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Translocation of the Aortic Arch with Norwood Procedure for Hypoplastic Left Heart Syndrome Variant with Circumflex Retroesophageal Aortic Arch

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Retroesophageal aortic arch, in which the aortic arch crosses the midline behind the esophagus to the contralateral side, is a rare form of vascular anomaly. The complete form may cause symptoms by compressing the esophagus or the trachea and need a surgical intervention. We report a rare case of a hypoplastic left heart syndrome variant with the left retroesophageal circumflex aortic arch in which the left aortic arch, retroesophageal circumflex aorta, and the right descending aorta with the aberrant right subclavian artery encircle the esophagus completely, thus causing central bronchial compression. Bilateral pulmonary artery banding and subsequent modified Norwood procedure with extensive mobilization and creation of the neo-aorta were performed. As a result of the successful translocation of the aorta, the airway compression was relieved. The patient underwent the second-stage operation and is doing well currently.

Key words: 1. Airway
2. Congenital heart disease
3. Hypoplastic left heart syndrome
4. Norwood procedures
5. Retroesophageal circumflex aortic arch

CASE REPORT

A 3.03-kg female neonate prenatally diagnosed with a hypoplastic left heart syndrome (HLHS) variant, which includes mitral atresia, hypoplastic aortic arch, and muscular ventricular septal defect without left ventricular forward flow, was delivered at 39 weeks of gestational age. Initially, a prostaglandin E-1 (PGE-1) infusion was begun immediately, and controlled ventilation was performed to prevent excessive pulmonary blood flow while waiting for an operation. The two-dimensional echocardiography revealed situs solitus, levocardia, and left aortic arch, which mainly originated from the left ventricle (LV) without a left ventricular outflow tract obstruction despite unrestrictive combined perimembranous and apical muscular ventricular septal defect (VSD) with aortic overriding. Mitral atresia with LV hypoplasia and a relatively normal-sized (6.04 mm, z-score: -0.8) ascending aorta with normal coronary arteries were also detected by echocardiography. Cardiac computed tomography (CT) revealed tubular hypoplasia of the aortic arch with a circumflex retroesophageal right descending aorta and an aberrant right subclavian artery, which encircled the central airway completely

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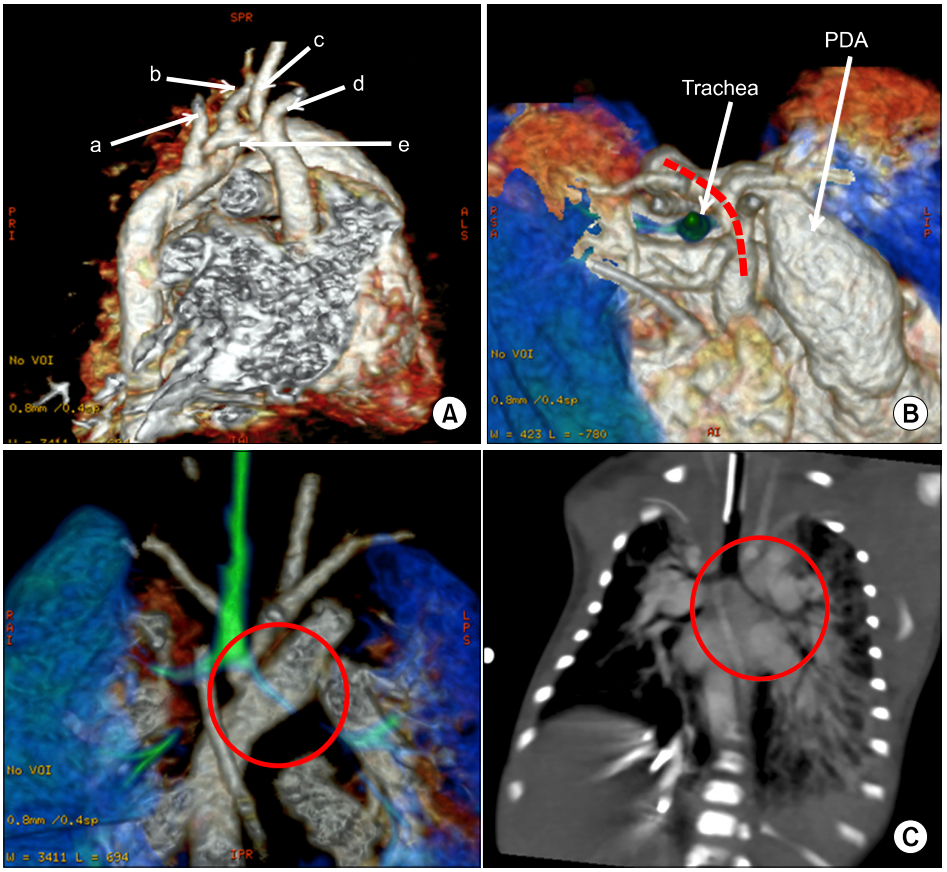

Fig. 1. Preoperative chest computed tomography findings. (A) Configuration of the aortic arch; (a) aberrant right subclavian artery, (b) left subclavian artery, (c) left common carotid artery, (d) right common carotid artery, and (e) hypoplastic arch. (B) Retroesophageal circumflex aortic arch (red dotted line) crosses the midline behind the trachea, causing airway compression. (C) Airway compression by the retroesophageal circumflex arch (red circle); the descending aorta crosses the midline behind the left main bronchus. PDA, patent ductus arteriosus.

and thereby led to a marked narrowing of the left main bronchus by mechanical compression (Fig. 1). Against these lesions, temporary bilateral pulmonary artery banding (PAB) was performed at 16 days of life. During bilateral PAB, fragments of a 3.5-mm Gore-tex graft were secured around the bilateral branch pulmonary arteries. The increase in the systemic blood pressure and the peripheral oxygen saturation under room air ventilation after PAB were 20 mmHg and 75% to 80%, respectively. On echocardiography, which was performed on postoperative day 2, the internal diameters of the right pulmonary artery and the left pulmonary artery were 2.5 mm and 2.2 mm, respectively. PGE-1 infusion was maintained until the Norwood procedure was performed. At 30 days of life, a subsequent modified Norwood operation was performed. The operation involved arch repair under selective cerebral perfusion, a 5-mm external ringed conduit from the right ventricle (RV) to the right pulmonary artery (PA) with the 'Dunk technique' [1], and atrial septectomy. Before arch reconstruction, the ascending aorta, aortic arch, aortic arch vessels, and descending aorta were extensively mobilized, particularly around the descending aorta. A single arterial cannula was inserted into the right common carotid artery via the 3.5-mm Gore-tex vascular graft. In the venous drainage, double venous cannulation via right atrial appendage and inferior vena cava was used. Considering the abnormality of the arch vessels (aberrant right subclavian artery), the arterial blood pressure during the selective cerebral perfusion was monitored via a right temporal artery catheterization. The schema of the operative procedures is depicted in Fig. 2. The cardiopulmonary bypass time, aorta clamping time, selective cerebral perfusion time, and the lowest rectal temperature were 237 minutes, 89 minutes, 37 minutes, and 24.4°C, respectively. On the intraoperative bronchoscopy, the left main bronchial stenosis was relieved. Delayed sternal closure was performed on postoperative day 2. The patient was extubated on postoperative day 5 and was discharged on postoperative day 16. A postoperative CT, which was carried out on postoperative day 8, showed the leftward translocation of

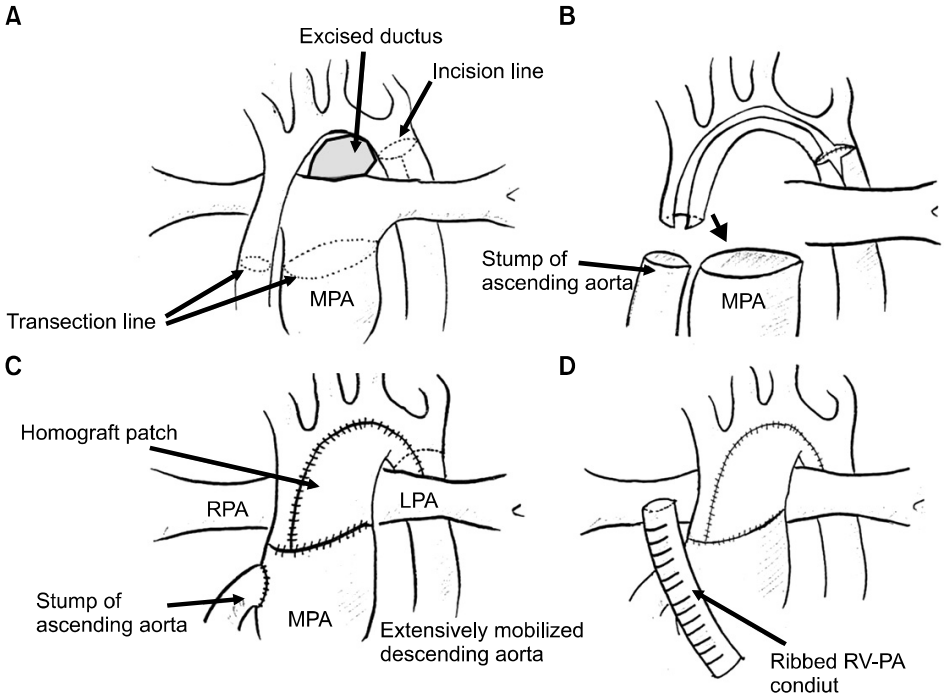

Fig. 2. Schema of the operation. (A) The left lateral side of the aortic arch is incised beginning at the divided aortic isthmus and extending proximally down to the transected ascending aorta. Further, the MPA was transected. (B) To obtain more space for the airway and the pulmonary vasculature, the repaired arch and the neo-aorta had to translocate leftward as much as possible. Therefore, (C) the ascending aorta was transected, and the proximal stump of the ascending aorta was anastomosed to the right lateral wall of the MPA. A pulmonary homograft patch was used for the completion of the arch reconstruction and the creation of the neo-aorta. (D) Then, a 5-mm external ringed conduit from the right ventricle (RV) to the right pulmonary artery (PA) with the "Dunk technique". MPA, main pulmonary artery; RPA, right pulmonary artery; LPA, left pulmonary artery.

the descending aorta that relieved the airway compression (Fig. 3). The patient underwent successful bidirectional cavo-pulmonary shunt at 6 months of age and is now waiting for the Fontan operation.

DISCUSSION

The vascular ring designates the abnormalities caused by a developmental failure of the paired aortic arches during embryonic life; these can present with the compression of the adjacent structures, including the central airway, pulmonary arteries, and the esophagus [2,3]. According to the vascular structures of the vascular ring, they have been classified into the following five distinct types: 1) double aortic arch, 2) right aortic arch with left ligamentum, 3) left aortic arch with aberrant right subclavian artery, 4) pulmonary artery sling, and 5) innominate artery compression [2,3]. Unlike these classifications, the retroesophageal circumflex aortic arch, in which the aortic arch crosses the midline behind the esophagus to the contralateral side, is a rare form. Furthermore, the condition of an HLHS variant combined with a vascular ring is extremely rare.

The retroesophageal circumflex aortic arch can cause several symptoms by compressing the esophagus and the central airway. Even if a concomitant intracardiac anomaly does not present, the treatment of the vascular anomaly can be challenging because excessive tension on the aorta caused by the surgical treatment can constrict the adjacent structures. Therefore, a compromise of the central airway is a major concern of the surgical treatment of the vascular anomaly. There is a report on the retroesophageal circumflex aortic arch, which was combined with the coarctation of aorta (CoA) and VSD [4]. The authors performed extensive dissection around the aorta, including the arch vessels for the re-

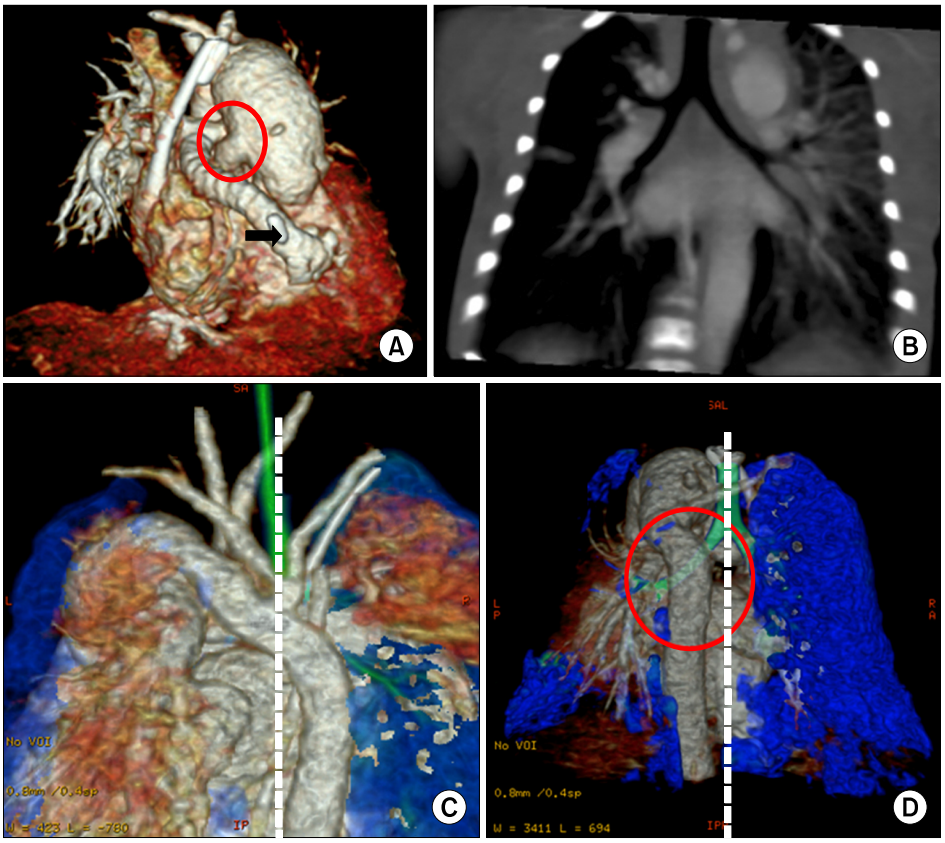

Fig. 3. Postoperative chest computed tomography findings and changes in the location of the descending aorta. (A) Transfer of the native aorta to the neo-aorta (red circle); ring-enforced right ventricle to the pulmonary artery conduit (black arrow). (B) Relief of airway compression. (C) Retroesophageal circumflex aortic arch crosses the midline, preoperatively. The white dotted line depicts the virtual line along the trachea. (D) After the operation, the descending aorta was located on the left side of the virtual line along the trachea.

pair of CoA and arch reconstruction by using the translocation of the descending aorta. In the presented case, however, the HLHS variant was combined with the retroesophageal circumflex aortic arch, and thus, we had to perform the Norwood procedure concomitantly. Therefore, we performed the modified Norwood procedure, including the translocation of the aorta combined with the full mobilization of the ascending aorta, aortic arch, arch vessels, and the descending aorta to relieve the airway compression.

To date, there have been three options for the Norwood procedure. The first option is the Norwood procedure with a Blalock-Taussig shunt or the RV to PA conduit. The second option is the 'rapid two-stage Norwood procedure,' which consists of temporary bilateral PAB and the maintenance of the ductus arteriosus by a ductal stent or PGE-1 infusion with a subsequent Norwood procedure. Temporary bilateral PAB has been performed for high-risk patients as an initial palliation of HLHS until the second-stage operation is undertaken [5,6]. The third option is a 'hybrid Norwood procedure' of bilateral PAB and ductal stenting. In the presented case, to minimize the perioperative risks of a Norwood operation by the stabilization of the pulmonary vasculature, we initially performed bilateral PAB with the maintenance of the PGE-1 infusion. With respect to the type of systemic-to-pulmonary shunt, as numerous previous studies have been reported, we selected the RV to right PA conduit, which was implanted with the 'Dunk technique' as the source of the pulmonary blood flow [1,5,7].

Because of the combined HLHS, the main presentations of our patient were congestive heart failure and excessive pulmonary blood flow rather than the typical symptoms of the vascular ring, which involve respiratory symptoms and dysphagia. Although the patient did not present airway symptoms, a markedly narrowed left main bronchus can cause significant airway problems, postoperatively. Therefore, efforts for the achievement of hemodynamic stability as well as the prevention of postoperative airway compression should be made during the operation. In this regard, to obtain adequate

space for the airway and to reconstruct the unobstructed systemic outflow tract, extensive mobilization of the descending aorta and arch reconstruction using techniques that facilitate the leftward translocation of the descending aorta were performed during the Norwood operation. As a result, successful aortic arch reconstruction and airway protection were simultaneously achieved. In addition, intraoperative bronchoscopy and perioperative CT scans are essential to evaluate the relationship of the airway and the surrounding vascular structures [8].

CONFLICT OF INTEREST

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

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