

## Adult Congenital Heart Diseases: Clinical Pattern, Management and Immediate Outcome in Ibn-Al-Bitar Center for Cardiac Surgery, Baghdad 2013

Dr. Jasim Nasir Alkhalidi <sup>1</sup> & Dr. Hassan Al-Farhan <sup>2</sup>

<sup>1</sup> M.B.Ch.B. C.A.B.P (Pediatric)

<sup>2</sup> Assistant Prof. Consultant Cardiologist FICMS (Cardio), DIC, FACC, DMe

### Abstract:

**Introduction:** Adult with congenital heart diseases (ACHD) are an important emerging group. The diagnosis and successful management of congenital heart disease represents one of the greatest triumphs of cardiovascular medicine and surgery in the 20th century. As a consequence, the number of adults with congenital heart disease both with repaired and unrepaired lesions has grown rapidly.

**Patients and Methods:** This is a descriptive cross sectional study that conducted in Ibn Al-Bitar Center for Cardiac Surgery during the period from January 2011 to December 2012 Baghdad-Iraq. In this study all adults' patients with CHD referred to Ibn Al-Bitar Center for Cardiac Surgery were recorded and evaluated to verify the pattern of ACHD in our center.

**Results:** A total of 789 patients, female to male ratio was 1.7:1, Acyanotic CHD constitute 92.9%, and whereas the cyanotic lesions form only 7.1%. The mean age of patients was  $29 \pm 7.300$  SD years. The most common defect was ASD followed in order of frequency by VSD, PS, TOF, PDA, AS and COA. Trans catheter intervention was the treatments of choice for many types of ACHD, but still surgery play an important role in managements of other defects.

**Conclusions:** Acyanotic ACHD are more common than the cyanotic one, more female predominance, Percutaneous intervention was became widely used as the first line of management of majority of ACHD with high successful rates, while surgical correction still the treatment of choice for certain types of ACHD.

**Key word:** Adult, congenital, heart disease, Iraq.

### Introduction:

Congenital heart disease (CHD), also called congenital heart defect, includes a variety of malformations of the heart and/or its major blood vessels that are present at birth. CHD is the most common birth defect the incidence of CHD in different studies varies from about 4/1,000 to 50/1,000 live births (1). About half of these cases require

medical treatment; the rest either resolve spontaneously shortly after birth or are as mild as to need no treatment or to go undetected. More than 1.4 million people with congenital heart defects were living in the United States in 2007(2). The number of adults with CHD either repaired or unrepaired lesions has grown rapidly. Congenital abnormalities in cardiac

structure that present in subjects older than 16years, either never having been diagnosed and treated or after palliative or corrective surgery in childhood. Successful treatments for even complex congenital heart defects in childhood have resulted in increasing numbers of young adults with congenital heart disease requiring continuing surveillance and management(3). Approximately 60% of all congenital heart disease is diagnosed in babies less than one year old, 30% in children and 10% in adults (those over 18 years of age). However, now there are more adults than children with congenital lesions and this has important implications for those practicing in any branch of adult medicine. The majority of adults with congenital heart lesions will make their way to the adult practitioner via the pediatric cardiologist (4). The population of adult CHD patients is growing at a rate of 5% per year. Survival to adulthood is better because of improved fetal diagnoses, advances in neonatal intensive care, improved surgical techniques, early complete surgical repair, lower perioperative mortality, and increased midterm and late survival. Over 85% of infants with CHD are now expected to reach adulthood: approximately 300,000 of these patients have complex adult CHD, 350,000 have moderately severe defects, and 350,000 have simple congenital cardiac defects (5). We have no sufficient data about adult patients with CHD. Up to our knowledge there is no national study deal with such subject. For that we conduct the study in order to define the frequency of various types of ACHD and to determine the management modalities and immediate outcome.

## Objectives:

1. To define the frequency of various types of ACHD.
2. To determine the management modalities and immediate outcome.

## Patients and Methods:

This is descriptive, cross-sectional study that conducted in Ibn Al-Bitar Center for Cardiac Surgery in Baghdad-Iraq during the period from January 2011 through December 2012. All cases of adult (patients more than 18 year of age) with congenital heart diseases referred to the Ibn Al-Bitar Center for Cardiac Surgery in Baghdad were included in this study.

**Case Definition:** are all patients with the diagnosis of CHD over 18 years of age (18, 19).

**Inclusion Criteria:** All cases of ACHD with diagnosis of one of the CHD

**Exclusion Criteria:** That all patients with insufficient data in their files.

A pre-designed form including name, age, sex, address, original diagnosis, types of management, whether it's medical, surgical, or Transcatheter interventions, and finally the immediate outcome is filled as it's reported. SPSS ver.19 software used for data entry and analysis. Different frequencies, percentage were calculated.

## Results:

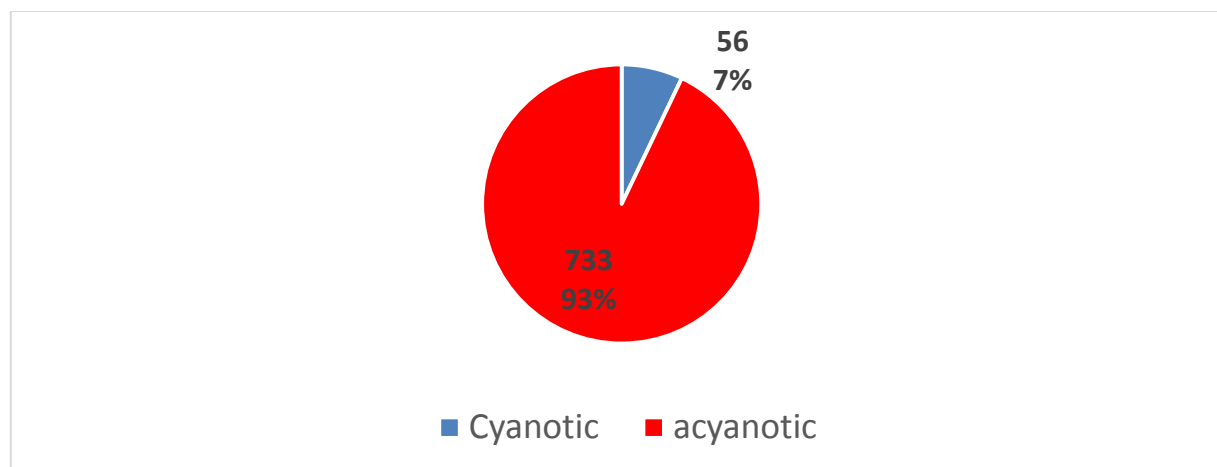
A total of 789 cases of ACHD referred to Ibn Al-Bitar Center for Cardiac Surgery, however the total number of all cases of CHD during the same period was 4442 patients. ACHD constitute about (17.76%).The mean age of patients was  $29 \pm 7.3$  SD years, the mean age of male patients was  $20 \pm 3.79$ SD years, while the

mean age of female patients was  $19 \pm 4.2$  SD years. The ages of the patients in this study range from 18-57 years, are divided into 4 major groups, as shown in table (1). Female was 499(63.2%) patients while male 290 (36.8%) patients, with female: male ratio is (1.7:1). It was divided according to the presence or absence of cyanosis into a cyanotic lesions 733 patients (92.9%) and cyanotic 56 (7.1%)

patients figure (1). ASD account for majority of cases of adult CHD 384 patients (48.7%) and followed in order of frequency by VSD 135 (17.1%) patients, PS 93 (11.8%patients), TOF 51 patients (6.5%), PDA 47 patients (6%), Aortic stenosis 42 patients (5.3%), and Coaractation of aorta 32 patients (4.1%) while complex CHD are constitute a very little number of cases, all these data are shown in table (2).

**Table (1): Gender and Age Group Distribution**

Category	Frequency	Percentage
<b>Gender</b>		
Male	290	36.8%
Female	499	63.2%
<b>Age groups</b>		
18-27 years	417	52.9%
28-37 years	272	34.4%
38-47years	93	11.8%
48-57 years	7	0.9%



**Figure (1): ACHD Classification according to Cyanosis**

**Table (2): Pattern of ACHD**

Diseases	Frequency	Percentage
ASD	384	48.7
VSD	135	17.1
PS	93	11.8
TOF	51	6.5
PDA	47	6
AS	42	5.3
COA	32	4.1
L-TGA	2	0.3
D-TGA	1	0.1
DORV	1	0.1
T A	1	0.1
<b>TOTAL</b>	<b>789</b>	<b>100</b>

Table (3): Analysis OF ASD Managements Data

Type of Treatment	Total Number	Successful Rate	Failure Rate
Transcatheter Closure	254 (66.1%)	246 (96.9%)	8 (3.1%)
Surgical Closure	71 (18.5%)	69 (97.2%)	2 (2.8%)
Medical	59 (15.4%)		

Table (4): Surgical Treatment OF VSD, TOF and AS

Disease	Number	Surgery	Successful	Failure
VSD	135	48 (35.6%)	46 (95.9%)	2 (4.1%)
TOF	51	49 (96.1%)	46 (93.9%)	3 (6.1%)
AS	42	35 (83.3%)	33 (94.2%)	2 (5.8%)

Table (5): Distribution of patients with PS according to the types of treatments.

Procedure	Number	Successful	Failure
Balloon Pulmonary Valvoplasty	86 (92.5%)	78 (90.7%)	8 (9.3%)
Medical treatment	7 (7.5%)		

**Table (6): Analysis of PDA Management's Data**

Procedures	Total	Successful
<b>Transcatheter Closure</b>	42 (89.3%)	42 (100%)
<b>Surgery</b>	2 (4.2%)	2 (4.2%)
<b>Medical Treatment</b>	3 (6.3%)	

**Table (7): Modes of Management of Coaractation of aorta**

Procedure	Native	Recurrent		Total
		Post-Balloon	Post Surgery	
<b>Balloon</b>	6(18.8%)			<b>6</b>
<b>Balloon + Stent</b>	14(68.8%)	5(62.5%)	3(37.5%)	<b>22</b>
<b>Surgery</b>	<b>4(12.5%)</b>			<b>4</b>

**Managements:****Atrial septal Defect (ASD):**

From 384 patients with ASD a Transcatheter closure was performed in 254 (66.1%) patients, which was successful in 246 patients (96.9%) (no significant shunt through ASD) while failed and patients referred to surgery in only 8 patients

(3.1%), Initial Surgical closure of ASD was performed in 71 (18.55%) patients, it was successful in 69(97.2%) patients and 2(2.8%) patients are died during the immediate post-operative period. The remaining 59 (15.4%) patients with ASD

are kept on medical therapy without intervention, this is shown in table (3).

#### **Ventricular septal defect (VSD):**

The frequency of adult patients with VSD was 135(17.1%) patients, from those 48 (35.6%) patients underwent surgical closure, and it was successful in 46 (95.9%)patients, while two (4.1%) patients are died post operatively, the remaining 87 (64.4%) patients are remain on medical therapy only, table (4).

#### **Pulmonary stenosis (PS):**

PS are found in 93(11.8%) patients in our study, a trial of balloon pulmonary valvoplasty was done for 86 patients (92.5%) which was successful in 78(90.7%), patients (pressure gradient across the pulmonary valve decline below 36 mmHg) and failure of this procedure in 8 (9.3%) patients, which may be due to dysplastic nature of pulmonary valve, the remaining 7(7.5%) patients are kept on medical therapy only. This is shown in table (5).

#### **Tetralogy of Fallot (TOF):**

The total number of cases of ACHD have TOF are 51 (6.5%) patients, 49(96.1%) patients are underwent total TOF correction with good results (93.9%) except three (6.1%) patients died, the other two (3.9%) patients remain on medical therapy only table (4).

#### **Patent ductus arteriosus (PDA):**

Account for 47 (6%) patients of adult with CHD, attempt of transcatheter closure was done for 42 patients and it has an excellent

result in all patients (no residual shunt through PDA by angiography) two (4.2%) patients are treated by surgical ligation without complication, and the last three (6.3%) patients are still on medical therapy, this is illustrated in table (6).

#### **Aortic stenosis (AS):**

Account for 42(5.3%) patients in our study, surgical aortic valve replacement done for 35 patients (83.3%) and it was successful in 33 patients (94.2%), while two (5.8%) patients are died in the post-operative period, table (4).

#### **Coaractation of aorta (COA):**

In our study COA was found in 32 (4.1%)patients, 22 patients (68.8%) are treated by balloon angioplasty with stent implementation, 14 (63.6%) patients of them for native COA, and eight (36.4%) patients for recurrent COA. three (37.5%) patients of them post-operative recurrence and five(62.5%) patients recurrence follow balloon dilatation few years ago. Balloon angioplasty alone were attempt in six (18.8%) patients and finally surgical treatment of COA was done in four (12.5%) patients with good results, these data shown in table (7).

### **Discussion:**

Adult with congenital heart diseases are an important emerging group. The modern era of surgery for CHD began 60 years ago, since then medical and surgical strategies have been devised and refined for even the most complex cardiac malformation. The spectrum of ACHD in Ibn- AlBitar Center

for Cardiac Surgery emerging from this study showed that the majority of patients have atrial septal defect (ASD) followed in order of frequency by ventricular septal defect (VSD), pulmonary stenosis (PS), tetralogy of Fallot (TOF), patent ductus arteriosus (PDA), aortic valve stenosis (AS), and Coarctation of aorta (COA), the complex ACHD in our study form minority of patients, this is similar to other study done by L Shamima Sharmin, et al(27) were they found that ASD (42.6%), VSD (18.3%), TOF (14.8%), PDA (7.8%). Also other study done by Carole A. Warnes, et al (9), and also similar to studied done by Abbag, F. et al (28) and, Jia B, et al(29) and also near the study done by Rahman F, et al.(30) A.C. Zomer, et al(31) in their study they found that the mean age of adults patient with CHD is 33.1 years while in our study we found that the mean age is 29.03 years. M A Gatzoulis, et al(32) found that there was steady fall in the mean age of patients seen in clinic with time 38.5, 33.6, 31.7 year in 1987, 1992, 1997 respectively, and this is also higher than that found in our study. The mean age of female patients was lower than that of male patients this finding could be explained by the fact that female tend to consult earlier than male due to psychosocial aspect regarding marriage and pregnancy. Rahman F, et al(29) found that female patients are more than male, this figure is near the figure emerging from our study, and also from studies done by P. Engelfriet1, et al.(33) The Acyanotic lesions form the majority of patients while the cyanotic lesion only in 7.1%, this result is also compatible to studies done by Kamata K, et al.(34).

### Management:

Majority of patients with ASD underwent transcatheter closure which was successful in (96.9%) and failed in (3.1%) & this is similar to other studies done by Gary Webb, et al.(35), while Behjati M, et al(36) found a lower incidence of successful transcatheter closure (89.6%) and a high failure rate (10.4%). Surgical closure of ASD was successful in (97.2%) with post-operative mortality rate (2.8%), which is lower than that found in other studies done by Suchon Elzbieta, et al(37) and Chatzis A, et al(38) who showed that the mortality rate is (0%), (1%) respectively. Surgical closure of VSD was successful in (95.9%) in our study, with (4.1%) mortality rate, which is higher (mortality rate) than that found in other studies done by Naser M Ammash, et al (39) Karamarie Fecho, et al (40) which reported that the operative and post-operative mortality rates was 0.8, 2, 1% respectively. Balloon pulmonary valvoplasty is the first line of management of adult patients with valvular pulmonary stenosis in our study and it was successful in (90.7%) and failure to achieve satisfactory result in (9.3%), which may be due to either dysplastic nature of the pulmonary valve or inefficient balloon dilatation during the procedure. This is similar to other studies done by Alan D. et al(41) Teupe C, et al(42) Lip G. et al.(43) Total correction of TOF was successful in (93.9%) with (6.1%) mortality rate, this result to some extent higher than that found by Shahid Madani et al,(44) were found that the mortality rate is (6.9%), while P. Engelfriet1 et al(39) and B.J.M. Mulder, et al(45) reported a lower mortality rates (3.1%), while Abdul Razzaq Naser Hussein found in his study the overall early mortality was (24.5%).(46) Transcatheter closure is the management of choice for



patient with PDA in our study and the successful rate was encouraging (100%), which is also compatible with result done by Eduardo E. et al(47) Ali Akbar Zeinaloo, et al(48), who found that the successful rate was (99%) and (98.6%) respectively. Surgical aortic valve replacement was successful in(94.2%) and (5.8%) post-operative mortality rate, this figure is higher than that obtained by Brad C. Astor, et al(49) and Lieberman EB, et al(50) Guerin F, et al(51) Mc Anulty JH et al(52) who found that the post-operative mortality rates are 3.5%, 2%, 1% respectively. Balloon angioplasty of Coaractation of aorta with stent implementation is the first line of management in our center which was successful in all patients (100%), these results are also compatible to other studies done by F Attie, et al(53) A Buendía-Hernández, et al(54) and also by Kphadke, et al.(55) Surgical repair of COA was performed in (12.5%) without complication, while Mohammed Mahmod Saleem found that the early mortality rate following surgical repair of COA was (5.3%).(56).

## Conclusions:

1. Simple CHD (Acyanotic) are most commonly seen in the outpatient department of adult and pediatric cardiology, while a very low percentage of complex (cyanotic) CHD.
2. CHD in adult female are more common than male.
3. Atrial septal defect are the most common type of CHD seen in the adult population.
4. Percutaneous intervention whether transcatheter closure of certain defects or

balloon dilatation of other defects was become the first line of management of many CHD in adults with high successful rates.

5. Still surgical correction is the treatment of choice for many types of CHD in adult.

## Recommendations:

ACHD need more focus attention, conduct more prospective study, registry and long term follow up, preferably to be done in specialized center for ACHD

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## تشوهات القلب الولادية عند البالغين, النمط السريري, طرق المعالجة والنتائج الاولية في مركز ابن البيطار لجراحة القلب في بغداد لسنة 2013

حسن علي الفرخان

جاسم ناصر عودة الخالدي

### الخلاصة :

**المقدمة :** ان الزيادة الحاصلة في اعداد المرضى البالغين (اكثر من 18 سنة) المصابين بتشوهات القلب الولادية جاءت نتيجة التطور الكبير الذي حصل في مجال علاج تشوهات القلب الولادية في هذا القرن والقرن الذي سبقه من الزمن عن طريق التداخل الجراحي او التداخل القسطاري او عن طريق العلاج الدوائي.

### الاهداف :

- لمعرفة اعداد المرضى البالغين المصابين بتشوهات القلب الولادية.
- لبيان نوع الطرق العلاجية للمرضى البالغين المصابين بتشوهات القلب الولادية والنتائج الأولية.

### طرائق البحث :

ان هذه الدراسة هي عبارة عن دراسة وصفية امتدة لمدة سنتان من كانون الثاني 2011 لغاية كانون الاول 2012 جرت في مركز ابن البيطار التخصصي لجراحة القلب في بغداد, العراق وتم خلالها جمع جميع الملفات الخاصة بالمرضى وتم تحليل النتائج بواسطة نظام (spss 19). كان عدد المرضى البالغين المصابين بتشوهات القلب الولادية الوافدين الى المركز 789 مريضاً بينهم 499 من النساء و 290 من الرجال اي بنسبة 1,7:1.

### الاستنتاجات :

ان اعداد المرضى البالغين المصابين بتشوهات القلب الولادية اصبحت في تزايد مستمر, نسبة التشوهات غير الازرقاقية اكثر من التشوهات الازرقاقية, الاناث شكلت نسبة اعلى من الذكور. علاج تشوهات القلب الولادية عند البالغين عن طريق القسطرة اصبحت من الطرق المهمة جداً في هذا المجال وبنسبة نجاح عالية, بينما لا يزال التداخل الجراحي يعد الخط الاول في العلاج لانواع اخرى من التشوهات.

### التوصيات :

لا بد من وجود دراسات اخرى جديدة طويلة الامد لهذه الشريحة من المرضى .  
استحداث عيادات استشارية متخصصة للعناية بهم والتعامل مع كل المضاعفات التي قد يتعرض لها المرضى البالغين المصابين بتشوهات القلب الولادية.