

Spectrum of Grown up Congenital Heart (GUCH) at Armed Forces Institute of Cardiology/ National Institute of Heart Disease: 10 years Experience

Hajira Akbar, Mohammad Amir Nawaid*, Tabassum Muzaffar**, Sana Imtiaz, Asma Ansari***

Armed Forces Institute of Cardiology/National Institute of Heart Diseases (AFIC/NIHD)/National University of Medical Sciences (NUMS) Rawalpindi, Pakistan, *Combined Military Hospital Bahawalpur/National University of Medical Sciences (NUMS) Pakistan, **Combined Military Hospital RWP/National University of Medical Sciences (NUMS) Rawalpindi, Pakistan, ***Combined Military Hospital Sialkot/National University of Medical Sciences (NUMS), Pakistan

ABSTRACT

Objective: To determine the spectrum of Grown up Congenital Heart (GUCH) at Armed Forces Institute of Cardiology/National Institute of Heart Diseases.

Study Design: Descriptive cross sectional study.

Place and Duration of Study: Paediatric Cardiac Surgery and Obstetric department of Armed Force Institute of Cardiology/National Institute of Heart Disease, Rawalpindi Pakistan, from 2011 to 2021.

Methodology: It was a descriptive cross sectional study conducted at Paediatric Cardiology, Paediatric Cardiac Surgery and Obstetric department of AFIC/NIHD. After taking informed consent, a total of 1344 patients fulfilling inclusion and exclusion criteria with non probability consecutive sampling were enrolled in study from 2011 to 2021. Diagnosis, transthoracic echocardiography findings, procedural details (cardiac catheterization/cardiac surgery/obstetrical intervention) and outcome of all patients were noted.

Results: Out of 1344 patients, 700 patients had cardiac catheterization. Among patients who underwent cardiac cath, 304(43%) were males and 396(56.6%) were females. 279(39%) diagnostic and 421(61%) cardiac interventional procedures were done. Most common procedure done was ASD device closure in 227(32%) patients. Complications were seen in 29(4%) patients and mortality in 3(0.4%) patients. There were a total of 188 patients who underwent congenital cardiac surgery. Out of patients who underwent congenital cardiac surgery procedures, 96(51%) were male and 92(49%) were female patients. Major complications were observed in 51(25%) patients and the mortality was 11(5%). There were total of 456 patients in obstetrics department. Out of 456 patients, 54(11%) were with congenital cardiac lesions. The most common defect was VSD 27(6%) patients and dilated cardiomyopathy (DCM) 27(6%) patients followed by ASD and TOF. The maternal and fetal mortality was 12(2.7%) and 22(5.5%) respectively.

Conclusion: With immense advancement in diagnosis and management of congenital cardiac diseases, a significant number of patients remain undiagnosed and untreated till the time they present as GUCH. Most common presentation is ASD which is mostly manageable by transcatheter intervention, however late presentation carries a risk of complications like pulmonary hypertension.

Keywords: Cardiac catheterization, Congenital cardiac surgery, GUCH.

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INTRODUCTION

Pediatric patients with congenital heart defects (CHD) are a large number of children which require extended medical care. With advances in cardiovascular management, operative and interventional procedures, greater number of children are surviving into adulthood. The definition of GUCH is the persistence of any structural abnormality present at birth that involves the heart and/or great vessels in adult life i.e., beyond 18 years of age.¹ There is a significant number of population among these children who acquire the age of 18 years. There are approximately 50 million GUCH patients world-wide and its prevalence is

expected to increase steadily until 2050 in projections.² This cohort of adolescents have specialized needs and in order to comprehend the requirements, there is an ongoing need to develop database and registry at tertiary care hospitals. Types of GUCH has been described as mild, moderate and complex.³ The current GUCH population reporting to congenital cardiac surgeon mainly comes from two groups. One is newly diagnosed and the other is who underwent surgery in childhood.⁴ These days, more than 90% of all children born with CHD reach adult life. Although initially considered to be cured, the majority of them continue to need specialised follow-up because they require re-do interventions or are at increased risk of cardiovascular complications like arrhythmias and heart failure.^{5,6} In general, GUCH patients have a high

Correspondence: Dr Hajira Akbar, Armed Forces Institute of Cardiology/National Institute of Heart Diseases (AFIC/NIHD), Rawalpindi, Pakistan

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utilisation of emergency resources, with emergency care utilisation increasing as age progresses.⁷ Another group is the one reporting for evaluation in congenital cardiac OPDs and being managed by catheter based interventions. GUCH female patients who get married and report to cardiovascular units with pregnancy are the most challenging group of patients requiring specialized care. CHD is the most common form of heart disease in pregnancy, and a leading cause of maternal morbidity and mortality.⁸ The aim of this study was to determine the types of defects in GUCH population, treatments offered, outcome analysis and morbidity faced by these individuals in tertiary care hospital.

METHODOLOGY

It is a descriptive cross sectional study conducted at Paediatric Cardiology, Paediatric Cardiac Surgery and Obstetric department of Armed Force Institute of Cardiology/National Institute of Heart Disease.

Sample Size: The calculated sample size was 139 by taking 10% prevalence of Adult Congenital heart disease,⁹ but we retrospectively collected data of 10 years from 2011 to 2021 at AFIC so our sample reached at 1344.

Inclusion Criteria: is all patients, both male & female, more than 18 years age presenting to AFIC outpatient and inpatient with congenital heart disease and GUCH patients undergoing cardiac cath procedures, cardiac surgery or cesarian section at AFIC/NIHD.

Exclusion Criteria: includes GUCH patients with mitral valve prolapse and non stenotic bicuspid aortic valve and GUCH patients coming for follow up with h/o procedures (cardiac cath intervention/cardiac surgery) in childhood.

It was a non probability consecutive sampling. After taking formal permission from institutional IERB (Ltr# 2/2/R&D/2022/14829), a total of 1344 patients fulfilling inclusion and exclusion criteria were enrolled in study from 2011 to 2021. Diagnosis, transthoracic echocardiography findings, procedural details (cardiac catheterization/cardiac surgery/obstetrical intervention) and outcome of all patients were noted.

RESULTS

Total 1344 patients were enrolled. Out of which, 700 patients underwent cardiac catheterization with mean height 160.9 ± 14.4 cm & mean weight 59.6 ± 14.6 kg. Mean age of the patients undergoing cardiac cath was 31 ± 11.4 years and average fluoroscopy time was 10.9 ± 13.6 mins. There were 309 male and 396 female

patients. 599 patients had local anesthesia(LA) and 101 patients had general anesthesia (GA). Total diagnostic catheterization were 279, out of which 175 were acyanotic heart disease and 104 were cyanotic heart diseases.

421 patients had interventional procedures done, further distribution of type of procedures is shown in Figure-1, most common being ASD device closure followed by PDA and VSD device closure.

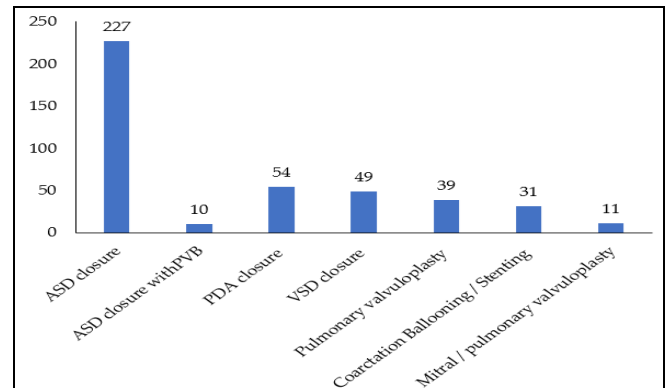


Figure-1: Distribution of Interventional Cath Procedures (n=421); ASD=Atrial Septal defect; VSD=Ventricular Spetal Defect; PDA=Patent Ductus Arteriosus; PVB=Pulmonary valve Ballooning

There were no complications in 669(95.5%) patients out of 700(100%) and complications were noted in 31(4.5%) patients. Major complications were procedure failure 10(1.5%). 9(1.3%) patients had device embolization which were retrieved and 2(0.3%) patients had dysrhythmia. Mortality was 3(0.4%) (Figure-2).

188 patients had congenital cardiac surgery of which mean age was 24.5 ± 16.9 years, mean height 150.7 ± 17.7 cm and mean weight was 56.2 ± 14.2 kg. Mean cardiopulmonary bypass (CPB) time was 126.5 ± 60.3 min and cross Clamp time 75.2 ± 40.5 min. Mean hospital stay was 11.5 ± 7.3 days and ICU Stay was 55 ± 41.7 hours. Average ventilation time was 15 ± 1.9 hours. Maximum GUCH patients who underwent surgical repair had acyanotic heart disease repair (VSD/ASD/Mid RV bands) followed by Tetralogy of fallot (TOF), Atrioventricular septal defect (AVSD) and coarctation repair. Distribution shown in Figure-3.

177 (94.1%) patients had successful repair.¹⁰ Patients didn't survive resulting in a mortality rate of 5.8%. Out of 1344, 456 patients reported in obstetrical department of AFIC /NIHD. The mean age was 28.3 ± 4.3 years with mean gestational age of 35.6 ± 3.5 weeks. Out of n=86, 54 (12.5%) patients who had congenital heart disease, the most common congenital cardiac

defect was VSD 27(6%) followed by ASD, TOF, pulmonary stenosis, pulmonary hypertension and PDA. 27(6%) patients had cardiomyopathies and 5(1.2%) patients had Eisenmenger physiology (Figure-4).

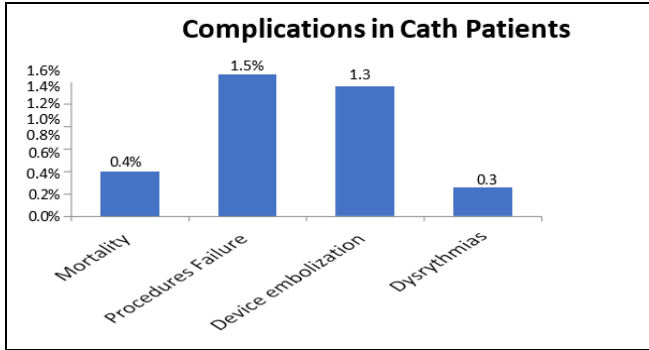


Figure-2: Complications During Cardiac Cath Procedures in GUCH (n=700)

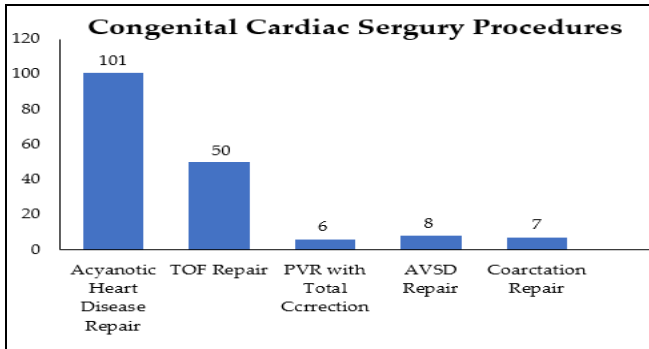


Figure-3: Distribution of Congenital Cardiac Surgery Performed in GUCH (n=188)

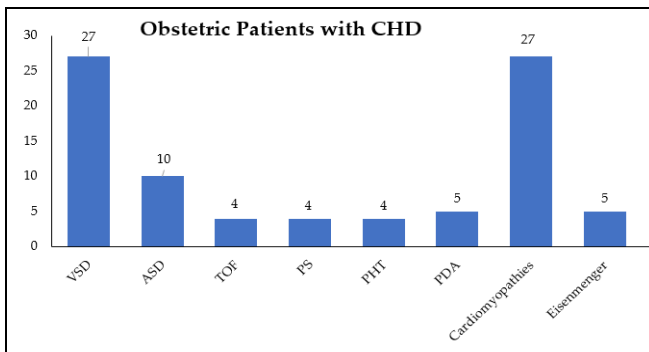


Figure-4: Frequency of obstetric patients with congenital heart disease (n=86)

VSD=Ventricular Septal Defect; ASD=Atrial Septal defect; TOF=Tetralogy of Fallot; PS=Pulmonary Stenosis; PHT= Pulmonary Hypertension; PDA=Patent Ductus Arteriosus

Mode of delivery was spontaneous vaginal delivery in 246(54%) patients, lower segment cesarian section (LSCS) in 168(37%), instrumental delivery in 8(9.4%) and assisted delivery in 4(1.5%) patients. There were no complications in 383(84%) patients. Major

morbidty was post partum hemorrhage/placental abruption, cervical tears and pre term labour. Maternal mortality was 12(2.7%) patients. Fetal outcome was uneventful in 314(89.9%) patients. Major morbidity was fetal growth retardation, congenital heart defects and multiple anomalies. Fetal mortality was 22(5.5%). Results are depicted in tabulated form in Table-I.

Table-I: Summary of Results in Obstetrical Patients

Obstetric patients year 2011-2021	n=456
Mean Age	28.3±4.3years
Mean GA	35.6±3.5weeks
Obstetric patients with CHD	n=54 (12.5%)
Mode of Delivery	
SVD	n=246 (54%)
LSCS	n=168 (37%)
Instrumental delivery	n=8 (9.4%)
Maternal mortality	n=12 (2.7%)
Fetal mortality	n=22 (5.5%)

GA=Gestational Age; CHD=Congenital Heart Disease; SVD=Small Vessel Disease; LSCS=Lower Segment Cesarian Section

DISCUSSION

Our study has revealed that a large number of GUCH patients can be treated with specialized interventional procedures in cardiac catheterization lab. The benefits were minimal hospital stay and early mobilization. Aleksandra,¹¹ reported pattern of GUCH in European Heart Journal 2016, in a span of 4 years. Most common surgical disease was ASD, TOF and aortic valve disease and in cath, most common procedure was device closure including ASD and PDA. Our study noted similar pattern of diseases and types of procedures being carried out by interventional cardiologist and congenital cardiac surgeon. British cardiac society in Heart 2002,¹⁰ observed the decrease in mortality and morbidity of infants with CHD and significant rise in the admissions of GUCH patients with complex issues and mortality in Royal Brompton Hospital. In comparison, our study has reflected the improvements in management and specialized care for GUCH population. We have reduced morbidity and mortality for GUCH population. Although our study is lacking psychosocial problems and surgical morbidity in GUCH patients on follow up. Shahbuddin,¹² in 2016 in 2nd annual surgery meeting provided the spectrum of GUCH patents for cardiac surgery reflecting a rise in numbers of population and most patients had ASD, TOF, VSD and concomitant valve surgeries with 2.1% mortality and 12% morbidity in Agha Khan Hospital. In another study by Horer published in Heart 2018, the immediate post operative mortality in GUCH patients was found to be 3.4%.¹³ Compared to both these

studies, we had higher mortality rate of 5.8%. Betro,¹⁴ published in *Circulation Research* in 2017 that significant improvements in cardiac catheterization for GUCH patients has increased the number of patients in cardiac cath lab with marked reduction in death and morbidity. The most important challenges are to address the specialized care for pulmonary hypertension, arrhythmias, infective endocarditis and heart failure. Most common age of presentation was from 25-45 years. Most common problems are of cognitive development, psychosocial rehabilitation and lifestyle adjustments. Advances in improved care for patients has led to a 20 times greater number of patients in OPD of obstetrical and surgical departments. Our study correlates with the fact that number of GUCH population has increased significantly over past few years. Anselm reported in *BMJ* 2006,¹⁵ the specialized needs of mothers with congenital heart disease which require higher vigilance and additional support. Extra care is needed for addressing pulmonary hypertension, arrhythmias, postpartum hemorrhages, intrauterine deaths (IUD) and congestive heart failure. The use of antifailure medications, anticoagulants and malnutrition predispose the obstetrical population towards maternal mortality. Our study revealed maternal mortality of 2.7% which is comparable to international outcomes. Most common defects were VSD, PDA, TOF & pulmonary stenosis followed by cardiomyopathies.

Another study published recently in *Ukrainian journal of cardiovascular surgery* has studied 896 GUCH pregnant women in a span of 7 years. Mean age of presentation was 27 years which is comparable to our results i.e. 28.3 years. Their maternal mortality was found to be 2.4%, almost similar to one found in our GUCH pregnant female population. However our fetal mortality is higher (5.5%) as compared to 3.8% found in *Ukrainian study*.¹⁶

Luh G.A.P reported in 2018 in *Medical Journal of Indonesia*,¹⁷ that in 6 years period there were 54 patients as compared to our study with 700 patients in 10 years. Most common intervention was ASD and PDA device closure just as our study concluded. Our institutional mortality was 0.4% as compared to their 3.7%. Their major complications were dysrhythmias as compared to ours which was device embolization.

Abdul Razzaq reported in 2019 in *Pakistan Journal of Medical Sciences*,¹⁸ that most common defects in GUCH population were ASD, VSD, TOF and PDA followed by Eisenmenger, pulmonary hypertension, rhythm disturbances and infective endocarditis.

In comparison, our study had a large cohort and multiple Interventions and outcomes are reported. The spectrum of diseases remains the same with lack of database for Eisenmenger, pulmonary hypertension, infective endocarditis and rhythm disturbances.

The shortfall of our study was that we did not conduct survey of psychological challenges faced by parents of GUCH population and their intelligence potential as reported by Carmen in 2017 in *Frontiers in Pediatrics*,¹⁹ by applying FSQ test to this specific group. Our study also did not encompass the comparison of surgical PVR versus Transcatheter pulmonary valve replacement in patients of TOF repair as reported by Ying,²⁰ in *Journal of thoracic disease* in 2019. It is becoming a major cohort of patients in GUCH population with advances in congenital cardiac surgery. Another major limitation of our study was failure to include GUCH patients on follow up with history of repaired congenital heart disease in childhood as done by Ruiz JMO.²¹ This was due to lack of database and registry of this particular group of patients. We have only mentioned immediate post procedure mortality in our study.

RECOMMENDATIONS

Congenital cardiac procedures in patients above 18 years of age are safe and carry low mortality and morbidity. A large number of patients can benefit from minimal invasive cardiac catheterization procedures & have improved quality of life. GUCH with pregnancy requires specialized care in tertiary care hospital.

Owing to the increasing population of GUCH patients, need of hour is to establish a separate registry for these patients. Specialized clinics need to be established specially for post congenital cardiac surgery repair follow up patients, patients with pulmonary hypertension and cardiomyopathies, so that in addition to their medical needs, appropriate means should be established to fulfil their psychosocial requirements as well. For this purpose, additional training is re-quired for Pediatric interventional cardiologists, Adult cardiologists with special interest in GUCH, obstetricians, nursing staff and psychologists.

LIMITATIONS OF STUDY

There was lack of proper registry for patients of GUCH diseases, difficult followup and limited resources.

CONCLUSION

With immense advancement in diagnosis and management of congenital cardiac diseases, a significant number of patients remain undiagnosed and untreated till the time they present as GUCH. Most common presentation is ASD which are mostly manageable by transcatheter intervention,

however late presentation carries a risk of complications like pulmonary hypertension.

Conflict of Interest: None.

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Author's Contribution

Following authors have made substantial contributions to the manuscript as under:

HA: Manuscript writing, drafting and editing

MAN: Intellectual contribution, concept and final approval

TM: Intellectual contribution, concept and final approval

SI: Proof reading, Intellectual contribution, final approval

AA: Formatting, critical review and final approval

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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