulmonary valve (15 patients), 12 patients had valvular PS, 2 patients had valvular PS associated with other lesions (ASD and VSD), and 1 patient had severe pulmonary regurgitation following repair of TOF. The tricuspid valve was affected in nine patients: three patients had tricuspid atresia, two had double outlet right ventricle and severe tricuspid valve regurgitation, two patients had partial atrioventricular canal defect with severe tricuspid regurgitation, one had Ebstein's anomaly, and one patient had significant tricuspid regurgitation following surgical closure of a VSD.

Six patients were proved, echocardiographically and by blood cultures, to have IE in the setting of CHD. The underlying cardiac defects were VSD (two patients), fibromembranous subaortic stenosis (one patient), PDA (one patient), bicuspid aortic valve (BAV; one patient), and secundum ASD associated with valvular PS (one patient, following normal vaginal delivery).

Investigations and Management

Transthoracic echocardiography identified all cardiac lesions in 99% of patients. Additional investigations were performed in 42 patients. Transesophageal echocardiography was performed in 20 patients (75% of them had ASD and underwent TEE for better delineation of the surrounding rims prior to planned percutaneous closure). Transesophageal echocardiography was also performed for assessment of four patients with IE and one patient with atrial flutter to rule out atrial thrombi prior to cardioversion.

Patients with aortic coarctation routinely underwent magnetic resonance aortography prior to any percutaneous or surgical intervention. Cardiac catheterization was performed in ten patients; three with TOF, and six with cardiac shunts and echocardiographically elevated pulmonary artery systolic pressure, for better assessment of the pulmonary vascular resistance. One patient with recurrent subaortic stenosis underwent aortography for better assessment of the severity of aortic regurgitation. Coronary angiography was performed in three patients prior to surgery; two were above 40 years and had severe aortic stenosis of a congenitally BAV and the third one (53 years) had TOF. Electrophysiology studies and ablation were performed in two patients. One had Ebstein's anomaly and atrial flutter. The second, who had previously undergone repair of TOF, presented with repeated episodes of ventricular tachycardia and had severe pulmonary regurgitation and right ventricular enlargement.

Forty-six patients were sent for surgical correction/repair, while percutaneous intervention was planned in 20 patients. Planned surgical and percutaneous interventions are summarized in Table 3. Patients with previously corrected lesions, those with mild to moderate valvular stenosis or regurgitation, and those with small left to right shunts (20 patients) were advised to attend regular follow-up visits in our center. Medical treatment was prescribed for the remaining 14 patients.

Table 3. Planned Management of the Enrolled Patients.

Plan	Number of Patients
Surgical closure of VSD (using Dacron graft)	10
Surgical closure of ASD (simple suture or using pericardial patch)	10
Percutaneous closure of secundum ASD	8
Total correction of TOF	7
Stenting of aortic coarctation	6
Balloon pulmonary valvuloplasty	5
Surgical repair of aortic coarctation	4
Surgery for fibromembranous subaortic stenosis	3
Surgical aortic valve replacement	2
Fontan procedure	2
Pulmonary valve replacement by tissue valve	2
Ablation of atrial flutter	1
Others	6

Abbreviations: ASD, atrial septal defect; TOF, tetralogy of Fallot; VSD, ventricular septal defect.

Medical treatment included drugs for pulmonary artery hypertension, systemic hypertension, and antiarrhythmic medications.

Discussion

Improvements in diagnosis, medical treatment, and surgical repair are changing the pattern of survival in patients with CHD.[5-7] Nowadays, 90% of children with serious CHD who have access to surgery survive to adulthood and 55% of them require long-term care by a specialist.[8] Unfortunately, data on the exact prevalence, management, and long-term outcome of CHD in adults (ie, adult congenital heart disease [ACHD]) in Egypt are lacking because of the absence of dedicated national registries. Accordingly, we established this clinical registry to produce a ''real-world'' picture of ACHD in our community, including current treatment practices and outcomes. This registry promotes the development of an efficient organizational structure with the ultimate aim being to improve clinical outcomes of ACHD. Equally important, it enables training of general cardiologists interested in ACHD, allows scientific research in this field, and facilitates the cooperation between specialist cardiologists in our center and the general cardiologists who do not specialize in the management of ACHD. This cooperation is highly recommended for proper management of complicated cases, especially those with heart failure, pulmonary hypertension, arrhythmias, IE, and high-risk pregnancies.

The staff involved in this registry included a senior adult cardiology consultant interested in the management of ACHD, two echocardiographers, two interventional cardiologists, and two additional cardiologists responsible for collection and analysis of data. All involved cardiologists and echocardiographers are adult cardiologists with special interest in ACHD. Moreover, only through personal communication, the team members were in contact with cardiac surgeons at Cairo University Hospital and Aswan Heart Center as well as with two expert electrophysiologists and an expert radiologist interested in cardiac magnetic resonance imaging. A high-risk pregnancy service is currently available at the hospital where pregnant women with CHD are managed. We don't have at present dental physician(s) responsible for routine and emergent dental care/procedures or a clinical nurse specialist or adequate numbers of medical secretaries. We currently face the following challenges: (1) the number of cardiologists interested in ACHD and willing to join the team remains small relative to the projected future workload, (2) the number of cardiac surgeons interested in the management of these patients is limited, especially with respect to the cyanotic population in which inconsistent results have been achieved in the past, (3) there is a limited supply of devices for interventional management of our patients due to lack of funds, (4) there is no formal organized connection between pediatric and adult cardiologists, as a result of which many patients are presumably lost to follow-up during this transitional period, and (5) there is uncertainty regarding continuous financial support to keep this costly registry going on.

Clinical Characteristics

Unlike some other CHD registries,[9,10] patients with Marfan syndrome and mitral valve prolapse were not enrolled in our work. On the other hand, patients with BAV were included, even in the absence of significant valvular dysfunction, as BAV may be associated with other CHD and with aortopathy and aortic complications even in the absence of noninvasively detected gross aortic abnormalities.

About two-thirds of our patients were classified as having complex lesions, making them susceptible to special risks and complications. These patients included those with cyanotic heart disease, some subsets of patients with acyanotic heart disease, and those with Eisenmenger syndrome.[5] Unfortunately, this figure is much higher than that detected in some previous studies of ACHD populations in other parts of the world, where prevalence of such lesions varied between 31% and 53%.[5,11,12] This suggests that a large percentage of our patients are in need of continuous care and may require costly management.

Compared with the patients enrolled in the CONgenital CORvitia (CONCOR) registry in the Netherlands, our patients were younger.[9] This could be explained by the higher prevalence of certain lesions, such as valvular PS and cyanotic lesions in our patients, which are usually diagnosed and treated before adulthood in developed countries. Atrial septal defect was the most prevalent lesion, followed by VSD, valvular PS, and TOF. The prevalence of VSD, aortic coarctation, and TOF in this registry was comparable to that observed in previous studies.[6,9,10] The prevalence of ASD, however, was much higher (43.3%) in a previous study.[13] Similar to previous reports,[9,10,16] this registry showed that ACHD was more prevalent in females and this is expected owing to the high prevalence of ASD among those patients. Unlike findings of the CONCOR registry,[9] aortic coarctation and documented

arrhythmias were more prevalent among females in our population.

Management

Among recruited patients, 66 patients were in need for intervention, 46 were sent for surgical correction, while percutaneous intervention was recommended in the remaining 20 patients. Treatment of our patients by means of surgery or an interventional approach is relatively expensive. The government subsidizes some of the patients who undergo surgical correction which is generally less expensive and, accordingly, more available than percutaneous interventions (especially for ASD). On the other hand, many nongovernmental organizations donate care and devices for the interventional treatment of patients with ACHD.

We refer our patients routinely to the cardiothoracic surgical department where surgical correction is performed by adult cardiac surgeons trained in pediatric cardiac surgery. Patients with aortic coarctation are referred to an adult cardiac surgeon who has expertise in the care of aortic lesions (both congenital and acquired). Occasionally, complicated cases are sent to the Aswan Heart Center, a specialized center with surgeons who specialize in treating CHD.

Unfortunately, not all patients sent for surgery were accepted by the surgeons. One of these patients (21-year-old female) had TOF and pulmonary atresia. Cardiac catheterization and magnetic resonance angiography revealed nonconfluent pulmonary arteries, and she was referred for surgical management (unifocalization). The patient was considered too high risk for surgical intervention, despite a history of presenting with headache and blurring of vision with a hematocrit level of 70% every 4 months. Although we try to avoid long-term reliance on this management strategy in our patients with cyanosis, the only treatment that we could offer her was therapeutic phlebotomy for relief of her symptoms related to hyperviscosity.

Gender disparities have been documented extensively in various fields of cardiovascular disease. Women may have distinctive clinical manifestations and outcomes and appear to be underdiagnosed, underinvestigated, and undertreated in some parts of the world.[14] In general cardiology, the suggestion that in similar physical conditions females are less likely to be referred, undergo diagnostic investigation, and receive treatment has been labeled the Yentl syndrome.[15] Engelfriet and Mulder have previously reported the existence of gender differences in morbidity, mortality, and medical management among adult patients with CHD.[16] Moreover, girls were less likely to undergo recommended surgical intervention, in part related to parental concerns about future matrimonial prospects and lack of social support.[17] In our center, we prefer to treat female patients with CHD as soon as possible to avoid pregnancy-related complications. Additionally, we try to provide an alternative percutaneous management to some of these patients who frequently refuse any surgical intervention because of social and cosmetic concerns.

Study Limitations

Our sample of patients does not represent the population of ACHD in Egypt but rather reflects a subset of patients seeking medical help at a major tertiary care center. Also, the lack of data on the pattern of CHD in children, median age of diagnosis, and the nature of treatment provided limit our ability to explain the present data.

Conclusion

Findings and pattern of CHD in the current registry suggest that in Egypt many patients born with CHD are not diagnosed and managed during their infancy and childhood, as would be the case in the more developed countries. Even those previously treated need costly management and follow-up. Sustained effort and financial support are challenging and require a major commitment from the government and Ministry of Health to provide better service for this growing population. A primary health care system where CHD diagnosis is not delayed beyond school admission is highly recommended. In countries without existing large database systems, the development of a clinical registry of ACHD provides important data that reflect real-world clinical practice, helps in the establishment of a dedicated multidisciplinary team, and encourages training of cardiologists specialized in the management of ACHD to achieve better outcomes in these patients.

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Declaration of Conflicting Interests

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