Korean J Thorac Cardiovasc Surg 2018;51:360-362 □ CASE REPORT □
ISSN: 2233-601X (Print) ISSN: 2093-6516 (Online) https://doi.org/10.5090/kjtcs.2018.51.5.360

A Rare Case of Tetralogy of Fallot Associated with Pulmonary Artery Sling

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Pulmonary artery sling is a rare congenital cardiac anomaly, in which the left pulmonary artery originates from the right pulmonary artery and courses leftward between the trachea and the esophagus. Tetralogy of Fallot associated with pulmonary artery sling is even rarer, and only a few cases have been reported in the literature. We present a case of tetralogy of Fallot associated with pulmonary artery sling that was repaired successfully.

Key words: 1. Tetralogy of Fallot
2. Vascular ring

Case report

A 6-month-old girl from another country was admitted to Seoul St. Mary's Hospital for the management of congenital heart disease. Echocardiography performed in her country had revealed tetralogy of Fallot (TOF) with an invisible left pulmonary artery (LPA). At admission to our hospital, her arterial oxygen saturation by pulse oximetry was around 85%, and stridor was heard on auscultation without respiratory symptoms. Echocardiography confirmed TOF and a pulmonary artery (PA) sling (Fig. 1A). The diameter of the pulmonary valve annulus was 5 mm (Z-score of −5.7) and there was a small patent ductus arteriosus (PDA) connected to the LPA. The diameter of the LPA was 4.5 mm and the diameter of the right pulmonary artery (RPA) was 5 mm. Computed tomography (CT) was performed to better delineate the anatomy of the PAs and to evaluate the airway (Fig. 1B). No tracheal stenosis associated with complete tracheal cartilage was found.

Surgery was performed under aorto-bicaval cardiopulmonary bypass. The main pulmonary artery (MPA) was hypoplastic and coursed rightward and posterior to the ascending aorta owing to the abnormal course of the LPA. The PDA was divided and the entire course of the LPA was carefully dissected to prevent injury to the trachea and the esophagus. Closure of the ventricular septal defect (VSD) and resection of the right ventricular outflow tract (RVOT) muscles were carried out through a right atrial incision. Then, the LPA was transected at its origin from the RPA and a cuff of the LPA was left on the RPA to prevent RPA stenosis and to shorten the LPA. The defect of the RPA was repaired primarily with a 6-0 polypropylene running suture. A short longitudinal incision was made along the RVOT, and the pulmonary valve was found to be bicuspid and dysplastic with a small opening. Transannular extension was carried out up to the distal MPA, and additional RVOT muscle resection was performed through the RVOT incision. The LPA was trimmed obliquely to

Received: April 9, 2018, Revised: May 14, 2018, Accepted: June 12, 2018, Published online: October 5, 2018

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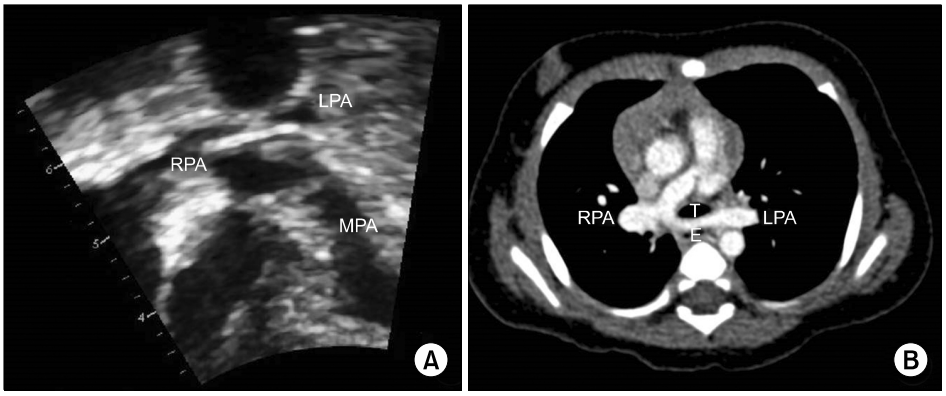

Fig. 1. (A) Subcostal coronal view of a preoperative echocardiogram showing the hypoplastic MPA and LPA originating from RPA. (B) Preoperative computed tomography showing the LPA originating from the RPA and coursing between the trachea and the esophagus. MPA, main pulmonary artery; LPA, left pulmonary artery; RPA, right pulmonary artery; T, trachea; E, esophagus.

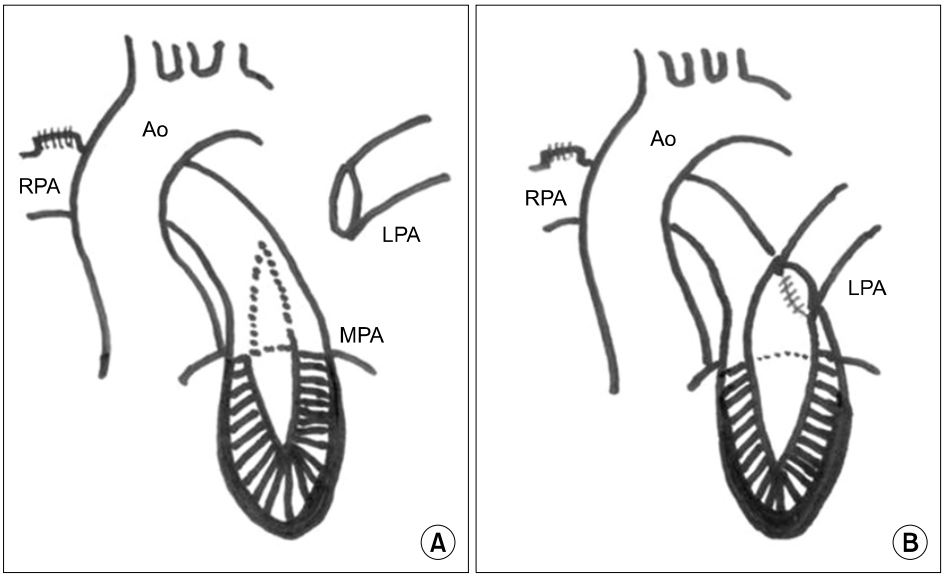

Fig. 2. Illustration of the operative procedure. (A) The transannular incision and the obliquely trimmed LPA. (B) The end-to-side anastomosis between the posterior half of the LPA and the left lateral aspect of the MPA. Ao, aorta; MPA, main pulmonary artery; RPA, right pulmonary artery; LPA, left pulmonary artery.

obtain a natural geometry and to prevent kinking after anastomosis to the MPA (Fig. 2A), and the posterior half of the LPA was anastomosed to the left lateral aspect of the MPA in an end-to-side fashion using a 6-0 polypropylene running suture (Fig. 2B). The transannular incision and the anterior aspect of the LPA were covered with a bovine pericardial patch secured with a 6-0 polypropylene running suture. The cardiopulmonary bypass time was 172 minutes and the aortic cross-clamp time was 114 minutes.

Postoperative echocardiography showed no residual VSD, no RVOT obstruction, severe pulmonary regurgitation, good flow in the branches of the PA, and good ventricular function. Postoperative CT also revealed well-reconstructed PAs (Fig. 3). The patient's recovery was uneventful and she was discharged on the ninth postoperative day.

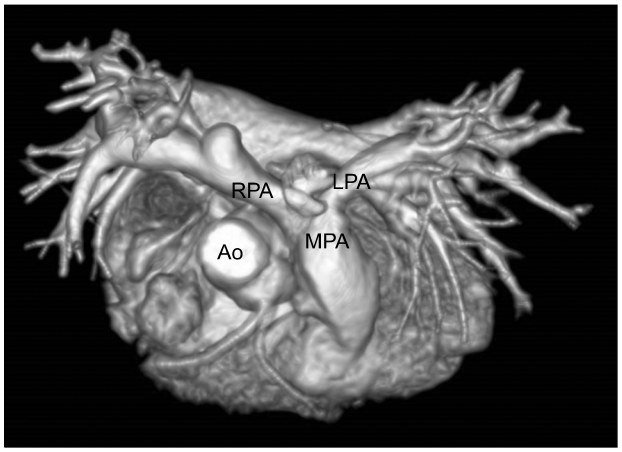

Fig. 3. Enhanced postoperative computed tomography showing the well-reconstructed pulmonary arteries. Ao, aorta; MPA, main pulmonary artery; RPA, right pulmonary artery; LPA, left pulmonary artery.

Discussion

PA sling is a rare congenital cardiac anomaly in which an anomalous LPA arises from the RPA and courses between the trachea and the esophagus. PA sling can be associated with various cardiovascular anomalies, including VSD, atrial septal defect, and PDA. However, PA sling associated with TOF is very rare. Gikonyo et al. [1] found that among 130 cases of PA sling, only 3 (2.3%) were associated with TOF, and concomitant repair of TOF associated with PA sling has rarely been reported [2-6].

The clinical manifestations of PA sling vary and depend on the severity of airway compromise. Our patient had stridor without other respiratory symptoms, and there was no tracheal stenosis. When associated tracheal stenosis is present, slide tracheoplasty can be a good surgical option [6]. Goldstein et al. [7] reported that the incidence of PA stenosis was relatively common, occurring in 79% of patients after the repair of PA sling or anomalous origin of 1 PA from the ascending aorta. However, Yong et al. [5] reported an incidence of PA stenosis of only 4.8% after PA sling repair in 21 patients. Similar results regarding the patency of the LPA were presented by Backer et al. [6]. Yong et al. [5] and Backer et al. [6] credited cardiopulmonary bypass and the implantation of the LPA to the proximal MPA as the surgical techniques that have lowered the incidence of LPA stenosis. In this case, we left a cuff of the LPA on the RPA to shorten the LPA, because the LPA is often redundant. In addition, we trimmed the LPA obliquely to obtain a natural geometry and to prevent kinking after it was anastomosed to the MPA. We chose to anastomose the posterior wall of the LPA to the MPA incision and to cover the anterior wall with a patch for 2 reasons. First, the diameter of the LPA was only 4.5 mm, and we expected that LPA stenosis would occur if the LPA was implanted to the separate MPA opening in an end-to-side manner. Secondly, the hypoplastic MPA was not ideally suited for re-implantation of the LPA at the separate MPA opening, and this drove our decision to make the MPA incision.

In conclusion, we successfully repaired a rare combination of TOF and PA sling. Although concomitant repair of these defects is technically straightforward, care should be taken to prevent post-repair LPA stenosis.

Conflict of interest

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

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