Anomalous origin of one pulmonary artery branch from the ascending aorta: experience of our center

Çıkan aorttan köken alan anormal pulmoner arter dalı: Merkezimizin deneyimi

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Objectives: Anomalous origin of one pulmonary artery branch from the aorta in the presence of separate aortic and pulmonary valves is a rare but important entity necessitating early diagnosis and surgery to prevent irreversible vascular pulmonary disease. We evaluated our experience with seven infants having this anomaly.

Study design: Between December 2003 and 2009, a total of seven infants (2 girls, 5 boys, age range 4 days to 84 days) were diagnosed as having anomalous origin of one pulmonary artery branch from the aorta. Clinical records were reviewed for clinical features, operative procedures, and postoperative follow-up.

Results: Common symptoms were dyspnea, tachypnea, and poor feeding. All the cases were diagnosed by echocardiography. The right pulmonary artery in six cases and the left pulmonary artery in one case originated from the ascending aorta. In addition, three patients had patent ductus arteriosus (PDA), five patients had patent foramen ovale, and one patient had interruption of the aortic arch and aortopulmonary window. All patients underwent surgical re-implantation of the anomalous pulmonary artery branch to the pulmonary trunk. Associated surgical procedures included PDA ligation in three patients, and total repair of interrupted aortic arch and aortopulmonary window in one patient. There were no in-hospital deaths. Two patients had prolonged intubation (10 and 16 days). All patients were discharged in good clinical condition. During a follow-up period ranging from two months to six years, only one patient developed stenosis at the site of anastomosis.

Conclusion: Prompt diagnosis at infancy, improved surgical technique, and good patient care decrease mortality and morbidity associated with anomalous origin of the pulmonary artery from the aorta.

Key words: Echocardiography; heart defects, congenital/surgery; infant, newborn; pulmonary artery/abnormalities/surgery.

Amaç: Aortik ve pulmoner kapakların ayrı ayrı varlığında, bir pulmoner arter dalının çıkan aorttan köken alması nadir rastlanan bir anomalidir ve geri dönüşsüz pulmoner vasküler hastalık gelişiminin engellenmesi için erken tanı ve cerrahi tamir gerektirir. Bu çalışmada bu nadir patolojinin görüldüğü yedi bebekle ilgili deneyimimiz değerlendirildi.

Çalışma planı: Aralık 2003 ile 2009 tarihleri arasındaki dönemde yedi hastada (2 kız, 5 erkek; yaş aralığı 4-84 gün) pulmoner arter dalının çıkan aorttan köken aldığı anomali tanısı kondu. Hastane kayıtlarından olguların klinik özellikleri, cerrahi işlemleri ve takip sonuçları incelendi.

Bulgular: Hastaların ortak başvuru semptomları nefes darlığı, hızlı soluma ve beslenme zorluğuydu. Tüm olgularda tanı ekokardiyografi ile kondu. Altı hastada sağ pulmoner arter, bir hastada sol pulmoner arter çıkan aorttan köken almaktaydı. Eşlik eden anomaliler üç hastada duktus arteriyozus açıklığı, beş hastada foramen ovale açıklığı, bir hastada aortik ark kesintisi ve aortopulmoner pencere idi. Tüm hastalarda cerrahi olarak anormal pulmoner arter pulmoner gövdeye taşındı. Ayrıca, üç hastada duktus arteriyozus açıklığı bağlandı ve bir hastada aortik ark kesintisi ve aortopulmoner pencere onarımı yapıldı. Hiçbir olguda hastaneiçi ölüm görülmedi. İki hastada entübasyon süresi uzadı (10 ve 16 gün). Tüm hastalar iyi durumda hastaneden taburcu edildi. Olguların iki ay ile altı yıl arasında değişen izlem süreleri içinde, sadece bir hastada anastomoz bölgesinde hafif derecede darlık gelişti.

Sonuç: Bebeklik döneminde erken tanı, zamanında cerrahi girişim ve ameliyat sürecinde iyi bakım, pulmoner arter dalının çıkan aorttan köken almasına bağlı morbidite ve mortaliteyi azaltmaktadır.

Anahtar sözcükler: Ekokardiyografi; kalp defekti, doğuştan/cerrahi; bebek, yenidoğan; pulmoner arter/anormallik/cerrahi.

Received: January 23, 2010 *Accepted:* March 24, 2010

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Anomalous origin of one pulmonary artery branch from the ascending aorta (hemitruncus) is a rare congenital heart disease, in which one of the pulmonary artery branches arises from the ascending aorta in the presence of separate aortic and pulmonary valves.[1,2] The pathology is frequently associated with other cardiac malformations and rarely presents as an isolated anomaly.[2-4] The condition is included in the group of aortic arch abnormalities and is caused by partial or complete developmental failure of the left sixth arch.[5]

Hemitruncus results in a large left-to-right shunt with the entire cardiac output from the right ventricle going to one lung while the other lung receives blood at systemic pressure from the aorta. Thus, the pulmonary vascular bed of both lungs may be vulnerable to the development of pulmonary vascular obstructive disease. Without surgery, one-year survival has been reported to be very low.[6] Early operation with good preoperative and postoperative care results in excellent survival.[7-9]

We evaluated surgical results of hemitruncus with separate aortic and pulmonary valves in seven infants with an emphasis on diagnostic evaluation.

PATIENTS AND METHODS

Between December 2003 and December 2009, a total of seven patients were diagnosed as having hemitruncus in normally connected hearts with two separate semilunar valves. Patients with a common arterial trunk with nonconfluent pulmonary arteries were excluded from the study. Clinical records were reviewed to document clinical features, operative procedures, and postoperative follow-up. Informed consent was obtained from the parents before operation, approval was obtained from the local ethics committee and hospital administration to review the files retrospectively.

RESULTS

Of seven patients, five patients were males, two patients were females. At the time of diagnosis, the age of the patients ranged from 4 to 84 days, and the weight from 3 to 4.8 kg. Common symptoms at presentation were dyspnea, tachypnea, and poor feeding. One patient presented with transient cyanosis because of a lung infection.

All the cases were diagnosed by echocardiography and only two patients underwent catheter-angiography for evaluation of cardiac morphology and associated anomalies (Fig. 1). The anomalous branch was the right pulmonary artery in six patients, and the left pulmonary artery in one patient. In all cases, the other pulmonary artery was in continuity with the main pulmonary artery originating from the right ventricle. All the patients had evidence for pulmonary hypertension detected by echocardiography. Associated anomalies included patent ductus arteriosus (PDA) in three patients, patent foramen ovale in five patients, and interruption of the aortic arch and aortopulmonary window in one patient.

Operative technique was direct implantation of the abnormal pulmonary artery branch to the main pulmonary artery in all the patients. The anomalous pulmonary artery was cut from its origin at the ascending aorta and anastomosed to the lateral aspect of the main pulmonary artery in end-to-side fashion (Fig. 2). The defect in the aorta was directly sutured. A median sternotomy was preferred in all the cases. Associated surgical procedures included PDA ligation in three patients, and total repair of interrupted aortic arch and aortopulmonary window in one patient. There was no intraoperative mortality. Two cases were extubated within 48 hours. Two patients developed early postoperative pulmonary hypertensive crisis that required prolonged ventilation (10 and 16 days) with the use of inhaled prostacyclin.

During a follow-up period ranging from two months to six years, none of the patients developed significant stenosis at the anastomosis line except one patient with mild stenosis. Periodical echocardiographic examination was suggested for this patient for a pulse wave gradient of 25 mmHg at the anastomosis site.

In one patient, a complex surgical procedure was performed in the same session, consisting of re-implantation of the right pulmonary artery to the main pulmonary artery, reconstruction surgery for type A interruption, closure of the aortopulmonary window, and PDA ligation. After a prolonged intubation period with several pulmonary hypertensive crises, the patient was discharged from hospital on the 43rd postoperative day.

DISCUSSION

Anomalous origin of one pulmonary artery branch from the ascending aorta is an extremely rare conotruncal malformation accounting for only 0.12% of all congenital heart diseases.[2,4,5] The disease was first described by Fraentzel in 1868.[10] Since then, there have been several case reports and case series. Anomalous origin of the right pulmonary artery is 5 to 6 times more frequent than the left.[2,4,5] In our se-

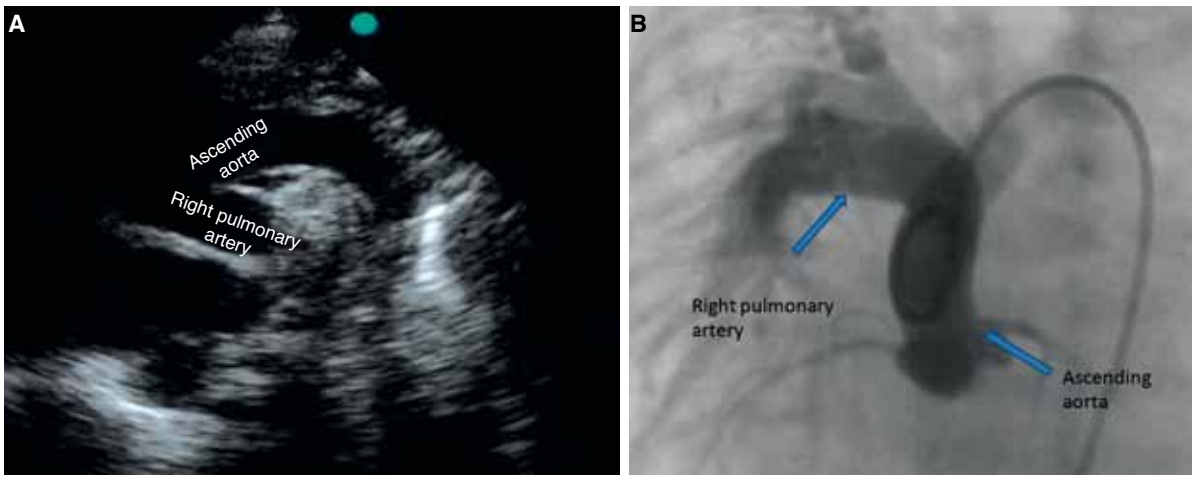

Figure 1. (A) Echocardiographic and **(B)** angiographic appearance of the right pulmonary artery originating from the ascending aorta.

ries, the anomalous branch was the right pulmonary artery in six cases. Usually the anomalous branch originates from the posterolateral wall of the ascending aorta, near the aortic valve. The pathology may be isolated or in association with other congenital heart defects, including tetralogy of Fallot, PDA, aortopulmonary window, arcus anomalies, and double outlet ventricle.[3,4,11-13] Several cases of isolated malformation have been reported, but its incidence is very limited. Left-sided anomalous origin of the pulmonary artery is frequently associated with tetralogy of Fallot and right aortic arcus.[14] In four of our cases, there was only a patent foramen ovale, which is a finding contrary to the literature.

Within days or a week after birth, tachypnea, dyspnea, and clinical symptoms of congestive heart failure become evident in these patients. Infants usually have a large left-to-right shunt without cyanosis. The pathology should be specifically investigated during echocardiographic examination in infants with heart failure without an intracardiac pathology.

Echocardiography is very important in detecting this rare malformation. Echocardiographic diagnosis relies on the presence of two concordant ventricular outflow tracts, absence of the usual bifurcation pattern of the pulmonary artery, and observation of the right or left pulmonary artery arising directly from the aorta, with the main pulmonary arterial tract continuing with the contralateral pulmonary branch.[15,16] We diagnosed the anomaly in all cases by transthoracic echocardiography and cardiac catheterization was performed in two patients to demonstrate the underlying pathology. One of the these patients had a complex pathology with interruption of the aortic arch and aortopulmonary window, the other had an isolated anomaly. Cardiac catheterization may sometimes be

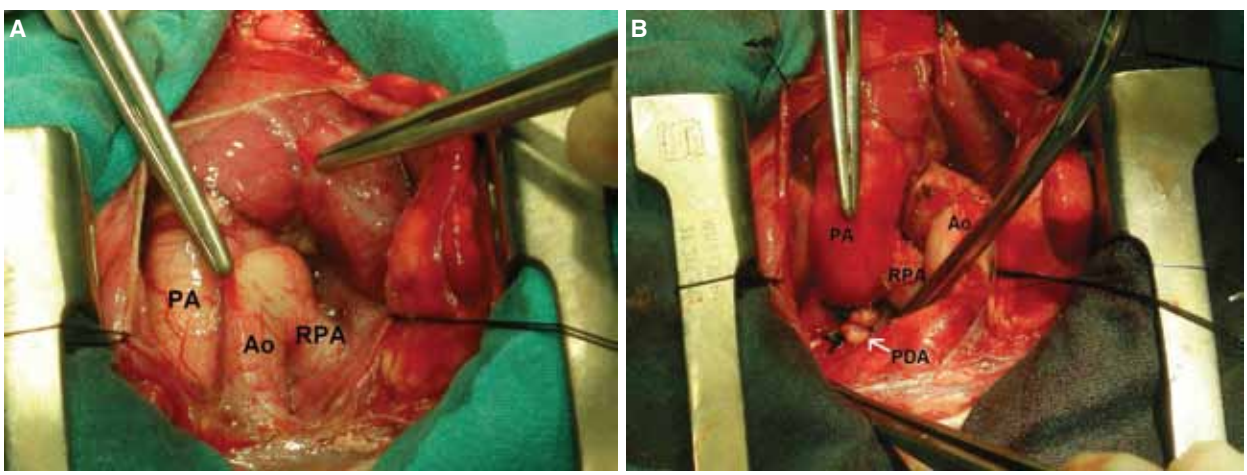

Figure 2. Macroscopic demonstration of an anomalous origin of the right pulmonary artery from the ascending aorta **(A)** before and **(B)** at the end of the operation. PA: Pulmonary artery; Ao: Aorta; RPA: Right pulmonary artery; PDA: Patent ductus arteriosus.

indicated preoperatively as a diagnostic procedure or as a therapeutic procedure for the treatment of surgical complications.[4,7,17-19]

Successful correction of this anomaly in the first days of life, even in premature babies, have been reported.[8] Early repair is preferred to avoid pulmonary hypertension and irreversible pulmonary vascular disease. Serious pulmonary vascular disease may develop as early as the third month of life due to several mechanisms including high pulmonary blood flow, circulating vasoconstrictor substances, neurogenic crossover from the unprotected lung, and left ventricular failure.[2,7,14] Our patients were diagnosed before three months of age, so a pulmonary vascular obstructive disease was not supposed to have developed before surgery.

Different surgical techniques have been employed but the most frequent is direct anastomosis of the anomalous pulmonary branch to the main pulmonary trunk.[7-9,20] End-to-end anastomosis with a synthetic graft, interposition of a homograft patch, aortic flap, or interposition of an autologous pericardial patch to increase the length of the anomalous branch have been successfully employed in specific cases where direct implantation is not possible.[7-9,21] We used direct implantation technique in our cases. Only in one patient, surgery was complicated with the development of mild stenosis at the site of anastomosis early after the procedure.

In conclusion, anomalous origin of one pulmonary artery branch from the ascending aorta is a rare disease that may cause pulmonary hypertension and irreversible pulmonary vascular disease if not treated in early infancy. Prompt diagnosis at infancy, improved surgical techniques, good perioperative and postoperative care decrease mortality and morbidity of this anomaly.

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