

Persistent Fifth Aortic Arch with Coarctation

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Persistent fifth aortic arch (PFAA) is a rare congenital anomaly of the aortic arch frequently associated with other cardiovascular anomalies, such as tetralogy of Fallot and aortic arch coarctation or interruption. We report the case of a neonate with PFAA with coarctation who successfully underwent surgical repair.

Key words: 1. Aorta, arch
2. Coarctation

CASE REPORT

Persistent fifth aortic arch (PFAA) is an exceedingly rare congenital anomaly of the aortic arch, frequently associated with other cardiovascular anomalies, such as tetralogy of Fallot, transposition of the great arteries, truncus arteriosus, and aortic arch coarctation or interruption [1]. PFAA originates from the distal ascending aorta, proximal and opposite to the ostium of the innominate artery, and is distally connected with the descending aorta [2]. We report a case of PFAA with coarctation, in which surgical repair was successfully performed.

An eight-day-old female neonate was referred to Cardiac Center of Seoul National University Children's Hospital due to PFAA with coarctation. She was born as the first twin by Caesarean section at 36+4 weeks of gestation after an uneventful pregnancy. Prenatal echocardiography revealed a perimembranous ventricular septal defect (VSD). Her birth weight was 2,230 g, and her Apgar score was 8 at one minute and 9 at five minutes. Her blood pressure was 95/59 mmHg in the right arm, 103/51 mmHg in the left arm, 60/44 mmHg in the right leg, and 55/33 mmHg in the left leg.

Echocardiography on admission revealed left PFAA with coarctation, secundum atrial septal defect (ASD), perimembranous VSD, and closed ductus arteriosus (Fig. 1A). Three-dimensional computed tomography confirmed the diagnosis of PFAA with coarctation (Fig. 1B).

One day after admission, the patient was operated on via a standard median sternotomy. Intraoperative findings revealed that the PFAA was connected to the descending thoracic aorta through the isthmus, which was closed by ductal tissue. Coarctation of the fourth aortic arch with a short and very tight stenotic isthmus was also observed (Fig. 2). The arterial cannula was inserted into the innominate artery, and the venous return cannulae were inserted into the right atrium. After the isthmus of the fourth aortic arch was divided, the fifth aortic arch and the ductal tissue were resected under regional perfusion. The coarctation of the PFAA was repaired by end-to-end anastomosis between the stump of the fifth aortic arch and the descending thoracic aorta (Fig. 3). The VSD and ASD were closed through a right atrial incision. The perioperative period was uneventful. Immediately after the operation, the patient's blood pressure was 82/52 mmHg in the left arm and 89/49 mmHg in the left leg. The patient

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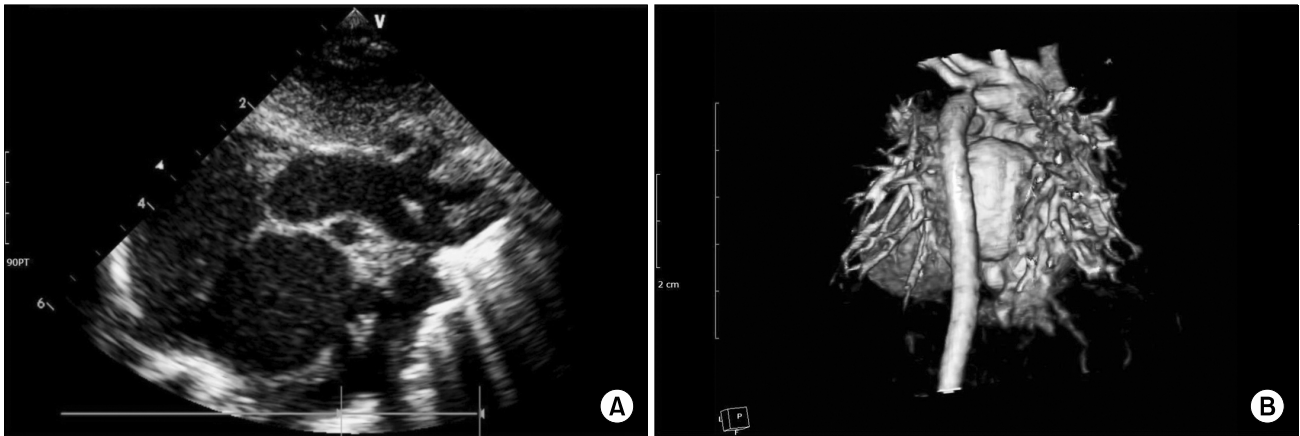


Fig. 1. (A) Echocardiogram and (B) three-dimensional computed tomography image showing the persistent fifth aortic arch (arrowhead) with segmental coarctation (arrow).

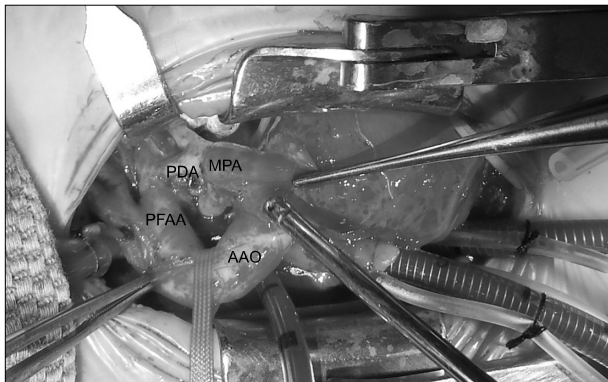


Fig. 2. Intraoperative view of the PFAA. AAO, ascending aorta; PFAA, persistent fifth aortic arch; PDA, patent ductus arteriosus; MPA, main pulmonary artery.

was discharged home 17 days after the operation without complications. Computed tomography performed before discharge showed that the anastomosis of the PFAA was wide and unobstructed (Fig. 4).

DISCUSSION

PFAA was first described by Van Praagh and van Praagh [2] in 1969. PFAA is located between the true aortic arch, which is a derivative of the fourth aortic arch, and the pulmonary artery, which is a derivative of the sixth aortic arch. The aortic arch is subdivided into superior and inferior parallel channels, each consisting of a double-lumen aortic arch.

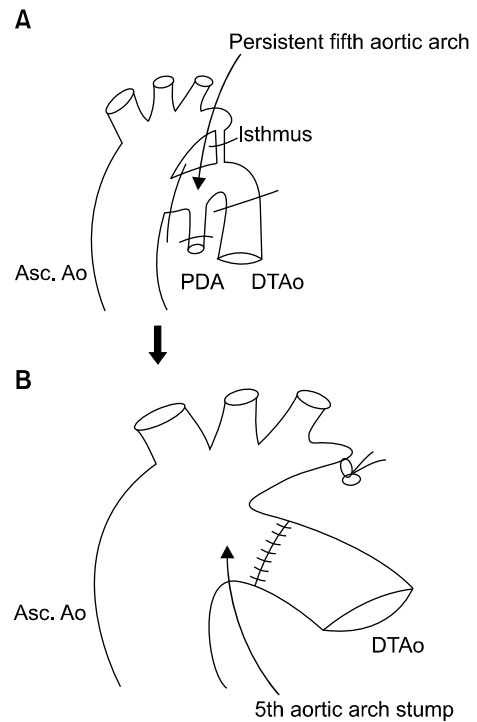


Fig. 3. Schematic drawing of persistent fifth aortic arch with coarctation. (A) The isthmus area and ductus arteriosus were divided. Dotted lines represent incisions. (B) Coarctoplasty was performed via end-to-end anastomosis between the orifice of the fifth aortic arch and the descending thoracic aorta. Asc. Ao, ascending aorta; PDA, patent ductus arteriosus; DTAo, descending thoracic aorta.

Previous studies of PFAA showed that patients with this malformation usually do not have hemodynamically significant



Fig. 4. Postoperative computed tomography image showing the wide and unobstructed anastomosis site (arrow).

symptoms and are diagnosed incidentally due to upper body hypertension and systolic murmur if the PFAA flow is sufficient [3]. According to a review of 26 cases of PFAA by Lambert et al. [1], 76% of cases demonstrated systemic-to-systemic connection via the fifth aortic arch, 38% of cases involved aortic coarctation, and 19% of cases presented with interruption of the fourth aortic arch. Echocardiography and angiography are used as tools to diagnose PFAA. However, magnetic resonance imaging and computed tomography provide more accurate information regarding the relationships with the surrounding structures and the vascular anatomy.

Previous reports have described a range of techniques for repairing PFAA associated with coarctation: (1) interposition of a Gore-Tex tube graft (W.L. Gore & Associates, Flagstaff, AZ, USA) [4], (2) patch aortoplasty augmenting both the fourth and the fifth aortic arch [5], (3) ligation of the fifth aortic arch used to reconstruct the fourth aortic arch [6], and (4) side-to-side anastomosis of the left common carotid artery and the left subclavian artery with patch repair of the coarcted segment [7]. In this case, after division of the fourth aortic arch isthmus and the ductal tissue, coarctoplasty was per-

formed using an end-to-end anastomosis between the proximal stump of the resected fifth aortic arch and the descending thoracic aorta. This procedure produced a good immediate result without complications.

In conclusion, this report describes a rare case of persistent fifth aortic arch associated with coarctation that was successfully surgically repaired.

CONFLICT OF INTEREST

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

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