

Right Aortic Arch with a Retroesophageal Left Subclavian Artery and an Anomalous Origin of the Pulmonary Artery from the Aorta

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We report the case of a newborn with a rare anatomic variation: a right aortic arch with a retroesophageal left subclavian artery and an anomalous origin of the pulmonary artery from the aorta. This variation was diagnosed using echocardiography and computed tomography, and we treated the condition surgically.

Key words: 1. Congenital heart disease
2. Aortic arch
3. DiGeorge syndrome

Case report

A 6-day-old boy was followed up for a right aortic arch on fetal echocardiography observed at 35 weeks of gestation. His mother underwent a cesarean section at 36 weeks of gestation due to labor pain and preterm rupture of the membranes. The infant's birth weight was 2,270 g, and the Apgar scores at 1 and 5 minutes after birth were 9 and 10, respectively. The infant was referred to Samsung Medical Center due to a right aortic arch with atrial septal defect (ASD) and patent ductus arteriosus (PDA) on postnatal echocardiography. On echocardiography performed at our hospital, severe pulmonary hypertension was observed and a double aortic arch, in which the left aortic arch was interrupted, was suspected. The right pulmonary artery was found to emerge from the ascending aorta. In addition, a large left PDA with a retroesophageal left subclavian artery (LSCA) and right-to-left shunt flow was observed. An intracardiac anomaly (moderate secundum ASD) was also present.

A 3-dimensional computed tomography (CT) heart scan showed an anomalous origin for the right pulmonary artery from the right aorta and retroesophageal LSCA from the left PDA; the right aortic arch and left PDA formed vascular rings, but tracheal stenosis was not clearly seen (Fig. 1, Fig. 2). The infant underwent surgical treatment at 7 days after birth due to aggravated heart failure symptoms. Median sternotomy was performed. The thymus was not visible. Cardiopulmonary bypass (CPB) was initiated with the standard cannulation technique in the ascending aorta and a bicaval venous cannula, at 34°C. The PDA was ligated and resected at the attachment site of the main pulmonary artery (MPA) and aorta through Kommerell diverticulum resection, followed by LSCA division. The right pulmonary artery, which originated from the posterior side of the ascending aorta, was divided and reimplanted into the MPA, and the ASD was closed with autologous pericardium. After rewarming, the patient was weaned from CPB. The CPB time was 144 minutes and the aortic cross-

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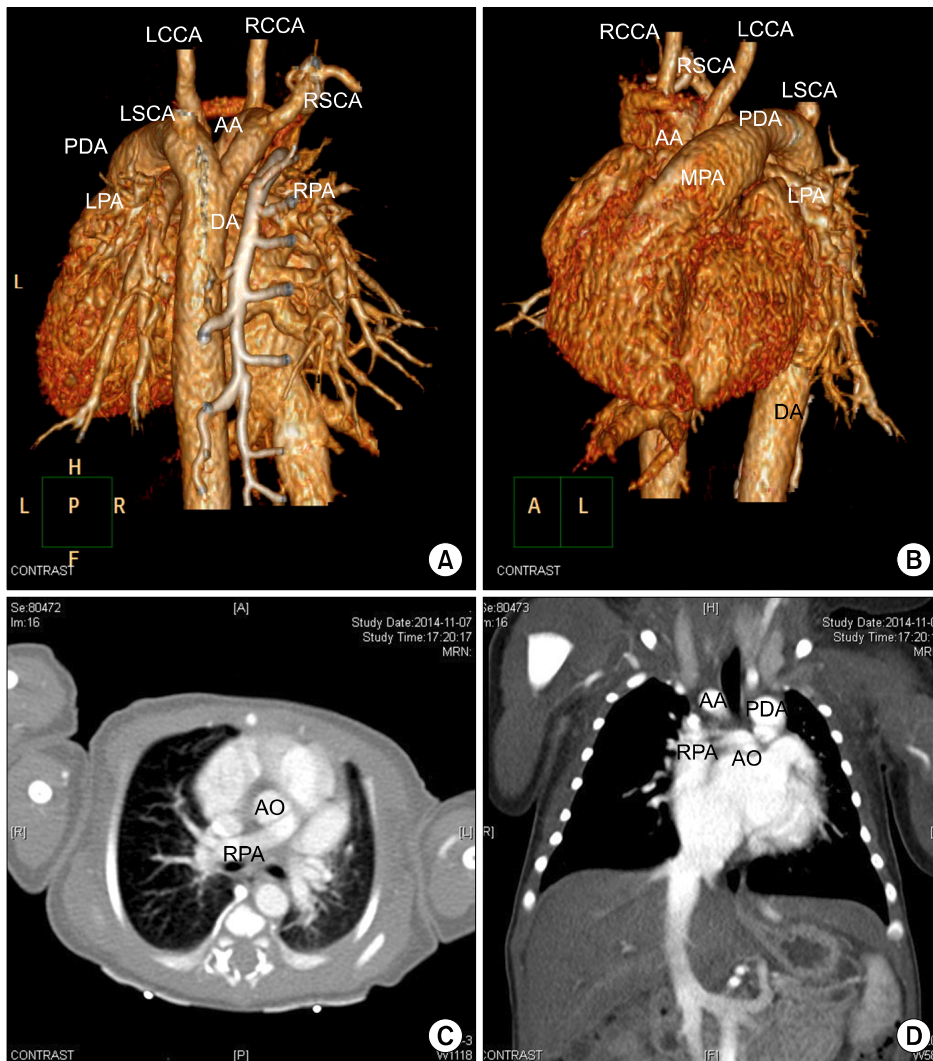


Fig. 1. (A-D) Computed tomography angiography showing right AA with retroesophageal LSCA and anomalous origin of pulmonary artery from aorta. AA, aortic arch; AO, aorta; RPA, right pulmonary artery; RCCA, right common carotid artery; LCCA, left common carotid artery; RSCA, right subclavian artery; MPA, main pulmonary artery; LPA, left pulmonary artery; PDA, patent ductus arteriosus; LSCA, left subclavian artery; DA, descending aorta; P, posterior; A, anterior; L, left; H, head; R, right; F, front.

clamping time was 67 minutes. After surgery, nitric oxide ventilation was necessary due to pulmonary hypertension. The patient was extubated on the 11th postoperative day. On postoperative echocardiography, the right aortic arch flow was good and the right pulmonary artery was found to be normally connected to the MPA. No residual ASD was found. Chromosomal analysis revealed 22q11 partial deletion, also known as DiGeorge syndrome. The infant was discharged on the 28th postoperative day.

Discussion

We diagnosed the neonate with right aortic arch with retroesophageal LSCA and anomalous origin of the pulmonary artery from the aorta using echo-

cardiography and CT, and we treated the condition surgically. This variant has been reported by Hamzeh et al. [1], but our case showed some differences in terms of the presence of an intracardiac anomaly (ASD) and DiGeorge syndrome. Congenital anomalies of the aortic arch range from asymptomatic normal variations in the arch vessel branch pattern to symptomatic vascular rings, stenosis, and arch interruptions. The frequency of aortic arch anomalies ranges from 0.5% to 3.0%. Right aortic arch occurs in approximately 0.1% of adults and is strongly associated with the presence of chromosome 22q11 deletion, also known as DiGeorge syndrome [2,3]. Right aortic arch with retroesophageal LSCA and left descending aorta is a rare configuration of the right aortic arch and may be a variant of the double aortic arch with

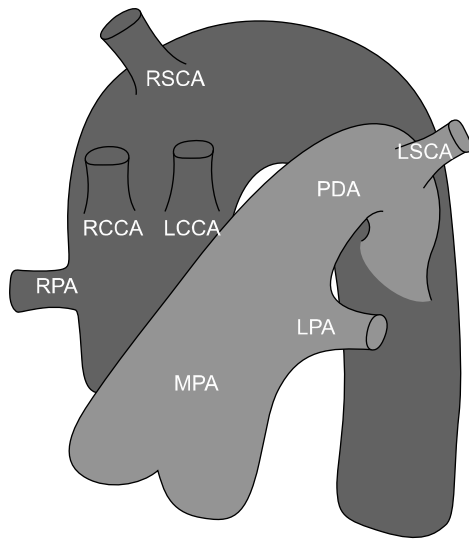


Fig. 2. Diagram. RPA, right pulmonary artery; RCCA, right common carotid artery; LCCA, left common carotid artery; RSCA, Right subclavian artery; MPA, main pulmonary artery; LPA, Left pulmonary artery; PDA, patent ductus arteriosus; LSCA, Left subclavian artery.

focal left arch atresia [2]. In this case, there was a left PDA, forming a vascular ring, but the vascular ring was relatively loose due to the anomalous origin of the right pulmonary artery. This rare variant accompanied by ASD, however, increased left-to-right shunting, resulting in aggravation of pulmonary hypertension and heart failure, which required early surgical treatment. Translocation of the retroesophageal LSCA to the left carotid artery with Kommerell diverticulum resection, PDA division, right pulmonary artery reimplantation to the MPA, and ASD closure is the preferred surgical approach and can be performed in most cases. Occasionally, however, translocation of the retroesophageal LSCA to the left carotid artery may be a less attractive option because of technical factors related to low-birthweight neonates.

In selected cases of right aortic arch with retroesophageal LSCA, especially in neonatal patients, division of the LSCA without reimplantation may be an alternative [4]. Right aortic arch anomalies are rare, but show diverse variations. Therefore, accurate diagnosis may not be possible by echocardiography alone. A CT scan can support further evaluation and cardiac catheterization, and magnetic resonance imaging may also be useful [5,6].

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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