Delayed Arterial Switch Operation in a Three Year Old Child with Transposition Of Great Arteries

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Abstract
Background: The incidence of transposition of the great arteries (TGA) is 0.5-1% of all congenital heart diseases. The aorta and pulmonary arteries exit inversely from the heart ventricles. In addition, 25% of TGA patients have ventricular septal defect (VSD). Some infants may have left ventricular outflow tract obstruction. Arterial switch (Jaten) operation is the accepted procedure for the treatment of TGA.

Case report: The patient had congenital severe cyanosis and was diagnosed as TGA, VSD and ASD after a pediatric cardiology examination. The patient went to follow-up for a short time and did not go to follow-up and admitted to our institute with complaints of severe cyanosis and decreased exercise capacity at the age of 34 months. According to echocardiography and angiography findings, arterial switch operation (AS) was planned for treatment despite delayed treatment.

Conclusion: In general; it is preferable to perform AS in the first month of life in patients with TGA. There is very rare information in the literature about the preferability of early childhood patients. In the light of this information, we present a case of successful atrial switch operation in a three-year-old patient.

Background
In order to maintain life in TGA, circulating blood must be mixed in the heart or elsewhere before complete correction. Blood mixing is critical to the protection of life. VSD, atrial septal defect (ASD) or patent ductus arteriosus (PDA) is very important in these patients to allow blood to be mixed naturally. If this mixing is not possible naturally, an emergency catheterization or atrial septostomy (AS) is required (1). It is generally preferred for the first 3 weeks in the neonatal period for AS surgery. It is often not possible to perform AS operation when this time is delayed. To perform delayed correction, palliative cardiac surgery should be performed first (2). In this study; having previously undergone pulmonary bunding; A 3-year-old boy with TGA diagnosed with VSD and ASD is presented.

Case Presentation
The patient, who had severe cyanosis since birth, was diagnosed as TGA, VSD and ASD by pediatric cardiology examination in another hospital. Palliative surgery was planned to the patient who was
followed up with medical treatment until eight months ago and then referred to our institute for further examination and treatment. Pulmonary bunding and surgical procedures were performed and the patient was discharged. When the patient was 34 months old, he was admitted to our institute again with complaints of severe cyanosis and decreased exercise capacity. Echocardiography and angiography were performed. The pulmynary gradient was 68 mmHg (figure 1). Atrioventricular intercourse was normal, cardiac cavities were normal and ventricular contractions were normal. No ventricular wall hypoplasia was detected. Valvular morphology was normal. No coronary abnormality was detected in angiography (3). Pulmonary catheterization did not show pulmonary hypertension (figure 2).

AS surgery was planned. Cardiopulmonary bypass (CPB) was performed at 32 degrees Celsius. During the cooling period of the CPB, the right and left pulmonary arteries were channeled towards the hilus entry and the patent ductus arteriosus was divisible. Antegrade route has been used in blood cardioplegia to protect myocardial tissue. Debunding of pulmonary arteries, arteriotomy and supravalvular fibrous membrane resection were performed respectively. VSD was continued with right arteriotomy and patch suturing technique. Valvular dilatation was performed with 17 plugs. Valvular leakage was tested after enlargement and no valvular insufficiency was detected. AS Leevte maneuver was performed. After neoaorta and coronary artery anastomosis, the second dose of cardioplegia was given. Before Neopulmunar anastomosis; anastomosis leakage, fullness and position of the coronary arteries were observed (Figure 3).

Perfusion adequacy was tested by observing coronary sinus-induced cardioplegia. After neoaortal and coronary anastomosis, bovine patch reconstruction of distal neopulmunar arteries was performed. After completion of the distal anastomosis, aortic bone was opened. Neopulmonary anastomosis was performed with 7/0 prolene suture, while cardiac functions returned (Fig. 4). ASD was fixed with large bovine patch. A small opening is left for the patent foramen ovale (PFO) mechanism to continue. The atrium was re-closed and the inotropic agents dopamine and adrenaline started intravenous infusion. The operation was completed after the sternum was closed again. The aortic cross-clamp time was 62 minutes and the CPB time was 112 minutes, respectively. Postoperative inotropic dopamine was
continued and extubation was performed after 5 hours. Sinus rhythm was sufficient in cardiac monitoring and ventricular functions were sufficient in echocardiography.

Discussion And Conclusions

In the treatment of TGA; first in 1959 and 1963, respectively, Senning and Mustard modifications were made. Various complications related to the use of these modifications have been limited. Currently Jaten operations are the main recommendation in these patients. Arterial switch operation is a globally preferred method for total anatomical correction of TGA and was first performed by Jaten et al. Later modified by Lecompte in 1975.

If the pathological condition is not suitable for performing AS operation; Pulmonary bunding, Rastelli operation, Atrial switch, Damus-Stansel-Key or Nikaidoh operations are also preferable. (4,5). Survival is not more than 1 year in patients without early surgical treatment. Left ventricular pressure and contraction are the main factors for success in simple TGA surgery with intact ventricular septum. Physiologically, pulmonary vascular resistances (PVR) are high in the first 2 weeks of life, so left ventricular functions remain normal. After 15 days of life, PVR drops and left ventricular function will be against low pressure. Over time, left ventricular function will be insufficient (6,7).

Therefore, in patients with delayed treatment, left ventricular function will be against high systemic vascular resistances, so heart output will be low and inotropic agents may be needed. In the literature, operation is recommended if left ventricular stroke volume and wall thickness is normal and left / right pressure ratio is higher than 0.70. In developed countries, delayed cases are rare, but are more common in developing countries for a number of reasons. As in our case, patients who did not undergo AS operation in the neonatal period; Pulmonary banding and shunting is recommended. This procedure provides better post-ventricular load performance to prepare the patient for subsequent AS surgery. The time taken to improve left ventricular performance is inversely proportional to the age of the patient. If palliative pulmonary banding is performed for 3 weeks, it will be sufficient even in older people up to 3 months ago. In developed countries, in recent years, direct surgery is recommended for patients under 3 months of age. In childhood; pulmonary hypertension is another factor related to morbidity and mortality of complex TGA patients (8,9).
In conclusion, although our case was 34 months old, ASO operation was performed successfully. If palliative operation was performed in early neonatal period and secondary left ventricular functions and dimensions were within normal limits, pulmonary hypertension did not develop and angiographic coronary artery anomaly was not observed in TGA cases; even at 3 years of age, arterial switch operation can be performed easily. In our case, arterial switch operation was successfully performed at 34 months of age. We present the successful outcome of surgical treatment in patients with delayed surgical treatment because it is very rare in the literature.

Abbreviations
TGA: Total anomalous of great arteries
VSD: Ventricular septal defect
ASD: Atrial septal defect
AS: Atrial switch operation
PDA: Patent ductus arteriosus
CBP: Cardiopulmonary bypass
PFO: Patent foramen ovalis
PVR: Pulmonary vascular resistance

Declarations
Ethics approval and consent to participate have been taken from Altınbas University Medicine School Ethical Committie
Consent for publication have been taken from the patients’ parents. Patient’s parents gave informed written consent for their personal or clinical details along with any identifying images to be published in this study.
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Figure 1

Total infrarenal aortiliac occlusion
Figure 2

Total infrarenal aortiliac occlusion
Figure 3

Laseration and hematoma of graft in anastomosis line

Figure 4

Hematoma and graft view of pektoral muscle

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