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Isolation of the Left Subclavian Artery with Right Aortic Arch in Association with Bilateral Ductus Arteriosus and Ventricular Septal Defect

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Right aortic arch with isolation of the left subclavian artery is a rare anomaly. The incidence of bilateral ductus arteriosus is sporadic, and a right aortic arch with isolation of the left subclavian artery in association with bilateral ductus arteriosus is therefore extremely rare. Since the symptoms and signs of isolation of the left subclavian artery can include the absence or underdevelopment of the left arm, subclavian steal syndrome, or pulmonary artery steal syndrome, the proper therapeutic approach is controversial. We report a case in which surgical reconstruction was used to treat isolation of the left subclavian artery with right aortic arch in association with bilateral ductus arteriosus and a ventricular septal defect.

Key words: 1. Aorta, arch
2. Congenital heart disease
3. Embryology

CASE REPORT

A girl weighing 3,190 g was delivered by a Caesarean section at 38 weeks and five days of gestation. At the time of birth, a systolic murmur was noted during a physical examination, and transthoracic echocardiography revealed a perimembranous ventricular septal defect (VSD) with a septal aneurysm, a small patent foramen ovale (PFO), and a small right-sided patent ductus arteriosus (PDA) from the innominate artery. The VSD was measured as having a diameter of 3.5 mm and a shunt flow less than 2.5 m/sec. Due to the presence of neonatal hyperbilirubinemia, echoencephalography was conducted, and no abnormalities were found. Five months later, the patient was referred to Konkuk University Medical Center for VSD and PDA. Her parents stated that she had shown shortness of breath while feeding. Her body weight was 6,700 g (25th percentile), and her height was 61.6 cm (10th percentile). She showed no cyanosis and no underdevelopment of any extremities. Her blood pressure, as measured at both upper extremities, was normal. A chest X-ray showed cardiomegaly. Transthoracic echocardiography revealed a VSD approximately 6 mm in size with a minimal aneurysm, a left-sided PDA 3.6 mm in diameter from the right aortic arch and an aberrant left subclavian artery. A subsequent computed tomographic scan demonstrated isolation of the left subclavian artery with a right aortic arch, a left PDA, and a VSD (Fig. 1A). Surgery was performed through median sternotomy. The intraoperative findings were a perimembranous VSD, a PFO,

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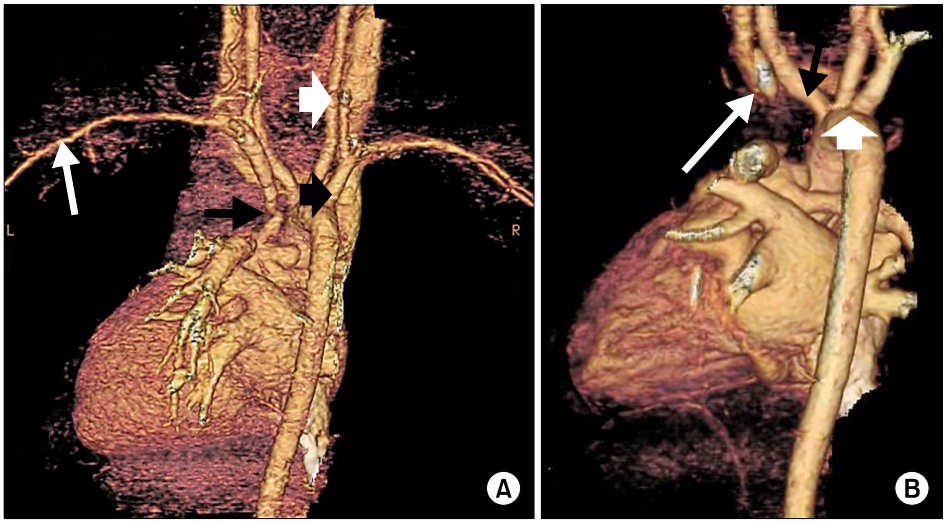

Fig. 1. (A) Preoperative computed tomography scan. The left subclavian artery (white arrow) is connected via the left patent ductus arteriosus (black arrow) to the left pulmonary artery. The white arrowhead indicates the right common carotid artery. The black arrowhead indicates the right subclavian artery. (B) Postoperative computed tomography scan. The left subclavian artery (white arrow) was reimplanted to the left common carotid artery (black arrow). The white arrowhead indicates the right common carotid artery and the right subclavian artery.

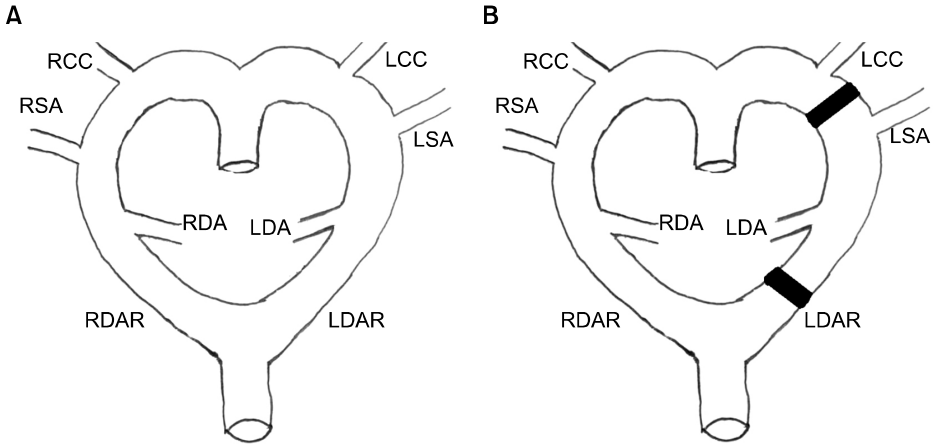

Fig. 2. (A) Schematic diagram on basic hypothetical aortic arch plan suggested by Edward. (B) Schematic diagram on isolation of left subclavian artery, bilateral ductus arteriosus and right aortic arch. Black shadow indicate the sites of regression. RCC, right common carotid artery. LCC, left common carotid artery. RSA, right subclavian artery. LSA, left subclavian artery. RDA, right ductus arteriosus. LDA, left ductus arteriosus. RDAR, right descending aortic root. LDAR, left descending aortic root.

a mildly patent right ductus arteriosus, and isolation of the left subclavian artery connected to the left pulmonary artery via a left PDA. Both PDAs were divided and oversewn. The left subclavian artery was disconnected from the left pulmonary artery and reimplanted to the left common carotid artery by end-to-side anastomosis with monofilament polypropylene 6-0 sutures (Fig. 1B). Under cardiopulmonary bypass, direct closure of the VSD and the PFO was performed via right atriotomy. The patient was discharged uneventfully.

DISCUSSION

Ductus arteriosus is usually located on the left side, between the descending aorta and the junction of the main pulmonary artery and left pulmonary artery. However, ductus arteriosus may also be present on the right side or, very rarely, may occur bilaterally in association with aortic arch anomalies or conotruncal anomalies [1]. In such aortic arch anomalies, isolation of the left subclavian artery with right aortic arch is also uncommon. Here, isolation refers to the fact that the left subclavian artery connects to the pulmonary artery via

either the ligamentum arteriosum or a patent ductus arteriosus without any connection to the aorta. Isolation of the left subclavian artery with a right aortic arch is known to be commonly associated with congenital heart disease [2,3], but may also occur with normal intracardiac anatomy, although few such cases have been described [1,4]. Isolation of the left subclavian artery with a right aortic arch may be related to the 22q11 deletion [3,4]. However, our patient showed a normal genotype in this regard.

Bilateral ductus arteriosus and isolation of the left subclavian artery with a right aortic arch can be explained through the hypothetical double aortic arch plan suggested by Edward [5]. Regression takes place on two levels in the double aortic arch plan: on one level, regression occurs between the left common carotid artery and the left subclavian artery; and on the other level, regression occurs at the left dorsal aortic root distal to the left ductus arteriosus. And then right ductus arteriosus remains persistent, left ductus arteriosus connects the left subclavian artery to the left pulmonary artery (Fig. 2). In our patient, both ductus arteriosi were patent at birth. However, the right ductus arteriosus regressed, and only the left ductus arteriosus remained patent. If the left ductus arteriosus is patent, blood may be supplied to the left subclavian artery via the left ductus arteriosus. If the left ductus arteriosus regresses, the blood supply to the left subclavian artery may involve a mediastinal, thoracic anastomosis, or vertebral pathway. Thus, pulmonary steal syndrome and/or subclavian steal syndrome may occur. Isolation of the left subclavian artery usually presents with no apparent symptoms in neonates, but it may present with congenital pulmonary steal syndrome, subclavian steal syndrome, or may even present in adults with late symptoms due to sporadic progression. Hayabuchi et al. [6] reported the case of a three-month-old girl with cerebral atrophy and an underdeveloped left arm. Jesudian et al. [7] reported the case of a 15-year-old boy with an underdeveloped left arm. Due to these symptoms and signs, the therapeutic management of isolation of the left subclavian artery remains controversial, especially when it is associated with complicated congenital heart disease. Some authors have suggested that adequate collateral circulation must be ensured, meaning that reconstruction of the isolated subclavian artery is optional, regardless of the symptoms and signs. Successful results have been reported after ligation or device closure of the PDA and ligation of the left subclavian artery [1,6]. However, reconstruction of the left subclavian artery due to pulmonary steal syndrome after right PDA closure in bilateral PDA has been reported. In one report, ischemic symptoms in the left arm and vertebrobasilar insufficiency occurred years after ligation of the left subclavian artery [1]. Hokari et al. [8] reported that a man with Peutz-Jeghers syndrome presented with his first vertigo attacks due to subclavian steal syndrome at 29 years of age. Our patient presented with no symptoms and signs related to subclavian or pulmonary steal syndrome, and had shown normal findings on an echoencephalography study conducted at our medical center due to neonatal hyperbilirubinemia. However, brain computed tomography angiography performed after surgery revealed hypoplasia of the left vertebral artery. We suggest that this hypoplasia would have led to vertebrobasilar insufficiency or underdevelopment of the left arm without surgical reconstruction. Since surgical reconstruction of the isolated left subclavian artery leads to antegrade flow in the left subclavian artery, it can prevent hypoplasia of the left vertebral artery and subclavian/pulmonary steal syndrome. Our case shows that early surgical reconstruction is reasonable, regardless of the symptoms, in cases of isolation of the left subclavian artery.

CONFLICT OF INTEREST

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

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