

## Pattern of Adult Congenital Heart Disease in a Tertiary Care Hospital in Saudi Arabia

Mi-Yeon Kim<sup>1</sup>,  
Mohammed Omar Galal<sup>2</sup>,  
Jameel Al-Ata<sup>2</sup>,  
Amjad Kouatli<sup>2</sup> and  
Milad El-Segaier<sup>3</sup>

### Abstract

**Background:** As the growing number of patients with Congenital Heart Disease (CHD) is surviving into the adulthood, the availability of regional centers that specialize in the treatment and follow-up care of adults with CHD bears a great importance. The need is especially pronounced for the population in the Middle East where the defect may not be detected in timely manner.

**Methods:** The clinical records of 30 patients referred to the Adult Congenital Heart Disease Program at a tertiary care hospital in Western Saudi Arabia were reviewed for the study. The patients were categorized into four groups based on their diagnoses: left to right shunt lesions; complex lesions; obstructive lesions; and arrhythmia. Following the diagnosis, four different type of treatments were provided to patients: 1) Interventional catheterization, 2) Surgery, 3) Medical treatment, or 4) No intervention.

**Results:** The patients in the complex lesions group exhibited the most serious and the highest number of symptoms. Fifteen patients underwent the cardiac interventional catheterization, eleven patients underwent surgery, one patient was placed on a medical treatment, and one patient was deemed not to be in need of any treatment.

**Conclusion:** Many of the patients with complex lesions were diagnosed relatively late in childhood and therefore had late first interventions. The regional facility equipped to early diagnose CHD and provide optimal treatment would increase patients' quality of life and reduce the ultimate health care costs. An awareness of the importance of the early diagnosis and treatment on the part of the primary physician, along with the education of patients and family affected with congenital heart disease on the importance of continued follow-up care in the center that specializes in adult congenital heart disease cannot be over-emphasized.

**Keywords:** Adult congenital heart disease; Interventional cardiac catheterization; Congenital heart disease; Heart defects; Health care

**Abbreviations:** ACHD: Adult Congenital Heart Disease; ASD: Atrial Septal Defect; AVSD: Atrioventricular Septal Defect; BTshunt: Blalock-Taussig Shunt; CHD: Congenital Heart Disease; CoA: Coarctation of Aorta; DORV: Double Outlet Right Ventricle; L-TGA: Levo-Transposition of Great Arteries; PAPVR: Partially Anomalous Pulmonary Venous Return; PDA: Patent Ductus Arteriosus; PS: Pulmonary Stenosis; PSVT: Paroxysmal Supraventricular Tachycardia; RPA: Right Pulmonary Artery; SAM: Systolic Anterior Motion; SOB: Shortness of Breath; TOF: Tetralogy of Fallot; TVR: Tricuspid Valve Repair; VSD: Ventricular Septal Defect

- 1 University of Calgary in Qatar, Doha, Qatar
- 2 Pediatric Cardiology, King Faisal Specialist Hospital, Jeddah, Saudi Arabia
- 3 Pediatric Cardiology, King Fahad Medical City, Riyadh, Saudi Arabia

**Corresponding author:** Omar M Galal

✉ ogalal@yahoo.com

Consultant, Pediatric Cardiology, King Faisal Specialist Hospital, Jeddah, Saudi Arabia.

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## Introduction

Congenital Heart Disease (CHD) causes the most frequent birth defect which, according to some researchers, has geographical variations in terms of the incidence rate and the type of heart defects. Although the most widely quoted incident rate of CHD in the world is 8/1000 live births, evidence from the literature indicates that the actual rate varies between 1.2–17 [1], with the rate in the Middle East being on the higher end at 12.23 [2].

As the growing number of patients with CHD is surviving well into their adulthood, the need for a regional center that specializes in the treatment of these patients who often require life-long follow up care has become imperative. The need for a specialized care facility for Adults with Congenital Heart Disease (ACHD) is especially urgent in the Middle East where patients often do not get diagnosed in a timely manner because of a late presentation to the health care system as well as a lack of availability to the medical resource. The treatment of individuals living with CHD is resource-intensive which causes a significant burden on health care costs. A careful planning in developing the necessary resources for this population, who often exhibit comorbidity, would ensure the maximum benefit of health care funding.

Examination of the type and pattern of patients with CHD being treated in Saudi Arabia would assist with collecting necessary information for addressing both the current and changing needs of this population and for developing health policies to provide direction in the health care system.

The purpose of this study was to describe the patient profile and the type of intervention provided to a group of individuals treated in a tertiary care facility in Saudi Arabia.

## Methods

The clinical records of 30 patients referred to the Adult Congenital Heart Disease Program in a tertiary care hospital in Jeddah, Saudi Arabia, were reviewed for the study. The demographic data collected from each patient includes: age, sex, nationality, height and weight. Clinical data was also collected on the patient's diagnosis, presenting signs and symptoms, type of intervention performed, and the outcome of the intervention.

## Results

The summary of the demographic characteristics of 30 patients is presented in **Table 1**. The subjects consisted of twelve males and eighteen females ranging in age from 14 to 59 years (mean: male=20, female=25). The mean height and weight were 164 cm and 55 kg, respectively, for males, and 154 cm and 51 kg for females. Although the patients were of diverse nationalities including Saudi, Yemen, Egyptian, Palestinian, Somalian and Sudanese, more than 50% of them were Saudis. The nationalities of two patients were unknown. The diagnosis and treatment options for each of the 30 patients were established following clinical examination, 2D transthoracic echocardiography, color Doppler, and cardiac catheterization. Each of the patient's diagnosis, presenting signs and symptoms and the type of the

intervention received in the hospital are presented in **Table 2**. The data pertaining to the patients' age at each time of the diagnosis, the initial presentation to the hospital for treatment and the first intervention, are illustrated in **Table 3**. It was interesting to note that, in a few patients whose diagnosis of the CHD had been established relatively early in childhood allowing for a timely intervention, it still took them a mean of 13 years to seek treatment despite the severe limitation in activities they had been experiencing from their symptoms.

Following the examination of clinical data, patients were categorized into four groups based on the type of their diagnoses: the first group consisting of left to right shunts lesions (n=16); the second group consisting of complex lesions (n=7); the third group consisting of obstructive lesions (n=5); and the fourth group consisting of arrhythmia (n=2).

### Left to right shunts

Sixteen out of thirty patients were affected with left to right shunt lesions. For some of the patients in this group, the congenital anomalies had been discovered either by accident or from a routine physical examination in school. Although not all patients were symptomatic, the primary presenting complaints of these patients were Shortness of Breath (SOB), exercise intolerance and fatigue. For example, a 25 year old male with a partial Atrioventricular Septal Defect (AVSD), who works as a National Guard Army Officer, had been suffering for fifteen years with easy fatigue that often forced him to take sick days from work. In two years prior to presenting to the hospital, his condition has progressively worsened and he had begun experiencing syncope, which would last approximately two minutes per episode. The episodes of syncope increased in frequency to the point where it would sometimes occur almost daily. Another patient, a 40-year old female diagnosed with an Atrial Septal Defect (ASD), had had previous surgery to close the defect. Her husband, however, reported that the ASD reopened shortly after the intervention. By the time she entered the ACHD program at a tertiary care hospital thirteen years later, she had developed a severe pulmonary hypertension with an echocardiographic finding of a tricuspid valve regurgitation peak systolic pressure gradient of 86

**Table 1:** Demographic characteristics of study participants.

Variables		Description
Sex	Male	12
	Female	18
Age (mean)	Male	20
	Female	25
Nationality	Saudi	19 (63.3%)
	Yemen	4 (13.3%)
	Egypt	2 (6.7%)
	Palestine	1 (3.3%)
	Somalia unknown	1 (3.3%)
Height* (mean)	Male	164
	Female	154
Weight‡ (mean)	Male	55
	Female	51

\*Measurement in cm

‡ Measurement in kg

**Table 2:** Diagnosis, presenting signs and symptoms, and the intervention/surgery provided for 30 patients.

Lesions	No. of subject	Signs & Symptoms	Intervention
<b>Left to Right Shunt Lesions</b>			
ASD	4	Easy fatigue, SOB, chest pain, right-sided weakness, syncope	Interventional catheterization with the Amplatzer device (n=3); medical treatment (n=1)
VSD	4	SOB, easy fatigue, palpitation	Interventional catheterization with the Amplatzer (n=1 treatment not required (n=1); no show (n=2)
PDA	4	Mainly asymptomatic	Interventional catheterization with the Amplatzer (n=2); Coil (n=2)
ASD, PAPVR	1	Easy fatigue, exercise intolerance, SOB, weight loss	Surgery (ASD closure & re-routing of PAPVR to LA)
Partial AVSD (ASD I, no VSD)	1	Easy fatigue, syncope, SOB, NYHA III	Surgery (ASD closure)
DCRV, VSD	1	110 mmHg gradient across mid-chamber obstruction, asymptomatic	Surgery (RV muscle resection & VSD closure)
Rupture of Sinus of Valsalva Aneurysm, SAM, VSD	1	SOB, easy fatigue, palpitation	Surgery (excision & repair of Sinus of Valsalva fistula; VSD patch closure; SAM resection)
<b>Complex Lesions</b>			
Ebstein's Anomaly, TVR (x2), S/P ASD repair	1	NYHA II-III, moderate-severe aortic regurgitation, severe dyspnea on min. exertion, palpitation, presyncope, chest pain	Medical treatment
DORV, PA, AVSD, Hypo LV	1	Severe exercise intolerance, cyanosis, SOB, SaO <sub>2</sub> 55%, palpitation	Interventional catheterization (dilatation of BT Shunt)
DORV, VSD, PS, Sub-PS	1	Chest pain, palpitation, NYHA II-III	Surgery (Rastelli & fenestrated Fontan)
DORV, RPA stenosis	1	Easy fatigue, orthopnea	Surgery (Rastelli & RPA stenting)
DORV, L-TGA, VSD, Valvar PS, PSVT, pulmonary situs inversus	1	Chest pain, cyanosis, SOB, palpitation	Surgery (Glenn & Fontan)
TOF, VSD	1	Cyanosis, clubbing of fingers & toes,	Surgery (total repair)
TOF, VSD, Valva & Subvalvar PS	1	Easy fatigue, diaphoresis, SaO <sub>2</sub> 89%, SOB, cyanosis, exercise intolerance	Surgery (total repair)
<b>Obstructive Lesions</b>			
PS	3	SOB, NYHA II, diaphoresis, fatigue	Interventional catheterization (balloon valvuloplasty)
CoA	2	SOB, NYHA II, palpitation, chest pain	Interventional catheterization (balloon dilatation)
<b>Arrhythmia</b>			
Atrial flutter (following removal of LA myxoma)	1	SOB, NYHA II, palpitation	Interventional catheterization (Radiofrequency ablation)
Wolf-Parkinson-White Syndrome	1	Palpitation, SOB, diaphoresis	Interventional catheterization (Radiofrequency ablation)

**Abbreviations:** ASD: Atrial Septal Defect; VSD: Ventricular Septal Defect; PDA: Patent Ductus Arteriosus; PAPVR: Partially Anomalous Pulmonary Venous Return; DCRV: Double Outlet Right Ventricle; SAM: Systolic Anterior Motion; TVR: Tricuspid Valve Repair; DORV: Double Outlet Right Ventricle; PA: Pulmonary Atresia; AVSD: Atrioventricular Canal Defect; PS: Pulmonary Stenosis; CoA: Coarctation of Aorta; L-TGA: Levo-Transposition of Great Arteries; RA: Right Pulmonary Artery; PSVT: Paroxysmal Supraventricular Tachycardia; TOF: Tetralogy of Fallot; SOB: Shortness of Breath

mmHg and bi-directional shunting across the atrial septal defect. Interventional cardiac catheterization was performed to close the defect in eight subjects; the Amplatzer device was used in six patients and the PDA Coil was deployed in two patients. The remaining four patients required surgical intervention.

For a 40-year old patient with ASD II, her existing condition of severe pulmonary hypertension and its associated bi-directional shunts precluded neither a cardiac catheterization nor a surgery. She was instead put on a medical treatment of Sildenafil and Furosemide-, with a possibility in future of surgery if she responded favorably to the treatment. One patient with a muscular Ventricular Septal Defect (VSD) did not require any

intervention as the defect was considered to be too small to cause any hemodynamic problems. Two patients, one in need of a surgery for a perimembranous VSD and another in need of a cardiac catheterization for a muscular VSD, did not show up for the procedures.

### Complex heart lesions

Seven patients belonged in the group of complex congenital heart lesions, which by definition, referred to individuals having three or more associated lesions. These individuals were severely limited in their activities by their CHD in comparison to patients in other categories. All patients exhibited cyanosis and suffered from SOB

**Table 3:** Comparison of time: age at the time of diagnosis, age at the time of first intervention, and age at the time of intervention received at the current in this study.

Subject	Diagnosis	Sex	age	Age at diagnosis	Age at 1st intervention	Age at intervention (at the time of this study)
<b>Left to Right Shunt</b>						
1	ASD II	M	34	33	34	34
2	ASD II	F	27	27	27	27
3	ASD II	M	18	18	18	18
4	ASD II	F	40	Unknown	27	40
5	VSD (perimembranous)	M	18	11	18	18
6	VSD (muscular)	F	22	“Childhood”	4	22
7	VSD (muscular)	F	18	18	N/A	Not required
8	VSD (muscular)	M	14	2	N/A	No show
9	PDA	M	22	Not known	22	22
10	PDA	F	14	Preschool	14 (asymptomatic)	14
11	PDA	F	31	31	31	31
12	PDA	F	14	14	14	14
13	Partial AVSD	M	25	infancy	25	25
14	ASD II & PAPVR	F	23	22	22	23
15	Double chamber RV & VSD	M	23	1 (asymptomatic)	was told not required	23
16	Rupture of Sinus Valsalva & SAM, VSD	F	23	13	23	23
<b>Complex</b>						
17	Ebstein’s Anomaly, S/P ASD repair, S/P valvuloplasty, S/P TVR (x2)	M	40	5	5	39 High surgical risk Medical treatment
18	DORV, AVSD, malposed great arteries, pulmonary atresia, S/P BT shunt	F	14	4	4	14
19	DORV, VSD, PS, Sub PS, S/P bi-directional Glenn	F	19	10	10	19
20	DORV, RPA stenosis	F	18	“early in life”	8	17
21	L-TGA, DORV, valvar PS, S/P Pulmonary valvuloplasty, PSVT	F	34	“childhood”	10	34
22	TOF, VSD (muscular), S/P BT shunt	M	15	2 months	4 months	15
23	TOF, VSD (subaortic), Valvar & Sub-valvar PS	F	16	8	8	16
<b>Obstructive</b>						
24	PS	F	17	17	17	17
25	PS	F	59	unclear	59	59
26	PS	M	16	7	16	16
27	CoA	F	24	15	23	23
28	CoA	M	15	Infancy (lost in follow up)	15	15
<b>Arrhythmia</b>						
29	Atrial flutter (following resection of myxoma in LA)	M	18	18	18	18
30	Wolf-Parkinson-White Syndrome	M	17	12	17	17

**Abbreviations:** ASD: Atrial Septal Defect; VSD: Ventricular Septal Defect; PDA: Patent Ductus Arteriosus; PAPVR: Partially Anomalous Pulmonary Venous Return; DORV: Double Outlet Right Ventricle; SAM: Systolic Anterior Motion; S/P: Status Post; TVR: Tricuspid Valve Repair; BTshunt: Blalock-Taussig Shunt; DORV: Double Outlet Right Ventricle; PA: Pulmonary Atresia; AVSD: Atrioventricular Canal Defect; PS: Pulmonary Stenosis; L-TGA: levo-Transposition of Great Arteries; RA: right Pulmonary Artery; PSVT: Paroxysmal Supraventricular Tachycardia; TOF: Tetralogy of Fallot; CoA: Coarctation of Aorta

on minimal exertion, with four of the patients experiencing an additional symptom of palpitation. Many of them also reported symptoms of dizziness, chest pain and easy fatigue. Most of the patients were in NYHA class II-III. Six patients had received previous interventions for their CHD. The patient with Ebstein's Anomaly had undergone two previous Tricuspid Valve Repair (TVR)s, ASD repair, Pulmonary Valvuloplasty and has now developed a severe aortic regurgitation. The patient with DORV with Pulmonary Atresia had a classical left BT Shunt performed at age 4 but it has since become stenosed. The third patient with Double Outlet Right Ventricle (DORV), VSD, pulmonary valve stenosis (PS) and sub-PS received the bi-directional Glenn procedure at age 10, however the follow-up history of the procedure was unknown. The fourth patient with levo- transposition of great arteries (L-TGA), DORV, VSD, and Valvar PS received a palliative Pulmonary Valvuloplasty at age 10, and although having had regular follow-up care, he has since been experiencing frequent episodes of symptomatic Paroxysmal Supraventricular Tachycardia (PSVT) that often required hospitalization to resolve it. The fifth patient with Tetralogy of Fallot (TOF) was diagnosed at the age of two months and subsequently had the BT Shunt performed at four months. The patient did not maintain regular follow-up care and, after seventeen years, has presented with a continuous and progressively worsening cyanosis. The sixth patient, also with TOF, was first seen at the age of eight months when apparently the first cardiac catheterization was performed. It is unclear, however, whether the procedure was diagnostic or interventional in nature. Nonetheless the patient fared relatively well until three years ago when she started to experience SOB with minimal exertion and easy fatigue.

The team members of the Adult Congenital Heart Disease Program agreed that all but two patients required surgical intervention. Of the two remaining patients, the patient with Ebstein's Anomaly who had developed a severe aortic regurgitation was considered to be too high a risk for a surgical procedure and therefore, was placed on a medical treatment instead.

### Obstructive lesions

The obstructive lesions observed in five patients were in the form of either a severe PS or a Coarctation of Aorta (CoA). While all patients suffered from SOB, one of the patients experienced an additional symptom of syncope. Most of the patients were in NYHA class II. The mean peak pressure gradient across the obstruction prior to the intervention was 86 mmHg, which decreased to 25.6 mmHg following the intervention. The interventional catheterization of balloon valvuloplasty was performed on all patients including one patient who refused surgery and opted for a catheterization treatment instead. The Cristal Balloon was utilized in three patients with PS and one patient with CoA, while the Double Lumen Balloon was utilized in one patient with CoA.

### Arrhythmia

Two patients were affected with arrhythmia. An 18-year old male developed an atrial flutter with 3:1 AV block following the resection of 8 × 5 cm myxoma in his left atrium. The other 17-year

old male was diagnosed with Wolf-Parkinson-White Syndrome associated with increasing episodes of palpitation. Palpitation was occurring a few times a month and often required hospital treatment to resolve. Electrophysiology study revealed an easily inducible AVRT (atrial ventricular re-entry tachycardia) and atrial fibrillation with rapid ventricular response in the range of 250-300 bpm. Both patients underwent successful Radiofrequency Ablation Therapy.

### Discussion

This study describes the clinical profile and treatment of selected patients referred to one of the few tertiary care centers capable of treating CHD in Saudi Arabia. The patients in this study were afflicted with congenital heart defects that varied in clinical complexity from mild to severe. All patients except one required either interventional catheterization or surgery to repair and/or hasten the symptomatic progression of the defects.

The most common types of CHD lesions observed in this study's participants were ASD and VSD of left-to-right shunt lesions. The high number of patients affected with ASD and VSD has previously been noted in different studies that reported the three most common defects as VSD, ASD, and TOF [1,3-5]. The actual prevalence rate of patients born with ASD and VSD in the Middle East may be much higher than the observed number given that the patients included in the study were limited to those who had access to the tertiary regional health center. The high incidence of CHD in the Middle East could partly be attributed to some of the genetic causative factors such as autosomal recessive mutations [6]. More specifically, evidence suggests that the parental consanguinity, a commonly observed cultural practice in people of the Middle East, is one of the factors that increase the risk of CHD by two to three times [7-9].

The type of heart defects reported to be significantly associated with first-cousin consanguinity is: VSD, ASD, AVSD, PS, and PA [6]. However, it was unknown whether, or how many, of the study participants were offspring of parental consanguinity.

The number of patients with complex heart lesions was also relatively high and accounted for almost as many cases as ASD and VSD combined. Many of the patients with complex lesions were delayed in receiving first intervention attributable to being diagnosed relatively late in childhood. Even in those patients whose diagnosis had been established early after birth and therefore were in a position to receive the most optimal treatment available, it took them almost 10 years to seek treatment. By this time, they were severely limited in functional capacities and their congenital heart lesions too advanced to obtain a maximum benefit from the intervention. Some of the reasons for the delay in establishing a diagnosis or seeking necessary treatment may be due to the lack of available regional facilities or the patient's inability to access the facilities for economic reasons [10]. One of the patients in particular, for reasons unknown, failed to seek the required treatment for a prolonged period. By the time she sought treatment, the progress of the disease was far too advanced for treatment.

## Conclusion

As individuals with CHD are now experiencing increased longevity as a result of advanced surgical and medical treatments, the issues of possible complications or residual lesions of repaired heart defects as well as the necessary long term follow-up care for those with complex congenital heart defect place a high demand

for tertiary care centers equipped with highly trained specialists and staff with complex knowledge of CHD. An awareness of the importance on early diagnosis and treatment by the primary physician, along with the education of patients and family affected with congenital heart disease on the importance of the continued follow-up care in the center that specializes in adult congenital heart disease, cannot be over-emphasized.

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