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CARDIOTHORACIC SURGERY

SHORT REPORT

Is Grown Up Congenital Heart (GUCH) disease different in a developing country?

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Abstract

In the current era grown up congenital heart disease (GUCH) patients undergoing surgical interventions are increasing. Most of the interventions in the developed countries are either complex or redo-operations in patients who had previously undergone repair, palliation or correction. However, in the developing countries most of the interventions are primary and corrective. This descriptive retrospective study comprised GUCH patients who underwent surgical intervention for congenital heart disease (CHD) at Aga Khan University Hospital, Karachi, from January 2006 to December 2015. A total of 195 patients had been treated surgically with a mean age of 31.0±13.5 years. Majority of the patients underwent surgical interventions for closure of atrial 109(55.3%) and ventricular 51(26.2%) septal defect. The most common complications were prolonged ventilation 16(8.1%). Overall mortality was 4(2.1%). GUCH in our practice is for primary procedure with simple diagnosis that should have been treated before reaching adulthood as is done in the developed countries.

Keywords: GUCH, CHD, Developing countries.

Introduction

Over the past century, massive breakthroughs have been achieved incardiovascular diagnostics and cardiothoracic surgery, leading to an increased survival of newborns with congenital heart disease (CHD).[1] Consequently, more patients with CHD reach adulthood, creating a completely new and steadily growing patient population with grown-up CHD (GUCH).[2] This is an inimitable challenge to cardiologists, anaesthetists and cardiac surgeons.[3] Careful, deliberate planning is crucial to transfer such patients from paediatric care to the adult cardiology and cardiothoracic surgery team. Birth defects are a global problem, but their impact is particularly severe in low and middle income countries (LMICs), where the conditions for prevention, treatment, and rehabilitation are more critical. Pakistan is LMIC in which approximately 40,000 babies are born with congenital cardiac defects annually. Most commonly, primary and corrective CHD repairs are done for atrial septal defect (ASD), ventricular septal defect (VSD), tetralogy of fallot (TOF) and patent ductusarteriosus (PDAs). On the other hand, literature suggests that most of the interventions in the developed countries are in the form of redo-operations in patients who had previously undergone repair, palliation or correction.[2] However, in a developing country like ours, most of the interventions are primary and corrective. Outcome data for GUCH surgery in a developing country like Pakistan is limited. We planned to study the frequency and spectrum of GUCH patients who underwent correction of congenital defect and to compare the GUCH burden with developed counties.

Methods and Results

This descriptive retrospective study was conducted at the Aga Khan University Hospital (AKUH), Karachi, and comprised data of 195 GUCH patients aged 16 years or above who underwent surgical intervention for CHD. Data was acquired from the dedicated cardiac surgery database over 10 years from January 2006 to December 2015. More than 175(90%) surgical procedures had been performed by paediatric cardiac surgeons as first-time surgeries.

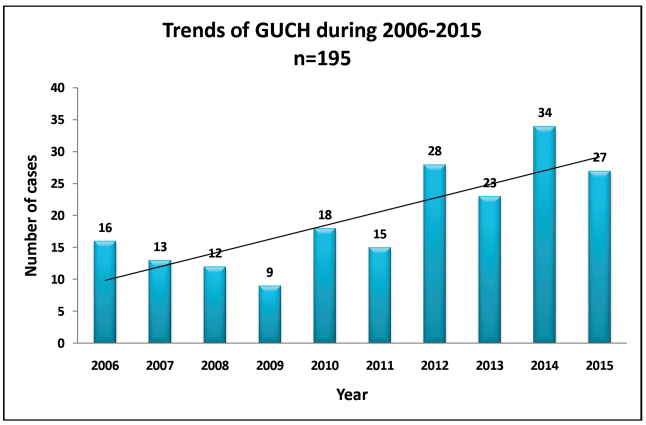

Figure: Trends of Grown Up Congenital Heart (GUCH) cases by year.

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Table: Pre and intraoperative clinical characteristics of GUCH patients by gender, n=195.

Characteristics	Total n=195	Male n=96 (49.2%)	Female n=99 (50.8%)	P value*
Age in years ±(SD)	31.0 ± 13.5	28.3 ± 12.0	33.6 ± 14.5	0.007
BMI kg/m^2	22.1 ± 5.0	21.3 ± 4.4	22.9 ± 5.6	0.028
Comorbidities				
Diabetes	8 (4.2)	3 (3.2)	5 (5.1)	0.722
Hypertension	36 (18.8)	14 (15.1)	22 (22.4)	0.202
Myocardial Infarction	9 (4.7)	5 (5.4)	4 (4.1)	0.742
Congestive Heart Failure	5 (2.6)	3 (3.3)	2 (2)	0.675
Arrhythmia	8 (4.1)	1 (1)	7 (7.1)	0.065
Ejection fraction				<0.001
≥ 50%	171 (87.7)	83 (86.5)	88 (88.9)	
30-50 %	18 (9.2)	9 (9.4)	9 (9.1)	
< 30%	6 (3.1)	4 (4.2)	2 (2.0)	
CHD anomalies†				
ASD	109 (55.9)	36 (37.5)	73 (73.7)	<0.001
VSD	51 (26.2)	39 (40.6)	12 (12.1)	<0.001
TOF	25 (12.8)	17 (17.7)	8 (8.1)	0.054
CoA‡	8 (4.1)	5 (5.2)	3 (3)	0.493
PDA	3 (1.5)	1 (1)	2 (2)	0.999
Glenn Shunt	1 (0.5)	0 (0)	1 (1)	0.999
Other congenital repair	3 (1.5)	1 (1)	2 (2)	0.999
Concomitant value replacement/repair				
Aortic value	14 (7.2)	9 (9.4)	5 (5.1)	0.278
Mitral value	15 (7.7)	4 (4.2)	11 (11.1)	0.105
Tricuspid value	3 (1.5)	0 (0)	3 (3)	0.246
Pulmonic Value	8 (4.1)	6 (6.3)	2 (2)	0.165
First time surgery	181 (92.8)	89 (92.7)	92 (92.9)	<0.001
Intra operative variables				
Bypass time median IQR (min)	81.5 (60-145)	105 (65-175)	70 (50-120)	<0.001
Cross clamp time median IQR (min)	55 (37-100)	70 (45-110)	48 (30-90)	0.002
Intra OP blood product used	82 (41.6)	37 (37.8)	45 (45.5)	0.273

*Independent samples t-test for continuous and Pearson Chi-square or Fisher's Exact test for categorical variables; mean and ± (SD) standard deviation or median (IQR) for skewed data where applied, p values relate to group differences. Coarctation of aorta; †More than one anomalies may be present
BMI: Body mass index
ASD: Atrialseptal defect
VSD: Ventricular septal defect
TOF: Tetralogy of fallot
‡CoA: Coarctation of aorta
PDA: Patent ductusarteriosus.

With regard to surgical strategy, ASD closure was performed with autologous pericardial patch and VSD closure with Dacron patch. The complete repair of TOF consisted of closure of VSD and relief of right ventricular outflow tract (RVOT) obstruction by muscle resection with pulmonary valve reconstruction or replacement. Repair of coarctation of aorta (CoA) was performed with end-to-end anastomosis. Bidirectional Glenn shunt was constructed for single-ventricle physiology.

The mean age of the patients was 31.0±13.8 years. There were 96(49.2%) male and 99(50.8%) female patients. The most prevalent procedure was ASD closure in 109(55.3%) patients, followed by VSD closure 51(26.2%), TOF 25(12.8%), CoA 8(4.1%), 3(1.5%) each for PDA and other congenital anomalies, and in 1(0.5%) case Glenn shunt was performed. ASD was more prevalent in females ($p<0.05$), whereas VSD and TOF were distinct in males ($p<0.05$ each). Overall, 181(93%) patients were operated for the repair of CHD for the first time. Hypertension was common comorbidity in 36(18.8%) patients. Concomitant repair or replacement of aortic valve was performed in 14(7.2%) patients, mitral 15(7.7%), tricuspid 3(1.5%) and pulmonic 8(4.1%) cases. Intraoperative Median bypass time was 81.5 minutes (intraquartile range [IQR]: 60-145), and median cross clamp time was 55 minutes (IQR: 37-100) (Table).

Most common complication was prolonged ventilation (>24 hours) in 16(8.1%) patients. Overall post-operative morbidity was 27(14%).There were 4(2.1%) deaths; 2(1%) each died of arrhythmia and septicaemia. Only 1(0.5%) patient required re-operative procedure.

Conclusion

History of CHD management is an inspiring tale of persistence, courage and ingenuity of pioneer cardiac surgeons. However, their attempts were thwarted with a mortality rate of about 85% for that era.[4] With the advancement in diagnostic imaging, preoperative evaluation, careful intra-operative management and improved postoperative care has led to an overall decrease in mortality and increase in the survival of CHD patients into adult hood.[1] Putamen et al. studied 830 adults with CHD undergoing 963 procedures over a 17 year period and concluded that surgery can be performed safely with low mortality and limited serious morbidity.

The trend showed a considerable increase in GUCH patients worldwide. In Netherland, the population estimated to be of GUCH disease patients will reach 20,000 in coming years.[5] Such an increasing trend is observed in other developed countries as well and reports from Canada and the United Kingdom (UK) have shown significant rise in GUCH compared to paediatric patients.[6] A survey in UK spanning over a decade predicted the annual rise of GUCH population to be 1600 with more than half of the patients having complex lesion, and thus demanding special care in follow-up by the attending physicians.[7] Another study from UK reported a decline in number of simple procedures from 46% to 28% and there is increase in number of re-operative surgeries in GUCH patients. Most of them are undergoing surgery for complex lesions or surgery for residual or sequel of previous corrective or palliative procedure.[8] Surgical procedures like coronary artery bypass grafting, aortic root surgery for connective tissue disorder, valve replacement and Maze procedure for ablation of refractory tachydysrhythmias are some of the complex procedures performed in GUCH patients.[9] We know now that no surgical "correction" exists for CHD and all such surgical modalities are palliative surgeries that require surgical sequel in near or far future, for example pulmonary regurgitation with right ventricular dysfunction is a common complication after reparative surgery of TOF.[10] In our country with the development of paediatric congenital cardiac surgical programmes, a number of adults with congenital lesion are being discovered and are seeking advice for treatment. In our series, similar rising trends have been observed over the years (Figure).

However, contrary to the west we are operating for simple procedure like ASD and VSD and we still have a population of patient who grow up with their lesions considering inoperable or with the false belief that it will go away with time. It is call of the time for countries like Pakistan to have well-defined guidelines and policies for developing dedicated centres in this regard. Such centres will be equipped with resources that will especially focus attention on the complex anatomy, physiology, biochemistry and more often psychiatry of GUCH patients. We at present do not have dedicated facilities for GUCH patients. This report highlights the fact that most of these simple lesions should have been treated before reaching adulthood.

Since most of the congenital lesions are not totally corrected, we should be ready to face them as GUCH patients with more complex issues as already faced in most of the developed countries.

Disclaimer: The study was presented as Poster at the 6th World Congress of Paediatric Cardiology and Cardiac Surgery (Feb. 17-22, 2013) at Cape Town, South Africa.

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