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Original Article

Cardiac surgery in adults with high-surgical complexity CHD: results of a network collaborative programme

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Abstract *Background:* Adults with CHD often exhibit complex cardiac abnormalities, whose management requires specific clinical and surgical expertise. To enable easier access of these patients to highly specialised care, we implemented a collaborative programme that incorporates medical and surgical specialists belonging to both paediatric and adult cardiovascular institutions. *Objectives:* The objective of this study was to review the experience gained and to analyse the surgical outcome of major cardiac surgery. *Methods:* We retrospectively reviewed all consecutive patients admitted for major cardiac surgery using our network between January, 2010 and December, 2013. Analysis of surgical outcome was performed in patients selected for major cardiac surgery with cardiopulmonary bypass. Early and late outcomes were evaluated. *Results:* Out of a total of 433 inward patients, 86 were selected for surgery. The median age was 25.5 years, –64 patients (74.4%) had previously undergone heart surgery, and –55 patients (64%) had been subjected to at least one sternotomy. Abnormalities of the left ventricular and right ventricular outflow tract were the most frequent (37.2% and 30.2%, respectively), and despite high-surgical complexity only one death occurred (in-hospital mortality 1.1%). On a median follow-up time of 4 years no deaths and no heart-failure events have occurred; one patient underwent further cardiac surgery programmed at the time of discharge. *Conclusions:* Low mortality and morbidity rates can be obtained in high-surgical complexity adults with CHD populations when paediatric and adult cardiac specialists operate in the same multidisciplinary environment.

Keywords: Adult; CHD; cardiac surgery; organisation model

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The estimated incidence of CHD ranges between 6 and 19 cases per 1000 live births[1,2] and surgical mortality rates that in patients with more complex abnormalities exceeded 50% in the fifties, have now been reduced to less than 15%. The significant increase in survival has produced a shift in the age distribution of the CHD population and adult patients now outnumber those in the paediatric age.[3–6]

Although cardiac surgery provides excellent results, very seldom does it allow full repair of complex CHD, and residual abnormalities or late complications are frequent. In fact, re-do operations are required in about 20–25% of patients with CHD,[7] owing to persistence of residual defects after partial repair, conduits degeneration and outgrowth, or native or prosthetic valve dysfunction.

As the management of adult patients with complex congenital cardiac abnormalities requires specific expertise and organisation, scientific associations worldwide have issued dedicated guidelines and recommendations emphasising the need for highly specialised centres and networks in which paediatric and adult cardiac specialists with specific experience in CHD can work together.[8–10] Although surgical mortality rates are reasonably low and range from 2 to 5% whenever such

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conditions are met,[7,11–13] the clinical and surgical expertise that are necessary to manage these patients are rarely found in general cardiology units and, often, even in tertiary referral centres.[10,13]

In 2010 we started a collaborative programme that was intended to team-up medical and surgical expertise, from both paediatric and adult cardiovascular centres, and aimed at promoting adults with CHD patients' access to highly specialised care and at optimising the quality of management and intervention. We report here the experience we gained over the first 4 years of activity in adult patients with complex CHD undergoing open-heart surgery.

Patients and methods

Patient population

We retrospectively reviewed all consecutive patients, aged 14 years and older, who underwent surgery for complex CHD between January, 2010 and December, 2013 at the "Istituto Clinico Ligure di Alta Specialita" (ICLAS), Rapallo, Genoa. The patients were part of a larger cohort recruited within the framework of a collaborative programme that was fostered by the Liguria Regional Government and implemented in 2010. The programme, which involved ICLAS, the Paediatric Institute G. Gaslini (Genoa), and the Genoa Local Health Authority, gathered medical and surgical expertise, from both paediatric and adult cardiovascular institutions. Recruitment and first-level clinical characterisation were conducted at ICLAS, in the Gaslini Institute, and in community outpatient clinics, whereas cardiac invasive and surgical procedures were all performed at ICLAS by a team of paediatric and adult specialists. Specifically, all open-heart cardiac surgeries were carried out by surgeons belonging to the Gaslini Institute (F.S.,E.R.) and experienced in both paediatric and adult cardiac surgery.

All information, including demographics, medical history, clinical, ECG, blood chemistry, echocardiography, magnetic resonance, cardiac catheterisation, and surgical data were taken from the patient's medical records, which had been previously made anonymous. Procedural and periprocedural data included the type of surgery performed, cardiopulmonary bypass, aortic cross-clamping, and ventilation times, as well as the duration of ICU and hospital stay. Surgeries were grouped into homogeneous categories based on the main intervention and analysed accordingly.

Assessment of surgical risk

Surgical risk was evaluated according to the Aristotle basic and comprehensive score systems.[14] Both scores have been previously validated in children[14,15] and have shown to predict postoperative mortality and morbidity[16,17] with a C-Index[18] of 0.70 and 0.860, respectively. These scores, moreover, were used successfully in preliminary reports conducted in adults with CHD[19,20] and shown to be better predictors of surgical outcome than other paediatric scores.

Outcomes

The 30-day composite of major adverse events was selected as the primary endpoint and included in-hospital mortality, acute renal failure, neurological events, major bleeding, major arrhythmias, need for permanent pacemaker implant, pleural effusion requiring thoracentesis, moderate to severe pericardial effusion, pneumothorax requiring drainage, as well as prolonged ventilation and ICU stay. Bleeding was defined as major when requiring 5 blood units or more, or when necessitating surgical revision. Renal failure was defined as a postoperative serum creatinine level greater than twice the preoperative level. Ventilation time and ICU stay were defined as prolonged when lasting for more than 24 and 72 hours, respectively. Late outcome including composite of overall mortality, further cardiac surgery, and heart failure requiring hospital admission over a time period of a minimum of 36 months was selected as the secondary endpoint.

Statistical analysis

Data are described as absolute and relative frequencies for categorical variables, whereas means, standard deviation, medians, and range are used for continuous variables. Categorical data were compared using the χ^2 test, or using Fisher's exact test when frequencies were expected to be lower than 5%. Comparisons of between-group quantitative variables were performed by means of two-tailed Student's t tests. A p-value of less than 0.05 was considered statistically significant. Univariate analysis was carried out to determine which of the potential risk factors were significantly associated with the risk for major adverse events. Logistic regression analyses were used and the results are reported as odds ratio with their 95% confidence intervals. The absence of exposure to the factor or the variable that was less likely to be associated with the outcome was used as the reference for each analysis. Multivariate logistic regression analysis was then performed, and only variables that proved to be statistically or borderline significant in univariate analysis were included in the model. In this case, a p value < 0.06 was used as the cut-off point. The model showing the best fit was based on backward stepwise selection procedures, and each variable was removed if it did not contribute significantly. In the final model a p-value < 0.05 was considered statistically significant, and all p-values were based on two-tailed tests. Statistical analysis was performed using SPSS for Windows (SPSS Inc., Chicago, Illinois, United States of America).

Results

Patients' characteristics

Out of a total of 433 inward patients between January, 2010 and December, 2013, 86 were selected for cardiac surgery with cardiopulmonary bypass. Of the remaining 347 patients, 273 were kept on medical follow-up observation and 74 underwent interventional procedures: 44 percutaneous closures of patent foramen ovale, 23 percutaneous secundum atrial septal defect closure, two permanent pacing implants, and five arrhythmia ablations.

Demographics, preoperative, and operative characteristics of the surgical cohort are summarised in Table 1. A total of 49 patients (57%) were male and 37 (43%) were female: the median age was 25.5 years (with a range from 14 to 65 years) and the median body mass index was 23.5 (with a range from 15.6 to 37.6). The majority of patients [n = 64 (74.4%)] had previously undergone heart surgery – via sternotomy, thoracotomy, or percutaneously – whereas the remaining 22 (25.6%) underwent the first cardiac operation. In all, 55 patients (64%) had undergone at least one sternotomy and 15 (17.4%) had undergone two or more.

Abnormalities of the left ventricular outflow tract were the most frequent alterations in 32 patients (37.2%) and included subvalvular, valvular, and supravalvular diseases such as aortic stenosis, bicuspid aortic valve, disease of the aortic root or ascending aorta, and conduit and autograft degradation. This group included two patients with severe subvalvular obstruction and annular hypoplasia undergoing the Konno operation and one patient who had replacement of a left ventricle–pulmonary artery conduit (Table 2).

The second most common group of interventions involved the right ventricular outflow tract in 26 patients (30.2%) and included pulmonary valve and conduit replacement. Both interventions were frequently associated with other procedures including infundibular aneurysmectomy, nine patients, removal of outflow tract obstruction, three patients, pulmonary artery enlargement, five patients, and tricuspid valvuloplasty, three patients (Table 2).

Of the remaining patients, 10 (11.6%) underwent atrioventricular valve replacement or repair, eight (9%) atrial septal defect closure, five (5.8%) bivalvular procedures, and five had other interventions including ascending–descending aortic bypass, aortic coarctation repair, and correction of anomalous pulmonary venous return (Table 1).

Surgical complexity, assessed by the Aristotle score system, was rather high and generally greater than that reported in published registries.[14] The mean Aristotle basic and comprehensive scores were 7.1 ± 1.7 and 9.6 ± 2.9, respectively (Table 1), with 48 patients (55.8%) having Aristotle comprehensive scores higher than 10. Operative performance (complexity score × hospital survival) was 7.01 and 9.48 for Aristotle basic and comprehensive scores, respectively.

Table 1. Patient's clinical and operative characteristics.

Male	n (%)	49 (57%)
Female	n (%)	37 (43%)
Age (years)	Mean ± SD	29.5 ± 13.3
	Median (range)	25.5 (14, 65)
BMI	Mean ± SD	23.7 ± 4.9
	Median (range)	23.5 (15.6, 37.6)
Previous cardiac surgery	n (%)	64 (74.4%)
Previous sternotomy		
No	n (%)	31 (36%)
1	n (%)	40 (46.5%)
⩾2	n (%)	15 (17.4%)
Genetic syndromes	n (%)	5 (5.8%)
Pulmonary hypertension	n (%)	5 (5.8%)
LV myocardial dysfunction	n (%)	5 (5.8%)
RV myocardial dysfunction	n (%)	21 (24.4%)
Arrhythmias	n (%)	7 (8.1%)
Haemoglobin (g/dl)	Mean ± SD	13.9 ± 1.4
ABC score	Mean ± SD	7.1 ± 1.7
	Median (range)	7 (3, 11)
ACC score	Mean ± SD	9.6 ± 2.9
	Median (range)	10.5 (3, 16.3)
Surgery type		
LVOT	n (%)	32 (37.2%)
RVOT	n (%)	26 (30.2%)
Atrioventricular valve	n (%)	10 (11.6%)
ASD	n (%)	8 (9.3%)
Bivalvular	n (%)	5 (5.8%)
Other	n (%)	5 (5.8%)
CPB time (minutes)	Mean ± SD	89 ± 48.3
	Median (range)	80 (21, 325)
ICU stay (hours)	Mean ± SD	39.4 ± 30.7
	Median (range)	35.5 (5, 190)
Postoperative hospital stay (days)	Mean ± SD	6.6 ± 3.48
	Median (range)	6 (3, 22)

ABC = Aristotle Basic score; ACC = Aristotle comprehensive score; ASD = atrial septal defect; BMI = body mass index; CPB = cardiopulmonary bypass; LV = left ventricle; LVOT = left ventricular outflow tract; RV = right ventricle; RVOT = right ventricular outflow tract.

Major adverse events

Major adverse events are listed in Table 3. The only hospital death occurred in a patient with Marfan disease and pulmonary atresia with an intact interventricular septum who had been previously operated upon using the one-and-a-half ventricular repair technique. He had subsequently developed aortic root dilatation and severe aortic valve regurgitation requiring re-intervention using the Bentall

Table 2. Primary cardiac defect and current surgical procedure (left ventricle outflow tract (LVOT) and right ventricle outflow tract (RVOT) groups).

Primary cardiac defect LVOT	n	Current surgical procedure LVOT	n
BAV	8	AVR	7
BAV + CoA	3	AVR + ascending aortic replacement	6
BAV + VSD	1	Bentall	4
Aortic valve dysplasia	3	AVR + aortic root reduction plasty	2
Aortic valve dysplasia + VSD	1	SubAS resection	3
SubAS	2	SubAS resection + Bentall	1
SubAS + Aortic valve dysplasia	4	SubAS resection + AVR + ascending aortic replacement	1
SubAS + CoA	1	Konno + AVR	2
TGA + VSD	1	LV–AO conduit replacement	1
TGA + VSD + CoA	1	LV–PA conduit replacement + PA enlargement	1
ccTGA	1	Bentall + right atrial reduction plasty	1
ccTGA + PS + VSD	1	Paraprosthetic leak suture	1
Pulmonary atresia with IVS	1	Ascending aortic replacement	1
Pulmonary atresia + VSD + MAPCAs	1	Aortic arch replacement	1
DORV + TGA + Pulmonary atresia	1		
CoA	1		
VSD	1		
Total	32	Total	32
RVOT		**RVOT**	
ToF	14	PVR	5
ToF with pulmonary atresia	1	PVR + infundibular aneurysmectomy	6
ToF absent pulmonary valve	2	PVR + infundibular aneurysmectomy + flutter ablation	1
ToF + tricuspid dysplasia	1	PVR + infundibular aneurysmectomy + PA enlargement	2
ToF absent left pulmonary artery	1	PVR + RVOT enlargement	1
ToF + atrioventricular canal	1	PVR + RVOT enlargement + VSD closure	2
AS	2	PVR + tricuspid valvuloplasty	2
SubAS + AS	1	PVR + tricuspid valvuloplasty + PA enlargement	1
Infundibular stenosis	2	RV-PA conduit replacement	4
PS	1	RV-PA conduit replacement + PA enlargement	2
Total	26	Total	26

AS = aortic stenosis; AVR = aortic valve replacement; BAV = bicuspid aortic valve; ccTGA = congenitally corrected transposition of the great arteries; CoA = coarctation of the aorta; DORV = double-outlet right ventricle; IVS = intact ventricular septal; LV-AO = left ventricle to aorta; MAPCAs = major aortic pulmonary collaterals; PA = pulmonary artery; PS = pulmonary stenosis; PVR = pulmonary valve replacement; SubAS = sub-aortic stenosis; TGA = transposition of the great arteries; ToF = Tetralogy of Fallot; VSD = ventricular septal defect

procedure. His postoperative course was complicated by uncontrollable bleeding due to severe deficit of coagulation factors. Major bleeding occurred in seven patients and required surgical revision in four. Overall, four patients developed supraventricular or ventricular tachyarrhythmia and two developed complete atrioventricular block requiring pacemaker implantation.

On univariate analysis the only factor significantly associated with major adverse events was haematocrit (42.8 ± 4% versus 40.8 ± 4%; p 0.05). No statistically significant associations were found on multivariate analysis (Table 4).

There was no apparent relation between surgical complexity as assessed by the Aristotle comprehensive score and incidence of major adverse events. In fact, in the low- (Aristotle comprehensive 1.5–5.9) and high- (Aristotle comprehensive 10–14.9) risk patients the incidence of major adverse events was, respectively, 25 and 27%.

Table 3. Major adverse events.

Adverse events	No.	%
In-hospital mortality	1	1.1
ICU stay >72 hours	6	6.9
Prolonged ventilation (>24 hours)	2	2.3
Major bleeding*	7	8.1
Major tachyarrhythmia	4	4.6
PM implant for advanced AV block	2	2.3
Acute renal failure	1	1.1
Thoracentesis	4	4.6
Pneumothorax	2	2.3
Pericardial effusion (⩾moderate)	2	2.3
TIA	1	1.1

AV = atrioventricular; PM = pacemaker; TIA = transient ischaemic attack

*Bleeding requiring surgical revision and/or >5 red blood cell units transfusion

Follow-up evaluation

After discharge, all surviving patients (85 patients) were evaluated by our team either in the outpatient clinic or

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Table 4. Factors associated with major adverse events (MAE).

Patients characteristics		No MAE (n = 66)	MAE (n = 20)	p value
Sex				0.52
Male	n (%)	38 (57.6%)	11 (55%)	
Female	n (%)	28 (42.4%)	9 (45%)	
Age (years)	Mean ± SD	29 ± 13.7	31.2 ± 12.2	0.52
	(Range)	(14, 65)	(16, 59)	
BMI	Mean ± SD	23.8 ± 5.4	23.1 ± 5.6	0.88
	(Range)	(15.6, 37.6)	(16, 29.9)	
Pulmonary hypertension	n (%)	4 (6.1%)	1 (5%)	1
LV myocardial dysfunction	n (%)	4 (6.1%)	1 (5%)	1
RV myocardial dysfunction	n (%)	14 (21.2%)	7 (35%)	0.24
Haemoglobin (g/dl)	Mean ± SD	13.7 ± 1.4	14.4 ± 1.3	0.06
	(Range)	(9.3, 16.3)	(12.1, 16.3)	
Haematocrit (%)	Mean ± SD	40.8 ± 4	42.8 ± 4	**0.05**
	(Range)	(28.6, 48.5)	(37.2, 49.7)	
Previous surgery	n (%)	47 (71.2%)	17 (85%)	0.25
Previous sternotomy	n (%)	39(59.1%)	16 (80%)	0.21
ABC score	Mean ± SD	7.1 ± 1.7	7.1 ± 1.4	0.96
ACC score	Mean ± SD	9.4 ± 2.8	10.2 ± 3.2	0.28
CPB time (minutes)	Mean ± SD	84.6 ± 45	103.3 ± 54	0.13
	(Range)	(39, 214)	(21, 325)	

ABC = Aristotle Basic score; ACC = Aristotle comprehensive score; BMI = body mass index; CPB = cardiopulmonary bypass; LV = left ventricle; RV = right ventricle

Bold value indicates statistically significant result

using telephone interview. Throughout a median follow-up period of 4 years (with a range from 3 to 5 years), no deaths and no heart failure requiring hospital admission have occurred. A single patient with Fallot tetralogy and atrioventricular canal who had undergone ventricular septal closure and mitral cleft repair during the study period, underwent pulmonary valve replacement that had been programmed at time of discharge.

Discussion

The present study reports the initial experience we obtained in a group of adult patients with high-surgical complexity CHD who underwent open-heart surgery. In agreement with previous investigations[7,8,11,12,21] our data confirm the extreme clinical and anatomical heterogeneity of this population that most often requires highly complex cardiac operations. In fact, over 75% of our patients had an Aristotle comprehensive score ⩾ 8, 74% had re-do surgery, and 64% had been previously undergone one (49.5%), two, or more (17.4%) sternotomies.

Despite the surgical complexity of our population, only one death occurred. This translates into a 1.1% rate, which compares favourably with the 2–5% mortality rates reported previously.[7,11–13] Undoubtedly, the relatively small size of our cohort does not allow to draw any definite conclusion on the quality of our surgical programme; however, in our patients, operative performance – that is, the product of the Aristotle basic and comprehensive scores times hospital survival – was 7.01 and 9.48, respectively, whereas previous series reported operative performances of 5.5 and 6.9.[14] Further, since the completion of this report, the number of cardiac surgeries conducted in our institution has increased considerably – 147 further cases from January, 2014 to June, 2016 – the case mix and mean surgical risk have remained practically unchanged, and no further deaths have been recorded. The excellent results observed during hospital admission in the study cohort, moreover, were confirmed during long-term follow-up evaluation, when no further deaths, nor hospital admission for heart failure or unplanned cardiac operations were recorded.

In our series, major adverse events were rather infrequent. This prevented us from obtaining meaningful correlations among different predictors of surgical risk and operative outcome. In fact, of the different factors assessed by logistic regression analysis, only greater haematocrit levels were significantly associated with major adverse events, confirming that cyanotic malformations are associated with increased morbidity in adult congenital cardiac surgery.[11]

In agreement with previous reports[11,22] most of our patients underwent surgery of the left and right ventricular outflow tracts, as residual abnormalities of these structures are extremely common in previously corrected, complex CHD. The majority of patients with left ventricular outflow tract abnormalities required combined valvular and aortic surgery, whereas correction of pulmonary valve incompetence and removal of recurrent obstruction were predominant in those with alterations of the right ventricular outflow tract. Indeed, significant pulmonary valve regurgitation is extremely frequent after primary correction of Fallot tetralogy and earlier correction appears to prevent irreversible right ventricular dysfunction as well as the occurrence of atrial and ventricular tachyarrhythmia.[23,24] Optimal surgical timing remains uncertain, although accepted criteria are emerging.[25]

As discussed in previous sections, the vast majority of patients with complex congenital malformations usually undergo primary cardiac repair in the paediatric age, but frequently require further surgery at a later stage. As patients' survival after primary surgery has considerably improved, a growing proportion of patients surviving the initial operation are left without a qualified clinical reference once they are past the paediatric age. In fact, the management of their condition, typical of the paediatric environment, requires specific expertise and skills, which are difficult to find in most adult cardiology units. In an attempt to provide a viable, effective solution to this ever-growing problem, we implemented a collaborative programme that allowed paediatric and adult cardiovascular specialists, belonging to different health organisations, to operate in the same adult cardiology institution (ICLAS). In keeping with guidelines recommendations, our surgical programme was conducted in a highly specialised, multidisciplinary environment where surgeries were performed by paediatric cardiac surgeons,[26] whereas anaesthesia, as well as ICU and postoperative care were carried out by a team of adult and paediatric cardiovascular specialists. This allowed to provide an efficient response fulfilling the needs of the local patients' population and to achieve low mortality–morbidity rates in a cohort of highly complex, high-risk patients.

Conclusions

Our programme fulfils the indications of the current Guidelines that recommend that surgical treatment of patients with complex congenital cardiac abnormalities should be conducted in highly specialised centres where paediatric and adult cardiac specialists operate synergistically.

The collaboration modality implemented in our programme involved different institutions, each contributing at different intervention levels with specific expertise. It appears to be suited for this patient population that, once past the paediatric age, is often left without a qualified clinical reference. It may provide an organisation model for those cardiology units that, although devoid of paediatric expertise, still serve a large catchment area.

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Conflicts of Interest

None.

Ethical Standards

The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation (GVM institutional committes) and with the Helsinki Declaration of 1975, as revised in 2008, and has been approved by the GVM institutional committees.

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