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Long-term outcomes after the arterial switch operation for D-transposition of the great arteries – a single centre experience

Dolgoročni rezultati po anatomski korekciji D-transpozicije velikih arterij – izkušnja enega centra

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Abstract

Background: Arterial switch operation (ASO) is the treatment of choice for D-transposition of great arteries (D-TGA). The purpose of the study was to evaluate the long-term outcome after ASO.

Methods: We either retrospectively reviewed the documentation or reassessed 38 patients (30 men, 8 women) during the transition period from adolescence to adulthood (age at the last clinical examination 17.1 ± 1.4 years) born from 2000 until 2005 with D-TGA and performed early ASO. We assessed: NYHA functional class, residual changes in neoaorta and neoaortic valve, neopulmonary valve, and pulmonary arteries, right and left ventricular function, physical performance, and signs of myocardial ischaemia.

Results: There was no mortality confidence interval (0.00-0.09). Thirty-two patients (84.2%) were in NYHA class I, and 6 patients (15.7%) were in NYHA class II. In 83.3% of patients, the aortic root was dilated (20.9 ± 2.8 mm/m^2, max. 27.7 mm/m^2). Neoaortic valve regurgitation was present in 27 patients (90%). There were no differences in bulbus width normalized to body surface area between groups without, mild or moderate neoaortic regurgitation (p = 0.6). Neopulmonary valve regurgitation was present in 58.1%. Due to an obstruction in the right ventricular outflow at any level, reoperation was required in one case and percutaneous dilatation of the neopulmonary valve in 1 case. Due to coronary artery complications, surgery was required only in one case because of myocardial infarction during exertion.

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Conclusion: Late results after complete correction in patients with D-TGA are good and comparable to larger centres. There was no mortality; most patients were asymptomatic, with normal systolic function of both ventricles and normal physical performance. Reoperations and percutaneous procedures were quite rare and successful in the long run.

Izvleček

Izhodišča: Anatomska korekcija (ASO) je zdravljenje izbire za D-transpozicijo velikih arterij (D-TGA). Namen raziskave je bil oceniti dolgoročno uspešnost anatomske kirurške korekcije.

Metode: Delno retrospektivno smo pregledali dokumentacijo ali ponovno ocenili zdravstveno stanje 38 bolnikov (30 moških, 8 žensk) v času tranzicije mladostnikov v odraslo dobo (starost ob zadnjem kliničnem pregledu 17,1 ± 1,4 let), ki so bili rojeni od leta 2000 do leta 2005 z D-TGA in so imeli ASO. Ocenili smo: funkcijski razred po NYHA, spremembe, ki so ostale, na neoaorti in neoaortni zaklopki, neopulmonalni zaklopki in pljučnih arterijah, funkcijo desnega in levega prekata, telesno zmogljivost in znake ishemije miokarda.

Rezultati: Nihče v skupini ni umrl (interval zaupanja (0,00-0,09). 32 bolnikov (84,2 %) je bilo v NYHA I, 6 bolnikov (15,7 %) je bilo v NYHA II. Pri 83,3 % bolnikov se je bulbus aorte razširil (20,9 ± 2,8 mm/m^2, max. 27,7 mm/m^2). Pri 27 bolnikih (90 %) je bila prisotna regurgitacija neoaortne zaklopke. Med skupinami brez, z blago ali zmerno neoaortno regurgitacijo ni bilo razlik v širini bulbusa, normaliziranih na telesno površino (p = 0,6). Regurgitacija neopulmonalne zaklopke je bila prisotna pri 58,1 % bolnikov. Zaradi obstrukcije v iztoku iz desnega prekata je bila potrebna ponovna operacija v enem primeru in perkutana dilatacija neopulmonalne zaklopke v enem primeru. Zaradi zapletov na koronarnih arterijah je bil potreben le en kirurški poseg zaradi miokardnega infarkta med naporom.

Zaključek: Pozni rezultati po anatomski korekciji D-TGA so dobri in primerljivi z večjimi centri. Nihče ni umrl, večina bolnikov je bila brez simptomov, z normalno sistolično funkcijo obeh prekatov in z normalno telesno zmogljivostjo. Ponovne operacije in perkutani posegi so bili dokaj redki, toda dolgoročno uspešni.

1 Introduction

D-transposition of the great arteries (D-TGA) accounts for 5–7% of congenital heart defects (1). Associated defects are common, the most common being the ventricular septal defect (VSD), about 50%.

D-TGA means that the aorta arises from the right ventricle and the pulmonary artery arises from the left ventricle. Transposition of the great arteries results in separate pulmonary and systemic blood circulation. Discordant ventriculo-arterial connection without a shunt between parallel circulations is not compatible with life. Therefore, a new-born needs a large enough shunt at the atrial level. If there is no associated major damage to the atrial septum, percutaneous surgery is performed via balloon atrial septostomy (the procedure pioneered by William Rashkind, a cardiologist). This is followed by arterial switch operation, (ASO), which should be performed as soon as possible, usually at the age of about two weeks.

An older method of correcting D-TGA is surgical atrial switch operation, introduced by Mustard and Senning. It allowed patients to survive into adulthood (2,3). The downside of this operation was that the right ventricle was a systemic ventricle. The long-term consequences of atrial correction of D-TGA were systemic ventricular dysfunction, regurgitation of the systemic atrioventricular valve (anatomically a tricuspid valve), supraventricular arrhythmias, baffle obstruction, and sudden cardiac death.

With the development of cardiac surgery, a new way of correcting D-TGA was introduced in the 1970s, namely by redirecting blood flow at the level of the great arteries. This way, anatomical correction of D-TGA is achieved (4). It involves switching the pulmonary artery and aorta above their sinuses and reimplanting the coronary artery into the neoaorta. To achieve this, the pulmonary artery bifurcation must be placed in front of the aorta using the LeCompte maneuver (5). With this procedure, the left ventricle remains the systemic ventricle and the supraventricular arrhythmias that occur during atrial diversion of blood flow due to scarring in the atria after surgery are avoided.

Although short-term and medium-term results after ASO surgery are usually favourable, there may be residues or complications that need to be identified as soon as possible. ASO means that the native pulmonary valve and the root of the pulmonary artery become the neoaortic valve and root. Therefore, neoaortic dilation and aortic regurgitation may occur.

Another consequence of ASO is supravalvular pulmonic stenosis, possibly branch pulmonary artery stenosis at the anastomosis site, or stenosis of the right ventricular outflow tract (6).

In rare cases, impaired left or right ventricular function occurs for a variety of reasons.

A very important consequence or complication after ASO is myocardial ischaemia, which is detected by a stress test. Possible morphological changes in the proximal segments of the coronary arteries are demonstrated by imaging methods. Myocardial ischaemia has several causes, from congenital anomalies of the coronary arteries to early and late consequences of reimplantation of coronary arteries during ASO.

Patients who underwent ASO for D-TGA are monitored according to existing guidelines (7), which were published only in 2017.

The aim of the study was to determine the late consequences of ASO in patients born with D-TGA at the end of growth before they are handed over for further follow-up to a cardiologist. To this end, we have decided to examine a group of patients, aged 15–20 years, after ASO for D-TGA in the transition period.

2 Methods

The study included all patients aged 15-20 years after ASO for D-TGA who were monitored by the transitional cardiology care clinic, Division of Paediatrics, University Medical Centre Ljubljana. These patients were born between 2000 and 2005. Originally, there were 40 such patients, of which two dropped out of the register (the first due to relocation, and the second refused check-ups), leaving 38 patients in the final treatment. Patients were monitored regularly once a year after surgery.

Patients were operated by the same method in two foreign centres: 22 patients in Bratislava and 16 patients in London.

Data were obtained partly retrospectively from documentation for the period up to the age of 15. The last treatment of patients after the age of 15 was in accordance with the guidelines for monitoring patients with D-TGA (7).

Patients were monitored on an outpatient basis. An extended history of symptoms and signs of heart failure, heart rhythm disorders, and chest pain was obtained.

An echocardiogram was performed to assess heart valve function as well as left and right ventricular function (8). To further define the function of the left and right ventricles and their sizes, magnetic resonance imaging (MRI) of the heart was made.

Patients underwent a stress test on a treadmill according to a customized Bruce protocol. The stress test was used to assess the patient's physical performance, cardiac rhythm disturbances, chest pain, and ECG changes characteristic of myocardial ischaemia. Myocardial perfusion scintigraphy was performed in three patients and stress echocardiography in one patient.

To assess coronary anatomy, computed tomography angiogram (CT) or MRI of the coronary arteries or coronary angiogram was performed. A blood sample was taken from 20 patients to determine NT-proBNP levels.

The research was approved by the Medical Ethics Committee of the Republic of Slovenia (No. 0120-193/2019/4, dated 23 May 2019).

2.1 Statistical treatment

The normality of the distribution of variables was tested by the Kolmogorov-Smirnov test. Values of variables that were normally distributed were shown by mean value and standard deviation, and values of variables that were not normally distributed were shown by median and the interquartile range. The confidence interval (CI) was calculated using the Clopper-Pearson method.

Analysis of variance (ANOVA) with the Bonferroni correction was used to compare the three groups. $P<0.05$ was considered statistically significant. Statistical analysis was performed with IBM SPSS version 26.0.

3 Results

The characteristics of the patients are shown in Table 1. Most of the new-borns were male. One was born at gestational age of 34 weeks and one at 36 weeks of age. All new-borns except the one born at only 34 weeks of age had a normal birth weight.

No one in the group died (CI (0.00–0.09).

There were no limitations in physical activity in 32 children (84%, CI 69–94%), who were classified into class I according to NYHA, and six patients were classified into class II according to NYHA (16%, CI 6–31%). No one was receiving medication for the underlying disease at the last clinical examination.

All patients were in sinus rhythm, 17 patients (45%, CI 29–62%) had a complete right bundle branch block (RBBB) and nine patients (24%, CI 11–40%) had an incomplete RBBB.

NT-proBNP values could only be measured in 20 patients (53%). As this was a partial retrospective study, some measurements were not made. The mean value was 46 (33.9) ng/L (max. 129 ng/L, min. 13.8 ng/L).

Table 1: Patient characteristics.

Patient characteristics	Number (%)
Age at ASO, days	14 (7.6)
Gender (M/F)	30/8 (79%, 21%)
Gestational age, weeks	39.4 (1.9) (min. 34, max. 42)
Birth weight (g)	3,480 (389.4) (min. 2,060, max. 4,320)
BSA (m^2) at last check-up	1,8 (0.2) (min. 1.51, max. 2.3)
D-TGA + VSD	9 (23.6%)
Associated anomalies: • ASD, • Coarctation of the aorta, • Muscular VSD, • interrupted aortic arch, • coronary artery anomalies.	 2 (5.2%) 5 (13.1%) 2 (5.2%) 1 (2.6%) 7 (18.4%)
Age at last cardiac examination (year)	17.1(1.4)

Legend: ASO – arterial switch operation; D-TGA – D-transposition of the great arteries; ASD – atrial septal defect; VSD – ventricular septal defect; min. – minimum; max. – maximum. The values are presented as the average (standard deviation).

Neoaortic root could be measured by echocardiography and MRI examination in 33 patients (87%). In only 5 patients the aortic root diameter was normal (17%, CI 4–28%) (15.6 (2.0) mm/m^2). In others, the aortic root was dilated (83%, CI 72–96%) (20.9 (2.8) mm/m^2), the largest measured dimension was 27.7 mm/m^2.

Neoaortic valve regurgitation was detected in 27 patients out of 38 (71%, CI 54–85%). In 21 cases the regurgitation was mild and in six it was moderate. Data on regurgitation and aortic root diameter are available for 30 patients. No differences in aortic root diameter with respect to body surface area were observed between groups without regurgitation, with mild regurgitation or with moderate regurgitation (analysis of variance, p=0.6).

The neopulmonary valve was shown in 31 patients (82%, CI 66–92%). In 18 cases the regurgitation was mild (58%, CI 31–64%), and in three (9%, CI 2–21%) moderate.

In four (11%, CI 3–25%) cases we observed mild neopulmonary valve stenosis, in six (16%, CI 6–31%) cases there was a mild stenosis of the pulmonary artery and in five (13%, CI 4–28%) cases there was a narrowing of the pulmonary artery branch after surgery. In one case (3%, CI 1–13%) reoperation was required due to the neopulmonary valve stenosis and in another case only percutaneous dilation of the valve was necessary.

Left ventricular systolic function was assessed by echocardiography (2D by Simpson's method, 3D) and by cardiac MRI (Table 2). Left ventricular ejection fraction was reduced in six patients (16%, CI 6–31%) out of 38.

It was not possible to accurately measure the diameter of the neoaortic root and to assess the degree of regurgitation in all patients due to technical reasons (poor ultrasound visibility, CT and MRI imaging difficulties due to tachycardia, artifacts, postoperative scars, poor patient co-operation).

Right ventricular systolic function could be numerically assessed only in 30 patients. In eight, it was assessed only as seemingly normal. Right ventricular systolic function was assessed by echocardiography or MRI in 30 patients (87.9%) (Table 3). Impaired systolic function was detected in eight patients (27%, CI 10–37%).

Table 2: Left ventricular ejection fraction (echocardiography - Echo and/or magnetic resonance imaging - MRI).

Method	Average ejection fraction %	Lowest ejection fraction %	Highest ejection fraction %	Number of evaluated subjects
2D echo	55.5 (7.3)	41	70	20
3D echo	51.2 (8.9)	42	68	10
MRI	56.8 (4.2)	50	61.3	20

Legend: Echo – echocardiography; 2D – two-dimensional echocardiography; 3D – three-dimensional echocardiography; MRI – magnetic resonance imaging. The values are presented as the average (standard deviation).

Table 3: Right ventricular systolic function (echocardiography - Echo and/or magnetic resonance imaging - MRI).

Method	Average	Min.	Max.	Number of evaluated subjects
TAPSE (cm)	1.7 (0.5)	1.1	3.3	25
S'_t (cm/s)	7.4 (1.8)	4.1	10	16
Ejection fraction (MRI) (%)	57.7 (4.3)	49.6	66.6	20

Legend: TAPSE – tricuspid annular plane systolic excursion; Min. – minimum; Max. – maximum; S't – S' tissue doppler velocity of lateral tricuspid annulus; MRI – magnetic resonance imaging. The values are presented as the average (standard deviation).

3.1 Stress testing

Stress testing was performed on 33 of 38 patients (86.8%). Of these, three patients underwent myocardial perfusion scintigraphy and one patient underwent additional stress echocardiography. All five patients who never underwent a stress test had normal coronary artery morphology. In 14 patients, the stress test was abnormal: decreased performance (10 patients), chest pain (two patients), ventricular arrhythmias (one patient), and ECG changes (one patient).

3.2 Coronary artery disease diagnosis

Coronary angiography was performed in 20 patients, CT coronary angiogram in 17 patients. The origins of coronary arteries and their proximal course were assessed by MRI in 19 patients. More than one imaging of coronary arteries was performed in 12 patients. The typical normal course of coronary arteries after ASO is shown in Figure 1.

Two of 38 patients did not have a morphologic imaging of coronary arteries. In both cases, the stress test was normal.

Prior to ASO, we were able to record only 13 descriptions of coronary artery course from the documentation. Different variants in the coronary artery course before ASO were identified in seven patients: left circumflex coronary artery (LCX) originating from the right coronary artery (RCA) (three patients), single coronary artery (one patient), RCA arising from sinus 1 (one patient), the left anterior descending coronary artery (LAD) from sinus 2 (one patient), and single origin of LCX and RCA (one patient). Only two

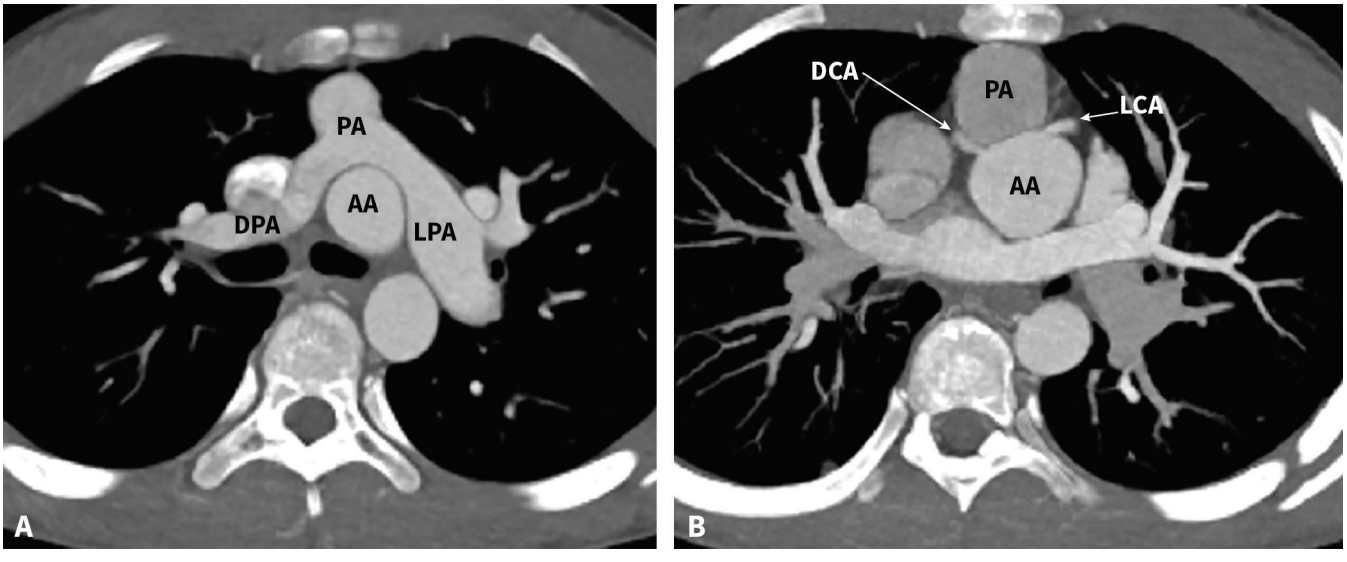

Figure 1: Computed tomographic angiogram (CTA) after the arterial switch operation (ASO) in a patient with D-transposition of the great arteries (D-TGA) - normal.
A – normal position of the pulmonary artery after surgery; B – normal right and left coronary artery origin.
Legend: AA – ascending aorta; DCA – right coronary artery; DPA – right pulmonary artery; LCA – left coronary artery; LPA – left pulmonary artery; PA – pulmonary artery; ASO – arterial switch operation; D-TGA – D-transposition of the great arteries.

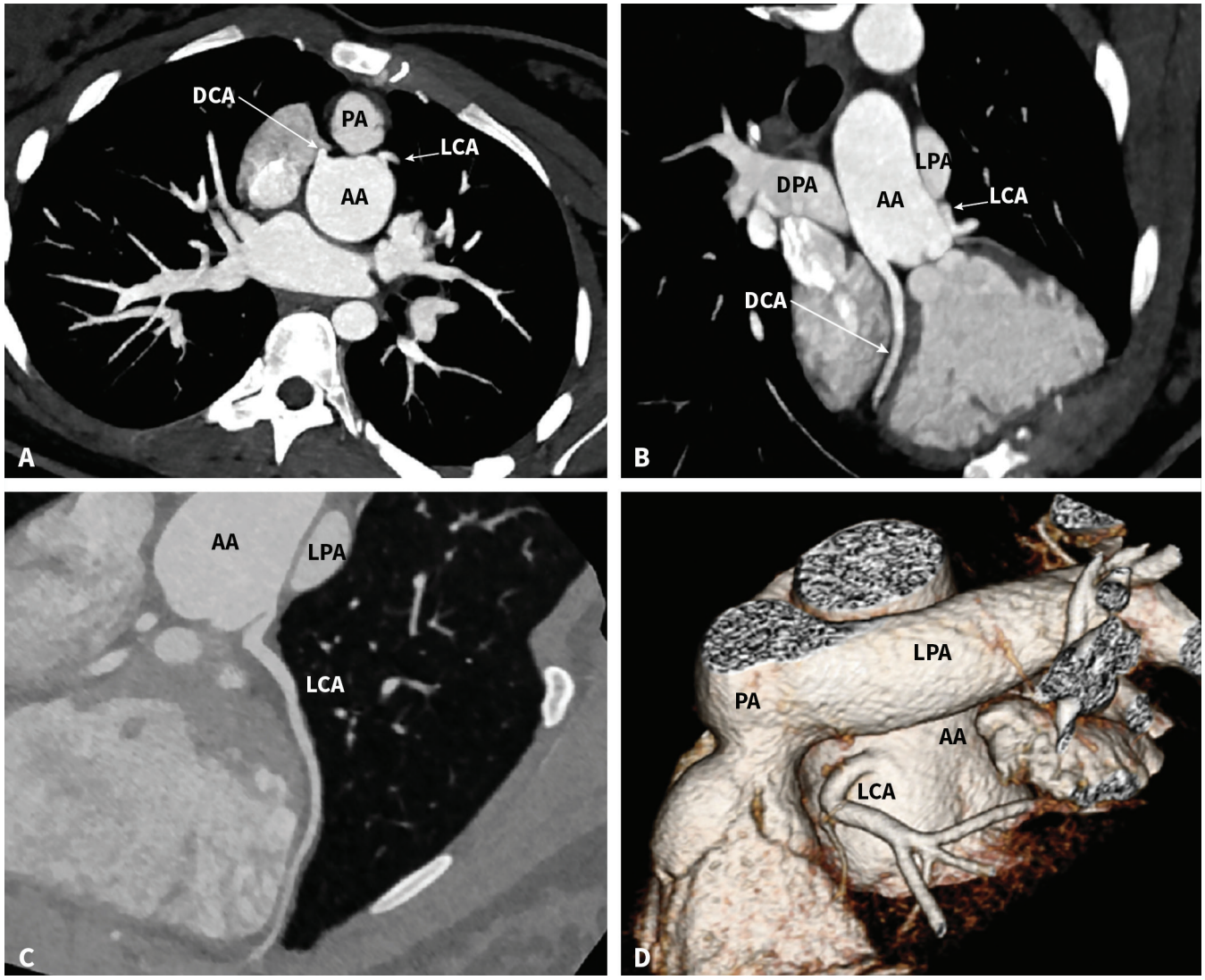

Figure 2: Computed tomography angiogram (CTA) after the arterial switch operation (ASO) in a patient with D-transposition of great arteries (D-TGA) - acute angle take-off of the anomalous left coronary artery, the initial part partly runs within the vessel wall.

A, B – multiplanar reconstruction (MPR); C – curved MPR of left coronary artery; D – 3D of the root of the left coronary artery.

Legenda: AA – ascending aorta; DCA – right coronary artery; DPA – right pulmonary artery; LCA – left coronary artery; LPA – left pulmonary artery; PA – pulmonary artery; ASO – arterial switch operation; D-TGA – D-transposition of the great arteries; MPR – multiplanar reconstruction.

of these patients had an abnormal stress test at a later period (a patient with a single coronary artery and a patient with a single LCX origin at birth).

Abnormalities in the coronary artery course after ASO were found in eight patients (21%, CI 10–37%): single coronary artery (one patient), LCX originating from RCA (three patients), high take-off of the left coronary artery (LCA), a further course at an acute angle (Figure 2) causing arterial stretch (three patients) and an intramyocardial course of the LAD (one patient). In seven (18%, CI 8–34%) of eight patients, the stress test was abnormal.

In these seven patients, for whom the stress test was abnormal and the coronary artery visualization was normal, we found other reasons that the stress test was positive:

residual coarctation of the aorta, moderate neoaortic valve regurgitation, impaired left ventricular function, and stenosis of the neopulmonary valve or pulmonary arteries.

3.3 Surgical and percutaneous procedures after arterial switch operation - ASO

Surgery was required in four patients (11%, CI 3–25%): surgical reconstruction of the LCA after non-ST-segment elevation myocardial infarction (one patient), dilation of the right ventricular outflow tract with insertion of a biological pulmonary valve (one patient), ASD closure (one patient), and pacemaker insertion (one patient).

Percutaneous coronary interventions were performed in five patients (13%, CI 4–28%): percutaneous ASD closure (one patient), insertion of a vascular stent in the area of aortic recoarctation (two patients), percutaneous closure of aortopulmonary collaterals (one patient), and balloon dilation of pulmonary valve stenosis (one patient).

We found that only three patients out of 38 (8%, CI 2–21%) had no complications or residual anomalies.

4 Discussion

Patients with D-TGA born in Slovenia had the arterial switch operation performed abroad until 2019, when we started with surgery on these patients at the University Medical Centre in Ljubljana. All patients included in the study were operated on abroad in two centres using the ASO method.

As abnormalities and complications may remain present after ASO, appearing at different ages, patients were monitored once a year in childhood. During the transition to adulthood, patients underwent clinical examination and non-invasive cardiac tests according to the latest guidelines, with special regard to possible residues and late complications of ASO, which should be monitored in adulthood due to possible progression of the defect and to plan any possibly necessary subsequent surgical correction.

The group of adolescents included in the study did not differ in basic characteristics from similar groups described in the literature. D-TGA is a rare congenital heart defect. According to our observations, the incidence in Slovenia is around 37 cases per 100,000 live births, which is similar to what other authors state (9). Regarding gender, our group also indicates a lower number of affected girls, which is in line with the literature (10).

In our group, two children were born prematurely, and one of them also had a low birth weight. Premature birth and lower birth weight are slightly more common in those with D-TGA than in the healthy population (11).

None of the patients born between 2000 and 2005 died after ASO. In principle, mortality after ASO is low, also depending on associated heart defects (12). The literature reports a mortality rate of 1.6% in the 19 years of follow-up, but in a 10-fold larger group of patients than ours (13). Our research is comparable to the latter, as the confidence interval in our research is 0–9%. The age of the patients at the last cardiac test was 17.1 years, that is, at the end of growth. This means we were able to consider adult values in imaging diagnostic methods.

Associated heart defects were the same as reported by other authors. Among the associated defects that required correction, interrupted aortic arch, coarctation of the aorta, and ASD were noted. ASD after atrial septostomy and an open Botallo duct were not included. The frequency of associated anomalies is difficult to compare with other authors due to the small size of our group.

The vast majority of patients did not report any heart problems, and some were even more active in sports than just recreationally. Problems were reported by six NYHA II patients, slightly more than in similar studies, where only 2.7% of NYHA II patients were reported (13). Everyone in the NYHA class II had residues or complications after ASO (aortic reconstruction, myocardial ischaemia, pulmonary artery stenosis). Four of them required repeated surgery or percutaneous procedures.

Regarding heart conduction disorders, our results were consistent with the literature; incomplete RBBB is described in 26% and complete RBBB in 21% (13).

No signs of heart failure were observed in any patient. Measurements of NT-proBNP were possible in only 53%. All values were normal. We believe that the determination of NT-proBNP during the transition of a patient to a cardiologist is an important parameter for further monitoring.

A common finding after ASO is neoaortic root dilation and neoaortic valve regurgitation, as after ASO, the pulmonary valve and the root of the neoaorta (formerly the pulmonary artery) are exposed to systemic pressure. In our study, we found neoaortic root dilation and neoaortic regurgitation in most patients, consistent with other major studies (13,14). Regurgitation was mild, moderate in only six cases, similar to what other authors have observed (mild regurgitation in 47%, moderate regurgitation in 3.4%) (13,14). So far, no one has required surgery due to the neoaortic dilation or neoaortic valve regurgitation, but regular

monitoring was required. Other authors also cite a low proportion of surgical correction of neoaortic regurgitation and aortic root dilation, especially in young people under 15 years of age (15,16,17). Longitudinal studies show that neoaortic dilation and regurgitant neoaortic valve occur within ten years in half of patients who underwent ASO, but the progression of neoaortic valve regurgitation and root dilation is slow and stabilizes in adulthood (6,18). Given that no differences were found between groups with different degrees of regurgitation and root diameter normalized to body surface area, it can be concluded that the latter does not affect the degree of regurgitation. However, all patients had mild neoaortic dilation. Other risk factors for the development of neoaortic regurgitation include the presence of VSD at birth, pulmonary artery banding, and neoaortic bicuspid valve (19). Mild changes in the neoaortic valve were also observed in patients with moderate aortic regurgitation, and some also had VSD at birth. However, no one has needed a neoaortic valve correction, so only monitoring is foreseen.

Obstruction in the right ventricular outflow after ASO occurs on several levels. Stenosis of the pulmonary artery and/or its branches is most common, less common is stenosis of the neopulmonary valve. The cause of stenosis is inadequate growth at the neopulmonary anastomotic site. It is also the most common reason for surgical or percutaneous corrections in early childhood (20). In our group, in almost half of the cases the obstruction occurred at one of the usual levels, but the stenosis was only mild. Other authors also report, similar to our study, mild to moderate neopulmonary valve stenosis in about 6% and stenosis of the branches of the pulmonary arteries in 22–24% (13). However, we have detected slightly fewer supravalvular aortic stenoses compared to others who reported supravalvular aortic stenoses in as many as 38% of patients (13).

In one case, the neopulmonary valve had to be replaced with an artificial one, and in one case, percutaneous dilation of the stenotic neopulmonary valve had to be performed. Both procedures were performed in early childhood. However, we found several cases of neopulmonary valve regurgitation. Regurgitation was moderate in only two cases, which is in line with expectations and data from the literature, which mention mild neopulmonary valve regurgitation in 54% and moderate regurgitation in 4.4% (13). The variability of neopulmonary valve regurgitation incidence after ASO is very wide (9–80%).

Left ventricular systolic function is rarely impaired. In our group, only six patients (16%) had a slightly reduced left ventricular ejection fraction, which is in line with data from other authors who mention a reduced left ventricular ejection fraction in 4.4% (13). Mildly reduced systolic function was attributed to moderate neoaortic regurgitation (two patients), reoperations (two patients), single coronary artery (one patient), and ischaemia due to anomalous LCA course in one patient. Compared to similar research, the share is larger (13). The difference can be explained by the small size of our group and is probably also due to the different methods of calculating the ejection fraction (Echo, MRI). However, the assessment of systolic function with the assessment of myocardial strain shows that the global longitudinal strain is also reduced in patients after ASO, who had the ejection fraction estimated as normal with other methods. The decrease in global longitudinal strain was correlated with the age of patients at ASO (21). In our group, such measurements were not performed enough for us to be able to comment on this.

No data are available on the right ventricle systolic function long after ASO. Reduced right ventricle systolic function was detected in eight patients in the study. Mild or moderate neopulmonary valve regurgitation was present in five, one underwent reoperation due to right ventricular outflow obstruction, and in two, no cause of poor right ventricular function was identified. Mildly impaired right ventricular systolic function is commonly seen after surgeries using the extracorporeal circulation, especially in patients who have had multiple surgeries or surgeries in the past.

One of the main and most dangerous consequences of ASO is myocardial ischaemia, which may be due to a congenital anatomical anomaly of the coronary arteries or a changed coronary artery course after coronary ostial reimplantation. After ASO, intimal thickening of the proximal region of the coronary arteries may occur. Due to the altered blood flow and the consequential shear stress, fibrocellular thickening of the intima occurs. Coronary artery occlusion due to ostial fibrosis at the suture line is possible. Other mechanical causes are: acute angle take-off of the coronary artery, proximal coronary artery course between aorta and pulmonary trunk, kinking of the coronary artery, pressure on coronary artery with high elliptical index, myocardial bridge, and coronary artery fistula. These variants of abnormal coronary artery disease after ASO may not cause symptoms, but may be dangerous for sudden death, cardiac arrhythmias, or acute

coronary syndrome (22). All of these complications do not occur immediately after surgery, but in later years. Therefore, periodic stress testing of children is required if they are already able to participate in the test (7). In young children, 24-hour ECG Holter monitoring is used for the purpose of ischaemia registration. Exercise testing alone is not sufficient, as morphological imaging of coronary arteries with CT coronary angiography or coronary angiogram is also required. MRI can only show coronary artery origins and their initial course. This is often sufficient, as changes after coronary artery surgery are expected right in this part.

Elderly patients in our group underwent coronary angiography and stress testing in childhood before the age of 10. However, when CT coronary angiography became a more accessible method, morphological changes were being detected only with this method. No additional tests were performed in patients who underwent cardiac MRI for other reasons and had a good visual of origins and proximal course of both coronary arteries.

Anomalies in the course of coronary arteries for various reasons were found after surgery in eight patients (21%), in seven of them the stress test was abnormal, which is comparable to the literature, which reports finding anomalous coronary arteries in 34% of cases (13). Among patients with an abnormal stress test (14 patients) and normal coronary imaging, other causes were found.

Coronary artery surgery was required in only one patient aged 18 years, who had suffered a myocardial infarction without non-ST elevation after a long run. She had a completely normal stress test four years earlier and had no problems with normal exertion, so she did not undergo imaging tests of the coronary arteries. The cause of ischaemia was an abnormal course of the LCA arising from the aorta at an acute angle, causing a narrowing of the lumen of the LCA.

The example suggests that when monitoring patients after ASO, stress testing, as well as one of the imaging methods to show coronary artery morphology, should be performed several times. In doubtful cases, stress myocardial perfusion scintigraphy is considered to show myocardial ischaemia.

Myocardial ischaemia after ASO is quite common. In our group, anomalous course of coronary arteries was found in 21% of patients. For most, the stress test was found to be not quite normal. For some, myocardial perfusion scintigraphy or stress echocardiography was also opted for. Additional tests did not show enough changes in myocardial blood flow to opt for an intervention, so these patients were only monitored. Imaging tests of the coronary arteries revealed an already known congenital single coronary artery and a benign anomalous course of the coronary arteries as a result of changes that occurred during surgery.

Myocardial ischaemia after ASO is unpredictable and much more dangerous than other residues after ASO. For this reason, attention should be paid to the patient's problems, such as chest pain on exertion or at rest, collapse, palpitations, arrhythmias, ECG changes. Both stress testing and coronary artery imaging are highly required tests in all patients after ASO. Both tests are also particularly important in asymptomatic patients prior to counselling them on their lifestyle, recreation, and sports.

4.1 Research limitations

The research has many limitations. This is a partial retrospective study, so patient follow-up could not be standardized in most cases. Great progress has been made, especially in the area of imaging methods. Guidelines for treatment and follow-up of patients after ASO had not been published until 2017, so the treatment had not been uniform until then. Descriptions of coronary anatomy before surgery are incomplete and therefore comparison with the coronary situation after surgery is impossible in many cases.

It was not possible to accurately measure all observed parameters (NT-proBNP values, neoaortic root diameter, and neoaortic of regurgitation severity) in all patients. These reasons were purely objective and partly related to the technical limitations of the studies (poor ultrasound visibility, challenges in CT and MRI imaging due to tachycardia, artifacts, postoperative scars), rarely poor patient cooperation and the fact that the study was partly retrospective.

Our group of patients is small compared to the groups in the literature.

Despite all these limitations, we have shown the status of our patients with D-TGA after ASO during the transition to adulthood.

5 Conclusion

A study on late consequences of ASO due to D-TGA at the University Medical Centre Ljubljana included adolescents before their transfer to cardiologist care. We identified the most common residues and late complications after surgery. Neoaortic root dilation and neoaortic valve regurgitation were the

most common. Due to the remaining obstruction in the right ventricular outflow and stenosis at the pulmonary artery anastomoses, surgery was required in one patient and percutaneous dilation of the pulmonary valve in the other. Abnormal coronary artery course was observed in eight patients. Coronary artery surgery was required in only one patient who had a non-ST-elevation myocardial infarction during a long run.

We believe that the results of the treatment of these children are good, completely comparable to the results of larger centres. We did not record any deaths after ASO. Most patients were asymptomatic, with normal systolic function of both ventricles and normal physical performance. Reoperations and percutaneous interventions were quite rare and successful in the long run.

In particular, it has been shown that patients need to be monitored regularly on an annual basis and attention should be paid to the symptoms and signs of unpredictable myocardial ischaemia. It manifests itself suddenly, so it can pose the worst threat to this group of patients in any period after ASO.

Conflict of interest

None declared.

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