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Results of Surgical Treatment of Congenital Heart Defects in Children with Down's Syndrome

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Abstract. We analyzed early and late results of surgical treatment of 100 consecutive children with Down's syndrome (DS) and congenital heart defect (CHD) who were operated on between 1990 and 1997. Fifty had common atrioventricular canal (CAVC), 24 ventricular septal defect, 8 the ostium primum atrial septal defect, 8 tetralogy of Fallot (TOF), 3 patent ductus arteriosus, 3 the ostium secundum atrial septal defect, and 4 CAVC coexisting with TOF. In 93 patients total correction was performed. The total death rate was 6%. Death in the CAVC group was 8%, but it decreased to 2.7% during the past 3 years. The children who were followed up (from 7 months to 6 years; mean, 39 months) are in NYHA class I or II. There were no reoperations. The postoperative course was complicated by pulmonary infections in 38% of patients, which converted to generalized infection in 10% and was the cause of death in 8% of patients. These results indicate that CHD in DS children can be repaired with a low death rate and low incidence of severe mitral atrioventricular valve regurgitation in the CAVC group. A high incidence of severe infections can influence the final results. Repair of CHD in infancy helps to eliminate problems connected with congestive heart failure and pulmonary hypertension.

Key words: Down's syndrome — Congenital heart defect — Total surgical repair

Down's syndrome (DS) is a chromosomal abnormality with a frequency of 1:800 live births [9, 13]. A congenital heart defect in children with this syndrome occurs in 40% of patients and results in a 35% death rate [19]. A variety of estimates also exist for the frequencies of specific cardiac lesions in affected children in the common atrioventricular canal, with the most frequent being endocardial cushion defects (approximately 40%), ventricular septal defect (VSD) in 31% of patients, atrial septal defect (ASD) II in 9%, and tetrology of Fallot (TOF) in 6%. For unknown reasons, certain defects, such as transposition of the great arteries, situs inversus, and situs ambiguous, appear less frequently than in the population of children with no chromosomal abnormalities; hence, a theory has been formulated on a "protective" effect of DS in the incidence of such defects [16]. Recently, the survival rates of children with DS and concomitant congenital heart defect (CHD) have considerably improved. In the group of children with endocardial cushion defects, 80% of patients live as long as 15 years [4]. The most frequent causes of death are respiratory tract infections and consequences of CHD such as congestive heart failure and pulmonary hypertension, a particularly dangerous condition which develops earlier in children with Down's syndrome and has a more violent course. The recent experience in surgical management of children employing extracorporeal circulation has resulted in a decrease of perioperative death rates, especially in the youngest children [5]. This article presents early and late results achieved in children with CHD and DS operated on at the Polish–American Children's Hospital.

Material and Methods

Between January 1990 and August 1997, 100 cardiac surgical procedures (Table 1) were performed in consecutive patients with DS. There were 92 (92%) total corrections and 8 (8%) palliative operations. The children ranged in age from 1.5 to 180 months (mean, 31.1 months) and in body weight from 2.3 to 58 kg (mean, 9.1 kg). Boys constituted 67% of the patients. Down's syndrome was diagnosed on the basis of clinical presentation and genetic examinations. The type of congenital heart defect was established on the basis of clinical observations, echocardiography, and hemodynamic studies performed when the degree of pulmonary hypertension had to be evaluated. A prerequisite for qualifying a patient for surgery was pulmonary resistance below 10 Wood's

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Table 1. Operated cardiac anomalies

Anomaly	No. of patients	Operative weight in kg (mean)	Operative age in months (mean)	Deaths
CAVC	50	2.3–47.2 (8.9)	1.5–180 (26)	4 (8%)
VSD	24	2.7–58 (8.9)	1.5–180 (31)	1(4.1%)
ASD I	8	6.3–27 (13.7)	10–120 (54)	0
ASD II	3	6.5–25.7 (24.6)	18, 60, 120	0
TOF total correction	3	14, 26, 36	36, 48, 108	0
TOF (RBTS)	6	6.3–15.2 (7.6)	9–60 (29)	0
PDA	2	5.3, 13.6	9 36	0
CAVC + TOF (RBTS)	2	2, 18.7	4.5 84	1 (50%)
CAVC + TOF (total correction)	3	8.3, 10.5, 17.7	26, 31, 94	0
Total	100	2.3–58 (9.1)	1.5–180 (31.1)	6 (6%)

CAVC, common atrioventricular canal; VSD, ventricular septal defect; ASD II, ostium secundum atrial septal defect; ASD I, ostium primum atrial septal defect; TOF, tetralogy of Fallot; PDA, patent ductus arteriosus; RBTS, right Blalock–Taussig shunt.

units/m^2, but the reactivity of the pulmonary bed in the course of the oxygen test (100% oxygen inhaled for 15 minutes) was a decisive factor in this respect. Intracardiac defects were operated on by employing extracorporeal circulation in mild hypothermia, with permanent flow of 2.4 L/min/m^2 of body surface or during deep hypothermic circulatory arrest. The method of extracorporeal circulation employed depended on the type of defect and age and body weight of the child. Cold single-dose crystalloid cardioplegic solution was injected to a total volume of 15 ml/kg, and topical hypothermia was added. When the postoperative course was uncomplicated, the patients were extubated on the day of the surgery or during the morning hours of the following day. In order to decrease the pressure in the pulmonary artery, mild hyperventilation was employed using high oxygen concentration. In children with pulmonary hypertension no pulmonary tissue sample was collected in view of a low correlation between hemodynamic parameters and histopathological changes. Since there were many patients with common atrioventricular canal (CAVC), we present a more detailed description of these patients.

The defect was corrected in 50 children aged 1.5 months to 15 years (mean, 26 months) and with body weight of 2.3 to 47.2 kg (mean, 8.9 kg). In 47 (94%) of them, deep hypothermia with circulatory arrest was employed. The duration of circulatory arrest ranged from 35 to 70 minutes (mean, 46 minutes). In all patients subjected to hemodynamic studies, the pulmonary pressure prior to the correction was at least two-thirds the systemic pressure. From the morphological viewpoint, the A and C type canals according to Rastelli were predominant. Except for 2 patients, the two-patch repair was employed. The VSD was primarily closed using Dacron patch and during suturing caution was maintained in order not to damage the chordal attachments of particular leaflets. In all cases, single sutures were used in the cleft in the left component of the atrioventricular valve beginning at its base and moving centrally. A mild degree of mitral incompetence was attempted rather than a maneuver resulting in a stenosis of the left atrioventricular orifice. The ASD was sutured using an autologous pericardial patch with the ostium of the coronary sinus drained to the right atrium. No anuloplasty was performed. Total corrections were in no case preceded by palliative procedures.

Table 2. Primary cause of death

No.	Anomaly	Cause	Early death	Late death
1	CAVC	Unknown	1	
2	CAVC	Unbalanced AV canal, hypoplastic left ventricle	1	
3	CAVC	Subaortic stenosis	1	
4	CAVC	Pulmonary hypertension (crisis)	1	
5	VSD	Sepsis, MOF	1	
6	CAVC + TOF	Sepsis, NEC	1	
7	CAVC	Retrocecal abscess, peritonitis, sepsis		1

CAVC, common atrioventricular canal; VSD, ventricular septal defect; TOF, tetralogy of Fallot; MOF, multiorgan failure; NEC, necrotizing enterocolitis.

The ASD I was corrected in 8 children (8% of the original group) aged 10 months to 10 years (mean, 54 months) and with body weight of 6.3 to 27 kg (mean, 13.7 kg). In all patients, single sutures were used to close the cleft of the anterior leaflet of the mitral valve. The ASD was closed with an autologous pericardial patch. Twenty-four children (24%) constituted a group of patients with VSD. They ranged in age from 6 weeks to 15 years (mean, 31 months) and in body weight from 2.7 to 58 kg (mean, 8.9 kg). In 9 children (37.5%), the pulmonary pressure before the surgery was at least 50% of the systemic pressure. The intraoperative morphological evaluation of VSD included conoventricular type in 11 patients (45.7%) and AV canal type in 13 patients (64.3%).

Additionally, surgical procedures were performed in three children with ASD II, two with patent ductus arteriosus (PDA), nine with TOF (three total corrections and six palliative procedures), and five with CAVC coexisting with TOF (three total corrections). Tetralogy of Fallot was repaired closing the VSD using the right ventricular approach and reconstructing the right ventricular outflow tract with a Gore-Tex or homogeneous patch (transanular in one case). Palliative procedures consisted in performing modified right Blalock–Taussig shunts employing a Gore-Tex vessel 3.5 to 5.0 mm in diameter depending on the age of the child and the size of the anastomosed vessels.

Results

In the CAVC group, 4 children (8%) died in the early postoperative period (Table 2); in the past 3 years, of 37 patients (65.9%), 1 died (2.7%) and the cause of death remains unknown. On the first postoperative day, low cardiac output developed suddenly. Despite extracorporeal life support (ECMO) employed for 3 days, no adequate cardiac function was achieved. Of the remaining 3 patients, 1 died due to pulmonary hypertension on day 1 postoperatively. In another child with an unbalanced AV canal, a biventricular correction was performed and the presence of a hypoplastic left ventricle was disclosed

only on autopsy. The third patient died due to a 60% obstruction of the left ventricular outflow tract which had not been diagnosed preoperatively.

In 5 (10%) children from the CAVC group, PDA coexisted with the underlying defect. In as many as 19 patients (34%), respiratory tract infections developed postoperatively and in 7 patients (14%) they converted to sepsis. This resulted in prolonged assisted ventilation and hospitalization in an intensive care unit.

Control echocardiography performed in the early postoperative period revealed the presence of residual VSD, with no hemodynamic significance in 3 patients (6%), mitral valve incompetence grade II in 4 patients (8%), and tricuspid valve incompetence grade II in 4 patients (8%). In 5 patients (10%), postoperative complications consisted of hydropericardium requiring pericardiocentesis. Concomitant duodenal atresia was corrected surgically in 2 patients (4%) in the first day of life. Late follow-up includes 38 patients (76%); the remaining ones failed to report and their fate is unknown. The follow-up duration ranged from 7 months to 6 years (mean, 39 months). One patient died in the late postoperative period (2 months following cardiac surgery) of noncardiac reasons (retrocecal abscess with perforation of the colon, sepsis, and multiorgan failure).

Control echocardiography performed in the previously mentioned 38 patients revealed mitral valve incompetence: grade III in 2 patients (5.2%), grade II/III in 4 patients (10.5%), and grade II in 3 patients (7.8%). Tricuspid valve incompetence grade II/III was demonstrated in 3 patients (7.8%) and grade II in another 3 patients (7.8%). In the remaining children either no incompetence was noted or it had a trivial character. Insignificant hemodynamically residual VSD was observed in 4 patients (10.5%), whereas 1 patient (2.6%) showed a residual defect in the lower part of the interatrial patch. In this group, 8 patients remain in NYHA class II, and the remaining ones are in class I. These 8 children were treated employing the following agents: digoxin, furosemide, and angiotensin-converting enzyme inhibitors. In no case were reoperations performed. No patient showed total atrioventricular dissociation.

Common atrioventricular canal concomitant with TOF was corrected in three children. No early postoperative complications were noted. Mitral valve incompetence was classified as grade I/II, II, and I, and the follow-up period is 3.5 years, 10 months, and 6 months, respectively.

Of 24 patients with VSD, 1 (4.2%) died due to sepsis and multiorgan failure. In 1 patient VSD was accompanied by ASD II and PDA. Another patient was diagnosed intraoperatively as having double-chamber right ventricle. Three patients (12.5%) displayed atrioventricular dissociation in the immediate postoperative period, but the condition subsided spontaneously within several days. In 1 patient, 6 months postoperatively a permanent pacemaker implantation duo to complete heart block was necessary. On follow-up, echocardiography showed mitral valve incompetence grade II in 2 patients (8.5%) and tricuspid valve incompetence in 2 patients. Hydropericardium requiring surgical intervention was encountered in 6 cases (25.5%).

In children who were operated on due to ASD I, no deaths occurred. Two patients (25%) showed in echocardiography mitral valve incompetence grade II. Pulmonary infection was a postoperative complication in three patients (37.5%). In the remaining patients one death occurred (a child with TOF subjected to a palliative procedure). The boy died due to noncardiac-related reasons (sepsis and necrotising enterocolitis).

Discussion

In view of their high frequency, congenital heart defects are believed to be clinical features of DS [18], but the cause of their frequent occurrence is still unknown [8, 11, 15]. A major factor influencing the survival of children with DS, especially in the first decade, is the presence of CHD. This is confirmed by studies performed in numerous centers in which pulmonary disease is the predominant cause of death (23–41%), followed by CHD (30–35%), infectious diseases (2–15%), and neoplastic diseases (2–9%). CHD is practically the only major defect that can be corrected [10]. Owing to the introduction of cardiac surgery in children with CAVC, 85% of them live to be 2 years old, whereas the analysis of a natural course of this defect shows a 54% survival rate up to 6 months of life and only 15% up to 2 years [3]. In the majority of children with large cardiac septal defects, normal postnatal regression of pulmonary resistance permits a left-to-right shunt and variable congestive heart failure to develop. Alternatively, pulmonary resistance fails to regress normally after birth and remains elevated. It prevents the development of a large increase in pulmonary blood flow and may lead to pulmonary obstructive disease. The development of pulmonary hypertension is generally considered to occur earlier and more severely in children with DS and CHD, although this conclusion has been disputed. According to Clapp et al. [6], in 12% of children with CAVC pulmonary hypertension can become irreversible within the first year of life. Hence, surgical treatment in defects with high-pressure shunts is recommended within the first year of life, or even within the first 6 months [12]. The young age of patients allows for the avoidance of problems related to pulmonary hypertension, especially in the postoperative period. When the mean age of surgical patients in our institution was decreased, no increase in death rates was observed, and it appears that no negative effect was exerted on late surgical results. The experience gained in our hospital confirms the effectiveness of early,

total elimination of high-pressure left-to-right shunts. Perioperative death in patients with DS is currently only slightly higher compared to that in children without chromosomal aberrations (mainly due to infections) [1]. The presented results (total death rate of 6.0% and 2.7% in children with CAVC in the past 3 years) are comparable with data reported by other centers [2]. Currently, the main risk factors are related to morphological features of CHD. In the case of CAVC, these risk factors are double-orifice left AV valve, single left-sided papillary muscle, and left-sided obstruction lesions. In our patients no morphological features related to poor prognosis were noted.

Late results are markedly affected by the competence of AV valves, especially mitral and tricuspid. It seems—this is also confirmed by other investigators—that in children with DS and CAVC with proper correction, the amount of tissue is usually sufficient to create competent valves [20] and the degree of valve incompetence prior to correction and the type of atrial canal are of secondary importance. It is also believed that the powerful stimulus of chronically elevated Q_p/Q_s could play a role in the onset of annular dilatation and secondary valve incompetence [17]. In other words, annular dilatation could represent the anatomical expression of heart failure in CAVC. Airway obstruction secondary to laryngobrochomalacia is frequently seen in patients with DS. Pulmonary vascular resistance is therefore higher in patients with DS, and Q_p/Q_s is relatively lower when compared with that of patients without DS. This could explain why annular dilatation is relatively uncommon. The confirmation can be found in the fact that to date there has been no necessity to implant valvar prostheses in the mitral position in our patients. In the case of congestive heart failure intensification in children on pharmacological treatment remaining in NYHA class II, a future reoperation will be considered, including implanting an artificial valve. A separate problem in children with DS is related to frequent infections, mainly of the respiratory system, complicating the postoperative course [14]. They are believed to result from frequent concomitant immunological defects, especially with respect to maturation and function of T lymphocytes and generalized muscular hypotonia. Pulmonary hypoplasia characterized by a diminished number of alveoli in relation to acini and in reduction in total alveolar surface area has been reported to be independent of the presence of heart disease. Reduction of airway branching and the thinning of the medial layer of the small pulmonary arteries have also been described [7]. In our patients, pulmonary infections, often progressing into sepsis, were the most common cause of early and late death, as well as prolonged hospitalization at intensive care units, and increased the costs of treatment. The role of frequent pulmonary infections in the development of fixed pulmonary hypertension has not been clarified.

Children with DS constitute a major percentage of patients operated on in our center, and their surgical results do not considerably differ from those achieved in patients without chromosomal aberrations.

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