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***Original Article*** / *Özgün Makale*

# Surgical options in complex transposition of great arteries

*Büyük arterlerin kompleks transpozisyonunda cerrahi seçenekler*

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***ABSTRACT***

***Background:*** In this study, we present our experience in selecting surgical approach for transposition of the great arteries and left ventricular outflow tract obstruction or aortic arch obstruction with ventricular septal defect and to report early and mid-term results.

***Methods:*** Between February 2007 and June 2012, a total of 18 patients (9 males, 9 females; median age 4.25 months; range, 12 days to 96 months) who were operated for transposition of the great arteries, ventricular septal defect, and left ventricular outflow tract obstruction or aortic arch obstruction were retrospectively analyzed.

***Results:*** Cardiac pathologies were transposition of the great arteries, ventricular septal defect and coarctation of aorta in four patients; transposition of the great arteries, ventricular septal defect and valvular pulmonary stenosis in two patients, and transposition of the great arteries, ventricular septal defect, valvular or subsubvalvular pulmonary stenosis in 12 patients. Arterial switch operation with ventricular septal defect closure and left ventricular outflow tract obstruction procedures were performed in nine patients, two of which were modified Konno operations. The other operations were arterial switch operation with ventricular septal defect closure and arcus reconstruction in four patients, Rastelli operation in three patients, and Nikaidoh operation in two patients. Median cardiopulmonary bypass and cross-clamp times were 228.5 min and 107 min, respectively. The median length of stay in the intensive care unit was 102.5 hours (range, 28 to 765 hours), while the median duration of intubation was 40.5 hours (range 17 to 275 hours). All patients were discharged within median seven days (range 5 to 55 days). The median follow-up was 37.7 months (range, 15 days to 74 months). Two patients who underwent Rastelli operation died due to low cardiac output in the intensive care unit. At the final echocardiographic examination, the median left ventricular outflow tract gradient was 12.4 mmHg (range, 2 to 38 mmHg) in the patients operated for left ventricular outflow tract obstruction, whereas the median descending aorta gradient was 13.5 mmHg (range, 7.8 to 28 mmHg) in the patients with arcus reconstruction. Only one patient with bicuspid neoaortic valve and posterior septal malalignment was reoperated due to a left ventricular outflow tract gradient of 38 mmHg.

***Conclusion:*** Our study results suggest that arterial switch operation is a preferable alternative, if the left ventricular outflow tract obstruction is resectable. Intraventricular re-routing procedures may be the choice in selected patients. We believe that choosing the optimal surgical technique demands appreciation of the particular anatomic features in each individual patient.

***Keywords:*** Surgical procedure; transposition of great vessels; ventricular outflow tract obstruction.

***ÖZ***

***Amaç:*** Bu çalışmada büyük arter transpozisyonu, ventriküler septal defekt ile birlikte sol ventrikül çıkım yolu darlığı veya arkus aort darlığında cerrahi yaklaşımın belirlenmesine yönelik deneyimimiz sunuldu ve kısa ve orta dönem sonuçlar bildirildi.

***Çalışma planı:*** Şubat 2007 - Temmuz 2012 tarihleri arasında büyük arter transpozisyonu, ventriküler septal defekt, sol ventrikül çıkım yolu veya arkus aort darlığı nedeniyle ameliyat edilen toplam 18 hasta (9 erkek, 9 kız; ortanca yaş 4.25 ay; dağılım 12 gün-96 ay) retrospektif olarak incelendi.

***Bulgular:*** Kardiyak patolojiler dört hastada büyük arter transpozisyonu, ventriküler septal defekt ve aort koarktasyonu, iki hastada büyük arter transpozisyonu, ventriküler septal defekt ve valvüler pulmoner darlık ve 12 hastada büyük arter transpozisyonu, ventriküler septal defekt, valvüler veya subvalvüler pulmoner darlık idi. İkisi modifiye Konno ameliyatı olmak üzere, toplam dokuz hastaya ventriküler septal defekt kapatılması ile birlikte arteriyel switch ameliyatı ve sol ventrikül çıkım yolu darlığına yönelik işlemler uygulandı. Diğer işlemler, dört hastada arteriyel switch ameliyatı ile birlikte ventriküler septal defekt kapatılması ve arkus rekonstrüksiyonu, üç hastada Rastelli ameliyatı ve iki hastada Nikaidoh ameliyatı idi. Ortanca kardiyopulmoner baypas ve kros klemp süreleri sırasıyla 228.5 dk. ve 107 dk. idi. Yoğun bakım ünitesinde kalış süresi ortanca 102.5 saat (dağılım 28-765 saat) iken, ortanca entübasyon süresi 40.5 saat (dağılım 17-275 saat) idi. Hastaların tümü ortanca yedi gün içinde (dağılım 5-55 gün) taburcu edildi. Ortanca takip süresi 37.7 ay (dağılım 15 gün-74 ay) idi. Rastelli ameliyatı yapılan iki hasta, düşük kalp debisi nedeniyle yoğun bakım ünitesinde kaybedildi. Son ekokardiyografik incelemede sol ventrikül çıkım yolu ameliyatı olan hastalarda ortanca sol ventrikül çıkım yolu gradyanı 12.4 mmHg (dağılım 2-38 mmHg) iken, arkus rekonstrüksiyonu yapılan hastalarda inen aort ortanca gradyanı 13.5 mmHg (dağılım 7.8-28 mmHg) idi. Neoaortik biküspid kapağı ve arka septal dizilim bozukluğu olan yalnızca bir hasta, sol ventrikül çıkım yolu gradyanı 38 mmHg olduğu için tekrar ameliyat edildi.

***Sonuç:*** Çalışma bulgularımız, arteriyel switch ameliyatının sol ventrikül çıkım yolu darlığı rezeke edilebilir ise, tercih edilen bir seçenek olduğunu göstermektedir. Seçilmiş hastalarda intraventriküler yeniden yönlendirme işlemleri seçenek olabilir. En uygun cerrahi tekniğin seçilmesinin her hastada spesifik anatomik özelliklerin anlaşılmasını gerektirdiği kanısındayız.

***Anahtar sözcükler:*** Cerrahi işlem; büyük damar transpozisyonu; ventrikül çıkım yolu darlığı.

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Patients with "transposition of the great arteries" (TGA) are included in a non-homogeneous group. Left ventricular outflow tract obstruction (LVOTO) is seen in 20 to 30% of these cases and is typically associated with ventricular septal defect (VSD).[1] Hypoplastic isthmus and arcus aorta with TGA are less common.[2] These pathologies are associated with noticeably higher surgical mortality rates than simple TGA.[2] There is still a considerable debate on the optimal surgical procedure which should be performed in this patient population. Resection of the LVOTO in combination with arterial switch operation (ASO) is favored, where applicable. If the LVOTO resection is unattainable, intraventricular re-routing operations are usually performed.[3] In this study, we aimed to present our experience in selecting surgical approach for TGA and LVOTO or aortic arch obstruction with VSD and to report early and mid-term results.

## PATIENTS AND METHODS

Between February 2007 and June 2012, a total of 18 patients (9 males, 9 females; mean age 4.25 months; range, 12 days to 96 months) who were operated for TGA, VSD, and LVOTO or aortic arch obstruction were retrospectively analyzed. The patients with Taussig-Bing anomaly, congenitally corrected TGA, or those with VSD without LVOTO were excluded.

The diagnoses and details of the cardiac anatomy were documented by reviewing the echocardiography and cardiac catheterization findings. The measurements of aortic, mitral, pulmonary, and tricuspid valve annuli were noted and Z scores for these measurements were calculated. The following variables were obtained from the intensive care unit (ICU) surveillance charts and surgery reports: age, weight, duration of cardiopulmonary bypass (CPB) and aortic cross-clamp, invasive blood pressure measurements, blood chemistry, and blood gas analyses, inotropic agent doses, duration of mechanical ventilation, and length of stay in the ICU.

*Inotropic score was calculated using the following formula:*

dopamine µg/kg/min x1 + dobutamine µg/kg/min x1 + milrinone µg/kg/min x15 + adrenalin µg/kg/min x100.[4]

The patients were divided into three groups based on applied surgical procedure. Those with TGA, VSD, and hypoplasia of the arcus aorta underwent ASO, VSD closure, and arcus aorta reconstruction (group 1). Those with TGA, VSD, and LVOTO either underwent direct procedures to remove LVOTO (i.e. valvotomy, myectomy, or modified Konno procedure) along with ASO and VSD closure (group 2), or underwent an intraventricular re-routing operation (i.e. Nikaidoh, Rastelli) (group 3).

The details of the cardiac anatomy of each patient were discussed among the pediatric cardiologists and cardiothoracic surgeons. Considering the echocardiographic and angiographic records, an operative decision was made based on the coronary and LVOT anatomy, size of the VSD, Z score of the pulmonary annulus and age of the patient. The decision for those with hypoplastic pulmonary annulus or LVOT (Z scores < –2), obstruction caused by a non-resectable tissue in the LVOT or coronary anomalies jeopardizing the safe transfer would be a type of intracardiac re-routing operation. Others were expected to benefit from the LVOT resection procedures.

The surgical option selected would be carried on during the operation, unless the anatomical details differed from that previously described. In that case, transesophageal echocardiography (TEE) would be performed to delineate the details and a new surgical decision would be made by the surgeon and the pediatric cardiologist in the operation room.

In four patients with TGA, VSD, and hypoplasia of the arcus aorta, the following operation technique was performed: The ascending aorta or the brachiocephalic artery was cannulated from the right lateral region. Under CPB, the patient was cooled to 22-24 °C. The pulmonary artery branches, arcus aorta, and descending aorta were dissected and the ductus arteriosus was divided. While cross-clamp was applied distally on the brachiocephalic or left carotid artery, a side-clamp was applied 2 cm distal to the coarcted segment of the descending aorta. Under selective cerebral and myocardial perfusion, the aortic reconstruction was done using glutaraldehyde treated autologous or bovine pericardium. The ascending aorta was reconstructed at the level of aortic cannula under short (5 to 10 min) total circulatory arrest (TCA). Subsequently, CPB was carried on with full flow-rate for 10 min. After application of cross-clamp and cardioplegic arrest, the VSD was closed mainly through the right atrium. Arterial switch was performed with coronary artery transfer and neoaortic anastomosis. The coronary arteries were transfered as described by Edward Bove.[5] A restrictive interatrial communication was, then, allowed.

There were 14 patients with TGA, VSD and pulmonary stenosis. Nine of them had operations for LVOTO along with ASO and VSD closure. Valvular lesions were repaired with valvotomy. The pulmonary valve incision was extended from the commissure towards the arterial wall to attain a 2-3 mm larger orifice.

Myectomy or membrane resection was done for simple resectable subvalvular LVOTO. All the muscular bands obstructing the outflow were resected intending an outflow tract which was at least 4 mm larger than the smallest aortic valve diameter consistent with the age of each patient. After weaning from CPB, left ventricular and neoaortic gradient were assessed through intraoperative TEE or by direct pressure measurements from the left ventricle and neoaorta.

For more complex (tunnel-shaped) LVOTO, modified Konno operation was performed.[6] Remaining five patients had intracardiac re-routing procedures such as Nikaidoh and Rastelli operations.

During these operations, VSD was closed through the right atrium in 10 patients and via right ventriculotomy in three patients. Transpulmonary approach was used in five patients. In the course of VSD closure, a Dacron patch was used in seven patients and an autologous pericardium was utilized in nine patients. Primary suture technique was applied to two patients.

### Statistical analysis

Statistical analysis was performed using SPSS for Windows version 15.0 software program (SPSS Inc., Chicago, IL, USA). As the continuous variables were abnormally distributed, the quantitative variables were expressed in median and 25 to 75 quartiles. Qualitative variables were represented in frequency distribution and percent. The results of the surgical groups (patients with LVOTO resection, intraventricular re-routing or arcus reconstruction) were compared using chi-square Fisher's exact test. A $p$ value of $<0.05$ was considered statistically significant.

## RESULTS

Demographic characteristics of the patients are given in Table 1. Aortic, pulmonary, mitral, and tricuspid annulus measurements and their Z scores of the surgical groups are listed in Table 2.

Selective cerebral perfusion was carried out in all patients with TGA, VSD, and arcus obstruction for median 20.5 min (range 16 to 30 min). In two of these patients, TCA (10 min and 6 min, respectively) was performed. The median splanchnic arrest time was 26 min (range 20 to 31 min). Autologous pericardium was used in three patients and bovine pericardium was used in one patient for arcus reconstruction.

Among patients with LVOTO, two patients underwent merely pulmonary valvotomy and four patients had myectomy combined with pulmonary valvotomy. Membrane resection from the LVOT was performed in one patient. Two patients had modified Konno operation, two patients underwent Nikaidoh procedure, and three of them had Rastelli operation (Table 1).

In both patients with modified Konno operation, VSDs were small and located in the muscular outlet region. In addition, LVOTOs were caused by the posterior malalignment of the outlet septum, whereas pulmonary annuli were normal, although one patient had bicuspid valve. In both cases, the septal incision was extended from the small VSD to the previously placed marking suture, just below the pulmonary valve. In Case 12, a patch of autologous pericardium treated with glutaraldehyde was sutured to the right ventricular aspect of the septal incision and the VSD. In Case 13, a Dacron patch was rather used.

Among all patients, the median CPB time was 228.5 min (range 164 to 309 min), cross-clamp time was 107 min (range 88 to 177 min). The median time for mechanical ventilation was 40.5 hours (range 17 to 275 hours), median duration of intensive care was 4.3 days (range 1.2 to 31.2 days), and total length of stay in hospital was seven days (range 5 to 55 days), excluding two patients who died in the early postoperative period. Surgical and postsurgical variables of the patient groups are shown in Table 3.

Only one patient with TGA, VSD, and hypoplastic arcus aorta needed mechanical ventilation during transfer preoperatively. Four patients were deemed ductus dependent (three patients with hypoplastic arcus and one with VSD + LVOTO) and received prostaglandin infusion for 29, 41, 12, and 21 days, respectively before operation (Table 1).

Two patients needed mechanical ventilation longer than 10 days. One was mentioned above (Case 3) who was already under ventilation support preoperatively during the transfer to our hospital. The other patient had TGA, VSD, LVOTO, and coronary artery anomaly, which required a particular transfer technique with a tunnel-type button (Case 10). These patients were ventilated for 23.7 days and 11.5 days, respectively. They also had the highest inotropic scores within the first 24 hours (72.5 and 62.5, respectively). The remaining patients were ventilated for median 36.6 hours (range 17 to 90 hours).

These two patients with long ventilation support also needed peritoneal dialysis due to oliguria postoperatively. Other two patients underwent dialysis:

**Table 1. Patient characteristics**

| Patient no | Age/Gender (month) | Weight (kg) | Cardiac pathology and coronary anatomy | Operation | Complication/Outcome |
|---|---|---|---|---|---|
| 1* | 1.4/M | 3 | TGA, VSD, CoAo | ASO + VSD closure + arcus reconstruction | Peritoneal dialysis residual VSD |
| 2* | 1.5/F | 3.5 | TGA, VSD, CoAo S1: L, Cx (2 ostia) S2: R | ASO + VSD closure + arcus reconstruction | Not remarkable |
| 3* | 0.4/M | 3.5 | TGA, VSD, CoAo | ASO + VSD closure + arcus reconstruction | Postoperative long ventilation, peritoneal dialysis, High IS |
| 4 | 3/M | 3.4 | TGA, VSD, CoAo | ASO + VSD closure + arcus reconstruction | Residual VSD |
| 5 | 96/F | 18.9 | TGA, VSD, PS (V) bicuspid PV | ASO + VSD closure + valvotomy | Subaortic membrane (LVOT gradient 16.3 mmHg) |
| 6 | 3/M | 5.4 | TGA, VSD, PS (V) bicuspid PV S1: L, R S2: Cx | ASO + VSD closure + valvotomy | Residual VSD |
| 7 | 21/F | 7.8 | TGA, VSD, PS (SV) | ASO + VSD closure + membrane resection | Not remarkable |
| 8 | 48/F | 11.9 | TGA, VSD, PS (V + SV) bicuspid PV | ASO + VSD closure + valvotomy + myectomy | Not remarkable |
| 9 | 4/F | 6.1 | TGA, VSD, PS (V + SV) bicuspid PV | ASO + VSD closure + valvotomy + myectomy | Subaortic membrane (LVOT gradient 18.2 mmHg) |
| 10 | 3/M | 3.7 | TGA, VSD, PS (V + SV) bicuspid PV S2: L, Cx, R | ASO + VSD closure + valvotomy + myectomy + tunnel type coronary button | Postoperative long ventilation, peritoneal dialysis, high IS |
| 11 | 0.8*/M | 3.5 | TGA, VSD, PS (V + SV) S1: L,R S2: Cx | ASO + VSD closure + valvotomy + myectomy | Diaphragm paralysis, reoperation: aortic valvotomy, LVOTR, RPA patch plasty |
| 12 | 5/F | 5 | TGA, VSD, PS (SV) | ASO + VSD closure + modified konno | JET |
| 13 | 3/F | 3 | TGA, VSD, PS (SV) | ASO + VSD closure + modified konno | Not remarkable |
| 14 | 4.5/F | 5.8 | TGA, VSD, PS (V + SV) bicuspid PV | Nikaidoh | Residual VSD |
| 15 | 24/M | 10.2 | TGA, VSD, PS (SV) | Nikaidoh | Residual VSD |
| 16 | 42/F | 10.6 | TGA, VSD, PS (SV) | Rastelli | Not remarkable |
| 17 | 30/M | 9.5 | TGA, VSD, PS (SV) S1: L,R S2: Cx | Rastelli | High IS, ECMO exitus |
| 18 | 30/M | 11 | TGA, VSD, PS (SV) S1: L, R, Cx (L intramural) | Rastelli | Residual LVOT gradient, peritoneal dialysis, High IS |

TGA: Transposition of great arteries; VSD: Ventricular septal defect; CoAo: Coarctation of aorta; S1: Coronary sinus 1; L: Left coronary artery; S2: Coronary sinus 2; ASO: Arterial switch operation; Cx: Circumflex coronary artery; R: Right coronary artery; IS: Inotropic score; V: Valvular; PS: Pulmonary stenosis; PV: Pulmonary valve; LVOT: Left ventricular outflow tract; SV: Subvalvular; LVOTR: Left ventricular outflow tract reconstruction; RPA: Right pulmonary artery; JET: Junctional ectopic tachycardia; ECMO: Extracorporeal membrane oxygenator. * Ductus dependent: prostaglandin E1 infusion was preoperatively administered. The coronary anatomy is as follows if not otherwise mentioned: Sinus 1: Left coronary artery and circumflex artery. Sinus 2: Right coronary artery.

one patient with Rastelli operation who died due to low cardiac output postoperatively (Case 18) and the other with arcus reconstruction and the lowest body weight (3 kg) (Case 1).

One patient (Case 12) had transient junctional ectopic tachycardia which responded to cooling and synchronized pacing. No anti-arrhythmic therapy was given, as there was no hemodynamic compromise.

**Table 2. Preoperative Z scores and valvular annulus measurements for different groups of operation**

| Operation | TGA+VSD+CoAo (n=4) | | TGA+VSD+LVOTR (n=14) | | TGA+VSD+rerouting (n=5) | |
|---|---|---|---|---|---|---|
| | Median (mm) | Range | Median (mm) | Range | Median (mm) | Range |
| Aortic valve | | | | | | |
| Annulus | 10.3 | 8.3-10.6 | 12.6 | 8.4-21.2 | 17.5 | 10.6-18.3 |
| Z score | 2.39 | 0.2-2.5 | 4.3 | 0.3-5.7 | 3.1 | 0.23-3.7 |
| Pulmonary valve | | | | | | |
| Annulus | 10.6 | 9.8-14.5 | 10.0 | 7.3-21.2 | 9.7 | 7.5-15.9 |
| Z score | 2.1 | 1.6-2.7 | 1.2 | –1.5-2.2 | 1.3 | –1.9-1.3 |
| Mitral valve | | | | | | |
| Annulus | 13.3 | 10.1-14.4 | 14.1 | 10.8-28.1 | 16.5 | 15.9-18.6 |
| Z score | 0.7 | –1.3-1.1 | 0.4 | 0.9-1.8 | 0.7 | 0.1-0.9 |
| Tricuspid valve | | | | | | |
| Annulus | 12.9 | 9.8-14.5 | 17.8 | 9.8-14.5 | 20.0 | 9.3-37.6 |
| Z score | –0.2 | –1.5-0.4 | 0.7 | –2.2-1.6 | 0.9 | –2.7-4.4 |

TGA: Transposition of great arteries; VSD: Ventricular septal defect; CoAo: Coarctation of aorta; LVOTR: Left ventricular outflow tract reconstruction.

The rhythm converted to sinus on the second day. Diaphragmatic plication was done in a patient (Case 11) on the third day postoperatively due to diaphragmatic paralysis.

The median follow-up was 37.7 months (range 15 days to 74 months), postoperatively. Two patients (Cases 17 and 18) died within 48 hours due to low cardiac output. Both of these patients had coronary artery anomalies which were not amenable to transfer, therefore they had Rastelli operation. Extracorporeal membrane oxygenation (ECMO) was used for one of them postoperatively (Table 1). No other mortality was observed during follow-up, including two patients with long ICU stays.

During follow-up, the last echocardiographic examination revealed median 12.4 mmHg gradient (range 2 to 38 mmHg) in the LVOT, in patients operated for LVOTO. Median descending aorta gradient was 13.5 mmHg (range 7.8 to 28 mmHg) in patients with arcus reconstruction. The median fractional shortening was 37.1% (range 28 to 46 mmHg). A small residual VSD was observed in five patients. Both patients whose defects were closed with primary suture technique had hemodynamically non-significant residual defects. The remaining three patients were those who had autologous pericardial patch for VSD repair. They were also hemodynamically non-significant.

The median maximum gradient of the neoaortic valve during follow-up was 12.3 mmHg (range 4 to 38 mmHg). The neoaortic valve was bicuspid in six patients. The maximum gradient of 38 mmHg was observed in a patient (Case 11) who had a bicuspid

**Table 3. Operative and postoperative details for different surgical groups**

| Surgical groups | TGA+VSD+CoAo (n=4) | | TGA+VSD+LVOTR (n=14) | | TGA+VSD+re-routing (n=3)* | |
|---|---|---|---|---|---|---|
| | Median (mm) | Range | Median (mm) | Range | Median (mm) | Range |
| Minimum body temperature in surgery (°C) | 20 | 17-24 | 26 | 24-32 | 26 | 26-28 |
| Cardiopulmonary bypass time (min) | 235 | 215-309 | 186 | 164-242 | 242 | 198-263 |
| Cross-clamp time (min) | 101 | 88-133 | 105 | 90-145 | 150 | 97-177 |
| Inotropic score | 28.8 | 25-72.5 | 24.3 | 10-62.5 | 22.5 | 10-63 |
| Ventilation time (hours) | 89 | 43-196 | 40 | 17-275 | 26 | 18-66 |
| Length of stay in the intensive care unit (hours) | 236 | 70-765 | 110 | 65-357 | 93 | 28-154 |
| Length of stay in the hospital (days) | 24 | 5-55 | 7 | 5-18 | 5 | 1.5-13 |

* Patients who died within 48 hours postoperatively are excluded.

neoaortic valve and posterior malalignment of the interventricular septum. This patient also had right pulmonary artery stenosis with a peak-to-peak gradient of 32 mmHg. The initial operation was pulmonary valvotomy and LVOT myectomy in addition to ASO and VSD closure. One year later, this patient was re-operated with neoaortic valvotomy, LVOT resection, and patch plasty to the right pulmonary artery. Discrete subaortic membrane developed in two other patients (Cases 5 and 9) during follow-up; however, in the final echocardiography, the gradients were measured as 16.3 mmHg and 18.2 mmHg, respectively. Therefore, no intervention has been planned yet. The maximum pulmonary gradient across the pulmonary conduit in patients undergoing intraventricular re-routing operations (2 Nikaidoh, 1 Rastelli) was 15 mmHg.

None of the patients with arcus aorta reconstruction experienced recoarctation. Three patients (Cases 15, 11, and 10) are still under follow-up due to increased neoaortic root diameters (Z scores are 4.1, 3.89, and 2.8, respectively) with no more than mild aortic regurgitation in any of the patients. Only one of them (Case 10) has still bicuspid neoaortic valve.

## DISCUSSION

A variety of surgical options are available for LVOTO in the setting of TGA. If the pulmonary valve annulus Z score is sufficient and the LVOTO is due to an accessory tissue, aneurysm of the membranous septum, septal malalignment, subpulmonary ridge or muscular conus, ASO with resection of the obstructive tissue may be preferred. Otherwise, more complex re-routing operations which connect the aorta to the left ventricle are carried out.[7] Similarly, we performed resection of the LVOTO with ASO in nine of the 14 patients with VSD and LVOTO. The Rastelli and Nikaidoh procedures were done in the remaining ones.

The selection of the optimal surgical procedure in a patient with TGA and LVOTO may be not always straightforward. Honjo et al.[8] proposed using the LVOT complexity scores to assist in the selection of an optimal operation. In this scoring system, the obstruction was classified as valvular and subvalvular; then, it was graded. They found that the complexity score was correlated with the surgical procedure utilized. However, the score was created retrospectively and the decisions were actually made conventionally. Therefore, it is questionable whether the score would influence the decision in the same manner, if it was considered prospectively. In the present study, the preoperative plan had to be altered in five patients due to unexpected intraoperative findings. Although the preoperative surgical decision was Nikaidoh procedure in four patients, modified Konno operation was applied to two of them, LVOT resection was done to another one, and Rastelli operation was done to the one with coronary artery anomaly which precluded the transfer. Preoperative surgical decision was LVOT resection in another patient; however, inspection during the operation revealed that LVOTO was tunnel-shaped, pulmonary valve was dysplastic and the coronary artery anomaly was not suitable for the transfer. Therefore, this decision was unable to be carried out and Rastelli operation was performed. In all the other patients, preoperative surgical decision was executed.

In patients with TGA, LVOTO and VSD whose LVOTO is not amenable to resection, intraventricular re-routing operations are Rastelli, Réparation à l'Etage Ventriculaire (REV), and Nikaidoh. Although the classical procedures such as Rastelli and REV are preferred in some centers, the application of Nikaidoh procedure has been recently increasing.[2,8-10]

On the other hand, the Rastelli procedure has several disadvantages which make it unfavorable. It provides a non-anatomical correction which cannot be performed readily in infants or newborns; it is associated with late morbidities such as conduit stenosis, residual or recurrent LVOTO, or arrhythmia.[10] Nearly half of the patients are reoperated within 10 years after the Rastelli procedure and reoperations are usually due to the right ventricular outflow tract obstruction (RVOTO). The mean survival of a conduit is reported to be five years.[10-12] Kreutzer et al.[13] suggested that the main reason for early mortality after the Rastelli operation was residual LVOTO and 20-year survival or transplantation-free survival was 52%.

Two of our patients who underwent Rastelli operation were 30 months old, and the third one was 42 months old. The younger ones died due to low cardiac output postoperatively. The intraoperative TEE revealed residual LVOTO due to restrictive VSD in the first patient. Although VSD was enlarged and the obstruction was alleviated, the patient developed left ventricular dysfunction and died 28 hours after he was put on ECMO. Venoarterial ECMO was applied to this patient to provide both cardiac and respiratory support. In general, no attempt to wean off ECMO is considered during the first 24 hours. Then, the left ventricular contractility is assessed with TEE. If systemic pressure, left ventricular contractility, central venous pressure, wedge pressure and mixed venous oxygen saturation does not alter significantly, ECMO can be removed. As our patient died within 28 hours, repeated TEE evaluation was unable to be done.

The other patient was evaluated echocardiographically in the ICU within first few hours postoperatively, when the low cardiac output syndrome ensued. The reason of low cardiac output was the dyskinesia in the LVOT caused by the intracardiac tunnel; however, there was no obstruction. The CPB times were 234 and 198 min in these patients, respectively. The remaining patient with Rastelli operation survived and no re-intervention was required or no RVOTO or LVOTO was seen during the five-year follow-up period.

Compared to other intraventricular re-routing operations, the Nikaidoh procedure yields the straightest connection from the left ventricle to the aorta. Therefore, the locations of the right and left ventricular outflow tracts are closer to their normal anatomical position. Although long-term results have not been determined yet, this aspect of Nikaidoh operation is considered as an advantage and fewer problems are expected, compared to REV and Rastelli.[14]

Including the ones going through Nikaidoh operation, none of our patients had pulmonary valve annulus Z score less than -2. Although the pulmonary valves were not hypoplastic in these patients, Nikaidoh operation was performed, since the valves were thick and dysplastic along with related posterior malalignment. No conduit stenosis was observed or no re-intervention was required during follow-up in these patients.

The main criterion which determines the selection of Nikaidoh or Rastelli operation is the coronary artery anomaly.[8,10] Any coronary artery anomaly precluding the transfer to the neoaortic root is a contraindication for Nikaidoh procedure.[14] The complexity of the procedure and potential late complications such as aortic regurgitation, coronary artery stenosis are other disadvantages of the Nikaidoh operation.

Nevertheless, not all coronary artery anomalies require Rastelli operation. In two of our patients, the coronary anatomy was unsuitable for a classical transfer technique, which would cause kinking and stretching of the coronary arteries. In these patients, a wide coronary button was harvested and attached to a pedicle from the pulmonary artery forming a tunnel.[15] Both patients were three months old during the operation. Coronary artery angiograms at one year showed patent coronary arteries and no sign of coronary artery obstruction was seen throughout the follow-up period.

If there is a diffuse subaortic obstruction and the pulmonary valve is normal, the conventional transpulmonary approach often fails to provide satisfactory results in patients with TGA. The main goal of the modified Konno operation is to allow adequate relief of diffuse subaortic obstructions, while preserving the pulmonary valve. We performed this operation in a couple of our patients who were three and six months old. Due to the proximity of RVOT to the aortic and pulmonary valves, the classical RVOT incision may be difficult during the Konno operation in infants and it may damage the aortic valve. In our modification of this operation, the aortic valve, which is in connection with the right ventricle, is partially excised similar to the Ross procedure. As a result, LVOT resection is easier and the aortic valve function is protected.[8]

The reconstruction of the hypoplastic arcus aorta in the TGA setting requires long TCA and CPB. The transatrial closure of VSD may be difficult in these patients due to frequently seen conal septal anomaly. Furthermore, the coronary anomalies are more common making the transfer challenging. Selective cerebral and myocardial perfusion enable shorter TCA and potentially minimize the morbidity and mortality in these patients.

Three of the four patients with hypoplastic arcus aorta had the most frequent coronary anatomy seen in TGA; namely left coronary and circumflex arteries arising from sinus 1, right coronary artery arising from sinus 2. The remaining patient had right and left coronary arteries arising in separate orifices from sinus 1 (the left coronary and circumflex arteries shared the same orifice). The coronary artery transfer was done without any difficulty in all these patients. The median time of selective cerebral and myocardial perfusion was 20.5 min. This obviated application of TCA during arcus reconstruction in two patients, and limited the TCA time to 10 min and six min in the remaining two patients. Only one of these patients had high inotropic score and needed peritoneal dialysis and prolonged intubation postoperatively. This patient was the only case who required preoperative ventilation support. Recoarctation was not observed during follow-up in any of our patients with arcus reconstruction.

In patients with TGA, VSD and LVOTO, larger aortic annulus is predicted due to decreased pulmonary flow. However, in TGA, VSD and arcus aorta obstruction, a larger pulmonary annulus is usually expected, since the blood flow favors the pulmonary artery through the VSD. When the aortic and pulmonary annulus measurements were compared among our patients with different surgical groups, the largest median pulmonary Z score was in patients with arcus reconstruction. However, the difference did not reach statistical significance and pulmonary Z score was not higher than the aortic Z score, as expected. Similarly, among patients with LVOTO, neither the pulmonary nor the aortic Z scores seem to be different between those undergoing intraventricular rerouting procedures and those with LVOT resection procedures. These results may be due to the small number of patients in different surgical groups which makes the statistical analysis unreliable.

As the discrepancy between aortic and pulmonary diameters increases, neoaortic root dilatation, regurgitation or stenosis may be more frequent.[8] However, none of our patients had more than mild regurgitation during follow-up. Seven patients had bicuspid neoaortic valve. Only one of them needed neoaortic valvotomy operation. This patient also had posterior malalignment of the outlet septum and myotomy was done additionally during the operation. Subaortic discrete membrane developed in another patient; however, the LVOT gradient was only 16.3 mmHg at the final follow-up visit. As Kalfa et al.[16] and Angeli et al.[17] reported in their studies, bicuspid neoaortic valve per se is not a significant risk factor for recurrent LVOTO, if the pulmonary valve Z score is within normal range.

On the other hand, there are some limitations to our study. First, our sample size was small. Second, we tailored a retrospective study design. The retrospective design of the study is a disadvantage in the interpretation of the results, in particular. The uneven distribution of different pathologies and small number of patients also precluded that. However, the study was not designed to compare the surgical outcomes; it was done to analyze our approach to surgical decision-making in these cardiac pathologies. Clearly, further large-scale prospective studies are required to compare the different surgical results and to establish a conclusion. Still, we believe that our study may provide invaluable advice on strategies in the evaluation of similar patient populations.

In conclusion, there are various surgical options in transposition of the great arteries associated with left ventricular outflow tract obstruction. As there is no ideal surgical procedure devoid of all disadvantages, deciding on the optimal operation is often challenging and should be done considering the specific cardiac anatomy in a selected patient. These priorities, however, may be kept in mind while considering the alternatives: If the pulmonary annulus is sufficient or there is a resectable tissue in the left ventricular outflow tract, valvotomy or resection of obstructive tissue is preferred. This would eliminate any reconstruction with a prosthetic material, and prevent the recurrent left ventricular outflow tract and right ventricular outflow tract obstructions in the most part. On the other hand, if the pulmonary annulus is too small or the obstruction in the left ventricular outflow tract is not amenable to resection, Nikaidoh operation, which can create a better right and left ventricular outflow alignment, may be favored among all other intraventricular re-routing operations. Finally, if there is coronary artery anomaly precluding safe transfer, Rastelli procedure may be essential. The arcus aorta reconstruction with arterial switch operation has been currently accomplished with low mortality and morbidity.

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