SURGICAL TREATMENT OF TRANSPOSITION OF THE GREAT ARTERIES

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CHIRURGICKÁ LIEČBA TRANSPOZÍCIE VEĽKÝCH CIEV

Abstract

Hraska V, Sojak V, Kostolny M, Nosal M, Sagat M, Petko M, Kantorova A, Nagi A, Kaldararova M, Siman J:
Surgical treatment of transposition of the great arteries
Bratisl Lek Listy 1999; 100 (6): 286–290

Background: **With regard to risk of the failure of systemic right ventricle after physiological correction of transposition of great arteries, anatomic repair is a current method of choice.**
Objective of study: **Analysis of results of surgical correction of transposition of great arteries performed between 1992 and October 1998.**
Method: **A total of 111 patients were operated on for transposition of the great arteries. In the 1st group of patients (n=21, mean age was 135±55 days), physiological correction according to Senning was performed. Patients of the 2nd group (n=90, mean age was 15.4±21.6 days) underwent anatomic repair.**
Re*sults:* **Early mortality was 6 % (7 patients). Mean follow-up is 2.95 years (1.9 SD) ranging from 0.2 years to 6.1 years. Actuarial 1-month survival in the whole cohort (n=111) is 94 %, and it remains unchanged at 1, 2, 3, 4, 5, and 6 years of follow-up. Patients, who underwent surgery after 1997, show significantly better survival compared to those operated before 1997 (p=0.0997). Thus, a date of operation (before 1997) is the only significant risk factor for death. Survival in patients operated after 1997 (n=40) is 98 %. All patients belonging to the 2nd group are in functional group NYHA 1.**
Concl*usion:* **Anatomic repair of transposition of the great arteries is a method of choice for treatment of this congenital heart defect. Left ventricle becomes systemic ventricle, which is essential in view of long-term performance. Psychomotor development of children, who underwent ASO, is comparable with that of healthy population.** ***(Tab. 3, Fig. 3, Ref. 18.)***
Key words: transposition of great arteries, physiological correction, anatomic correction.

Abstrakt

Hraška V., Soják V., Kostolný M., Nosáľ M., Šagát M., Peťko M., Kántorová A., Nagi A., Kaldarárová M., Siman J.:
Chirurgická liečba transpozície veľkých ciev
Bratisl. lek. Listy, 100, 1999, č. 6, s. 286–290

Pozadie problému: Vzhľadom na riziko zlyhávania systémovej pravej komory po fyziologickej korekcii transpozície veľkých ciev je v súčasnosti metódou voľby anatomická korekcia.
Cieľ sledovania: Analýza výsledkov chirurgickej korekcie transpozície veľkých ciev od roku 1992 do októbra 1998.
Metóda: 111 pacientov bolo operovaných pre transpozíciu veľkých ciev. V 1. skupine pacientov (n=21) s priemerným vekom 134 dní (55 SD) bola realizovaná fyziologická korekcia podľa Senninga. V 2. skupine (n=90) sa pacienti s priemerným vekom 15,4 dňa (21,6 SD) podrobili anatomickej korekcii.
Výsledky: Včasná mortalita bola 6 % (7 pacientov). Priemerná dĺžka sledovania je 2,95 roka (1,9 SD) od 0,21 do 6,1 roka. Aktuariálne prežívanie v prvom mesiaci je v celej skupine (n=111) 94 % a zostáva identické v 1., 2., 3., 4., 5. a 6. roku sledovania. Pacienti operovaní po roku 1997 majú signifikantne lepšie prežívanie (p=0,0997) v porovnaní s pacientmi operovanými pred rokom 1997. Rok operácie (pred 1997) je jediným rizikovým faktorom pre úmrtie. U pacientov operovaných od roku 1997 (n=40) je prežívanie v sledovanom období 98 %. Všetci pacienti v 2. skupine sú vo funkčnej skupine NYHA I.
Záver: Metódou voľby chirurgickej liečby transpozície veľkých ciev je anatomická korekcia, ktorá anatomicky i funkčne koriguje patofyziológiu transpozície. Ľavá komora sa stáva systémovou komorou, čo je rozhodujúce z dlhodobého funkčného hľadiska. Psychomotorický vývoj detí po anatomickej korekcii je porovnateľný so zdravou populáciou. *(T*ab. 3*, obr.* 3*, lit.* 18.*)*
Kľúčové slová: transpozícia veľkých ciev, fyziologická korekcia, anatomická korekcia.

Complete transposition of great arteries (D-TGA) is a congenital heart defect with concordant atrio-ventricular and discordant ventriculo-arterial connection. Aorta (Ao) arises from the right ventricle (RV) and pulmonary artery (PA) comes from the left ventricle (LV) (Van Praagh, 1971). Without surgery this congenital heart defect is fatal due to a parallel type of circulation. The ob-

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Tab. 1. Clinical characteristics of D-TGA (n = 111).
Tab. 1. Klinická charakteristika D-TGA (n=111).

	n	Age (days) Vek (dni)	Weight (g) Hmotnosť (g)
1st Group Skupina 1	21		5500±850
D-TGA+IVS	20	139±52.5	
D-TGA+VSD	1	44	
2nd Group Skupina 2	90		3476±420
D-TGA + IVS	68	10.8±5.02	
D-TGA + VSD	20	29.9±40.1	
Total Celkovo	111		

Key:
D-TGA: D-transposition of the great arteries
IVS: Intact ventricular septum
VSD: Ventricular septal defect
Vysvetlivky:
D-TGA: D-transpozícia veľkých ciev D-TGA: D-transposition of great arteries
IVS: Intaktné komorové septum IVS: Intact ventricular septum
VSD: Defekt komorového septa VSD: Ventricular septal defect

Tab. 2. Coronary anatomy in the 2nd group of patients (n=90).
Tab. 2. Typ koronárnej anatómie v II. skupine pacientov (n = 90).

Coronary anatomy Typ koronárnej anatómie	n	%
1 LCA; 2 RCA	70 7	8
1 RCA, LCA	1	1
1 LAD; 2 RCA, Cx	7	8
2 RCA, LCA	7	8
1 RCA; 2 LCA	2	2
1 LAD, RCA; 2 Cx	2	2
Intramural LCA Intramurálna LCA	1	1

Key:
LCA: Left coronary artery
RCA: Right coronary artery
LAD: Left anterior descending artery
Cx: Circumflex artery
1: left posterior coronary sinus
2: right posterior coronary sinus
Vysvetlivky:
LCA: Ľavá koronárna artéria
RCA: Pravá koronárna artéria
LAD: Ramus interventricularis anterior
Cx: Ramus circumflexus
1: ľavý zadný koronárny sínus
2: pravý zadný koronárny sínus

jective of this study was to analyze results of surgical treatment of D-TGA accomplished between 1992 and October 1998, and to identify risk factors for survival.

Material a methods

Patients: Between 1992 and October 1998, 111 patients were operated on D-TGA without left ventricle outflow tract obstruction at Department of Cardiovascular Surgery, Children's University Hospital in Bratislava (Table 1). Eighty-eight patients had D-TGA with intact ventricular septum (D-TGA+IVS) and 23 patients had D-TGA in combination with hemodynamically significant ventricular septal defect (D-TGA+VSD). Twenty-one patients underwent physiological correction according to Senning (1st group) between 1992 and October 1993. Since October 1993, all patients (2nd group) underwent anatomic repair so called arterial switch operation (ASO). Mean age at operation in the first and second group was 134±55days and 15.4±21,6 days, respectively.

Preoperative management: In both groups, preoperative protocol included balloon atrioseptostomy, and in the 2nd group also intravenous application of prostaglandin E1 (PGE 1) (0.01 g/kg/min). Patients with D-TGA+VSD were not administered PGE1. Echocardiography established a complete diagnosis in 95 % patients. In case of unclear coronary anatomy, patients underwent angiography. Preoperative LV competence with respect to performance of systemic function after ASO was evaluated according to geometry of LV and interventricular septum. The pressure gradient at the level of arterial duct served as another evaluation criterion. Coronary anatomy was assessed according to Quaegebeur (Quaegebeur et al., 1986). Most commonly, the left coronary artery (LCA) arises from aortic sinus 1 and the right coronary artery comes off aortic sinus 2 (1LCA; 2RCA). LCA further gives origin to the left anterior descending artery (LAD) and circumflex artery (Cx). These coronary arteries may, however, arise separately from any sinus creating thus numerous combinations of origin and course of coronary vessels. Table 2 summarizes variability of coronary anatomy in the 2nd patient group.

Technique of operation and perfusion protocol: General principles of extracorporeal circulation in the youngest children were applied (Castaneda et al., 1994).In the 1st group, all patients were operated in deep hypothermia (rectal temperature 18 ˚C) and total circulatory arrest (Jonas, 1996). Physiological correction according to Senning creates two venous tunnels through which systemic and pulmonary venous returns are redirected. Blood from superior and inferior venae cavae returns through the central tunnel to mitral valve (MV) and LV. PA arises from LV, hence, the latter serves as a pulmonary ventricle. Blood from pulmonary veins returns through the complex tunnel to tricuspid valve and RV, which serves as a systemic ventricle connected to Ao (Senning, 1959). A detailed description of surgical technique falls beyond the scope of this paper.In the 2nd group, hypothermia between 21 ˚C—18 ˚C was employed. Circulatory arrest was used during closure of ventricular and atrial septal defects and at preference of surgeon, respectively. ASO encompasses positional change of great arteries and translocation of coronary arteries. Following transection of patent arterial duct, Ao and PA are divided just above valvar commissures. Both PA branches are mobilized extensively to pulmonary hila. PA is transferred anteriorly and above ascending Ao. After creation of coronary buttons and sufficient mobilization of coronary arteries, the latter are transferred to corresponding sinuses in neoarta (previous PA trunk). Neoarta is then connected with Ao using end-to-end anastomosis. A detailed descrip-

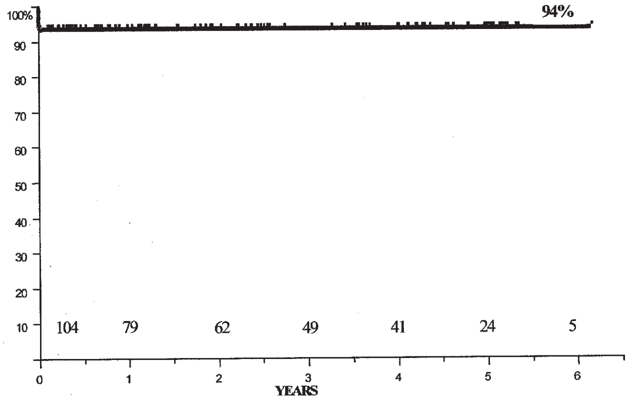

Fig. 1. Survival in patients after repair of transposition of the great arteries (n=111; mean follow-up 2.95 years).
Obr. 1. Prežívanie pacientov po korekcii transpozície veľkých ciev (n=111; priemerná dĺžka sledovania 2,95 roka).

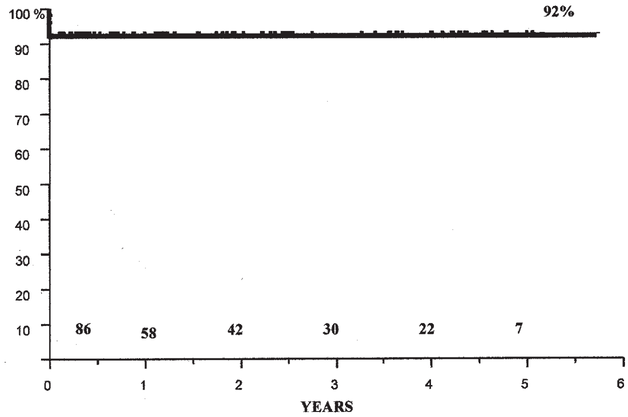

Fig. 2. Survival with anatomic correction of transposition of the great arteries (n=90).
Obr. 2. Prežívanie pacientov po anatomickej korekcii transpozície veľkých ciev (n=90).

tion of surgical technique falls beyond the scope of this paper (Castaneda et al., 1984; Castaneda et al., 1994).

Follow-up: Clinical examinations, EKG, x-ray, and echocardiography were accomplished at regular 3-month intervals. At the age of 1-3 years, patients underwent catheterization and angiography. *Statistical analysis:* Data was analyzed using a statistical program (JMP Statistical Analysis, Cary, NC). The primary outcome variable was survival after operation. Early failure was defined as death within 30 days of operation and late failure was defined as death 30 days after operation. Multiple clinical parameters were analyzed for their possible impact on survival using univariate analysis with Pearson's chi-square for categorical variables and Student's t-test for continuous variables ($p<0.05$). The Kaplan-Maier method was used for actuarial survival analysis. Subgroups were compared with the use of the log-rank test ($p<0.1$). All results are expressed as mean standard deviation.

Tab. 3. Causes of operative mortality (n=7).
Tab. 3. Príčiny včasnej mortality (n=7).

Cause of death Príčiny úmrtia	n
Coronary artery related Koronárny transfer	3
Preoperative LV incompetence Nekompetentnosť LV pred operáciou	1
LV dysfunction Dysfunkcia LV	3

Key:
Left ventricle
Vysvetlivky:
LV: Ľavá komora

Results

Mortality

Early mortality (Table 3). There was no death in the 1st group. In the 2nd group, 7 patients (7.7 %) died. Of these, 5 patients (7.35 %) had D-TGA+IVS and 2 patients (9.1 %) had D-TGA+VSD. Univariate analysis of several variables (age, weight, septal geometry, presence of VSD, coronary anatomy) did not reveal any risk factor for death.

Late mortality: We have not lost any patient during follow-up.

Functional condition

In the 1st group, 20 patients are in a functional group NYHA 1. One patient is in NYHA 2 due to a gradual development of tricuspid regurgitation. This patient is being prepared for conversion of atrial correction (Senning) to anatomic one (ASO). Therefore, PA banding was performed in order to prepare LV for systemic function.

In the 2nd group, all patients are in a functional group NYHA 1, they have sinus rhythm and no changes on ST segment. Echocardiography found normal LV function in entire group. In two patients (2.2 %), pressure gradient >30 mmHg was noted in RV outflow tract (RVOT). One patient has a significant pressure gradient at anastomosis between neoarta and original aorta. Moderate aortic regurgitation was found in 1 patient (1 %). Patients do not receive digoxin and diuretics, and redo has not yet been required.

Long-term survival

No patient was lost to follow-up. Mean follow-up in entire group is 2.95±1,9 years ranging from 0.2 to 6.1 years. Average follow-up in the 1st and 2nd group is 5.1±1,17 years and 2.41±1.66 years, respectively. Actuarial 1-month survival in the whole cohort (n=111) is 94 % and remains unchanged at 1, 2, 3, 4, 5, and 6 years of follow-up (Fig. 1). Survival in the 1st and 2nd group is not statistically different ($p=0.193$). In the 2nd group (n=90), a 1-month survival is 92 % and remains identical in the 1st, 2nd, 3rd, 4th, and 5th year of follow-up (Fig. 2). In this group, there is no

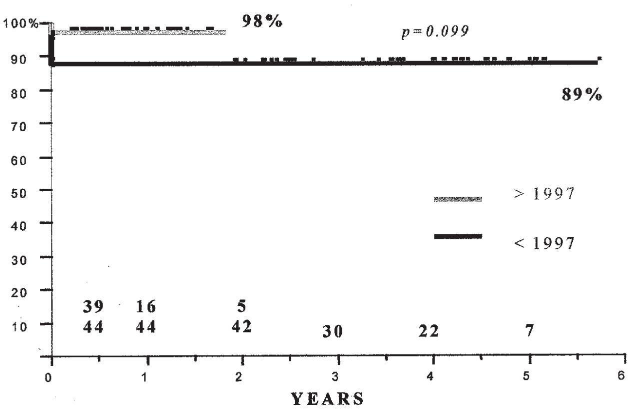

Fig. 3. Survival with anatomic correction of D-TGA before and after 1997 (n=90).
Obr. 3. Prežívanie pacientov po anatomickej korekcii D-TGA pred a po roku 1997 (n=90).

statistically significantly difference in survival between subgroups D-TGA+IVS and D-TGA+VSD (p=0.783). Survival in patients operated on after 1997 (n=40) is 98 % (Fig. 3). Patients, who underwent surgery after 1997 show significantly better survival compared to those operated on before 1997 (p=0.0997) (Fig. 3). Thus, a date of operation (before 1997) is the only significant risk factor for death.

Discussion

Physiological correction of D-TGA achieved excellent immediate and intermediate results. Initial mortality in best centers varied around 5 % and 15 % in a subgroup with D-TGA+IVS and D-TGA+VSD, respectively. Overall survival was, however, significantly lower as a result of mortality due to delayed surgery. Surgery is performed standardly at the age of 3—6 months of life. Thus, it is not surprising that a 5-year survival in patients after Senning correction is only 80 % (Rubay et al., 1988; Williams et al., 1988). In a long perspective, this type of repair is burdened by high incidence of cardiac arrhythmias and gradual failure of systemic RV. In a 10-year follow-up, only 55 % patients have a sinus rhythm. This issue can be solved. However, the failure of systemic RV is almost not negotiable. In a 10-year follow-up, incidence of RV failure varies between 10—20 %. It applies particularly to a subgroup with D-TGA+VSD (Williams et al., 1988). RV and TV are morphologically not built up for systemic function as was demonstrated by the failure of systemic, anatomical RV in patients with corrected transposition of great arteries (Sano et al., 1995; van Son et al., 1995). Only two options are available for children with failing RV after physiological correction: heart transplantation, or conversion of physiological correction to anatomic one after LV preparation for systemic performance (Chang et al., 1992; Cochrane et al., 1993). In a 6-year follow-up, survival of our group of patients after a physiological correction according to Senning is 100 %. In one patient, conversion to anatomic repair is required due to incompetent TV. RV failure in a long-term follow-up was critical for a change of operation strategy also at our department. ASO corrects D-TGA both anatomically and functionally. LV becomes systemic ventricle and RV pulmonary one. The only difference is that morphologically PV is left in systemic circulation (Jenkins et al., 1991).

Initial mortality was around 20—30 % even in the best centers (Kirklin et al., 1992; Norwood et al., 1988; Serraf et al., 1993). The success of surgery is related to postoperative LV competence and quality of operation. LV competence to perform systemic function immediately after operation is determined by a degree of pulmonary vascular resistance (PVR), i.e. preoperative LV afterload. Soon after the birth newborns with D-TGA+IVS experience PDA closure and pronounced decline in PVR. LV pressure decreases to "normal values", i.e. 1/3 of systemic RV pressure. As a result, stimulus for LV growth adaptation attenuates. Gradually, LV wall significantly thins out, and LV compliance increases at the same time. These changes occur 2—4 weeks after the birth. After this time, LV is no longer able to perform systemic function (Castaneda et al., 1984). This moment should be considered in respect to the timing of surgery. Ideally, operation should be performed within 2 weeks of life. This is not true in patients with D-TGA+VSD, where left-to-right shunt at ventricular level preserves LV competence. On the other hand, there is a risk of developing fixed pulmonary hypertension. Therefore, also these children are operated on in a newborn period.

Success of this type of operation depends on coronary anatomy, associated lesions, VSD type, relationship of great arteries, their size discrepancy, and gained experience. Difficulties associated with translocation of coronary arteries are related to anatomy of their origin and course. Groups at highest risk are those with common origin of the right coronary artery (RCA), with the left coronary artery (LCA) and circumflex artery arising from RCA, those with inverted coronary anatomy, and patients with intramural course of LCA (Kirklin et al., 1992; Serraf et al., 1993). In our material, coronary anatomy was not found as a risk factor for death. Analogically, the presence of VSD did not serve as a risk factor. The date of operation (before 1997) proved as the only risk factor for death. The above data indicate a high-standard method of operation and overcome of "learning curve", i.e. gaining adequate experience. Currently, leading centers achieve a long-term survival of 90—92 % and mortality below 5 % after learning curve has been overcome (Serraf et al., 1993). A 5-year survival in our ASO patients is 92 %, and survival over past two years is 98 %. Incidence of RVOT obstruction in the literature is below 10 % (Serraf et al., 1993; Nogi et al., 1998) and in our center, it is 2 %. Freedom from reoperation in our cohort is 100 % so far, and literature suggests a 5-year freedom from reoperation 90 % (Serraf et al., 1993). The fate of neoartic, morphologically pulmonary valve (PV) in systemic circulation remains unclear (Jenkins et al., 1991; Serraf et al., 1993). The frequency of PV regurgitation varies between 5 % and 50 %, and in our file, it is 1 %. Functionally, all children after ASO receive no medicines, and their psychomotor development is consistent with that of their peers.

Conclusion

Despite excellent immediate and short-term results of physiological correction of D-TGA, serious rhythm disturbances and particularly the failure of systemic RV were noticed in long-term follow-up. Therefore, anatomic repair has become a method of choice. ASO corrects pathophysiology of D-TGA both anatomically and functionally. Psychomotor development of children, who underwent ASO, is comparable with that of healthy population.

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Received December 14, 1998.
Accepted April 30, 1999.