Spontaneous Closure of Ductus Arteriosus in Interrupted Aortic Arch with Ventricular Septal Defect

A 2-month-old boy diagnosed with interrupted aortic arch type B was treated with a two-stage procedure. His ductus arteriosus had closed spontaneously. Collaterals via both vertebral arteries developed. A 15-mm stenotic segment existed between the left subclavian artery and the descending aorta. The direct anastomosis between the common carotid artery and the descending aorta was performed as a first palliation at the age of 3 months. The left subclavian artery was reconstructed by end-to-side anastomosis to the descending aorta. The postoperative course was uneventful. The closure of ventricular septal defect and pulmonary artery debanding were performed as a second operation 4 months after the first palliation. The patient is alive and well 7 months after the second operation.

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Interrupted aortic arch (IAA) is characterized by the absence of an anatomic continuity within the aortic arch or at the aortic isthmus. It is commonly associated with ventricular septal defect (VSD) and patent ductus arteriosus (PDA). The natural history is that, about 76% of patients die of progressive heart failure following ductus arteriosus (DA) closure within 1 month after birth.

We successfully performed a two-stage operation on a 2-month-old boy with a type B IAA complex with spontaneous DA closure.

Case

A 2-month-old boy, 56 cm in height, and 4.1 kg weight, was referred to us. His systolic blood pressure in the right upper extremity was 130 mmHg, and in the lower extremities 90 mmHg. A chest roentgenogram showed prominence of the pulmonary vasculature, and a cardiothoracic ratio of 70%. Echocardiography confirmed a type B IAA with a 10-mm perimembranous VSD. The aortic valve was bicuspid without subvalvular stenosis. The PG across the aortic valve was 15 mmHg. No blood flow was detected in the DA. Interventricular PG was 23 mmHg consistent with pulmonary hypertension (PH). Angiocardiography confirmed the diagnosis of type B IAA and showed collateral flow through the vertebral arteries bilaterally and other collateral circulation as well (Fig. 1). The left subclavian artery contrasting with the left vertebral artery showed retrograde flow. It was identified that a 15-mm segment had narrowed from the origin of the left subclavian artery (LSCA) to the descending aorta (Desc. Ao.).

In this case, the anastomosis site of the Desc. Ao. would be far via sternotomy. Therefore we decided to perform a staged operation. Because the respiratory rate was increasing, and the blood pressure was becoming unstable, an emergency operation was performed. Thoracotomy was carried out on the left chest with the patient under general anesthesia. After isolation of the aortic arch and Desc. Ao., the narrowed segment of the Desc. Ao. was excised. Continuity was reestablished by direct anastomosis between the aortic arch at
Radial injection

![Radial injection image]

**Fig. 1.** Preoperative angiography.
*Asc. Ao., Ascending aorta; Desc. Ao., descending aorta; LCCA, left common carotid artery; RSCA, right subclavian artery; LSCA, left subclavian artery.*

the left carotid artery root and the Desc. Ao.. Because the origin of the LSCA was stenotic, the stenotic segment was removed and the LSCA was reconstructed by end-to-side anastomosis to the Desc. Ao.. Pulmonary artery banding (PAB) limiting the pulmonary artery circumference to 28 mm was performed, and the chest was closed.

The patient was extubated the next day, and discharged from the ICU the following day. The hospital stay was 24 days. Postoperative angiography showed no stenosis of the aortic arch, and good flow in the LSCA (Fig. 2). Postoperative pressure data were, ascending aorta (Asc. Ao.) 111/61 (83) mmHg, Desc. Ao. 104/60 (79) mmHg, LV-Ao. PG 15 mmHg, MPA PG 52 mmHg. The Qp/Qs was 1.6, and the diameter of the perimembranous VSD was 6.8 mm. Four months later, we closed the VSD and debanded the pulmonary artery (PA). The patient is alive and well after 7 months of follow-up.

**Discussion**

IAA is a disorder of early branchial arch artery development, and represents about 1% of congenital cardiac disease.\(^2\)\(^3\) This lesion usually is accompanied by cardiac malformations such as PDA or VSD. Blood flow to the lower extremities usually is via a PDA. Celoria and Patton\(^*\) classified IAA into three types: type A, stenosis distal to the LSCA; type B, stenosis between the left common carotid artery and the LSCA; type C, stenosis between the left and common carotid arteries. Type A accounts for 40% of cases, type B for 55%, and type C for 5%.\(^5\) With no treatment, the mortality rate within the first month of life is 76%,\(^1\) and the average survival is less than 1 week.

We believe this is the first report of IAA with sponta-