Available online at www.sciencedirect.com

ScienceDirect

journal homepage: http://www.elsevier.com/locate/crvasa

Original research article

Operations of adults with congenital heart disease – Single center experience with 10 years results

Jana Rubáčková Popelová [a,*], Roman Gebauer [a], Štěpán Černý [a], Petr Pavel [a], Ferdinand Timko [a], Pavel Jehlička [a], Petr Plášil [a], Jakub Tomek [b], Markéta Tomková [a,c], Ivo Skalský [a]

[a] Department of Cardiac Surgery Hospital Na Homolce, Roentgenova 2, 15030 Prague 5, Czech Republic
[b] Department of Anatomy, Physiology and Genetics, University of Oxford, United Kingdom
[c] Nuffield Department of Medicine, University of Oxford, Oxford, United Kingdom

ARTICLE INFO

Article history:
Received 18 November 2015
Received in revised form
24 December 2015
Accepted 29 December 2015
Available online 25 January 2016

Keywords:
Congenital heart disease in adults
Operations
Surgical mortality
Risk factors
Heart reoperations

ABSTRACT

During the last 10 years, 844 operations of 805 adults with congenital heart disease (CHD) were performed in Hospital Na Homolce in Prague, Czech Republic. The median age was 37 (interquartile range 25–49, full range 16–81) years. Operations of complex and rare CHD represented 47%. Forty-four percent of patients (354) underwent previous cardiac surgery in childhood or adulthood. Three and more operations were performed in 14% (113 patients). Combined surgical procedures were performed in 70% of operations.

Thirty-day mortality was 1.36%, hospital mortality 1.7% and 5-year survival probability 97%. The risk factors for early and late mortality were NYHA class III and IV symptoms ($p < 0.0001$; OR 30.8), history of heart failure ($p = 0.001$; OR 6.7), cyanosis ($p < 0.0001$; OR 60.5), number of previous operations ($p = 0.00033$), presence of mechanical prosthetic valves ($p = 0.0032$; OR 3.7) and univentricular circulation ($p = 0.0276$; OR 5.4). The difference was not significant for arrhythmias ($p = 0.078$), pulmonary hypertension ($p = 0.072$), age at operation ($p = 0.372$) and gender ($p = 0.48$).

Centralization of adult CHD care in a high volume center carries very good surgical results with low early and late mortality. It is important to perform the operations in time and to eliminate all residual lesions by combined surgical procedure. The presence of pediatric cardiac surgeon is necessary for the operations of complex CHD.

* *Corresponding author*. Tel.: +420 605 513 540; fax: +420 2 5727 2941.
E-mail address: jana.popelova@homolka.cz (J.R. Popelová).

http://dx.doi.org/10.1016/j.crvasa.2015.12.005

Introduction

The number of adults with congenital heart disease (CHD) is rapidly increasing with the estimated prevalence of 2800–3000 per one million inhabitants; that means altogether 30 000 adults with CHD for Czech Republic [1–3]. Nowadays, approximately 90% of children born with CHD are surviving to adulthood [2,4]. Even patients with critical and complex CHD may reach adulthood due to the progress in pediatric cardiology, cardiac surgery and intensive care. The contemporary population of adults with CHD comprises a broad spectrum of CHD with different previous surgical procedures and different significance of residual lesions. Regular follow-up is considered necessary in approximately half of the patients with CHD reaching adulthood [5,6]. Some authors recommend at least one examination in a specialized center for all adults with the history of CHD for further stratification [6,7].

However, besides children with regular follow-up until adulthood, there are adults operated for CHD many years ago in the 60th to 80th years of the 20th century. They often have no follow-up by a cardiologist, even though they have important residual findings. Moreover, some adults may have unoperated CHD which was asymptomatic or even unrecognized until older age.

The long lasting abnormal hemodynamics with pressure or volume overload of the ventricles and chronically increased or decreased pulmonary flow leads to functional and structural changes in myocardium and pulmonary vessels. Another important factor is the interaction of left and right ventricle under pathological conditions. We may find dilatation, hypertrophy, increased fibrosis, arrhythmias or heart failure, both systolic and diastolic. These patients may come in adulthood with problems related to their unsolved or residual congenital lesion.

It is important to decide who should take care of these patients and who should perform the surgery. The requirements for centers specialized at adults with CHD were defined [6,7]. The best results in operations of adults with CHD were reached in experienced adult cardiac surgeries with the participation of pediatric cardiac surgeon [8,9]. This was our strategy since 2005 [10]. In this article we would like to present our 10-year experience and results.

Patients and methods

We analyzed the results of all operations of adults with CHD performed from May 2005 to June 2015. Long term results including the mortality were evaluated from the records of the outpatient clinic for adults with CHD. In the case that patient did not come for control we contacted his family, his local cardiologist or general practitioner.

We included patients with bicuspid aortic valve (BAV) or morbus Barlow only in the case they were followed-up for this diagnosis since childhood or had BAV combined with COA or other CHD. Therefore, most adults with BAV and morbus Barlow were not included in our study. We did not include patent foramen ovale. We included patients with CHD who were reoperated for acquired valve disease as a consequence of CHD. Patients with atrial septal defect (ASD) type sinus venosus and partial anomalous pulmonary venous connection (PAPVC) were included among ASD. The diagnosis of PAPVC comprised patients with scimitar syndrome and isolated PAPVC. The presence of mechanical heart valves was evaluated regardless of the time of implantation and regardless of the cause of death.

Statistical analysis

The age of patients is given as the median and interquartile range (IQR), since the age was not distributed normally in our dataset. The statistical significance in Table 1 was evaluated by Wilcoxon non-parametric rank-sum test (age and number of operations) and by Fisher exact test (other data, i.e., binary risk factors). The value of $p < 0.05$ was considered a statistically significant difference. Furthermore, odds ratio was computed for binary risk factors. Survival analysis of simple (common) versus complex CHD was performed using Kaplan–Meier curves and log-rank test to compare the significance of the survival difference of the two groups.

Results

During the 10-years period 2005–2015 a total of 844 operations of 805 adult patients with CHD were performed. Forty-nine

Table 1 – Comparison of the frequency of risk factors among survivors and deceased.

Risk factor	Survivors N = 780	Deceased N = 25	*p*	OR
NYHA III–IV before operation	150 (19%)	22 (88%)	<0.0001	30.8
Cyanosis before operation	6 (0.77%)	8 (32%)	<0.0001	60.5
History of congestive heart failure	35 (4.5%)	6 (24%)	0.001	6.7
Presence of mechanical valve prosthesis	120 (15.4%)	10 (40%)	0.0032	3.7
Univentricular circulation	19 (2.4%)	3 (12%)	0.0276	5.4
Pulmonary hypertension	106 (13.6%)	7 (28%)	0.072	2.4
Arrhythmias	238 (30%)	12 (48%)	0.078	2.1
Men	419 (54%)	9 (36%)	0.103	0.48
Age at operation	39	40	0.636	NA
Number of previous operations	538	35		
Previous operations per patient	0.63	1.4	0.00033	NA

OR: odds ratio.