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Transposition of the great arteries with ventricular septal defect and interrupted aortic arch: a successfull surgical correction with one stage and two separate incisions

Büyük damarların transpozisyonu, ventriküler septal defekt ve aortik ark interapsiyonu bulunan hastada tek oturum ve iki ayrı insizyonla cerrahi tedavi

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Transposition of great arteries with ventricular septal defect and interruption of aortic arch is a rare malformation. We present a nine-day-old cyanotic male neonate whose echocardiographic assessment revealed transposition of great arteries with ventricular septal defect and interruption of aortic arch. The patient had indirect hyperbilirubinemia and was operated 3 days after admission. Complete repair was performed by a single stage operation with two incisions. With this technique the need for deep hypothermia and total circulatory arrest was abolished.

Key words: Heart defects, congenital/surgery.

Büyük arterlerin transpozisyonu ile birlikte ventriküler septal defekt ve aorta kavsinin interapsiyonu nadir olarak görülmektedir. Siyanoz bulunan altı günlük erkek bebeğin ekokardiyografik incelemesinde büyük damarların transpozisyonu, ventriküler septal defekt ve aortik ark interapsiyonu saptandı. İndirekt bilirubini yüksek olan hasta üç gün sonra ameliyata alındı. Tek oturumda posterolateral torakotomi ve midsternotomi insizyonu yaklaşımlarıyla tam düzeltme yapıldı, derin hipotermi ve total sirkülatuvar arreste gerek duyulmadı.

Anahtar sözcükler: Doğumsal kalp defektleri/cerrahi.

Transposition of great arteries (TGA) with ventricular septal defect (VSD) and interruption of aortic arch (IAA) is a rare malformation. Repair of this complex form of TGA is still a surgical challenge.[1,2] Complete repair in one stage operation through midsternotomy during neonatal period has been shown to be a safest and most effective management protocol with better results as compared to the two-stage technique.[2-6] However, it necessitates deep hypothermia and total circulatory arrest (TCA) for arch reconstruction which has a potential effect of cerebral damage.

Single stage complete repair may also be done by two incisions; a left thoracotomy for direct reconstruction of arch and median sternotomy for subsequent repair of TGA. We report herein a patient with TGA associated IAA type A operated by a single stage operation with two incisions.

CASE REPORT

A 6-days-old cyanotic male neonate, weighing 3.5 kg was admitted to our institution with a prostaglandin infusion. Echocardiographic examination demonstrated TGA with anteroposterior position of the great arteries, narrow Patent Ductus Arteriosus (PDA) and aortic arch interrupted distal to the left subclavian artery (IAA, type A). Balloon atrial septectomy and selective angiocardiography were performed (Fig. 1a, b). Coronary arteries showed usual pattern. Pulse oxymetry saturation was 65% and no acidosis was detected. Surgery was performed three days following the admission due to the elevated indirect bilirubin levels.

Surgery. A left posterolateral thoracotomy was performed and thorax was opened through the fourth intercostal space. Aortic segments, pulmonary artery

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and arch vessels were all identified and extensively mobilized without damaging or sacrifying any intercostal arteries. Descending thoracic aorta was mobilized down to the diaphragm. Large and multiple collateral vessels were observed. Topical hypothermia was achieved at 34 °C. PDA was ligated and PGE1 infusion was stopped. Thoracic aorta and distal archus aorta were clamped using side clamps. A part of origin of left subclavian artery was included within the proximal site. Following transsection of descending thoracic aorta, all ductal tissue was completely excised from the distal segment of aorta. An incision was performed in proximal clamped area and elongated to the under surface of the left subclavian artery. The two vascular clamps were then approximated allowing proximal and distal segments to be anastomosed using 7-0 polyprolene suture without any tension. After completion of anastomosis, vascular clamps were removed and thoracotomy was closed in a routine manner. After proper stabilization, patient was repositioned for the second incision and midline sternotomy was performed. Thymus was removed partially. Ascending aorta was cannulated just below the truncus brachiocephalicus. Both venae cavaes were cannulated. During cooling period, pulmonary trunk was separated from ascending aorta, right and left pulmonary arteries were dissected out to their first branches. At 28 °C the aorta was clamped and blood cardioplegia was administered to the aortic root, and direct cardioplegia was repeated in every 20 minutes. The left ventricle decompensated by the use of a suction which inserted through fossa ovalis. The ascending aorta transsected 5 mm above the coronary artery ostiums, and pulmonary trunk transacted just proximal to its bifurcation. Both coronary artery ostiums excised with buttons of aortic wall and reimplanted to the pulmonary artery using continuous 7-0 polyprolene sutures. Following Lecompte manoeuvre, proximal neoaorta and distal aortic segment were anastomosed with 6-0 PDS suture. The neopulmonary trunk was reconstructed using a glutaraldehyde treated pericardial patch (single patch) and anastomosed with pulmonary bifurcation using 6-0 PDS. VSD was repaired with a patch of dacron graft via transatrial approach. Bypass and cross clamp times were 245 and 147 minutes, respectively. The patient was brought to ICU with standard dose of dopamine (6 µg/kg/min) and adrenaline (0.03 µg/kg/min) perfusion. He was extubated on postoperative 48th hour and observed for 4 days at ICU. On 14th day of the postoperative period, the patient was discharged from hospital in good condition. Two years following operation the patient is still doing well in NYHA class I without any medication and with good femoral pulses. Echocardiographic examination showed mild aortic and pulmonary regurgitation, 16 mmHg peak systolic gradients between left ventricle and aorta. There is no transaortic pressure gradient in the archus and descending thoracic aorta.

DISCUSSION

Single stage repair for TGA with IAA was first reported by Pigott et al.[3] Planche and associates demonstrated the superiority of single stage repair over two-stage repair for TGA, VSD and IAA.[2]

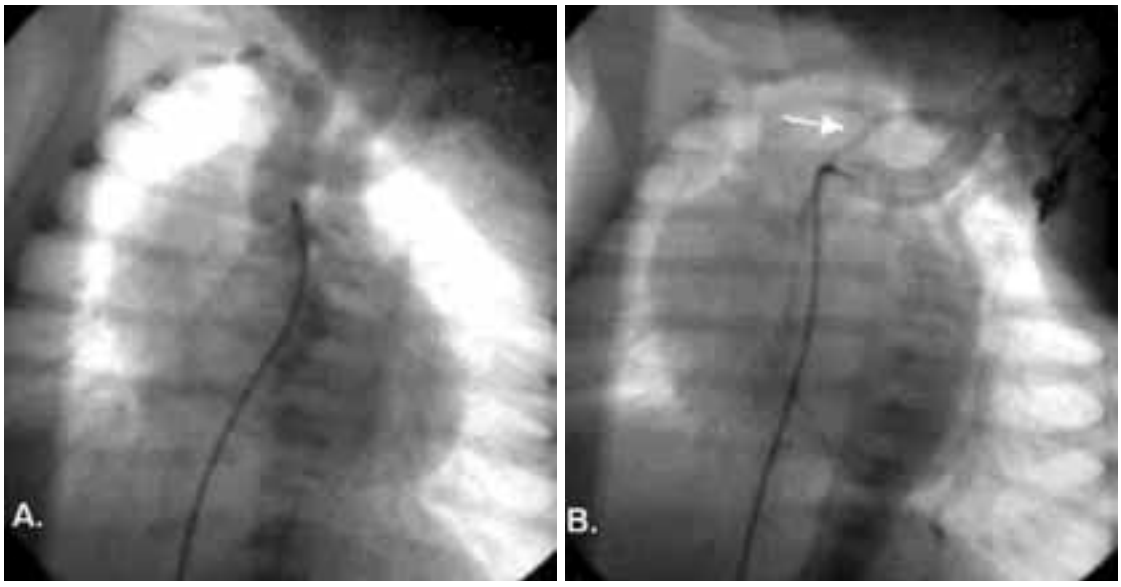

Fig. 1. Preoperative aortogram showing **(a)** type A aortic arch interruption, **(b)** a collateral artery originating from ascending aorta supplying descending aorta.

Although there is some uncertainty regarding the best method for repair of TGA with VSD and IAA, the most preferred approach has been the primary definitive repair of all the malformations at once for years.[2,7] But, for complete repair in one stage operation through midsternotomy, in order to construct a large anastomosis between the arch and descending thoracic aorta, surgeons generally prefer open repair with deep hypothermic total circulatory arrest. There are lots of reports in the literature related with harmful side effects of deep hypothermia and TCA on neonates and infants.[6,8] By correcting IAA through a limited left thoracotomy prior to CPB, the deep hypothermic total circulatory arrest and so the harmful effects of it are avoided as occurred in our case.

An extended end-to-end direct anastomosis is always the first choice for repair of IAA. This can be possible if adequate and extensive mobilization of ascending and descending thoracic aorta is achieved.[9,10] This may be done in a better and easier way through left thoracotomy as compared to midsternotomy.

Through left thoracotomy repair of arch may be complicated either by poor exposure or difficulty in replacement of clamps especially in type B and C interruptions, but type A interruption which resembles more a severe coarctation than a type B interruption would be most amenable to direct repair by lateral approach. In our clinical experience, we observed that limited thoracotomy is well tolerated by neonates and infants like our patient who was weaned from ventilator within 48 hours after the operation without any pulmonary problems.

We believe single stage correction of patients with TGA, VSD and IAA with two subsequent incisions is a good alternative surgical method with multiple advantages for such patients.

Patients with complex type coarctation associated with intracardiac defects could be operated via anterior approach by a single operation. In the very near future, we believe that a only single procedure with median sternotomy will be enough to repair these complex lesions including IAA without the need for deep hypothermia and TCA.

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