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Case Report / Olgu Sunumu

A successful surgical repair of truncus arteriosus in a three-year-old boy

Üç yaşında erkek çocukta trunkus arteriozusun başarılı cerrahi onarımı

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ABSTRACT

Truncus arteriosus is an extremely rare congenital heart disease in which a single great artery arises from the heart and supplies systemic, coronary, and pulmonary blood flow presenting with a ventricular septal defect. If left untreated, the majority of cases die within the first year of life. Herein, we report a three-year-old male case with type 1 truncus arteriosus who was successfully operated.

Keywords: Congenital heart defect; nitric oxide; truncus arteriosus.

ÖZ

Trunkus arteriozus, kalpten çıkan tek bir büyük damarın sistemik, koroner ve pulmoner kan akımını sağladığı ve ventriküler septal defekt ile birlikte seyreden çok nadir bir doğuştan kalp hastalığıdır. Tedavi edilmediğinde, olguların büyük bir çoğunluğu yaşamın ilk yılı içerisinde kaybedilmektedir. Bu yazıda, başarılı bir şekilde ameliyat edilen tip 1 trunkus arteriozuslu üç yaşında bir erkek olgu sunuldu.

Anahtar sözcükler: Doğuştan kalp defekti; nitrik oksit; trunkus arteriozus.

Truncus arteriosus (TA) is an uncommon congenital cardiac malformation presenting for less than 3% of all congenital heart malformations.[1,2] The features of TA include a single great artery originating from the heart, supplying systemic, coronary and pulmonary blood flow, presenting with a ventricular septal defect (VSD). Without surgical treatment, 80% of patients die within the first year of life, mostly during early infancy.[2,3]

The first successful correction of truncus arteriosus was performed in 1965 by McGoon et al.[4] Since then, there have been many advances in the surgical management with an evident trend to perform primary repair in early infancy, thereby, avoiding the complications of pulmonary hypertension (PH). Although timing of TA repair is advocated by most surgeons during the first three months of life,[2] advanced anti-PH treatment modalities and increased surgical experiences encourage us to perform these challenging operations in all age groups.

Herein, we present a boy with type 1 truncus arteriosus treated surgically with the aid of nitrous oxide which was initiated after tracheal intubation and continued during the early postoperative period with the highest dose at the termination of the cardiopulmonary bypass (CPB).

CASE REPORT

A three-year-old boy with a weight of 10.8 kg was referred to our clinic with complaints of shortness of breath and fatigue on exertion. He had a history of lower respiratory tract infections since infancy. There was central cyanosis and clubbing of the fingers and toes. On auscultation, both heart sounds were single and there was a long Grade 3/6 systolic murmur and a Grade of 3/6 early diastolic murmur which were both loudest at the lower left sternal border. A chest X-ray showed cardiomegaly with a cardiothoracic ratio of 60% and increased pulmonary vascularity. Preoperative saturation was about 70 to 75%. Echocardiography showed a single arterial trunk arising from the base of the heart which was the origin of the aorta, pulmonary, and coronary arteries with a large perimembranous

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outlet ventricular septal defect. The truncal valve had a gradient of 60 mmHg with a moderate to severe insufficiency (4.6 m/s). The cardiac catheterization with a mean pulmonary artery pressure of 55 mmHg and saturation from the ascending aorta of 79% confirmed the diagnosis. After positive vasoreactivity test (pulmonary artery resistance decreased by nasal oxygen from 7.87 wood units to 3.4 wood units), surgery was planned.

The procedure was performed through a median sternotomy with standard aorto-bicaval cannulation and under hypothermic CPB. The pulmonary arteries were excised from the ascending aorta by careful dissection to prevent possible injuries to the truncal valve or left coronary artery (Figure 1). The pulmonary arteries were snared at the onset of bypass and the heart was arrested with antegrade cold cristalloid cardioplegia and it was repeated in every 15 minute while cooling to 26 ºC. The truncal valve was, then, inspected and found to be quadricuspid and stenotic. We performed commissurotomy and tricuspidization of the truncal valve which resulted in mild stenosis and insufficiency during intraoperative transesophageal echocardiography (TEE). The defect in the ascending aorta was, then, closed directly without distorting the truncal valve and coronary ostia. A vertical right ventriculotomy was made and the ventricular septal defect was closed with a Dacron patch. As a standard approach, the continuity between the right ventricle and pulmonary arteries was established with a 14 mm valved conduit (Contegra, Medtronic Inc., Minneapolis, MN, USA). After weaning from CPB, the right ventricular pressure was 68/7 (mean pressure 29) mmHg, while systemic arterial pressure was 80/47 (mean pressure 62) mmHg. The patient was transferred to the intensive care unit (ICU) with dopamine (5 µg/kg/min), adrenalin (0.05 µg/kg/min), nitroglycerin (0.25 µg/kg/min). Postoperative transcutaneous oxygen saturation was 88 to 92%. Postoperative echocardiographic evaluation (at four hours) showed moderate aortic insufficiency and minimally tricuspid insufficiency with normal ventricular functions and a patent right ventricle to the pulmonary artery conduit.

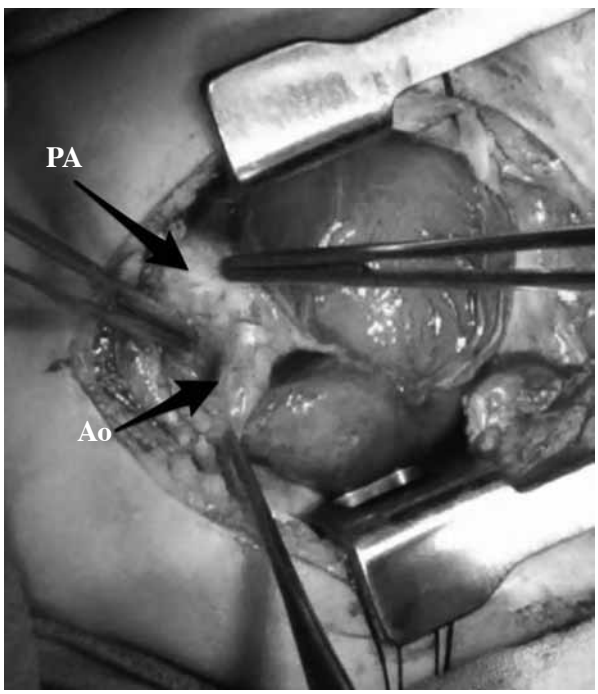

Figure 1. An intraoperative view of the truncus arteriosus.
PA: Pulmonary artery; Ao: Aorta.

Since PH is an important cause of morbidity and mortality in patients with TA beyond three months of age,[3] we attempted to initiate inhaled nitric oxide upon weaning from CPB. Nitric oxide was given between 10 to 20 ppm and continued 24 hours postoperatively. Then, the patient was extubated. Meanwhile, we administered bosentan (Tracleer, Actelion Pharmaceuticals Ltd., Allschwil, Switzerland) via a nasogastric tube with a dose of 0.1 mg/kg at 16 hours postoperatively. After extubation, we continued bosentan together with inhaled iloprost (Ventavis, Bayer Schering Pharma AG, Spain) for further management of PH. This protocol was continued for the first month after the operation. The length of intensive care unit and hospital stays was three and eight days, respectively.

Repeated echocardiography at one month showed minimal insufficiency at the truncal valve and a progressive decrease in the pulmonary artery pressure (mean pressure: 16 mmHg) without any other complication.

DISCUSSION

Although the surgical management of truncus arteriosus has evolved over the past decade to include primary correction in the neonatal period, timing of TA repair is still controversial.[1] Although favorable results have been achieved to date, certain challenges still remain: the management of complex anatomy including truncal valve regurgitation, and the optimum method of establishing right ventricular to pulmonary artery continuity. Current opinion suggests that reconstructing of the right ventricular outflow tract (RVOT) with a valved allograft conduit is probably the optimal method of choice.[5] Elective repair of TA without major associated cardiac anomalies during the first three months of life is also advocated by some surgeons, while others prefer primary neonatal

correction.[5,6] Of note, delayed operation beyond 100 days increases the risk of pulmonary hypertensive crises.[6] Repair of the truncal valve is an option which has been increasingly adopted with favorable outcomes in recent years, both at the time of primary repair and at reoperation.[7] We repaired truncal valve insufficiency by suturing rudimentary leaflet to the adjacent leaflet, making the valve trileaflet which resulted in mild to moderate regurgitation by TEE. A mild to moderate regurgitation is usually well-tolerated postoperatively, after the volume overload is able to be reduced, as in our case.

Furthermore, PH is one of the major causes of morbidity and mortality in pediatric patients with congenital heart diseases.[5] Congenital heart defects which are associated with significantly increased pulmonary blood flow and pulmonary venous obstruction are most likely predisposing postoperative PH. Truncus arteriosus, particularly, is one of the major pathology leading to PH early in life; therefore, early surgical correction is the most appropriate approach. As in our case, truncal valve stenosis may prevent PH which may also cause late diagnosis.

In our patient, after a corrective surgery was decided, we planned to use nitric oxide for the postoperative period. Despite all disadvantages of total correction in this patient population, nitric oxide was very useful to lower the pulmonary vascular resistance. Inhaled nitric oxide is the standard pharmacological therapy per protocol for postoperative pH at our clinic and is usually utilized in conjuction with 100% moderate oxygen-controlled hyperventilation with reduced carbon dioxide to maintain lung volumes near the functional residual capacity and to decrease metabolic support through paralysis and sedation.

In conclusion, delayed truncus arteriosus can be treated successfully thanks to novel surgical techniques and pharmacological methods.

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