COMPLETE TRANSPOSITION OF THE GREAT ARTERIES – SURGICAL RESULTS AND PROGNOSIS

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ABSTRACT

OBJECTIVES: The aim of this study was to analyze the surgical results and the prognosis in patients with complete transposition of the great arteries (TGA).

STUDY DESIGN: A group of 64 children with complete TGA out of 272 patients with critical congenital heart disease (CCHD) from the region of North-East Bulgaria was retrospectively followed for a period of 25 years (1987–2011 year). Methods of investigations include: clinical examination, electrocardiography, conventional radiography, echocardiography and cardiac catheterization. Surgical results were based on a scale, elaborated by us.

RESULTS: Complete TGA is the most frequent CCHD in the region of North-East Bulgaria – 23%, and in 64.1% of the cases it was associated with other cardiac malformations. Surgical correction was performed in 41 of the children (64.1%) with TGA - an average 2.17 interventions on each patient. The overall postoperative mortality rate was 43.9% and it was highest if concomitant lesions were presented, such as pulmonary stenosis or atresia, criss-cross atrioventricular connection and others – up to 50-60%. The balloon atrioseptostomy was the most common initial palliative procedure, performed in 83% of the newborns at a mean age of 7.1days and a mortality rate - 8.8%. In 31 of the children (75.6%) a complete corrective intervention was made. The most frequent was arterial switch operation (ASO) - 16 children (51%). Six of the children died – 37.5%, but the mean age of which the surgery was performed was significantly higher – 32.2 days in contrast to 16.8 days for the whole group ($p<0.05$). The surgical results on the 5th year were assessed as good/very good in 4 of the children and satisfactory in 1 child. Senning procedure was performed in 8 of the patients (26%) with a mortality rate of 25%. Four of the children (13%) undergone Rastelli or REV procedure and 3 of them (10%) had one-chambered Fontan procedure.

CONCLUSIONS: Complete TGA is the most frequent CCHD. The associated cardiac malformations have significant influence upon the prognosis and the surgical results. On different reasons still significantly high number of children with TGA remains without corrective interventions. The fundamental corrective procedure is ASO and the delayed intervention leads to unsatisfactory postoperative results.

Key words: *complete transposition of the great arteries, critical congenital heart disease, surgical results, prognosis, evolution*

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Complete transposition of the great arteries (TGA) is the most frequent critical congenital heart disease (CCHD) and counts for 5-7% of all the congenital heart defects (13). It is described as atrioventricular concordance with ventriculoarteral discordance, leading to pulmonary and systemic circulation function in parallel, rather than in series

and inefficient circulation. The natural evolution of TGA is unfavorable – the mortality rate is 28.7% during the first week of life, 51.6% in the first month, and 89.3% till the end of the first year. Associated heart malformation can significantly influence the prognosis (6). According to Campbel the mean lifespan of the patients with TGA is 10 months. In isolated TGA it is barely 1 month, if a ventricular septal defect (VSD) is presented – 22 months, and in cases with VSD and pulmonary stenosis (PS) the lifespan can reach 5 years (2). The prognosis has significantly changed after development of surgical atrial septectomy in the 1950s, balloon atrial septostomy (BASS) in the 1960s and the following physiological procedure (atrial switch operation) and anatomic repair (arterial switch operation).

The purpose of this research was to analyze the surgical results and the prognosis in patients with complete TGA.

MATERIALS AND METHODS

A group of 64 children with TGA out of 272 patients with CCHD from the region of North-East Bulgaria was retrospectively followed for a period of 25 years (1987–2011 year). Methods of investigations include: clinical examination, electrocardiography, conventional radiography, echocardiography and cardiac catheterization. Surgical correction was performed in 41 of the children with TGA. The surgical results are described as very good, good, satisfactory and unsatisfactory based on the clinical condition of the patients, hemodynamic indices, rhythm and conduction disorders, other postoperative complications and the necessity of reoperation.

RESULTS AND DISCUSSION

Complete TGA is the most frequent CCHD in the region of North-East Bulgaria – 23%. Isolated TGA was established in 23 of the children – 35.9% of all the patients and 41.5% of the operated ones. The most common associated heart malformations were VSD and PS. Rarely presented defects associated with TGA were coarctation of the aorta (CoA) – 5 children, criss-cross atrioventricular connection – 2 children, aortopulmonary fenestration – 1 child. According to Martins and coauthors concomitant heart malformations are presented in 50% of the cases with TGA and the leading ones are VSD, often with anterior or posterior malalignement, and subpulmonic and subaortic obstruction (10).

The surgical activity in the patients with TGA in the region is 64.1% (41 children out of 64). Total performed 89 corrective procedures or 2.17 operation for each child. The most surgical operation undergone the children with CTGA with VSD and PS or atresia – 2.7 on each patient, and even 6 in some children (Table 1).

Table 1. Surgical results, lethality and survival rate in TGA

Type of TGA	All	Operated ones		Number operations	Operated and died		Patients alive	
		Count	Percentage		Count	Percentage	Count	% of all
Isolated	23	17	73,9%	32	8	47%	9	39,1
TGA/VSD	18	10	55,6%	21	2	20%	8	44,4
TGA/VSD/ PS	13	10	76,9%	27	6	60%	4	30,8
TGA/PS	2							
TGA/CoA	5	2	40,0%	5	1	50%	1	20
Other TGA	3	2	66,7%	4	1	50%	1	33,3
All	64	41	64,1%	89	18	43,9%	23	35,90%

The overall postoperative mortality rate in our patients with TGA was 43.9 % and the mean time of follow-up was 7.8 years. The lowest postoperative lethality was presented in patients with associated VSD - 20% and the highest was in: concomitant PS/ atresia – 60%, CoA, criss-cross atrioventricular connection and others - 50%. The overall survival rate in

the whole cohort, operated and unoperated patients with TGA, was 36%. The highest survival showed the cases of TGA associated with VSD – 44.4%, and the lowest survival - when CoA was presented – 20%. According to de Koning and coauthors the survival rate in patients with TGA, who undergone surgery, reaches 90% for the last 15 years with a low frequency of reoperation – 6% for a 10-year period and 88% survival without any clinical accidents (3).

The number of ***initial palliative surgical interventions***, which 41 newborns with TGA undergone, was 45 and they include: balloon atrial septostomy (BASS) – 34, atrial septectomy (ASE) – 1, aorto-pulmonary anastomosis (AoPA) - 6, banding of the pulmonary artery (BPA) – 2 and correction of the aortic arch (CAoA) - 2 (Fig.1).

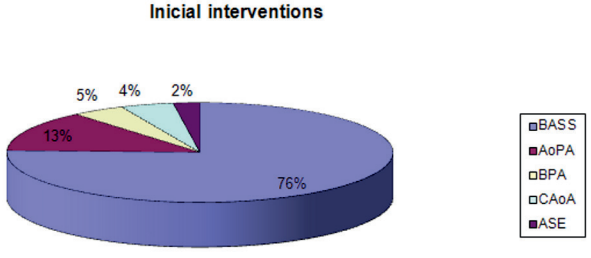

Fig. 1. Initial palliative interventions in TGA

BASS is the most common initial palliative procedure, performed in 34 of the newborns with TGA – 77% at the mean age of 7.1 days (0 days – 2 months). In 76% the intervention was made during the first week of life. The primary complications were: acute circulatory failure, septic arthritis and restriction of the newly created communication. Three of the newborns died – 8.8% at the mean time of intervention/exitus – 8 days. BASS is an established procedure (14), performed in Bulgaria in 1972-73, under echocardiographic control, as well as in many other centers in order to avoid radiation and for quick assessment of the results (7, 12).

Complete corrective surgical interventions were performed in 31 of the patients with TGA – 75.6% and included: atrial correction (Senning operation) - 8 (26%), arterial switch operation (ASO) – 16 (51%), Rastelli or REV operation – 4 (13%) and one-chambered Fontan procedure – 3 (10%) (Fig. 2).

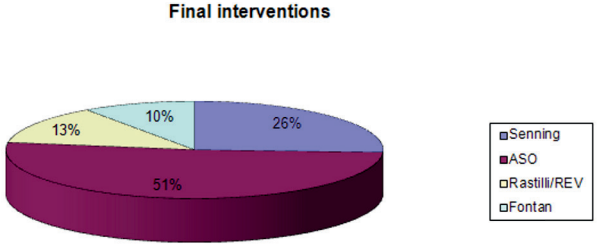

Fig. 2. Complete corrective surgical procedures in TGA.

In more than a half of the patients with TGA was performed ***ASO*** – 51% (16 children) at the mean age of 16.8 days (4 days - 1 month 22 days). The early postoperative complications were: infections, unstable hemodynamic and heart failure, respiratory failure, rhythm and conduction disorders and acute renal failure. Six of the children died – 37.5%, but the mean age of which the surgery was performed was significantly higher – 32.2 days ($p<0.05$). The mean time operation/exitus was 18 days (3-52 days). Unfavorable factors for the children who died after ASO were low birth weight, coronary anomalies, late diagnosis and intervention (Table 2).

Table 2. Age at surgical intervention and lethality in patients with ASO

Type of TGA	ASO Count	Age at the time of operation		Died	
		Borders	Mean	Count	Percentage
Isolated	10	6d.-1m.9d.	15,7 days	5	50%
TGA/VSD	6	4d.-1m.22d.	18,5 days	1	16,70%
All	**16**	**4d.-1m.22d.**	**16,8 days**	**6**	**37,50%**

The mean time of follow–up of the patients, who undergone ASO (n=10) was 5.5 years (1,2 years – 12.8 years). The surgical results on the 5th year after the procedure in 5 of the children were good/very

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good in 4 of the patients and satisfactory in 1 child with sinus bradicardia and supravalvular pulmonary stenosis, with elevated end-diastolic pressure in right ventricular, border EF – 59% and mild diastolic dysfunction. In 2 of the children the results in the 10th year after the operation were assessed as good and very good. Insignificant postoperative complications were observed: mild supravalvular pulmonary stenosis, PI – I-II gr., dilatation of the aortic root and minimal residual ventricular left to right shunt. Postoperative 15-year survival rate, according to literature database, reaches 88% (8), and reoperations were rarely performed (4.5 – 18%) mostly because of the development of supravalvular pulmonary stenosis at the place of the vessels reconstruction (1). The success of the operation is based on the corrective elimination of the obstructive lesions and coronary anatomy (9). In 1993 in Bulgaria was performed the first anatomic correction of TGA.

Senning procedure was performed in 8 of the children with TGA during the first years of the research - 26% at the mean age of 13.1 months (8 month – 1.9 years). The most frequent postoperative complications were: pleural effusions, rhythm and conductions disorders (4 children, in 3 of them a pacemaker was implanted), obstruction of the systemic venous atria and subpulmonary obstruction. Two of the children died – 25% (Table 3).

of the newly-created atriums, residual shunt at the atrial level, dysfunction of the right ventricular or pulmonary obstructive disease. Predecessors of unfavorable outcome are arrhythmias and advanced heart failure (4).

Three of the children with TGA undergone ***Rastelli procedure***, and in one child was performed unsuccessfully ***REV operation***. The mean age of the patients was 22 months (9 month - 4,5 years). Early postoperative complications includes: unstable hemodynamic with continuous mechanic ventilation, complete AV- block, and acute renal failure. Two of the children died – 50%, immediately after the operation and in the child who undergone REV operation was made an attempt to perform one-chambered correction. Based on the literature database the priority of REV operation over Rastelli is in the absence of extracardiac conduit and in better positioning of the intracardiac tunnel, because of resection of the muscular septum (5, 15). The two alive patients, who undergone Rastelli operation are at a mean age at the closure of the investigation of 13.5 years and postoperative time – average 10 years and 9 months. The final surgical results were assessed as satisfactory and unsatisfactory.

Palliative Fontan procedure was performed in 3 of the children with TGA at mean age of the bidirectional Glenn anastomosis - 1.9 years and

Table 3. Senning operation in TGA

Type of TGA	Count	Mean age at which the operation was performed (month)	Died	
			Count	Percentage
Isolated	5	11,4	1	20
TGA/VSD	3	24	1	33,3
All	8	13,1 month	2	25%

The mean age of the survivors after Senning procedure (n=10) was 16.4 years with postoperative time 15.2 years. The final surgical results were evaluated as very good/good in 2 of the patients, satisfactory in 2, and unsatisfactory, again in 2 of the children. According to literature database the early death-rate in these operations reaches 16 %, and 30-year survival rate is 67.2% (11). The most frequent complications are damage of the sinus node, supraventricular tachycardia, obstruction in TCPC – 4.4 years. One child with D-TGA, VSD and PS died (33%) on the 8th day after TCPC and two unsuccessful reoperations. In two of the survived children, age 7 and 8 years, at mean time of last operation 3.1 years, the surgical results were evaluated as good and unsatisfactory.

The overall mortality rate in our patients with complete TGA after different surgical correction is presented in Fig. 3.

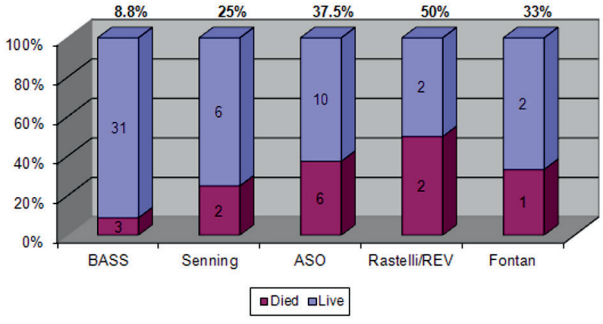

Fig. 3. Lethality rate after basic intervention in patients with TGA

CONCLUSION

Complete TGA is the most common CCHD – 23%. The associated heart malformations have significant influence on the prognosis and the surgical results. On different reasons still significantly high number of children with CTGA remains without corrective interventions. The fundamental corrective procedure is ASO and the delayed intervention leads to unsatisfactory postoperative results.

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