

# Pregnancy in Adult Congenital Heart Disease Maternal and Fetal Outcome, a Single Centre Study in a Developing Country

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## Abstract

**Background:** Women with congenital heart disease represent the largest group of women with cardiac disease in the child bearing period. This poses additional burden on the medical system as these mothers and their infants require special care.

**Methods:** This was a cross sectional observational study involving 90 female patients in the child bearing period who came for regular follow up visit in the adult congenital heart disease clinic in our hospital over a period of 12 months. A custom-made sheet was done including maternal diagnoses, age of the patient, age during the first pregnancy, and maternal morbidity during pregnancy and fetal outcome.

**Results:** The mean age of the patients was  $35.4 \pm 12.2$  years. 97.7% of these patients report at least one successful pregnancy; the mean age at time of first pregnancy was  $22.77 \pm 2.97$  years. The average number of offsprings was 1.6. Forty-one (41) of our patients sought medical advice because of being pregnant, 24 of which were pregnant for the first time. The patients most common underlying heart disease was ASD (n=39), VSD (n=11) and AVCD (n=10) but still more complex diagnosis was present; TOF (n=4) L-TGA (n=3), Truncus (n=1) Ebstein anomaly (n=2). 62.2% of the patients did not report any maternal morbidity during pregnancy. The most common morbidities were complete heart block (n=5), abortion (n=5), progressive heart failure (n=3), stroke (n=3), progressive pulmonary hypertension (n=4). Among the 143 offsprings delivered only 12 were abnormal, 8 of which had ASD and none of them had complex CHD.

**Conclusion:** Although pregnancy in adult patients with congenital heart disease is feasible especially in patients with simple lesions, special attention is needed to guard against maternal morbidity.

**Keywords:** Pregnancy; Adult congenital heart disease

**Abbreviations:** ASD: Atrial Septal Defect; AF: Atrial Fibrillation; AV: Aortic Valve; AVSD: Atrioventricular Septal Defect; CHD: Congenital Heart Disease; DCRV Double Chambered RV; DILV: Double Inlet Left Ventricle; ACHD: Adult Congenital Heart Disease; ICD-10 International Classification of Diseases 10<sup>th</sup> Revision; IUGR: Intrauterine Growth Retardation; LBW: Low Birth Weight; L-TGA: L-Transposition of the Great Arteries; LVOTO: Left Ventricular Outflow Tract Obstruction; NCHS: National Center for Health Statistics; PAPVD: Partial Anomalous Pulmonary Venous Drainage; PDA: Patent Ductus Arteriosus; PS: Pulmonary Stenosis; PVCS: Premature Ventricular Contractions; SD: Standard Deviation; SPSS: Statistical Package Version Sixteen; SVT: Supraventricular Tachycardia; TAPVD: Total Anomalous Pulmonary Venous Drainage; TOF: Tetralogy of Fallot; U.S: United States of America; VSD: Ventricular Septal Defect; WHO: World Health Organization

## Introduction

There is an increased prevalence of ACHD (adult congenital heart disease) in the last decades, as advances in surgery and medical management have greatly improved, those born with cardiac anomalies are now living far into adulthood. Currently, adults make up the largest population of people living with CHD (congenital heart disease [1]) The incidence of congenital heart disease in pregnancy has increased over the last 30 years which is attributed to the improving results of surgical and catheter interventions, which lead to survival of this female into the reproductive age group [2].

Since pregnancy is associated with additional risks in the ACHD patients and due to the diversity of the lesions in patients with CHDS It is not possible to give a list of lesions with precise risks attached,

because few women have the same lesion, and the risk is also strongly modulated by complications that may or may not develop during pregnancy [3].

Pregnancy is an additional stress on an already compromised cardiovascular system. There is a 50% expansion in blood volume, mostly due to retention of fluid and relative enlargement of the plasma volume, which is necessary to provide appropriate blood flow to the uterus and fetoplacental unit. This increase in volume is associated with increase in the cardiac output by 30%, decrease in the peripheral resistance and increase in the uterine blood flow, increase in blood volume by 40%-50%, and in heart rate by 10%-20% [4]. In addition to other threats as thromboembolic complications, arrhythmias and infections [5]. Since women now a day give birth to their first child at an older age as compared to the previous decades, there is an increase in the mean maternal age at the time of the first delivery which is associated with increased maternal and fetal risk [6].

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The number of the off springs can add to the evaluation of pregnant women in that women with multiple pregnancies are at a higher risk of developing cardiac and extra-cardiac comorbidities. This can correlate with the severity of the lesion of CHD [7].

As Pregnancy carries both maternal and fetal risks. Maternal risks can increase in these mothers due to a compromised cardiovascular system, abnormal coagulation profile, increased thrombogenicity, infection (infective endocarditis) arrhythmias and hemorrhage. They also have the additional risk of therapeutic abortions and miss carriages [4]. The fetal risk comprises IUGR (intrauterine growth retardation), Low birth weight and the risk of genetic transmission of the CHD from the mother to the fetus. the incidence of recurrence depends on the type of the defect [8].

In the current study we evaluated the relation between the maternal age at the first delivery, number of offspring's, pregnancy outcomes, maternal morbidity and mortality and risk of CHD recurrence in offspring's born to mothers with congenital heart disease [9].

## Methods

The current study included 90 female patients with adult congenital heart disease who presented to Ain shams university hospitals, which is a large tertiary center, with a structural and congenital heart disease unit, a cath. Lab and a cardiothoracic surgery hospital.

The patients included in the study were either following up or referred to our hospital for medical treatment, catheter or surgical intervention.

The study population duration was 1 year starting from 1<sup>st</sup> of May 2015 till the end of May 2016.

We included all female patients with ACHD defined as those patients who diagnosed, treated, and followed up during their pediatric years. We also included *de novo* adults, previously undiagnosed in the childhood.

We excluded female patients with poor mental functions, those with acquired heart diseases or non ACHD diagnosis.

This was a cross sectional observational study, it included 90 female patients in the child bearing period who came for regular follow up visits in the adult congenital heart disease unit.

Data was taken from the patients regarding their current or previous pregnancies Maternal and fetal outcome during or pre-dating our registry all patients enrolled in the study signed an informed consent.

After the informed consent all patients were subjected to:

Detailed history taking: personal history, history of the present illness, past history, with especial emphasis on the gestational history, number of previous pregnancies, current pregnancy status, miscarriages, abortions, cardiac and obstetric complications. Gestational age at delivery, mode of delivery.

1. Cardiac diagnosis, symptoms, medications and previous echocardiographic data.
2. Physical examination: general and local examination.
3. 12 lead surface ECG
4. Transthoracic Echocardiography, Transesophageal echocardiography or 3D echocardiography when indicated.
5. Fetal echocardiography was done for those patients with current pregnancy during the study.

6. Routine lab investigations including: full blood picture.

7. Other imaging modalities when indicated.

8. According to WHO organization. The congenital heart diseases were translated from the International Society of Cardiology (1970) classification to the International Classification of Diseases 10<sup>th</sup> revision (ICD-10) lastly updated by the National Center for Health Statistics (NCHS) under the authorization of World Health Organization in 2017.

9. Maternal complications were defined as cardiac complications (heart failure (systemic ventricular failure), brady or tachy arrhythmias, thromboembolic complications, syncope, infective endocarditis or death) and obstetric complications (as post-partum hemorrhage, low birth weight (LBW), still birth or birth defects)

## Statistical Analysis

Data were collected, verified, revised then edited on the data base computer system. Categorical variables were expressed as absolute and relative frequencies (percentage) while continuous variables were presented as mean values  $\pm$  standard deviation (SD). Comparisons were made between the two groups using t-test for continuous variables and chi-square test and Pearson correlation coefficient for categorical variables.

Statistical analysis was performed using SPSS (statistical package version sixteen). Difference was considered statistically significant at a P value<0.05 and highly significant at P value<0.01.

## Results

The mean age of the patients was  $35.4 \pm 12.2$  years. 97.7% of these patients report at least one successful pregnancy; the mean age at time of first pregnancy was  $22.77 \pm 2.97$  years. The average number of offsprings was 1.6. 41 of our patients sought medical advice because of being pregnant, 24 of which were pregnant for the first time and 14 patients of the whole study group underwent 1 pregnancy, 21 patients underwent two pregnancies while 23 patients underwent 3 pregnancies (Table 1). Out of 62 patients who underwent previous pregnancies 35 underwent vaginal delivery and 27 patients underwent caesarian section.

The patients' most common underlying heart disease was ASD (n=39), VSD (n=11) and AVCD (n=10) but still more complex diagnosis was present; TOF (n=4) L-TGA (n=3), Truncus (n=1) Ebstein anomaly (n=2). 62.2% of the patients did not report any maternal morbidity during pregnancy (Table 2) (Figure 1). The most common morbidities were complete heart block (n=5), abortion (n=5), progressive heart failure (n=5), stroke (n=3), progressive pulmonary hypertension (n=4) (Table 3), (Figure 2). Among the 143-offsprings delivered only 12 were abnormal, 8 of which had ASD and none of them had complex CHD (Figure 3).

## Discussion

In our single tertiary center registry. A one-year retrospective review of maternal and fetal outcome in pregnant women with CHDs. All patients were in the child bearing period with mean age of patients during the study  $35.4 \pm 12$  years. and the mean age at the time of the first pregnancy 22 years, also when we divided the patients into two

Number of patients	Number of pregnancies
24	Pregnant during the study
14	Pregnant once before
21	Pregnant twice before
23	Pregnant thrice before

Table 1: Showing number of pregnancies for each patient.

groups with 35 years the cutoff value, we found that we had 36 patients above the age of 35 years with non-statistically significant difference in the occurrence of cardiac complications between the two age groups however those above 35 years had all simple lesions or were following post-operative only 2 patient had severe pulmonary hypertension, 1 patients with L-TGA heart failure and 1 patient was post Glenn.

Diagnosis	N	Percentage
ASD	38	42.20%
PAPVD	3	3.30%
VSD	8	8.90%
AVCD	10	11.10%
PDA	4	4.40%
Interrupted ARCH	1	1.10%
LVOTO	6	6.70%
PS	4	4.40%
TOF	4	4.40%
L TGA	3	3.30%
DILV	1	1.10%
TRUNCUS	1	1.10%
EBSTEIN	2	2.20%
VSD DCRV	3	3.30%
ASD PS	1	1.10%
PAPVD PS	1	1.10%
Total	90	100.00%

Table 2: Showing patient diagnosis.

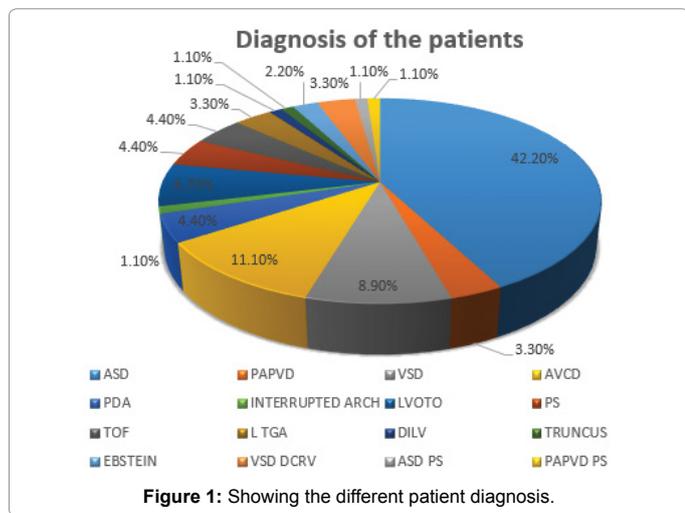


Figure 1: Showing the different patient diagnosis.

Cause	N	Percentage
Normal	56	62.20%
Complete heart block	5	5.60%
Heart Failure	5	5.60%
Cerebro-vascular stroke	3	3.30%
Infective endocarditis	1	1.10%
Valve regurgitation	6	6.70%
Pulmonary hypertension	4	4.40%
AF	1	1.10%
ABORTION	5	5.60%
Pulmonary edema	2	2.20%
PVCS	1	1.10%
Supraventricular arrhythmia	1	1.10%
Total	90	100.00%

Table 3: Showing the maternal morbidity.

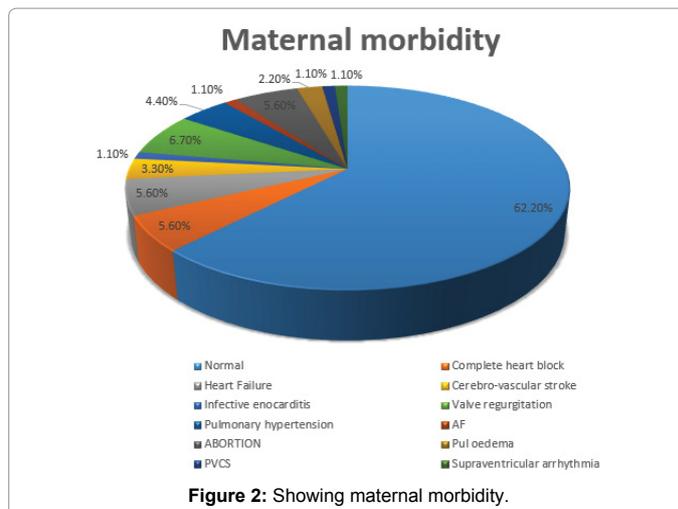


Figure 2: Showing maternal morbidity.

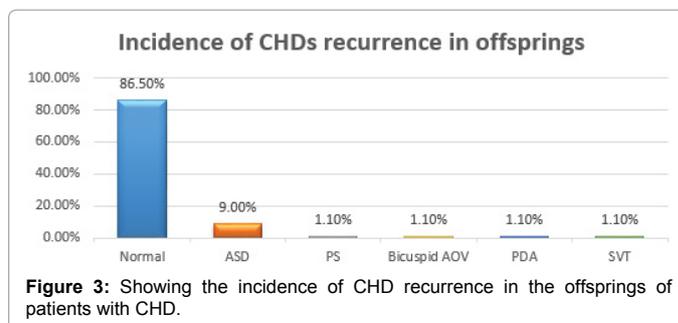


Figure 3: Showing the incidence of CHD recurrence in the offsprings of patients with CHD.

However, this is relatively lower than the mean of age in the western countries [10], where E. Furenas and his coworkers mentioned that the mean of age in his registry was 29 years and the mean age at the time of the first pregnancy was 28 years, also age above 35 years was not a significant risk factor for the cardiac complication [11].

### Pregnancy Outcome

41 patients of the total patients sought medical advice for their first pregnancy, 4 patients sought medical advice, but they haven't been pregnant yet and 45 patients had previous successful 45 pregnancies. A total of 97.7% from the whole cohort of patients report at least one successful pregnancy, where vaginal deliveries represent 56% of the cases with only one patient with maternal cardiac event developed acute pulmonary edema during the delivery.

Similar to our study Hidano et al reported that 128 pregnant women with CHDs had 151 successful pregnancies, 56% had successful vaginal delivery with only one patient with unrepaired tetralogy of Fallot developed congestive heart failure and hemoptysis from major aortopulmonary collateral during the post-partum period [12].

The most commonly coded congenital heart lesion was ASD (42.2%), and VSD (8.9%), AVSD (11.1%), PDA (4.4%), LVOTO (6.7%), PS (4.4%), TOF (4.4%), L-TGA (3.3%), DILV, truncus arteriosus (post truncal repair) 1 patient each. Comparing our findings to a national U.S study published in 2015 reporting that more women with CHD are now getting pregnant leading to increased number of hospitalizations before deliveries, the most common diagnosis were as follows ASD (22.6%), VSD, (14.5%), LVOTO (13.8%), right sided congenital valvular lesions (7.5%). less common coarctation of aorta (2.9%), TOF (2.6%), TGA (1.5%) Ebstein anomaly (1.4%), TAPVR (0.1%) and truncus arteriosus (0.1%) [13].

## Cardiac Outcome

Our study demonstrated that there was no maternal mortality, however the maternal morbidity was considerable representing 52.2% of the whole study group including heart failure (5 patients), arrhythmias as (CHB, AF, SVT, PVCs) 8 patients, infective endocarditis however those with pulmonary hypertension or Eisenmenger syndrome (4 patients) had the worst outcome.

In contrast to our results Furends and his colleagues reported less incidence of cardiac complications in pregnant women with CHDs at a rate of 14%, however arrhythmia was the commonest complication (6%) followed by heart failure (5 patients) [11].

Similar to our results Thompsons et al. reported that among all 655 women with CHD the incidence of cardiovascular complications was 4.1% mostly due to arrhythmia, heart failure, stroke and myocardial infarctions, especially those with CHD and pulmonary hypertension [13].

The Registry of Pregnancy and Cardiac disease investigators recently published on outcomes of 872 women with congenital heart disease, 8% of which had cardiovascular complications, mostly heart failure [14], while Karamlou et al. found a rate of composite cardiac complications (cardiac arrhythmia, cardiac arrest, or cardiac events related to anesthesia) of 2.3% in women with CHD compared with 0.2% in those without CHD [15].

## Obstetric Outcome

Only 2 patients (5.5%) in our study had therapeutic abortions. In contrast to other registries with higher rate of obstetric complications, in ZAHARA registry 20% of pregnancies ended with miscarriages [16], while Furenas reported 12.6% as a total fetal loss [11].

## Fetal Outcome

In the general population, about 1% of all children are born with CHD. However, the risk increases when either parents has CHD, where a polygenic multifactorial inheritance is involved in causing birth defects so in our registry we studied the risk of recurrence of CHD in offsprings of mothers with CHD. We found that 12 (13.4%) out of 90 patients had offsprings with CHDs, the most common inherited defect was ASD 9.0%, where PS, bicuspid AV, PDA 1.1%, a single case of a neonate 1.1% had SVT born to a mother with Ebstein anomaly, exact concordance of CHD was found in 8% and a partial concordance in 2% of cases.

In concordance to our findings, Velde et al. reported that 15% of the patients with CHD reported an affected family member with CHD, 6% of which were from a first degree relative [17]. In another multi-center clinical study carried out in Italy, Vlasta et al. reported that Total recurrence rate of CHD was 5.2% from the mother to her own fetus. exact concordance of CHD was found in 21.5% and a partial concordance in 20% of cases [18].

## Study Limitations

This study represents one of the initial attempts to establish an adult congenital heart disease registry, the relative small number of patients enrolled in the registry, a longer period of time should overcome this limitation.

The absence of a universally accepted system to follow up ACHD patients, this makes it more difficult to compare the current registry to others.

## Conclusion

Although pregnancy in adult patients with congenital heart disease is feasible especially in patients with simple lesions, special attention is needed to guard against maternal morbidity like complete heart block, progressive heart failure, stroke or pulmonary hypertension.

There is a considerable risk of CHD recurrence in the offspring's of the affected mother, father or a first degree relative which dictates the need for further genetic testing and future microbiological studies which makes gene therapy a possible option for prevention of CHD.

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