Clinical Research

Successful Surgical Correction of Congenital Heart Disease in Adults: Seven Years' Experience

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Congenital heart disease, adults, surgical treatment.

Introduction: The long term consequences of untreated or residual/recurrent lesions pose unique challenges in the growing population of adults with congenital heart disease (CHD). This study summarizes the results of the surgical treatment of these patients.

Methods: From October 1997 to October 2004, 289 patients with CHD, aged 18-72 (35 ± 13.6) years, 143 men and 146 women, presented for surgery to our unit. Thirty eight patients (13.15%) had 1 to 3 prior surgical procedures. Although 137 patients (47%) were asymptomatic (NYHA I), 117 (40.5%) had mild (NYHA II), 34 (11.7%) moderate (NYHA III) and 1 (0.3%) severe (NYHA IV) symptoms. Thirty four patients had an established arrhythmia, requiring treatment in 25 (8.6%). Chromosomal anomalies were identified in 10 (3.4%) and diagnostic catheterization was required in 178 (61.5%) patients. All patients underwent complete surgical correction and in 275 (95%) of them this was performed with cardiopulmonary bypass of 107 ± 74 min mean duration.

Results: There was 1 early death (0.34%) due to embolic stroke related to atrial fibrillation (AF). Complications occurred in 50 patients (17%) and included re-operation for bleeding (5), stroke (3), pneumothorax (12), AF (22), complete heart block requiring permanent pacemaker implantation (2), wound dehiscence (1), pericardial (7) or pleural (3) effusion requiring drainage and peripheral neuropathy (1). Median intensive care unit and hospital stay was 1 (range 1-10) and 8 (range 5-42) days respectively. Two late deaths (0.7%) occurred in patients with AF and pulmonary hypertension. At mean follow-up of 45 ± 24 (range 1-82) months all other patients are well with resolution or significant improvement in their symptoms.

Conclusion: Despite the long term deleterious effects of CHD in adult patients, surgical correction can be achieved with low mortality and acceptable morbidity. All deaths and most significant complications are related to arrhythmias.

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Despite the early recognition and, by and large, surgical treatment of congenital heart disease (CHD) in neonates, infants and children, there is a significant number of patients who reach adulthood without having undergone surgical or other correction, for various reasons. In addition, the development and improvement of surgical techniques, along with the accumulated experience in the treatment of CHD, have significantly reduced the surgical and overall mortality of CHD patients and improved their long term survival. This has resulted in the development of a population of adults with corrected or uncorrected CHD that is significant in size.[1] The variety of cardiac anomalies, the presence of residual or recurrent lesions from prior therapeutic interventions and the particularity of each individual case pose unique challenges

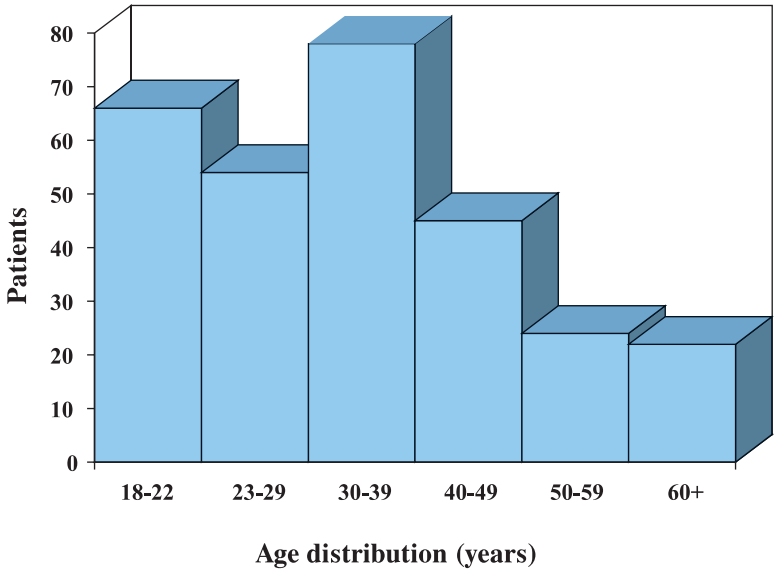

Figure 1. Age distribution of patients.

in the monitoring and management of this growing population of adults with CHD.[2]

Herein we present and analyze the results of the surgical treatment of adult CHD patients in our department.

Material and methods

During the seven-year period from October 1997 to September 2004, 289 patients with CHD, aged 18-72 years (mean 35 ± 13.6, Figure 1), 143 men and 146 women, mean weight 71 ± 16 kg and body surface area 1.78 ± 0.23 m^2, presented to our department for surgical treatment (Table 1). Chromosomal abnormalities existed in 10 patients (3.4%, Table 2). One hundred fifty two patients (53%) were symptomatic (Figure 2). Diagnosis was reached by a combination of echocardiography and cardiac catheterization with coronary angiography in 178 cases (61.5%), while in the remainder echocardiography alone proved sufficient. Most patients presented for the first time, while 38 (13.15%) had undergone one or more prior operations in the past for the same lesion (Figure 3). Thirty four patients had an established rhythm disorder and some form of treatment was required in 25 of them (8.6%, Table 3). All patients underwent complete surgical correction, which in several cases included multiple surgical procedures (Table 4). In 275 patients (95%) surgery was performed under cardiopulmonary bypass of 107±74 minutes mean duration.

Results

There was one early death (0.34%) caused by embolic stroke in a patient with chronic atrial fibrillation (AF). A total of 55 complications occurred in 50 patients (17%) who, apart from the case mentioned above, were treated successfully with no long term consequences (Figure 4). The median duration of mechanical ventilation was 8 hours, while the median intensive care unit and hospital stays were 1 and 8 days, respectively. During a postoperative follow up period of 1-82 (44.5 ± 24) months, two late deaths occurred (0.7%) in patients with AF and pulmonary hypertension. All the remaining patients are in excellent clinical condition with elimination of their symptoms or significant improvement (Figure 2).

Discussion

"Pediatric Congenital Cardiac Becomes a Postoperative Adult: The Changing Population of Congenital Heart Disease." This article was written 30 years ago by Joseph Perloff, well known authority in adult congenital heart disease, in order to stress the impending rapid increase in the number of patients with corrected CHD reaching adulthood.[3] In addition, a large number of patients with CHD reach adulthood without having undergone surgical repair. All this has led to the development of a sizeable population of adult patients with repaired or untreated heart lesions. In a clinical study of 1000 CHD patients treated surgically or medically in the 1950s, actuarial survival at 40 years was 72%.[4] Currently, it is estimated that around 85% of children who are born with CHD and undergo surgical correction will survive at least until adulthood.[1]

A large number of adults with CHD present either for primary surgical repair or for the surgical management of residual, recurrent lesions or expected degen-

Table 1. Diagnoses.

Diagnoses					Patients
Septal defects	Atrial septal defect	Secundum		127	**193**
		Sinus venosus with partial anomalous pulmonary venous return		25	
		Coronary sinus		1	
	Ventricular septal defect	Simple		16	
		With double outlet right ventricle		2	
	Partial atrioventricular canal			22	
Anomalous pulmonary venous return	Partial anomalous right upper venous return			1	**2**
	Partial anomalous right lower venous return (Scimitar)			1	
Left heart lesions	Aortic valve	Stenosis	Subvalvular	16	**47**
			Valvular	6	
			Supravalvular	2	
		Insufficiency		11	
		Mixed		4	
		Aneurysm of Valsalva sinus		1	
	Mitral valve (insufficiency)			7	
Right heart lesions	Tetralogy of Fallot			12	**20**
	Tricuspid valve			4	
	Pulmonary valve			4	
Single ventricle					**3**
Thoracic arteries and veins	Aorta	Coarctation of the aorta		16	**23**
		Hypoplastic aortic arch		1	
	Ductus arteriosus			3	
	Coronary arteries	Arteriovenous fistula		1	
		ALCAPA		1	
	Vascular ring			1	
Complex heart disease					**1**
Total					**289**

ALCAPA: Anomalous left coronary artery from pulmonary artery

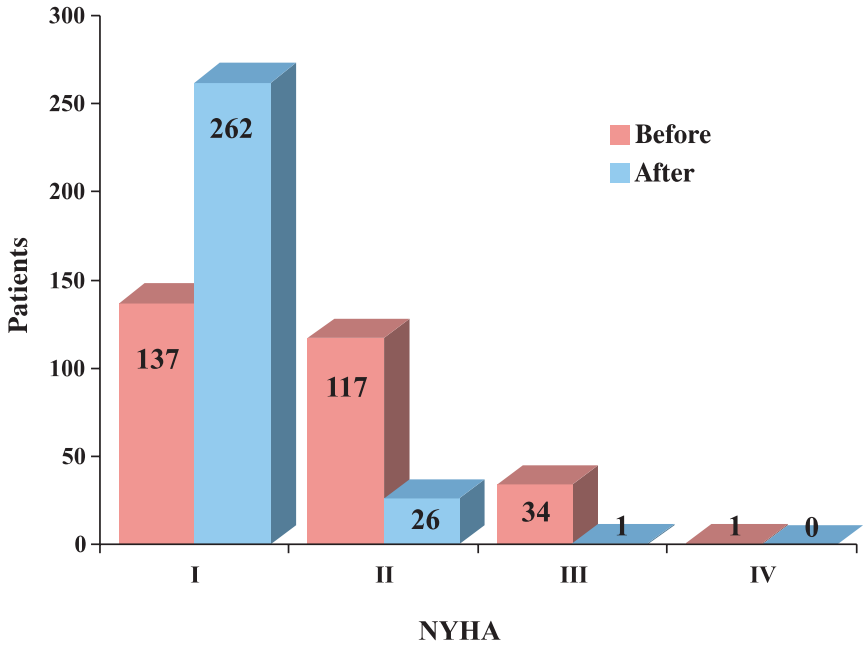

Figure 2. NYHA classification of patients before and after surgical treatment.

Table 2. Chromosomal anomalies.

Syndrome	Patients
Down	2
Kleinefelter	1
Noonan	1
Turner	1
William – Beuren	2
Other	3
Total	**10**

Table 3. Preoperative rhythm disorders.

Arrhythmia	Patients
Supraventricular tachycardia	2
Atrial fibrillation	15
Atrial flutter	3
1st. degree atrioventricular block	2
2nd. degree atrioventricular block	1
Complete heart block	5
Left bundle branch block	1
Right bundle branch block	4
Wolff-Parkinson-White syndrome	1
Total	**34**

erative changes, such as, for example, the failure of previously implanted biological valves.[3,5,6]

The most common types of uncorrected, non-cyanotic CHD seen in adults are, in decreasing order of frequency, atrial and ventricular septal defects, patent ductus arteriosus and coarctation of the aorta. Of the cyanotic type the most frequently occurring is tetralogy of Fallot. However, rarer, complex congenital heart anomalies may also be encountered, such as single ventricle, Ebstein's anomaly, or congenitally corrected transposition of the great vessels.[7]

Surgical treatment is considered to be curative in cases such as the ligation of a ductus arteriosus, closure of an atrial or ventricular septal defect, or repair of pulmonary valvular stenosis.[3,8] In most cases, however, surgery is considered corrective, while in others it is just palliative. In these cases there is a high probability of a follow-up procedure, which is ever more often carried out during adult life. The most commonly encountered residua, recurrences and sequelae after reparative surgery are valvular lesions, residual or recurring defects, myocardial dysfunction, vascular disorders (stenosis, aneurysms), degeneration of prosthetic materials, pulmonary hypertension and arrhythmias.[3]

In corrected tetralogy of Fallot a common sequela is pulmonary insufficiency, which necessitates valve replacement if severe regurgitation is present, preferably before the right ventricle is compromised.[9] If arrhythmias are also present, valve replacement results in stabilization of intraventricular conduction, and when this is combined with a cryoablation procedure a significant reduction in the appearance of ventricular and supraventricular arrhythmias can be achieved.[10] In cases where pulmonary stenosis is treated successfully by valvotomy or valvuloplasty there is usually no need for reoperation, except in a few cases of restenosis with a pressure gradient ≥50 mmHg.[8]

Aortic valve repair is feasible in young age with relatively good results; however, eventually these patients will usually require a valve replacement, most likely in adulthood.[11] Patients with aortic stenosis should be operated early because of the risk of arrhythmias and sudden death.[4,12]

Actuarial survival for the Fontan procedure (e.g. for tricuspid atresia) is 79% at 20 years. Patients who

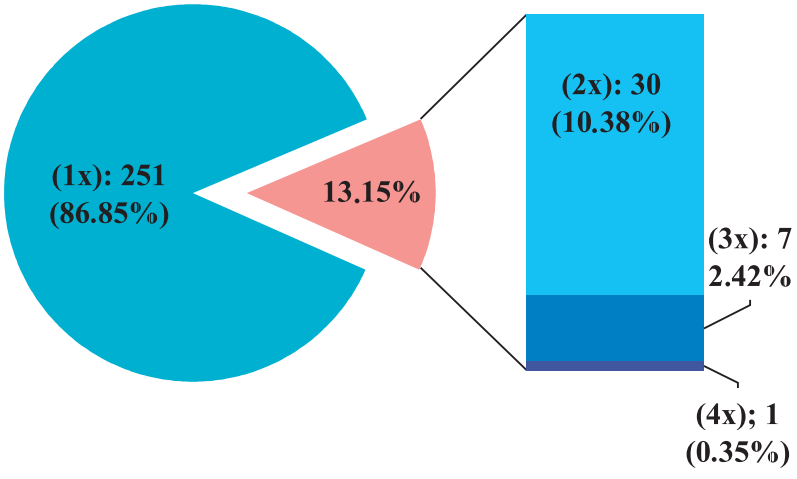

Figure 3. Total number of procedures per patient. x=times

Table 4. Surgical procedures.

Procedures			Patients
Closure of atrial septal defect (secundum)	With pericardial patch	Simple 113 With fossa ovalis closure 3 With aortic valve replacement 2 With mitral valve replacement 1	**125**
	Primary closure	6	
Closure of atrial septal defect (sinus venosus) & correction of anomalous pulmonary venous return	Simple 23 With tricuspid valvuloplasty 1 With replacement of aortic root and valve 1		**25**
Repair of coronary sinus defect			**1**
Closure of ventricular septal defect	With Dacron patch	Simple 12 With fossa ovalis closure 2 With enlargement of right ventricular outflow tract 2 With repair of subvalvular aortic stenosis 1	**18**
	Primary closure	1	
Combined closure of atrial and ventricular septal defects			**2**
Repair of partial atrioventricular canal	Simple 21 With ligation of ductus arteriosus 1		**22**
Repair of anomalous pulmonary venous return	Upper right 1 Lower right (scimitar) 1		**2**
Repair of subvalvular aortic stenosis (removal of membrane & myectomy of interventricular septum)	Simple 13 With aortic valve repair 1 With aortic valve replacement 1 With ligation of ductus arteriosus 1		**16**
Repair of supravalvular aortic stenosis (widening with Dacron patch)	Simple 1 With enlargement of aortic arch 1		**2**
Repair of aortic stenosis and/or insufficiency (aortic valve replacement)	With mechanical prosthetic valve	Simple 15 With repair of ascending aortic aneurysm 2 With ligation of coronary arteriovenous fistula 1 With repair of anomalous right upper pulmonary venous return 1	**21**
	With bioprosthetic valve	2	
Rupture of Valsalva sinus aneurysm	Repair and closure of ventricular septal defect		**1**
Mitral valve	Repair	1	**7**
	Replacement	Simple 5 With closure of atrial septal defect 1	
Repair of tetralogy of Fallot	Typical 5 With right ventricle → pulmonary artery graft 1		**6**
Reoperation for tetralogy of Fallot	Repair of residual ventricular septal defect & right ventricle → pulmonary artery graft 1 Repair of residual ventricular septal defect & pulmonary valve replacement 1 Repair of residual ventricular septal defect & repair of pulmonary valve stenosis 1 Repair of residual ventricular septal defect & aortic valve replacement & tricuspid valvuloplasty 1 Repair of residual ventricular septal defect & aortic valve replacement & right ventricle → pulmonary artery graft 1 Aortic valve replacement & right ventricle → pulmonary artery graft 1		**6**
Tricuspid valve replacement	Simple 2 With closure of atrial septal defect 2		**4**
Surgery of the pulmonary valve and right ventricular outflow tract	Pulmonary valvotomy 1 Pulmonary valve replacement with surgical reduction of large pulmonary vessels 1		**4**
	Enlargement of right ventricular outflow tract	Typical 1 With tricuspid valvuloplasty and coronary bypass grafting 1	

→

→ Table 4 (Continued).

	Procedures			Patients
Single ventricle surgery (Fontan procedure, with fenestration)	Lateral tunnel Extracardiac repair (graft)		1 2	**3**
Coarctation of the aorta	Excision and end-end anastomosis	Simple With aneurysm repair	8 1	**16**
	Excision and tubular graft interposition		4	
	Bypass with graft	Simple With aortic valve replacement	2 1	
Surgery of the aortic arch	Aortic arch enlargement			**1**
Patent ductus arteriosus	Left thoracotomy, external ligation and dissection Sternotomy and internal closure using Dacron graft under cardiopulmonary bypass		2 1	**3**
Coronary arteriovenous fistula	Ligation of the connection and normal diversion of arterial blood with Dacron patch			**1**
ALCAPA	Transfer of coronary artery ostium to aorta			**1**
Vascular ring	Ligation and dissection of accessory aortic arch			**1**
Repair of complex heart disease	Repair of residual ventricular septal defect & replacement of right ventricular → pulmonary artery graft			**1**
	Total			**289**

ALCAPA: Anomalous left coronary artery from pulmonary artery

underwent the procedure reach adulthood having a good quality of life.[13] However, sooner or later many of them will develop severe arrhythmias and progressive heart failure. In many of these cases a review and modification of the repair (e.g. converting it to a hemodynamically more efficient "extracardiac" Fontan) may improve the situation; otherwise, a transplant remains the final solution.[14,15]

Adult patients with CHD are at increased risk of complications and death from both cardiac and extracardiac complications and diseases.[16] It is well known that CHD (apart from atrial septal defects) is a risk factor for the development of infective endocarditis and therefore appropriate antibiotic prophylaxis is recommended. Reparative surgery, apart from closure of atrial septal defects, ligation of a patent ductus arteriosus or pulmonary stenosis repair, does not appear to alter this risk.[4,17] Also, there is an increased risk for thromboembolic episodes or more infrequent, yet characteristic complications, such as pro-

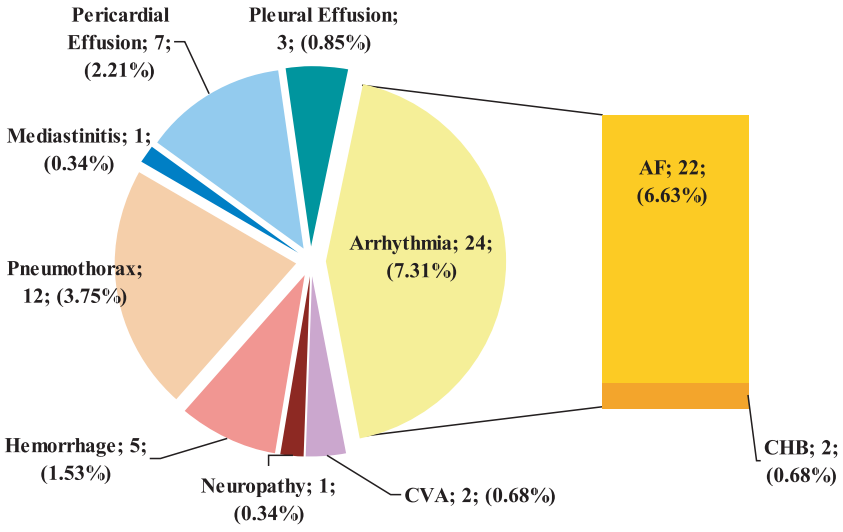

Figure 4. Postoperative complications. AF – atrial fibrillation; CHB – complete heart block; CVA – cerebrovascular accident.

tein losing enteropathy following procedures of the Fontan type.[3,18]

The surgical and medical management of adult CHD patients is a relatively new challenge for the medical community. Surgical treatment seems to be undertaken by experienced surgeons, mostly on an individual basis in both pediatric and adult cardiac surgery units. This has resulted in a lack of large series reported in the international literature. In one of the few published series originating from a children's hospital the results appeared to be good. For 112 surgically treated cases mortality was 6% with acceptable postoperative complications, including a 6% incidence of postoperative arrhythmias.[19] Our own results are comparable or even better, and this may be attributed largely to our unit's experience in both pediatric cardiac surgery and surgery in adults with acquired heart disease. The view that adult CHD patients should be treated in centers with experience in both congenital anomalies and acquired disorders that usually affect adults appears to be gaining increasing support.[2,15,19,20] Also, the fact that many patients with successfully corrected conditions (e.g. atrial septal defects) who are considered to have been completely cured have a lower level of physical activity compared to the general population, may be attributed, apart from the deleterious long term consequences of CHD, to the lack of relevant experience in the centers where they are followed.[7,21]

Adults with CHD make up a special group of patients with unique problems, both cardiac and extracardiac. Their surgical treatment has proved to be feasible, with exceptionally low mortality and acceptable morbidity. All the deaths in our series and many of the complications are related to arrhythmias and might thus perhaps be avoided if the patients were referred sooner for treatment.[19,22,23] Surgical intervention for the treatment of established arrhythmias has proved to be effective in a limited number of cases, mainly patients with a Fontan operation resistant to medical therapy.[24]

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