

## Surgery of Grown up Congenital Hearts (GUCH) in Pakistan; what is different from the developed countries?

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### ABSTRACT

**Background:** Outcome data for Grown up congenital heart (GUCH) surgery in a developing country like Pakistan are limited. We describe our experience of GUCH surgery at Rawalpindi Institute of Cardiology in comparison with the results from developed countries.

**Methods:** The data of patient who underwent GUCH surgery from Feb 2013 to Feb 2015, was retrieved from our dedicated Database. Patient characteristics, morbidity and mortality were described. We examined our data to evaluate the scope of GUCH surgeries in a predominantly Adult Cardiac Surgery center in a developing country.

**Results:** A total of 170 patients of GUCH surgery were found in the database. Median age, body weight and body surface area (BSA) were 19 years, 42.75 kg and 1.37 m<sup>2</sup> respectively. The operative procedures included Isolated ASD Repair (n=71), ASD with valve Repair (n=8), total correction of Fallot's Tetralogy (n=29), Isolated VSD repair (n=20), VSD repair with valve Repair/Replacement (n=7), Isolated PDA ligations (n=12), and repair of Anomalous Pulmonary Venous Connections was done (n=3). Nine patients underwent miscellaneous operations including Coarctation repairs, Modified BT shunts, Glenn Shunt, AVSD repair, Pulmonary valvotomy and resection of sub-aortic membrane. One patient underwent Redo TOF with Pulmonary valve replacement. The median CPB time was 58 mins, median ICU stay was 24 hrs and median length of hospital stay was 6 days. Overall hospital mortality was 4.7 %.

**Conclusion:** In a developing country like Pakistan where the services for congenital heart surgery are not fully developed, operating upon grown up congenital hearts in an adult cardiac setting is a challenge worth taken.

**Keywords:** GUCH, fallot's tetralogy, pulmonary valvotomy

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### INTRODUCTION

A vast majority of developing nations are struggling to achieve satisfactory standards of adult cardiac surgery and therefore access to the congenital cardiac surgery is an even more arduous task<sup>1</sup>. Being an expensive specialty, requiring highly individualized care and multi-disciplinary involvement makes congenital cardiac surgery a difficult task even in the developed countries<sup>2</sup>. There is a direct link between the hospital cost and the complexity of the congenital cardiac surgical procedure<sup>3</sup> which makes it an uphill task to run congenital cardiac surgery services. In addition to economic limitations there are several other reasons for slow progress in this speciality including political apathy, non structured healthcare system and paucity of skilled man power<sup>4,5</sup>. Pakistan is a low middle income country where it is estimated that around 40,000 babies are born with congenital cardiac defects annually<sup>6</sup>. Despite the odds, over the years several congenital cardiac surgery programs

have been started in the developing countries including Pakistan<sup>7,8</sup>. Developed countries have made progress in congenital cardiac surgery through research, surgical innovations and by strengthening of the supporting disciplines like intensive care and interventional cardiology<sup>9</sup>. Early referrals and timely surgical interventions have made a gross difference in the spectrum of congenital cardiac surgery<sup>10</sup>. Survival of these patients to adulthood has led to the emergence of a new clinical sub-set of patients variously labelled as Adults with Congenital Heart Defects (ACHD) or Grown Ups with Congenital Heart Defects (GUCH)<sup>11,12</sup>.

There is a lack of data on the spectrum GUCH patients in the developing countries, but the available data suggests that the majority of the lesions need primary and corrective surgeries which in the developed countries are done at a much younger age.

In this article we compare the spectrum of our GUCH population and their surgical outcome data of GUCH surgery with the available data from the other countries.

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**PATIENTS & METHODS**

After approval from the hospital ethics committee, this descriptive study was conducted at the Department of Cardiac Surgery, Rawalpindi Institute of Cardiology (RIC). The cut-off limit for age of GUCH was set 18-years. GUCH patients comprised 9% of our total workload. The data of all the GUCH patients (n=170) undergoing surgery at Rawalpindi Institute of Cardiology, from February 2013 to February of 2015 was retrieved from our dedicated electronic database. Data was analyzed on SPSS version 15.0. Quantitative variables e.g., age, weight, height, body mass index (BMI), body surface area (BSA), cardiopulmonary bypass time, cross clamp time, post operative blood loss (chest drainage), ICU stay, ventilation duration, duration of inotropes and the hospital stay were expressed as mean and standard deviation (SD). Frequency and percentages were used to describe categoric variables like gender, type of disease and surgery. Patient characteristics, morbidity and mortality were described.

**RESULTS**

**Patient Characteristics:** The mean age was 22.85 yr (SD±11.6), mean weight was 42.58kg (SD ±19.53), mean height was 151.24cm (SD±20.45), mean BMI was 21.20 kg/m<sup>2</sup> (SD ±45.82) and BSA was 1.312 m<sup>2</sup> (SD ±0.36). The gender distribution consisted of 91(53.5%) male and 79(46.5%) female.

**Diagnostic Spectrum:**

GUCH patients consisted of 82(48.2%) atrial septal defects (ASDs), 33(19.4%) tetralogy of fallot (TOF) patients, 27(15.8%) patients had ventricular septal defects (VSDs), 14(8.2%) patients had patent ductus arteriosus (PDAs), 4(2.4%) patients had anomalous venous connections, 3(1.7%) patients had Coarctation of aorta (CoA), 3(1.7%) patients had ruptured sinus of valsalva (RSOV), 2(1.1%) patients had pulmonary atresia with single ventricle, 1(0.5%) patient had atrio-ventricular canal defect and 1(0.5%) patient had pulmonary stenosis (PS).

**Surgical Procedures:** Consequently, 82 patients underwent ASD repairs; 74 were isolated repairs, 7 had associated valve repairs and 1 patient had a concomitant mitral valve replacement. Of the 33 patients diagnosed with tetralogy of fallot 32 patients came for primary corrective surgery with one patient undergoing surgery with modified Blalock-Taussig shunt due to small pulmonary arteries, whereas only one patient underwent redo surgery with pulmonary valve replacement, he was operated at a young age at another center. There were 27 patients undergoing surgery for VSD repairs, 20 had isolated VSD repairs, 6 patients had concomitant valve repairs and only

one patient underwent Aortic and Mitral valve replacements. There were 29 patients who were categorized in the miscellaneous surgical procedure category; 13 patients underwent PDA ligations, 1 patient underwent PDA ligation with sub aortic membrane excision, 3 Coarctation of aorta repairs, 3 repairs for ruptured of sinus of valsalva repairs, 2 partial and 2 total anomalous pulmonary venous connection repairs. Both the patients with the single ventricular pathologies with pulmonary atresia underwent surgery for Glenn’s shunt. One patient had an atrio-ventricular canal defect repair and one patient had a pulmonary valvotomy for pulmonary stenosis.

**Outcome:** The main outcome variables analyzed were mean Cardio-pulmonary bypass time was 65.26 min (SD ±30.4), mean cross clamp time was 44.7 min (SD ±27.04), mean ICU stay was 24.22 hr (SD ±12.64), mean ventilation duration was 4.8 hr (SD ±6.8), the mean duration of inotropes was 16.11 hr (SD ±19.28), the mean chest drainage was 506 ml (SD ±516.27) and the mean hospital stay was 7.06 days (SD ±3.5). In the post operative complications, 21 (12.35%) patients had Arrhythmias, 2 (1.17%) patients had low cardiac output syndrome, 3 (1.7%) patients had AV block requiring pacing, 8 (4.7%) patients had cardiac arrest, 9 (5.3%) patients had pulmonary complications, 12 (7%) patients required re-intubation and ventilation, 5 (2.9%) patients required mechanical ventilation more than 7 days, 4 (2.35%) had renal failure requiring dialysis, 3 (1.76%) patients under went unplanned re operation and 6 (3.5%) patients underwent re-operation for bleeding. The overall hospital mortality was 4.7% (8 patients).

Table 1: Patients characteristics

Characteristics	Mean	Median	Standard Deviation
<b>Pre-operative</b>			
Age (years)	22.85	19	11.607
Weight (kg)	42.58	42.75	19.53
Height (cm)	151.24	156	20.45
BMI (kg/m <sup>2</sup> )	21.20	17.01	45.82
BSA (m <sup>2</sup> )	1.312	1.37	0.36
<b>Operative &amp; Post-operative</b>			
CPB Time (mins)	65.26	58	30.4
Cross Clamp time (mins)	44.7	37	27.04
ICU stay (hrs)	24.22	24	12.64
Ventilation time (hrs)	4.8	4	6.8
Inotropes (hrs)	16.11	8	19.28
Chest Drainage (mls)	506.043	360	516.27
Total hospital stay (days)	7.06	6	3.5

**Surgeon Experience vs. Preference:** There were 4 operating surgeons in the cardiac surgery department. The most experienced surgeon with a post fellowship experience of 16 years had operated upon 111 patients, where as the second most experienced surgeon with a post fellowship experience of 13 yrs had only operated upon 14 patients. The two junior surgeons both having a post fellowship experience of 2.4 years had operated upon 24 and 21 patients respectively.

Table 2: Operative Procedures

Procedure	n	%
<b>ASD &amp; Related Procedure (n=82)</b>		
Isolated ASD Repair	74	43.5
ASD + MV Repair	4	2.35
ASD + MV Replacement	3	1.76
ASD + MV & TV Repair	1	0.59
<b>VSD/TOF &amp; Related Procedures (n=59)</b>		
Isolated VSD Repair	20	11.76
Isolated TOF Repair	31	18.24
Re-do TOF Repair	1	0.59
VSD + AVR	4	2.36
VSD + AV Repair	1	0.59
VSD + MV Repair	1	0.59
VSD + MVR + AVR	1	0.59
<b>Miscellaneous (n=29)</b>		
PDA Ligation	13	7.76
Coarctation of Aorta Repair	3	1.77
Rupture Sinus of Valsalva	3	1.77
PAPVC	2	1.18
TAPVC	2	1.18
Subaortic Membrane	1	0.59
BT shunt	1	0.59
Glenn Shunt	2	1.18
AVSD	1	0.59
Pulmonary Valvotomy	1	0.59

Table 3: Operative Outcomes

Variable	RIC	STS
Arrhythmia	21(12.35%)	496(9.4%)
Low cardiac output	2(1.17%)	87(1.7%)
AV block requiring pacing	3(1.7%)	69(1.3%)
Cardiac arrest	8(4.7%)	52(1.0%)
Pulmonary complications	9(5.3%)	286(5.4%)
Reintubation	12(7%)	71(1.4%)
Mechanical Ventilation > 7days	5(2.9%)	52(1.0%)
Unplanned reoperation	3(1.76%)	111(2.1%)
Bleeding requiring reoperation	6(3.5%)	100(1.9%)
Renal failure (temporary dialysis)	4(2.35%)	56(1.1%)
Death	8(4.7%)	109(2.07%)

## DISCUSSION

The term GUCH was proposed by Jane Somerville<sup>12</sup> and refers to adults  $\geq 18$  years with congenital heart defects (CHDs). It is categorized into the following groups;

- Patients with congenital heart defects (CHD) presenting for the first time as adults,
- CHD patients with previous surgical procedures presenting for further correction or palliation,
- CHD patients coming for anticipated re-operations,
- CHD patients requiring repair of residual lesions,
- CHD patients coming for heart/ heart-lung transplant and
- CHD patients with acquired heart disease .

The vast majority of GUCH patients belong to the first group in the developing countries. Our study showed 48.2% ASDs, 19.4% TOFs, 15.8% VSDs, 8.2% PDAs, 2.4% patients with anomalous venous connections, 1.7% patients with CoA, 1.7% patients with RSOV, 1.1% patients with pulmonary atresia with single ventricle, 0.5% patients had atrio-ventricular canal defect and 0.5% patient had Pulmonary Stenosis. This was similar to the trend seen in the studies from the developing countries where ASDs, VSDs, PDAs were the most common acyanotic CHDs whereas TOF was the most common cyanotic CHD<sup>13,14,15</sup>.

A different spectrum was seen in the data analyzed by Mascio CE et al<sup>9</sup> from STS database representing 68 centers from the developed countries, where the vast majority of the patients belonged to the second, third and fourth categories of GUCH patients. Majority of the GUCH patients undergoing surgeries have already undergone surgery at a young age and had now presented for a redo surgery for further correction/palliation, anticipated surgeries and repair of residual lesions. Primary corrective surgery represented only a small proportion.

In the developing world, the most common primary and corrective CHD repairs were done for ASDs, VSDs, TOF and PDAs. These procedures constituted 74.7%, 77.8%, 82% the series by De Mello GA et al<sup>13</sup>, Hannoush et al<sup>14</sup> and Edwin F et al<sup>15</sup> respectively. However, in our series it was 91.6%, thus, showing that the surgical procedures for GUCH patients in the developing countries are primary and corrective, unlike the developed countries where the surgeries in these patients are mainly redo and palliative.

Regarding the patient characteristics, there was a slight male preponderance in our study with 53% being males and 46.5% being female. The reverse was seen i.e., a female preponderance was seen in other studies from the developing countries. The mean age of our patients was 22.85 years (SD±11.6) which showed that our patients were 5-10 years younger as compared to the other GUCH surgery series from the developing world<sup>13,14,15</sup>. The mean age of patients from the STS congenital database analysis was 25 years and as the data showed that the majority of these patients were presenting for a redo surgery rather than a primary corrective procedure<sup>9</sup>.

A comparatively large number of GUCH patients underwent surgery at our center which was accumulated over the course of only two years. Hannoush et al from Lebanon had 206 patients in their study over the course of 20 years<sup>14</sup>. In the study by Klčovansky J et al from Denmark who had a comparatively similar GUCH surgery profile albeit with a gradually increasing redo surgeries, operated 225 patients over a course of 7 years<sup>16</sup>. At Rawalpindi Institute of Cardiology, the cardiac surgery department caters primarily for the Adult cardiac surgery patients. The 170 GUCH patients undergoing surgery comprised 9% of our work load, which is a huge number for an adult cardiac center in a developing or a low middle income country like Pakistan when compared with other countries with a similar economic profile<sup>13,14,15</sup>.

In our study we described a variety of operative parameters which have not been cited by other similar articles from the developing countries. However, when we compared our operative parameters with the series of Klčovansky J et al from Denmark, we found that our mean CPB time 65.26 mins (±30.4) vs. 104 mins (±64), our mean cross clamp time 44.7 mins (±27.04) vs. 65 mins (±39), our mean ICU stay was 24.22 hours (±12.64) vs. 2 days (± 4) and our patients had a total hospital stay of 7.06 days (±3.5) vs. 12 days (±10). Although our figures appear much more impressive but the difference is due to the fact that their surgeries were much more complex and 44% of their GUCH surgery cases were redo cases who previously had 1 to 5 surgeries [16]. When we individually compared these parameters for ASD, VSD, PDA and TOF surgeries our results were remarkably similar. This was quite encouraging for an adult cardiac center like ours in a country with limited resources.

We also compared the post operative complications in our patients with those of the STS database analysis. The post operative complication rates for Arrhythmias, Low cardiac output syndrome, heart block requiring pacing, unplanned re-operation and

pulmonary complications were similar. We had higher rates of Re-intubations 1.4% vs. 7 %, Mechanical ventilation more than 7 days 1% vs. 2.9%, Re-operation for bleeding 1.9% vs. 3.5 % and Renal failure requiring dialysis 1.1% vs. 2.35%<sup>9</sup>.

The mortality at our center for GUCH surgery was 4.7% which was comparable with the figures from STS database which ranged from 2.1% to 5%. We had slightly higher mortality rates when compared with those from the developing world which was around 3%<sup>13,14,15</sup>.

An interesting finding that we noted was the surgeon preference, out of the two senior surgeons only one was operating upon the bulk of GUCH patients where as the second surgeon was operating only upon a fraction. It was encouraging to see that the two young surgeons were operating upon a sizable proportion of GUCH patients at RIC with good results. As noted in USA by Karamlou T et al, the surgical outcomes were improved with increasing number GUCH surgeries per surgeon per year<sup>17</sup>. The current estimates suggest that there will be a shortage of dedicated Cardiothoracic surgeons by 2020<sup>18</sup>, and with the growing number of GUCH patients there will be an ever more need for cardiothoracic surgeons in the developed countries let alone developing countries. In the light of this, it stands true for adult cardiac surgeons in developing countries to develop interest in pediatric cardiac surgery and operate upon GUCH patients as an obligation

## CONCLUSION

In a developing country like Pakistan where the services for congenital heart surgery are not fully developed, operating upon grown up congenital hearts in an adult cardiac setting by properly trained surgeons, is a challenge worth taken.

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