Turk Gogus Kalp Dama 2016;24(2):350-355
doi: 10.5606/tgkdc.dergisi.2016.12166

Case Report / Olgu Sunumu

Does late primary arterial switch operation with extracorporeal membrane oxygenator support change the surgical approach in simple transposition of the great arteries?

Basit büyük arter transpozisyonunda ekstrakorporeal membran oksijenatör destekli geç primer arteriyel switch ameliyatı cerrahi yaklaşımı değiştirir mi?

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ABSTRACT

Currently, arterial switch operation appears as a standard surgical management for patients under three weeks of age with transposition of the great arteries with an intact ventricular septum, while, beyond three weeks of age, there is no such standard approach and surgical procedures may vary among the health care centers. Low cardiac output and left ventricle failure may also develop after three weeks due to the progressive involution of left ventricle after arterial switch operation. Therefore, the Senning or Mustard procedure, or two-stage repair arterial switch operation are optional surgical management modalities in this patient population. However, due to the potential complications of these procedures in the short- and long-term, there has been an increased interest in performing primary arterial switch operation with extracorporeal membrane oxygenator support in patients older than three weeks of age. This report presents two cases in whom primary arterial switch operation with extracorporeal membrane oxygenator support was performed at the age of 110 days and 60 days, respectively. Primary arterial switch operation with extracorporeal membrane oxygenator support appears to be a more effective option in the short-term than alternative surgical management modalities.

Keywords: Extracorporeal membrane oxygenator; late arterial switch; transposition.

ÖZ

Günümüzde ventriküler septumu intakt, üç haftadan küçük basit büyük arter transpoziyonu olan hastalarda standart cerrahi tedavi primer arteriyel switch ameliyatı iken, üç haftadan büyüklerde bu tür bir standart yaklaşım yoktur ve cerrahi işlemler merkezler arasında değişiklik gösterebilmektedir. Üç haftadan sonra sol ventrikülün ilerleyici involüsyonu nedeniyle, arteriyel switch ameliyatı sonrası düşük kalp debisi ve sol ventrikül yetmezliği de gelişebilir. Bu nedenle, Senning veya Mustard işlemi veya iki aşamalı arteriyel switch ameliyatı, bu hasta popülasyonunda alternatif cerrahi tedavi yöntemleridir. Bununla birlikte, bu işlemlerin kısa ve uzun dönemde muhtemel komplikasyonları nedeniyle, üç haftadan büyük olan hastalarda ekstrakorporeal membran oksijenatörü desteği altında primer arteriyel switch ameliyatına karşı ilgi giderek artmaktadır. Bu yazıda, ekstrakorporeal membran oksijenatörü desteği altında primer arteriyel switch ameliyatı yapılan 110 günlük ve 60 günlük iki olgu sunuldu. Kısa dönemde ekstrakorporeal membran oksijenatörü desteği ile yapılan arteriyel switch ameliyatının, diğer seçeneklere kıyasla, daha etkili bir seçenek olabileceği düşünülmektedir.

Anahtar sözcükler: Ekstrakorporeal membran oksijenatörü; geç arteriyel switch; transpozisyon.

While traditionally Senning[1] or staged switch operation used to be major treatment options in patients with simple transposition of the great arteries (TGA) who are ineligible for arterial switch operation (ASO) during the initial weeks,[2,3] with the introduction of increased use of left ventricle-supporting devices and the level of experience, primary ASO has recently become a promising treatment option for these patients,

Available online at
www.tgkdc.dergisi.org
doi: 10.5606/tgkdc.dergisi.2016.12166
QR (Quick Response) Code

Received: July 08, 2015 *Accepted*: November 08, 2015

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as well.[4] In this article, we present two cases who we performed extracorporeal membrane oxygenator (ECMO)-supported primary ASO in the late period.

CASE REPORT

Case 1– A three-month and 20-day-old male infant with a weight of 5.5 kg was admitted to our clinic. He had no pathological signs in his physical examination except a saturation level of 70% and existence of central cyanosis. Electrocardiography (ECG) showed normal sinus rhythm with right axis and right ventricle hypertrophy. Transposition of the great arteries and two small secundum atrial septal defects (ASD) with shunts from the left-to-the right side were observed in transthoracic echocardiographic (TTE) examination. There was no left ventricle (LV) systolic dysfunction, while there was a banana-shaped LV involution (Figure 1). Left ventricle posterior wall end-of-diastole thickness was 3 mm (z score: -1.69), end-diastolic interventricular septum thickness 3 mm (z score: -1.29), end-diastolic LV diameter 13 mm (z score: -4) and left ventricular mass index (LVMI) 14.7 gram/m^2 (z score: -4) (Figure 2).

A written informed consent was obtained from the parents of the patient. Extracorporeal membrane oxygenator-supported primary ASO operation was performed in the patient with a circumflex artery originating from the right coronary artery and the ASDs were closed leaving only a small opening in the inter-atrial septum (IAS). Due to insufficient LV contractions and hypotensive progression following weaning from cardiopulmonary bypass (CPB), the patient was switched to ECMO support. Central cannulation was performed. He was cannulated from the ascending aorta for arterial flow and from right atrium for venosum flow. Extracorporeal membrane oxygenator support was initiated with a flow rate of 2.4 L/m^2/min and the rate was gradually increased to 2.8 L/m^2/min according to the lactate level. The sternum was not closed, while the skin was closed with a patch. During ECMO support, ventilation variables were lowered. According to our protocol, the parameters were set as PEEP: 4 cmH_2O, FiO_2: 40%, frequency: 20/min, tidal volume: 6 mL/kg. Both hemithoraces were opened and the drainage tubes were placed. We routinely drained the anterior mediastinal space via suction systems. Dopamine, milrinone, and adrenalin were used for inotropic agents for cardiac support. Activated clotting time was measured in a-two-hour interval and the target range for ACT values was predefined to be 180 to 200 sec. Heparine was used for anticoagulation (300 IU/kg bolus before cannulation and 20 IU/kg/hour infusion).

The patient was monitored with daily echocardiographic examinations and hemodynamic findings.

He needed for revision on postoperative day zero due to the drainage (>100 cc/hour for 3 hours). No major bleeding was observed in the surgical area. He was revised for mediastinal irrigation of drainage tubes and exploration of infection signs. There was no mediastinitis findings. Clinical and hemodynamic variables such as dimensions and ejection fraction of the left ventricular, aortic velocity, and time integral were used for weaning. The ventilation and inotropic support were maximized and ECMO flow rate was gradually lowered. Urine output (1 mL/kg/h) was accepted as sufficient and the lactate levels are primary variables for clinical observation. The chest was left open for additional 24 hours. Then, the sternal and skin closure was performed. The ECMO support was reduced gradually and the patient was

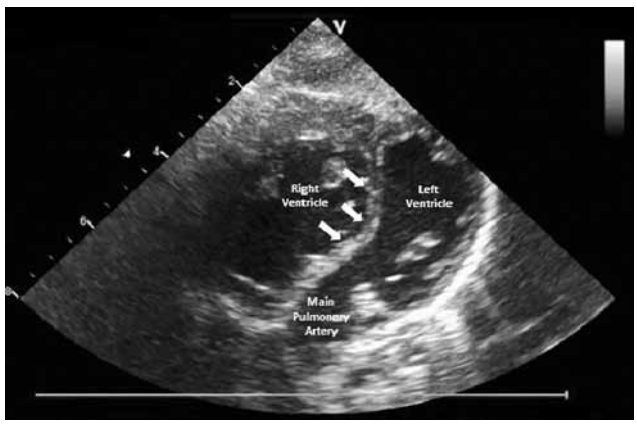

Figure 1. A two-dimensional echocardiographic image showing significant left ventricle (banana-shaped left ventricle).

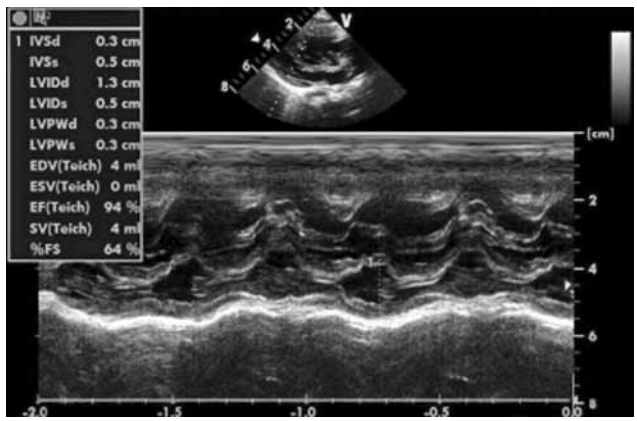

Figure 2. A M-mode echocardiographic image showing left ventricle involution.

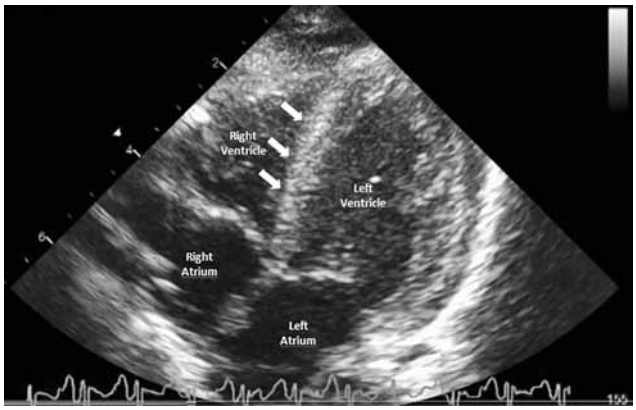

Figure 3. A two-dimensional echocardiographic image showing left ventricle following surgery.

disconnected from the ECMO support at the end of one week.

On the following days, the overall condition of the patient was good and LV returned to normal shape from the original banana shape (Figure 3). There was no systolic dysfunction, as confirmed by the ECG. However, the patient received necessary treatment for possible bacterial ocular keratitis infection. As the patient's hemodynamic variables progressed to normal values, medical inotropic support was gradually reduced and discontinued completely. Echocardiographic examinations in the 24th postoperative day showed that systolic functions were normal; however, there were mild insufficiency in the neo-aorta and the neo-pulmonary artery and a small left-to-right shunt through the opening path left in IAS. The end-diastolic LV posterior wall thickness was 5 mm (z score: +1.38) and the end-diastolic interventricular septum thickness 5 mm (z score: +1.57), end-diastolic LV diameter 30 mm (z score: +4.9) and LVMI 76.6 g/m^2 (z score: -4), as assessed by TTE. On ECG, there were no signs of arrhythmia or ischemia and the patient was discharged from the hospital with a good overall condition in the 35th postoperative day. At 11 months of follow-up, there was no vision related problem found on keratitis examination. Echocardiography showed trace neo-pulmonary and neo-aortic insufficiency. There is no any developmental or neurological disorder to date.

Case 2– A two-month-old female patient who was diagnosed with simple TGA and received emergency balloon atrial septostomy (BAS) due to cyanosis (saturation 45%) and metabolic acidosis in an external center was transferred to our hospital with a body weight of 3400 g (25th to 50th percentiles), and a height of 60 cm (50th percentiles) at her physical examination. The saturation was measured within the range of 75 to 80%. Cardiac auscultation signs and system examination results were normal.

Transthoracic echocardiography showed TGA and post-BAS non-restrictive inter-atrial shunt. The patient was in normal sinus rhythm. Electrocardiography revealed right axis and right ventricle hypertrophy. There was no systolic dysfunction; however, LV involution was significant and LV was banana-shaped. Left ventricular end-diastolic posterior wall thickness was 3 mm (z score -0.92), end-diastolic interventricular septum thickness 3 mm (z score: -4), LV end-diastolic diameter 15 mm (z score: -4), and LVMI 24.9 gr/m^2 (z score: -4) as seen in TTE.

A written informed consent was obtained from the parents of the patient. An ASO operation was performed on the patient who did not have any coronary artery abnormalities. The patient was taken into the intensive care unit to receive ECMO support due to the development of LV insufficiency signs in the operation room, when CPB support was disconnected; however, decannulation was not performed. Cannulation, hemodynamic support, open chest follow-up, details of ECMO support, coagulation, ventilation, and surgical protocols were applied similarly.

There was no event about cardiopulmonary instability or bleeding. She was revised for mediastinal irrigation of the drainage tubes and exploration of the infection signs. There was no mediastinitis findings, either.

The ECMO support was gradually reduced and the patient was disconnected from the ECMO support in the seventh postoperative day.

On the following days, her overall condition was good. The LV returned to its normal shape and no systolic dysfunction was present, as confirmed by the TTE [LV end-diastolic posterior wall thickness 5 mm, end-diastolic interventricular septum thickness 5 mm, LV end diastolic thickness 19 mm and LVMI 68.8 g/m^2 (z score: -0.92, -4, -4 and -4, respectively)]. Hemodynamic signs were stable and medical inotropic support was gradually reduced and discontinued finally. According to TTE in the 15th postoperative day, systolic functions were normal, while there were mild insufficiencies in the neo-artery and neo-pulmonary artery. There were no arrhythmia and ischemia signs. The patient was discharged from the hospital in the 20th postoperative day with a good overall condition. During the echocardiographic examination at 12 months, there was no abnormality detected except mild neo-aortic insufficiency and left proximal pulmonary artery

stenosis. There is no developmental or neurological disorder to date.

DISCUSSION

Today, ASO is performed as a standard treatment modality in the treatment of simple TGA in younger than three-week infants, while for infants older than three weeks, atrial switch, two-stage ASO, and recently primary ASO have become the preferred interventions.[1,2-4]

Undoubtedly, the most important factor affecting the selection of treatment modality is the involution degree of LV.[5] The long-term results of operations and the relevant experience of health centers also play an important role in the preference of health centers. In both of our cases, LV involution was significant and LV was banana-shaped. While LV can be trained via stent implantation in ductus arteriosus in patients who developed LV involution,[5] LV can be prepared for systemic afterload in simple TGA patients with no ductus by means of shunted or shunt-less pulmonary banding operation before ASO surgery.[6] However, those undergoing two-stage ASO may develop deterioration in the LV function, insufficiencies in the neo-aortic valve, and obstructions in the right ventricular outflow tract and this intervention results in higher mortality and morbidity rates, longer stays at the intensive care unit, increased nosocomial infections, and increased hospital costs compared to ASO.[7-9]

Despite all these complications, two-stage ASO seems to be a better option compared to ASO, which causes LV dysfunction and frequently atrial arrhythmias.[9]

The performance of ASO on older infants was originally reported by Foran et al.[10] The authors assessed 37 patients aged 21 to 61 days and 156 patients younger than 21 days in their study and reported that the mortality rates were 8.3% and 2.7% in the younger and older groups, respectively. They observed no significant differences between the two groups in terms of ventilation and hospital stays.

In the following years, late ASO performance was reported at an increasing rate.[11] Furthermore, a study comparing the results of infants older than three weeks who underwent two-stage ASO and primary ASO suggested that the results were in favor of primary ASO.[11]

In the literature review, it can be concluded that late ASO can be performed successfully in the older months and even at around the age of eight.[4] On the other hand, it was reported that performing primary ASO with ECMO support was a safe and appropriate approach to ensure postoperative LV support in infants aged above 21 days.[10]

It was reported that the need for ECMO was higher in patients undergoing late ASO.[9] In recent years, mechanical support has been required more often in patients aged above 10 weeks undergoing primary ASO; however, no increased mortality was reported.[10,12,13]

Indeed, Bisoi et al.[4] reported that ECMO support was used at a rate of 20% in 109 patients with simple TGA undergoing primary ASO and their early mortality rates were 3.7%, while their late mortalities were due to reasons other than the primary disease. However, we have not performed late primary ASO without ECMO support on any patient yet thanks to the increase of our experience in ECMO recently; we preferred to perform primary ASO with ECMO support in our two simple TGA patients aged three months and 20 days (80 days) and two months who were diagnosed late and presented to us with cyanosis symptoms. The reason why we used ECMO in our patient aged lower than two months was that the patient, who was not decannulated but disconnected from CPB support, developed LV insufficiency while still in the operating room. As for our older patient, LV geometry was shaped as a banana due to the significant LV involution. This patient was scheduled for ECMO support following surgery. As ventricular functions improved, ECMO support was, then, tapered out. Nearly one week later, he was disconnected from the ECMO support.

Following primary ASO performed in the late period, the patients need to be followed for complications which may require re-intervention such as previously reported pulmonary arterial anastomosis or peripheral stenosis, coronary artery insufficiency, neo-aortic valve insufficiency, disposition to atherosclerosis, and neo-aortic root dilation.[14]

Although mild aortic insufficiency is an extremely frequent sign following ASO operations when performed in a timely fashion and it rarely progresses to a clinically significant level, the most important risk factor in the development of aortic insufficiency is the performance of VSD and ASO at later ages.[15,16]

Furthermore, data are reported that patients undergoing ASO in the late period have prolonged ventilator process, hospital stays, more LV mechanical support need, and delayed closure of the chest.[17] However, a recent study reported that there were

no statistically significant differences in early complications including the mean time of ventilation, intensive care unit, and hospital stay, inotropic agent need, mortality, diaphragm paralysis, acute renal failure, pericardial effusion, pleural effusion, arrhythmia, and chylothorax.[18] Today, ECMO is used in several conditions for the treatment of cardiac surgery patients such as recurrent cardiac arrest requiring continuous resuscitation, uncontrollable hypotension, despite the sufficient inotropic support, uncontrollable ventricular tachycardia and ventricular fibrillation episodes, myocarditis with persistent low cardiac output, despite all kinds of medical treatment, and support requirement before and after transplantation.[19] On the other hand, these patients who receive ECMO support also need to be followed for complications such as hemorrhage, micro-embolization, neurological problems, developmental, and sensorineural disorders, hemolysis, arrhythmia, pneumothorax, mechanical complications (i.e., oxygenator degradation, deterioration of the pump, and tear in ECC (i.e., extra-corporeal circulation) lines, cannula problems), which may develop during ECMO application or become apparent in the follow-up period.[19,20] While our two-month-old patient did not have any complications, our older patient had bacterial ocular infection and secondary keratitis. Systolic dysfunction following ECMO was not observed in neither of the two patients. On the other hand, we have not yet had adequate follow-up for both of our patients for other potential complications which may develop due to both late primary ASO operation and ECMO support. Both patients are currently alive and they attend to scheduled follow-up visits.

In conclusion, primary arterial switch operation with extracorporeal membrane oxygenator support appears as the best option among alternatives for patients with late diagnosis in terms of the long-term life quality and service. Although primary arterial switch operation without extracorporeal membrane oxygenator support can also be carried out in these patients; it should always be kept in mind that these patients may potentially still need extracorporeal membrane oxygenator support later on, and, therefore, stand-by extracorporeal membrane oxygenator support should be provided to these patients.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

REFERENCES

1. Sarkar D, Bull C, Yates R, Wright D, Cullen S, Gewillig M, et al. Comparison of long-term outcomes of atrial repair of simple transposition with implications for a late arterial switch strategy. Circulation 1999;100:176-81.
2. Däbritz S, Engelhardt W, von Bernuth G, Messmer BJ. Trial of pulmonary artery banding: a diagnostic criterion for 'one-stage' arterial switch in simple transposition of the great arteries beyond the neonatal period. Eur J Cardiothorac Surg 1997;11:112-6.
3. Jonas RA, Giglia TM, Sanders SP, Wernovsky G, Nadal-Ginard B, Mayer JE Jr, et al. Rapid, two-stage arterial switch for transposition of the great arteries and intact ventricular septum beyond the neonatal period. Circulation 1989;80:I203-8.
4. Bisoi AK, Ahmed T, Malankar DP, Chauhan S, Das S, Sharma P, et al. Midterm outcome of primary arterial switch operation beyond six weeks of life in children with transposition of great arteries and intact ventricular septum. World J Pediatr Congenit Heart Surg 2014;5:219-25.
5. Celebi A, Demir H, Aydemir NA, Saritas T, Erdem A. Successful arterial switch procedure following ductal stent implantation for left ventricular conditioning in a four month-old case with simple transposition of the great arteries. Turk Gogus Kalp Dama 2013;21:451-4.
6. Yacoub MH, Radley-Smith R, Maclaurin R. Two-stage operation for anatomical correction of transposition of the great arteries with intact interventricular septum. Lancet 1977;1:1275-8.
7. Boutin C, Wernovsky G, Sanders SP, Jonas RA, Castaneda AR, Colan SD. Rapid two-stage arterial switch operation. Evaluation of left ventricular systolic mechanics late after an acute pressure overload stimulus in infancy. Circulation 1994;90:1294-303.
8. Colan SD, Boutin C, Castañeda AR, Wernovsky G. Status of the left ventricle after arterial switch operation for transposition of the great arteries. Hemodynamic and echocardiographic evaluation. J Thorac Cardiovasc Surg 1995;109:311-21.
9. Kang N, de Leval MR, Elliott M, Tsang V, Kocyildirim E, Sehic I, et al. Extending the boundaries of the primary arterial switch operation in patients with transposition of the great arteries and intact ventricular septum. Circulation 2004;110:123-7.
10. Foran JP, Sullivan ID, Elliott MJ, de Leval MR. Primary arterial switch operation for transposition of the great arteries with intact ventricular septum in infants older than 21 days. J Am Coll Cardiol 1998;31:883-9.
11. Bisoi AK, Chauhan S, Khanzode SD, Hote MP, Juneja R, Venugopal P. D-transposition of great vessels with intact ventricular septum presenting at 3-8 weeks: should all go for rapid two stage arterial switch or primary arterial switch? IJTCVS 2006;22:5-9.
12. Nathan M. Late arterial switch operation for transposition with intact septum. World J Pediatr Congenit Heart Surg 2014;5:226-8.
13. Edwin F, Kinsley RH, Brink J, Martin G, Mamorare H, Colsen P. Late primary arterial switch for transposition

of the great arteries with intact ventricular septum in an african population. World J Pediatr Congenit Heart Surg 2011;2:237-42.

14. Fulton DR, Fyler DC. D-Transposition of the great arteries. In: Keane JF, Lock JE, Fyler DC, editors. Nadas' Pediatric Cardiology. Philadelphia: Saunders Elsevier; 2006. p. 645-62.
15. Schwartz ML, Gauvreau K, del Nido P, Mayer JE, Colan SD. Long-term predictors of aortic root dilation and aortic regurgitation after arterial switch operation. Circulation 2004;110:128-32.
16. Losay J, Touchot A, Capderou A, Piot JD, Belli E, Planché C, et al. Aortic valve regurgitation after arterial switch operation for transposition of the great arteries: incidence, risk factors, and outcome. J Am Coll Cardiol 2006;47:2057-62.
17. Lacour-Gayet F, Piot D, Zoghbi J, Serraf A, Gruber P, Macé L, et al. Surgical management and indication of left ventricular retraining in arterial switch for transposition of the great arteries with intact ventricular septum. Eur J Cardiothorac Surg 2001;20:824-9.
18. Ismail SR, Kabbani MS, Najm HK, Abusuliman RM, Elbarbary M. Early outcome for the primary arterial switch operation beyond the age of 3 weeks. Pediatr Cardiol 2010;31:663-7.
19. Cingoz F, Tatar H. Çocuklarda ekstrakorporeal membrane oksijenatör kullanımı. Turk Gogus Kalp Dama 2008;16:50-7.
20. Kolovos NS, Bratton SL, Moler FW, Bove EL, Ohye RG, Bartlett RH, et al. Outcome of pediatric patients treated with extracorporeal life support after cardiac surgery. Ann Thorac Surg 2003;76:1435-41.