The Cardiology 1 (2): 87-94 , 2005

Early Outcome in Guch Surgery

[1]A. Pascal, Berdat, [2]Nicolas Trost, [2]Mladen Pavlovic, [2]Jean-Pierre Pfammatter, [1]Thierry Carrel,
[1]Clinic for Cardiovascular Surgery, [2]Division for Pediatric Cardiology, Swiss Cardiovascular Center Bern,
University Hospital, Bern, Switzerland

Abstract: To assess early results and identify perioperative risk factors in Grown-ups with Congenital Heart Disease (GUCH) needing primary or redo cardiac surgery. Perioperative outcome of 296 consecutive GUCH patients (mean age 35±16 (14-72) years) needing primary (230 pts, Group 1) or redo surgery (66 pts, Group 2) was analysed. Most frequent initial cardiac defects were ASD (49%), LVOT pathology (23.3%), Marfan syndrome (7.4%) and coarctation (6.1%). Closure of ASD or VSD (54.7%), aortic valve procedures (34.5%) and aortic replacement (12.2%) were the most frequent performed. Durations of surgical repair, CPB, aortic cross-clamping and hypothermic circulatory arrest (HCA) were all significantly longer and core temperature lower in Group 2 than 1 (all p<.0001). Early mortality was 3.4% but was significantly higher in Group 2 than 1 (7.6% vs. 2.2%; p=.04). Significant postoperative complications occurred in 30% and were more frequent in Group 2 (42.4%) than 1 (26.5%; p=.01), especially multi-organ failure (MOF) (p=.015), low output failure (LOF) (p=.04), respiratory complications (p=.0009), renal failure (p<.0001) andtotal av-block (p=.04). MOF, LOF, HCA and respiratory complications were independent predictors for fatal outcome. Surgery in GUCH patients is mostly performed due to ASD, outflow tract lesions, Marfan syndrome and coarctation. First operations have an excellent outcome, whereas re-operations still have increased mortality and morbidity and particular risk factors. Therefore, close follow-up, timely referral for re-intervention and aggressive perioperative monitoring are mandatory to reduce perioperative risks and improve results in these patients.

Key Words: Adult , congenital heart disease, cardiac surgery, early outcome

INTRODUCTION

Grown-ups with Congenital Heart disease (GUCH) are becoming an important and still increasing group of patients, since quality of current surgical repair allow the majority of newborns with congenital heart disease to survive to adulthood. A smaller proportion of them reach a dulthood without I ntervention and usually present with symptoms, the majority however, are those requiring reoperations for various reasons, i.e. residual defects, long-term sequelae, degeneration or outgrow of implants, or terminal heart failure. Surgery in these patients may be challenging, because of long-standing defects, the diversity of underlying pathologies, the presence of sequelae and residual defects from previous operations and the multi-organic nature of many complex cardiac malformations associated with congenital syndromes and cyanosis. Although operations in GUCH patients become more and more frequent today, limited knowledge exists on their perioperative risk factors and only a few studies have been published so far focusing on these operations and their inherent risks.

MATERIALS AND METHODS

All records from patients with congenital heart disease 14 years or older needing their first operation (Group 1) or a reoperation (Group 2) between July 1987 and December 2000 were retrospectively analysed. The initial congenital cardiovascular pathology, as well as the indications for surgery and redo-surgery were assessed. Outcome of all operations and previous interventions was recorded. Early mortality was analysed for first and reoperations in order to identify perioperative risk factors for fatal outcome.

Statistical analysis: Data are presented as Mean±SD and range. Percentages are given where appropriate. For statistical analysis, Statview 5.0.1 for Windows (SAS Institute Inc., Cary, NC, USA) was used. A Mann-Whitney-U test and Fisher's exact test for contingency tables were used for analysis of between-group differences. Variables identified in univariate analysis as risk factors for early mortality were entered into a multivariate logistic regression model to assess the independent impact of each risk factor on early mortality.

RESULTS

296 patients with a mean age of 35±16 (14-72) and a mean EF of 64.2±13.5 (10-92) were operated at our institution between July 1987 and December 2000. 230

Corresponding Author: Berdat PA, Clinic for Cardiovascular Surgery, Inselspital, CH-3010 Bern, Switzerland
Tel: ++41316329268 Fax: ++41316321929

Table 1: Demographic data

	Total		Group 1		Group 2		
Variable	(No.)	(%)	(No.)	(%)	(No.)	(%)	p- Value
Age (y)	35±16	14-72	37±16	14-72	28±12	14-64	<0.0001
Sex (m:f)	192:104	65:35	145:85	53:47	47:19	71:29	0.0111
Ejection fraction (%)	64±14	10-92	66±10	30-87	59±19	10-92	ns
Initial pathology							
ASD	145	48.7	141	61.3	4	6.1	<0.0001
LVOT pathology	69	23.3	52	22.6	17	25.8	ns
Marfan syndrome	22	7.4	15	6.5	7	10.6	ns
Coarctation	18	6.1	8	3.5	10	15.2	0.0016
VSD	14	4.7	6	2.6	8	12.1	0.0004
Tetralogy of fallot	11	3.7	2	0.9	9	13.6	0.0001
PDA	8	2.7	8	3.5	-	-	ns
d-TGA	7	2.4	-	-	7	10.6	0.0006
PAPVC	2	0.7	2	0.9	-	-	ns
Partial / complete AV-canal	2	0.7	1/1	0.9	-	-	ns
Truncus arteriosus	2	0.7	1	0.4	1	1.5	ns

PAPVC: Partial anomalous pulmonary venous connection; PDA: Patent Ductus Arteriosus; d-TGA: d-Transposition of the Great Arteries

Table 2: Previous procedures in Group 2

Inital cardiac defect n=66	Previous interventions	No. of interventions
Isolated LVOT pathology (17)	Total	25
	Commissurotomy	12
	SAS resection	2
	Konno	1
	Composite homograft	3
	Aortic vale repair	5
	BVAD	1
Coarctation (10)	Total	13
	End-to-end anastomosis	8
	Dacronpatch	1
	Graft	2
	Ascending-descending Bypass	2
Tetralogy of Fallot (9)	Total	15
	Blalock-taussig shunt	8
	Aorto-pulmonary shunt	1
	Repair	6
VSD (9)	Total	10
	Closure	7
	Pulmonary artery banding	1
	SAS resection	1
	Aorto-pulmonary shunt	1
Marfan syndrome (7)	Total	15
	Composite graft	6
	Arch replacement	3
	Ascending aortic replacement	3
	Aortic valve repair	1
	Aorta other replacement	4
d-TGA (7)	Total	12
	Blalock-taussig shunt	3
	Atrioseptectomy	4
	Rastelli repair	2
	Atrial switch procedure (Mustard) TCPC	2
ASD (4)	Total	5
	Sondergaard repair	2
	Direct closure	2
	device closure	1
Truncus arteriosus Type I (1)	Truncus repair	1

BVAD: Biventricular Assist Device; DORV: Double Outlet right Ventricle; SAS: Subaortic Stenosis; TCPC: Total Cavo-pulmonary Connection; d-TGA: d-Transposition of the Great Arteries

patients (78%) were operated for the first time (Group 1) and66 of 296 patients (22%) needed 95 reoperations (Group 2). Patients referred for their first operation were significantly older (p<.0001) and more frequently females (p=.0111) compared to patients for reoperations(Table 1).

Cardiac defect, indication and surgical treatment: The largest subgroup (Table 1) were patients with ASD (145 pts), including all types, accounting for 48.7% of all patients andsignificantly more frequent in Group 1 than 2 (61.3% vs. 6.1%; p<.0001). Coarctation (15.2% vs. 3.5%;

Table 3: Surgical procedures

	Group 1		Group 2		
n=296	(No.)	(%)	(No.)	(%)	p- value
Aortic valve	71	30.9	31	47	0.0187
-mechanical/bio prosthesis	37/2		17		
-composite mechanical prosthesis	20		3		
-composite homograft	1		4		
-valve repair	3		1		
-SAS resection	8		6		
+ ascending aortic enlargement	2		4		
Aortic replacement	19	8.3	17	25.8	0.0004
-ascending/ Yacoub	9/1		6/1		
-arch	6		3		
-descending/ thoracoabdominal	-/2		4/2		
-abdominal (infrarenal)	1		1		
Closure	146	63.5	16	24.2	<0.0001
-residual VSD	-		6		
-VSD	5		6		
-residual ASD	-		2		
-ASD/ PFO/ SVD±PAPVC	141	61.3	2		
Conduits			12	18.2	<0.0001
-replacement			5		
-new RV-PA			7		
RVOT procedures	6	2.6	8	12.1	0.0004
Coarctation repair	9	3.9	5	7.6	ns
-in situ replacement	6		3		
-ascending-descending bypass	3		2		ns
Mitral/ Tricuspid valve procedures	11/-	4.8	1/3	6.1	ns
Miscellaneous others	10	4.3	4	6.1	ns
Heart transplantation	-		3	4.5	0.0107
Coronary artery bypass grafting	5	2.2	-		ns
Total repair	135	58.7	4	6.1	<0.0001

HTX: Heart Transplantation; PA: Pulmonary Artery; PAPVC: Partial Anomalous Pulmonary Venous Connection; RV: Right Ventricle; RVOT: Right Ventricular Outflow tract; SAS: Subaortic Stenosis

Table 4. Operations data

	Total		Group 1		Group 2		
n=296	mean±SD	Range	mean±SD	Range	mean±SD	Range	p-value
Operation time (min)	184.5±81.3	60-630	166.0±63	60-390	246.0±102	110-630	0.0001
CPB time (min)	71.2±48.6	00-261	61.0±41	00-234	105.0±58	000-261	0.0001
AXC time (min)	42.6±36.8	00-165	37.0±33	00-165	62.0±42	000-154	0.0001
HCA time (min)	1.5±5	00-32	0.8±3.6	00-24	4.0±8	000-32	0.0001
Body core temperature (°C)	28.9±5.8	17-36.5	29.7±5.5	17-36	2.0±6	17.3-36.5	0.0001
ICU stay (d)	1.7±1.3	00-7	1.6±1.1	01-7	2.0±1.7	000-7	ns
Postoperative LOS (d)	9.9±4.8	00-27	9.6±4.6	01-27	10.9±5.5	000-24	ns

AXC: Aortic cross Clamp; CPB: Cardiopulmonary Bypass; HCA: Hypothermic Circulatory Arrest; ICU: Intensive Care Unit; LOS: Length Of Stay

Table 5. Early mortality and morbidity

	Total		Group 1		Group 2		
	(No.)	(%)	(No.)	(%)	(No.)	(%)	p-value
Mortality	10	3.4	5	2.2	5	7.6	0.04
Arrhythmias	63	21.3	43	18.7	20	30.3	ns
supraventricular/ ventricular	51/ 14		39/9	17/ 3.9	12/ 5	18.4/7.6	ns
total av-block	11		5	2.2	6	9.2	0.04
(Pacemaker insertion)	(5)		(2)	(0.8)	(3)	(4.6)	
Low output failure	8	2.7	3	1.3	5	7.6	0.04
Intraaortic balloon pump3	3	1	0	0	3	4.6	0.01
Neurological deficit	13	4.4	10	4.3	3	4.6	ns
transient/ permanent	8/5		6/ 4		2/1		
Respiratory complications	13/ 5	4.4	7/ 2	3.9	5/ 3	12.2	0.0009
Renal failure	12	4.1	3	1.3	9	13.6	<0.0001
dialysis	3		1	0.4	2	3.1	
Multi-organ failure	8	2.7	3	1.3	5	7.6	0.015
Revisions	6	2.0	3	1.3	3	4.6	ns
Total	89	30.0	61	26.5%	28	42.4%	0.01

Table 6: Early mortality

Nr	Age (y)	Diagnosed	Operation	Mode of death	Survival (d)
1	65	Marfan; A dissection	Composite graft	CVA	7
2	47	Marfan; A dissection	Composite graft	CVA	6
3	17.5	Marfan; AAE; AR, MR	Composite graft, MVR, TVR	MOF	1
4	36	ASD II, PHT	ASD closure	LOF	4
5	70	ASD II, LV perforation	ASD closure, LV repair	Bleeding	2
6	29	Marfan; Chronic dissection	Thoraco-abdominal aortic replacement	Bleeding	0
7	19.5	VSD, PA	Central aorto-pulm onary shunt	MOF	3
8	32.5	Rastelli; DORV, TGA, PS, degenerated TVP	Conduit exchange, TVR	LOF/MOF	1
9	15.5	DORV, TGA, PS	Rastelli repair	MOF	6
10	24	TGA, VSD, PA	Rastelli repair	LOF/ MOF	1

AAE: Anuloaortic Ectasia; AR: Aortic Regurgitation; CVA: Cerebro-vascular Accident; DORV: Double Outlet Right Ventricle; LOF: Low Output Failure; MOF: Multi-organ Failure; MR: Mitral Regurgitation; MVR: Mitral Valve Repair; PA: Pulmonary Atresia; PHT: Pulmonary Hypertension; PS: Pulmonary Stenosis; TGA: Transposition of the Great Arteries; TVP: Tricuspid Valve Prosthesis; TVR: Tricuspid Valve Repair

Table 7: Risk factors for early mortality in univariate analysis

Variable	Group 1 (p-value)	Group 2 (p-value)
Cyanosis	ns	0.0021
Coarctation	ns	0.0424
Marfan syndrome	0.0452	ns
Transposition of the great arteries	ns	0.0041
Pulmonary atresia	-	0.0135
Number of operations	-	0.051
Operation time (min)	ns	0.0012
CPB time (min)	0.0236	0.0012
Aortic cross-clamp time (min)	ns	0.0015
Use of hypothermic circulatory arrest	<0.0001	ns
Core temperature (°C)	ns	0.0121
Total correction	ns	0.0262
Composite graft	0.0059	ns
Aortic surgery	0.0495	ns
Low output failure	0.0044	<0.0001
Use of intra-aortic balloon pump	ns	0.0164
Use of epinephrine	<0.0001	0.0014
Use of dobutamine	0.006	ns
Perioperative myocardial infarction	0.0243	ns
Multi-organ failure	0.0011	<0.0001
Respiratory complications	0.0192	0.0316
Renal failure	ns	0.0013
Dialysis	ns	0.0065
Stroke	0.0188	0.0169

Table 8: Independent predictors of early mortality in logistic regression analysis

Variable	Odd ratio	95%CI	P-Value
Multi-organ failure	14.5	1.1-177.4	0.0363
Low output failure	157.6	13.3-1865.8	<0.0001
Hypothermic circulatory arrest (min)	1.148*	1.0-1.3	0.0332
Respiratory complications	12.788	0.2-690	0.210

Pearson >0.9999, Deviance 0.9992, Likelihood ratio <0.0001 *OR relates to increases in the unit measured, CI: Confidence Interval; Coef: Coefficient;

(p=.0016), VSD (12.1.% vs. 2.6%; p=.0006), TOF (13.6% vs. 0.9%; p=.0001) and TGA (10.6% vs. 0%; p=.0006) were all significantly more frequent in Group 2 than 1. (Table 2)

Most frequently, repair of ASDs (145/296; 49%), including PFOs and SVDs, followed by interventions on the aortic valve (102/296; 34%) or aortic replacement (36/296; 12%) were necessary. Closure of septal defects (63.5% vs.24.2%; p<.0001) and total repair (58.7% vs. 6.1%;p<.0001) were significantly more frequent in Group 1 than 2, whereas aortic valve procedures (47% vs. 30.9%; p=.018), aortic surgery (25.8% vs. 8.3%; p=.0004), RVOT- (12.1% vs.2.6%; p=.0004) or conduit procedures (18.2% vs. 0%; p<.0001) andheart transplantation (4.5% vs. 0%; p=.01) were significantly more frequent in Group 2 than 1. 34 patients had their first reoperation 15.7±9 (0.04 – 39.4) years after the initial procedure, 20 their second, 10 their third and 2 patients 4 or more reoperations (Table 3). Duration of operation (246±102 vs. 166±63min; p<.0001), CPB-(105±58 vs. 61±41 min; p<.0001), aortic cross-clamp- (62±42 vs. 37±33 min; p<.0001) and deep hypothermic

circulatory arrest times (DHCA) (4±8 vs. 0.8±3.6 min; p<0.0001) were all significantly longer and minimal body core temperature lower (26±6 vs. 29.7±5.5°C; p<0.0001) in Group 2 than 1. However, ICU-and hospital stay were similar in both groups (Table 4).

Postoperative mortality and morbidity: Ten patients (3.4%) died within 3±2.5 (0-7) days following surgery. Seven of these ten deaths were cardiac-related (Table 5). Early mortality was significantly higher in Group 2 than 1 (p=0.04).

Five patients (2.2%) died in Group 1 within 4±2.5 (1 – 7) days, four of them with Marfan syndrome with three patients presenting with acute type-A aortic dissection. Marfan patients (7.4%) presented with a high proportion of acute aortic dissection (Table 6). Stanford Type A (9/ 22 pts, 41%), mainly contributing to a high mortality rate of 18.2% in this subset of patients. Main reason for fatal outcome was postoperative cardiac low output and/ or Multi-organ Failure (MOF). Marfan syndrome, CPB time, use of DHCA, aortic root replacement with a composite graft and aortic surgery were significantly different in non-survivors compared to survivors of Group 1 in univariate analysis (Table 7). In Group 2, 5 patients (7.6%) died within 2.2±2.4 (0 – 6) days postoperatively. Four of these 5 early deaths were cardiac-related. Presence of cyanosis, coarctation, TGA, pulmonary atresia, operation time, cardiopulmonary bypass time, aortic cross-clamp time, core temperature and total correction were significantly different in non-survivors compared to survivors of Group 2 (Table 7).

Complication rate was significantly lower in Group 1 than 2 (26.5% vs. 42.4%; p=0.01). Especially MOF (p=0.015), cardiac low output (p=0.04), use of IABP (p=0.01), need for permanent pacemaker (p=0.04), respiratory complications (p=0.009) andrenal failure (p<0.0001) were all significantly lower in Group 1 than 2. Arrhythmias were the most frequent postoperative complications (21.3%) in both groups with total av-block being significantly more frequent in Group 2 than 1 (9.2% vs. 2.2%; p=0.04). MOF, cardiac low output, use of epinephrine, respiratory complications and stroke were significant risk factors for fatal outcome in both groups (Table 7). In Group 1 perioperative myocardial infarction and use of dobutamine andin Group 2 use of IABP, renal failure and dialysis were additional risk factors for fatal outcome.

In logistic regression analysis MOF, cardiac low output failure, DHCA and respiratory complications were identified as independent predictors for fatal outcome (Table 8).

DISCUSSION

In recent years, GUCH patients have emerged as a growing and special group of patients and their unique medical and surgical problems have lead to creation of new units providing special care necessary. Although surgery and especially redo surgery are a dominant feature of adult congenital heart surgery[1-4], not much data exist on perioperative risk factors affecting early outcome in GUCH patients.

Proportions of major cardiac pathologies comparable to our series were reported by Dore *et al.*[5] with LVOT pathology, RVOT pathology and coarctation being the largest subgroups andby the Mayo Clinic, with ASD and bicuspid aortic valves being the most frequent major cardiac diagnoses[6]. In present and other[5] series, the spectrum of pathology and the surgical procedures performed were considerably different between the groups. First operations were mainly for ASD closure, aortic valve and aortic disease. It is not surprising, because these malformations are common[7-9] and often do not cause symptoms in early life[7,8,10,11]. Furthermore, aortic disease, except coarcations, needs time to manifest itself, as aortic aneurysms may grow slowly. Reoperations were mainly for aortic valve disease, aortic disease, closure of residual septal defects and conduit procedures as in other series as well[5,6]. This spectrum of reoperations may be typical for GUCH patients, because children having had surgery for complex lesioins now survive and return with new or residual lesions or long-term complications of failed replaced valves or conduits. Typically, reoperations on the LVOT were mainly necessary in patients with congenital aortic stenosis, with two-third of these patients having had surgical aortic valve commissurotomy a mean of 16.4±5.6 years earlier in their lives andin patients with surgically closed VSD and progressive AR. Although commissurotomy may lead to reoperations of the LVOT within 10 years in up to 80% of patients[12-16], it is mostly effective to postpone valve replacement to adulthood in a significant proportion of patients[17,18]. 6.1% of all patients had coarctation, an important proportion of GUCH patients in other series as well[5,19] and was significantly more frequent in Group 2. However, recoarctation was the cause for reoperation in only 5 out of 10 patients having had coarctation as primary lesion repaired at a mean age of 9.3±6.1 (0.03 – 16) years. The other 5 had reoperations due to associated lesions like bicuspid aortic valve or dilation of the ascending aorta, or long-term hypertensive complication like Stanford A aortic dissection, reflecting the fact, that 50% of coarctations are associated with other lesions[20] and as much as 60% of corrected patients suffer from re-coarctation[20], which may not all be amenable to interventional balloon angioplasty. A similar spectrum of reoperations after repair of coarctation was found by others as well[21,22]. Patients with VSD, TOF and TGA were significantly over-represented in Group 2. TOF patients are known for their need of reoperations[23,24], mainly because of recurrent problems concerning the right

ventricular outflow tract or pulmonary arteries. In the series from Toronto[25] long-term complications of the RVOT, mainly severe pulmonary regurgitation and conduit failure lead to reoperations in 45.75% of TOF patients.

The overall early mortality of 3.4%, with 2.2% after first and 7.6% after redo surgery compares favorably to the mortality rates of 3-6.8% in published series[4,5,26-28]. Considering reoperations alone, mortality is acceptable compared to the rare data published: Isomura and co-workers had 8.6% early mortality in their study of reoperations in GUCH patients[23]. In a small series of 18 patients with reoperations, one died early postoperatively due to bleeding (5.6%)[27]. However, mortality was significantly higher after reoperations as in other published series as well[5,26], but could be significantly reduced in the last third of patients (0/22; 0%) without any death in our experience. The Toronto group has also reported a reduction in their overall postoperative mortality of 4.7% over almost three decades to 1.9% in the later half of their experience[4]. Death was mainly due to multi-organ failure and/ or cardiac low output failure. Not surprisingly, both complications were identified as independent risk factor for early death and conditions which may have a negative impact on ventricular function such as cyanosis, number of previous operations and aortic cross-clamp time were identified as risk factors for early death as well. Inadequate intraoperative myocardial preservation and longer aortic cross-clamp times may also negatively affect ventricular function, especially when repetitively applied during numerous surgical procedures. This may partly explain the significantly higher proportion of cardiac low output in Group 2 compared to 1 and the relatively high percentage of low output failure in this compared to other series of mainly first operations[5,26,27]. Myocardial management may be especially difficult in GUCH reoperations, because of systemic-to-coronary collaterals, coronary anomalies, risk for distension and incomplete de-airing of the systemic ventricle. Low output has been identified as risk factor for early mortality by others as well[5]. Interestingly, preoperative impaired ventricular function did not emerge as a risk factor for early death in uni or multivariate analysis, although 4 of the 5 deaths in Group 2 occurred in patients with preexistent impaired ventricular function. One may speculate, that systolic function is a poor predictor of the functional myocardial reserve to tolerate cardiac surgery and only partially reflects myocardial function. Some of these patients might have benefited from earlier interventions before irreversible myocardial deterioration was established, as proposed by others[29]. However, most of these patients were not thought to be suitable to repair in the eighties and inadequate follow-up in the early era of this study may have delayed timely re-interventions in these cases.

DHCA has also been identified as an independent predictor of early mortality in other subsets of patients[30]. Although most surgeons are trying to reduce or omit its use because of the potential negative effects on cerebral functions, or adjunct it with selective cerebral perfusion techniques, brief periods of DHCA of less than 40 min are considered sufficiently safe[31] and may still be necessary in aortic surgery or in cyanotics with excessive aorto-pulmonary and mediastinal collaterals. Most of the variables identified for adverse outcome in univariate analysis are known as risk factors in surgery for acquired cardiac or aortic disease as well[32-34] but cyanosis, coarctation, TGA andpulmonary atresia are specific for GUCH patients and reflect their complex pathology. Cyanosis is known to compromise postoperative outcome and has been identified as a risk factor for death by others as well[5], but good surgical results may be obtained in adult cyanotics[35].

Although ASD closure is considered very safe with low mortality[36,37], mortality was 1.4% in this subgroup with one patient dying after elective surgical ASD closure due to persistent fixed Pulmonary Arterial Hypertension (PAHT) with consecutive RV failure and another dying of intractable bleeding from LV perforation during an attempt of elective transcatheter ASD device closure. The first patient demonstrates, how poorly any surgical interventions are tolerated in fixed PAHT and Eisenmenger physiology. Mortality of the second patient may be attributed to the interventional procedure attempted in the early era of transcatheter ASD device closure[38].

Postoperative morbidity remains important in GUCH surgery with significant complications occurring in up to 30% of all patients. The complication rate was significantly higher after redo than first operations, despite significantly younger age and fewer female patients, probably reflecting the more complex pathology of reoperations, making longer operation, CPB and myocardial ischemia times and more extensive use of DHCA necessary. However, this had no impact on the duration of ICU and hospital stay. Despite the younger age, stroke rates of both groups were fairly high (4.4%) in comparison to those in contemporary adult cardiac surgery for acquired diseases[39] with much older patients presenting with more risk factors for perioperative stroke. Fortunately, two-third were fully reversible, but mortality (40%) was high in patients with severe cerebral deficits and stroke was identified as risk factor for early death in univariate analysis in both first and redo surgery. Renal and respiratory complications, also significantly more frequent after reoperations, are the main source of perioperative morbidity in other series of first and reoperations as well[26,27] and the latter have even emerged as independent predictor for early mortality in our patients.

Limitations of the study: This study has several limitations inherent to the retrospective analysis. Also, the patient population is very heterogeneous in nature, leading to small numbers in certain subgroups. Therefore, a meaningful multivariate analysis could not be performed separately for the reoperation group with only 5 deaths, but for the study population with 10 deaths. Furthermore, this study covers a long period of time, during which concepts of treatment, follow-up, surgical techniques and perioperative management have changed considerably. However, only few studies, specifically addressing perioperative risks of operations and reoperations in GUCH, have been published so far. This is even more astonishing, since operations of GUCH patients are becoming more frequent these days.

CONCLUSIONS

Main cause of surgery in adolescents and adults with congenital heart disease are ASD, outflow tract lesions, Marfan syndrome and coarctation. First operations have an excellent outcome, similar to most procedures in adult cardiac surgery, whereas re-operations have increased mortality and morbidity and particular risk factors. Deep hypothermic circulatory arrest, multi-organ failure, cardiac low output andrespiratory complications are independent predictors for early death after GUCH surgery. Close follow-up, timely referral for re-intervention and adequate perioperative management are mandatory to reduce perioperative risks and improve results in this demanding group of patients.

REFERENCES

1. Thilen, U., 2001. Adults with congenital heart defects: A growing patient group. Lakartidningen, 98: 656-60.
2. Webb, GD., 2001. Care of adults with congenital heart disease: A challenge for the new millennium. Thorac. Cardiovasc. Surg., 49: 30-4.
3. Somerville, J., 1997. Management of adults with congenital heart disease: An increasing problem. Ann. Rev. Med., 48: 283-93.
4. Williams, W. and G. Webb, 2000 . The emerging adult population with congenital heart disease. Pediatric Cardiac Surgery Annual of the Seminars in Thoracic and Cardiovascular Surgery, 3: 227-233.
5. Dore, A., D.L. Glancy, S. Stone, VD. Menashe and J. Somerville, 1997. Cardiac surgery for grown-up congenital heart patients: Survey of 307 consecutive operations from 1991 to 1994. Am. J. Cardiol,. 80: 906-13.
6. Warnes, C.A., R. Liberthson, G.K. Danielson, A. Dore, L. Harris and J.I. Hoffman *et al.*, 2001. Task force 1: The changing profile of congenital heart disease in adult life. J. Am. Coll. Cardiol., 37: 1170-5.
7. Hoffmann, J., 1987. Incidence, Mortality and Natural History. In: R. Anderson, F. Macartney, E. Shinebourne, M. Tynan (Eds.). Pediatric Cardiology. Edinburgh: Churchill Livingstone, pp: 4-14.
8. Kitchiner, D., M. Jackson, K. Walsh, I. Pear and R. Arnold, 1993. The progression of mild congenital aortic valve stenosis from childhood into adult life. Intl. J. Cardiol., 42: 217-23.
9. Roberts, W.C., 1970. The structure of the aortic valve in clinically isolated aortic stenosis: An autopsy study of 162 patients over 15 years of age. Circulation, 42: 91-7.
10. Glancy, D.L., AG. Morrow, A.L. Simon and W.C. Roberts, 1983. Juxtaductal aortic coarctation. Analysis of 84 patients studied hemodynamically, angiographically andmorphologically after age 1 year. Am. J. Cardiol., 51: 537-51.
11. Horvath, K.A., R.P. Burke, J.J. Collins and L.H. Cohn, 1992. Surgical treatment of adult atrial septal defect: Early and long-term results. J. Am. Coll. Cardiol., 20: 1156-9.
12. Champsaur, G., J. Ninet, J.P. Frieh, A. Bozio, F. Sassolas and M. El-Kirat *et al.*, 1985. Congenital aortic valve stenosis. Long-term results after valvulotomy. Arch. Mal. Coeur. Vaiss., 78: 220-4.
13. Keane, J.F., D.J. Driscoll, W.M. Gersony, C.J. Hayes, L. Kidd and W.M. O'Fallon *et al.*, 1993. Second natural history study of congenital heart defects. Results of treatment of patients with aortic valvar stenosis. Circulation, 87: I16-27.
14. Kugelmeier, J., L. Egloff , F. Real , M. Rothlin and M. Turina, 1982. Senning A. Congenital aortic stenosis. Early and late results of aortic valvulotomy. Thorac. Cardiovasc. Surg., 30: 91-5.
15. Presbiteo, P., J. Somerville, R. Revel-Chion and D. Ross, 1982. Open aortic valvotomy for congenital aortic stenosis. Late results. Br. Heart J.,47: 26-34.
16. Tveter, K.J., J.E. Foker, J.H. Moller, W.S. Ring, C.W. Lillehei and R.L. Varco, 1987. Long-term evaluation of aortic valvotomy for congenital aortic stenosis. Ann. Surg., 206: 496-503.
17. Haydar, H.S., G.W. He, H. Hovaguimian, D.M. McIrvin, D.H. King and A. Starr, 1997. Valve repair for aortic insufficiency: surgical classification and techniques. Eur. J. Cardio. thorac. Surg., 11: 258-65.

18. Chartrand, C., E. Saro-Servando and J.S. Vobecky, 1999. Aortic valve stenosis in children. Surgical valvuloplasty long-term results. Ann. Chir., 53: 717-22.
19. Somerville, J., 2001. Grown-up congenital heart disease: Medical demands look back, look forward 2000. Thorac. Cardiovasc. Surg., 49: 21-6.
20. Amato, J.J., W.I. Douglas, T. James and U. Desai, 2000. Coarctation of the aorta. Semin. Thorac. Cardiovasc. Surg. Pediatr. Card. Surg. Ann., 3: 125-141.
21. Stewart, A.B., R. Ahmed, C.M. Travill and C.G. Newman, 1993. Coarctation of the aorta life and health 20-44 years after surgical repair. Br. Heart. J., 69: 65-70.
22. Jost, C.H., H.V. Schaff, H.M. Connolly, G.K. Danielson, F.J. Dearani and Pug *et al.*, 2002. Spectrum of reoperations after repair of aortic coarctation: Importance of an individualized approach because of coexistent cardiovascular disease. Mayo. Clin. Proc., 77: 646-53.
23. Isomura, T., K. Hisatomi, F. Andoh, T. Kawara, A. Hiran and K. Kosuga *et al.*, 1991. Reoperation following total repair of congenital heart disease. Jap. Circ. J., 55: 453-8.
24. Sohn, S. and Y.T. Lee, 2000. Outcome of adults with repaired tetralogy of Fallot. J. Korean. Med. Sci., 15: 37-43.
25. Oechslin, E.N., D.A. Harrison, L. Harris, E. Downar, G.D. Webb and S.S. Siu *et al.*, 1999. Reoperation in adults with repair of tetralogy of fallot: Indications and outcomes. J. Thorac. Cardiovas. Surg., 118: 245-51.
26. Geissler, H.J., M. Sudkamp, J. Nowak and E.R. de Vivie, 1996. Congenital heart defects in adolescence and adulthood: Fatalities and morbidity in primary and reoperation. Z. Kardiol., 85: 782-9.
27. Berdjis, F., I.D. Brand, F. Uhlemann, G. Hausdorf, L. Lange and Y. Weng *et al*, 1996. Adults with congenital heart defects: Clinical spectrum and surgical management. Herz, 21: 330-6.
28. Pass, H.I., F.A. Crawford, R.M. Sade, M.E. Assey and B.W. Usher, 1984. Congenital heart disease in adults. Am. Surg., 50: 36-9.
29. Therrien, J., S.C. Siu, P.R. McLaughlin, P.P. Liu, W.G. Williams and G.D. Webb, 2000. Pulmonary valve replacement in adults late after repair of tetralogy of fallot: Are we operating too late? J. Am. Coll. Cardiol., 36: 1670-5.
30. Yagdi, T., Y. Atay, M. Cikirikcioglu,M. Boga, H. Posacioglu and M. Ozbaran *et al*, 2000. Determinants of early mortality and neurological morbidity in aortic operations performed under circulatory arrest. J. Cardiol. Surg., 15: 186-93.
31. Ergin, M.A., E.B. Griepp, S.L. Lansman, J.D. Galla, M. Levy and R.B. Griepp, 1994 . Hypothermic circulatory arrest and other methods of cerebral protection during operations on the thoracic aorta. J. Cardiol. Surg., 9: 525-37.
32. Roques, F., S.A. Nashef, P. Michel, E. Gauducheau, C. De Vincentiis and E. Baudet *et al.*, 1999. Risk factors and outcome in European cardiac surgery: Analysis of the EuroSCORE multinational database of 19030 patients. Eur. J. Cardiothorac. Surg., 15: 816-22; discussion 822-3.
33. Coselli, J.S., S.A. LeMaire, C.C. Miller, Z.C. Schmittling, C. Koksoy , J. Pagan *et al.*, 2000. Mortality and paraplegia after thoracoabdominal aortic aneurysm repair: A risk factor analysis. Ann. Thorac. Surg., 69: 409-14.
34. Calafiore, A.M., M.D. Mauro, G. Teodori, G.D. Giammarco, S. Cirmeni, M. Contini *et al.*, 2002. Impact of aortic manipulation on incidence of cerebrovascular accidents after surgical myocardial revascularization. Ann. Thorac. Surg., 73: 1387-93.
35. Mohanty, S.R., B. Airan, A. Bhan, R. Sharma, A.S. Kumar, S.S. Kothari *et al.*, 1999. Adult cyanotic congenital heart disease: Surgical experience. Ind. Heart J., 51:186-92.
36. Galal, M.O., A. Wobst, Z. Halees, L. Hatle, A.A. Schmalt and F. Khougeer *et al.*, 1994. Peri-operative complications following surgical closure of atrial septal defect type II in 232 patients: A base line study. Eur. Heart J., 15: 1381-4.
37. Byrne, J.G., D.H. Adams, M.E. Mitchell and L.H. Cohn, 1999. Minimally invasive direct access for repair of atrial septal defect in adults. Am. J. Cardiol., 84: 919-22.
38. Berdat, P., T. Chatterje, J. Pfammatter, S. Windecker, B. Meier and T. Carrel , 2000. Surgical management of complications after transcatheter closure of an atrial septal defect or patent foramen ovale. J. Thorac. Cardiovasc. Surg., 120: 1034-9.
39. Bucerius, J., J. Gummert, M. Borger, T. Walther, N. Doll and J. Onnasch *et dl.*, 2003. Stroke after Cardiac Surgery: A Risk Factor Analysis of 16184 Consecutive Adult Patients. Ann. Thorac. Surg., 75: 472-8.