

# Left Hemitruncus Treated Along with Ventricular Septal Defect in a Neonate

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Hemitruncus arteriosus is a rare cardiovascular malformation in which one of the pulmonary arteries anomalously originates from the aorta. Left hemitruncus arteriosus, defined as the origination of the left pulmonary artery from the aorta, is less common than right hemitruncus arteriosus. In this study, we report the case of a neonate diagnosed with left hemitruncus arteriosus, ventricular septal defect, and atrial septal defect who underwent successful surgical treatment.

**Keywords:** Truncus arteriosus, Heart septal defects, Ventricular heart septal defects

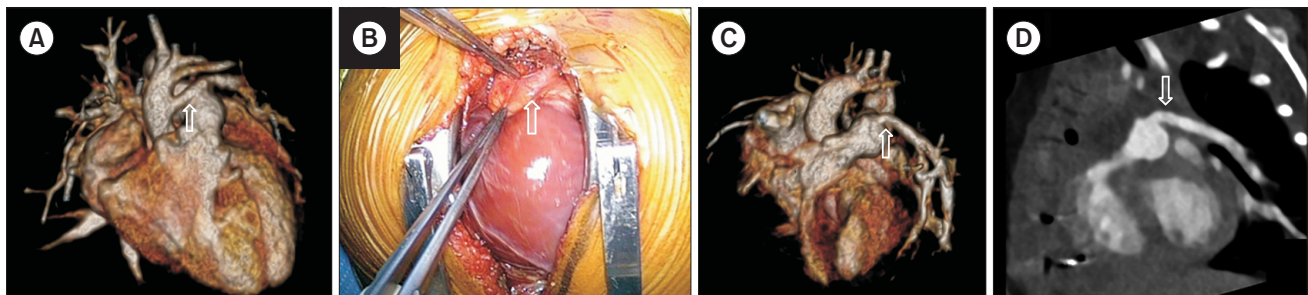
## Case report

In a female neonate, prenatal ultrasonography showed a parallel course of 2 great arteries and other malformations, including left first toe polydactyly and hemivertebra. The neonate was delivered at 38 weeks and weighed 3.6 kg.

The patient was admitted to the neonatal intensive care unit at birth due to respiratory distress. Chest radiography revealed segmentation anomalies in the vertebral bodies, from T10 to the sacrum. Echocardiography showed atrial septal defect, ventricular septal defect (VSD), and suspected patent ductus arteriosus (PDA) between the right pulmonary artery and the descending aorta. A 3-dimensional

reconstruction computed tomography (CT) scan was performed on postnatal day 7 to evaluate the great vessels, and this scan confirmed the anomalous origin of the left pulmonary artery (LPA) from the ascending aorta (Fig. 1A).

The patient underwent surgical repair on day 18 after birth. The operation was carried out through a median sternotomy. After pericardial tenting, careful dissection was performed to encircle the aorta, LPA, and PDA (Fig. 1B). After routine ascending aortic bicaval cannulation immediately prior to the initiation of cardiopulmonary bypass, PDA ligation and LPA division were performed, with the aortic end repaired with 6-0 polypropylene suture and the other end ligated with black silk.

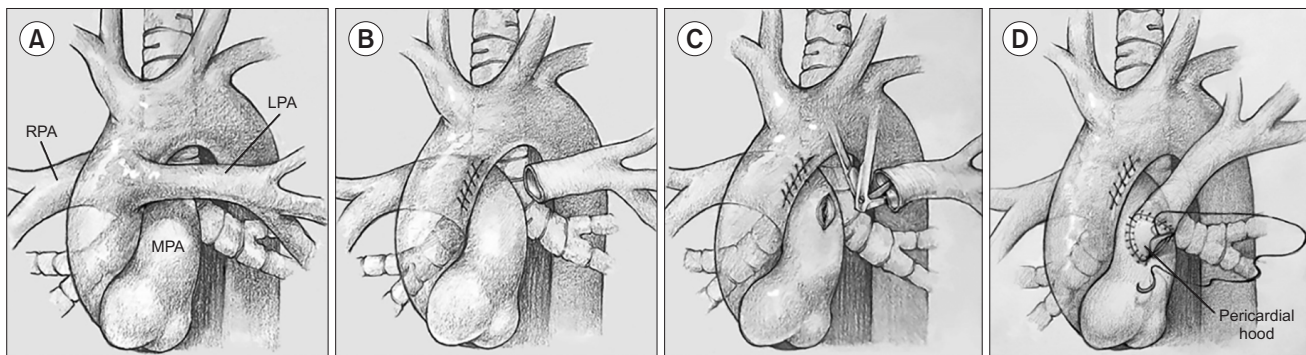


**Fig. 1.** (A) Preoperative computed tomography image and (B) operative findings showing the left pulmonary artery originating from the aorta. Postoperative computed tomography image showing (C) the reimplemented left pulmonary artery and (D) no anastomotic site stenosis. The white arrow indicates the left pulmonary artery.

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**Fig. 2.** Reimplantation technique for the left pulmonary artery. (A) Abnormal anatomy of left hemitruncus arteriosus: anomalous origin of the left pulmonary artery from the ascending aorta. (B) The left pulmonary artery was divided, and the aortic wall was repaired with a 6-0 polypropylene running suture. (C) The main pulmonary artery and the posterior wall of the left pulmonary artery were excised. (D) The left pulmonary artery was re-implanted into the main pulmonary artery using an autologous pericardial hood patch. LPA, left pulmonary artery; MPA, main pulmonary artery; RPA, right pulmonary artery.

After aortic cross-clamping, VSD closure with an autologous pericardial patch and direct atrial septal defect closure were performed via right atriotomy. The divided LPA was then re-implanted into the main pulmonary artery with an autologous pericardial hood patch to avoid anastomotic stenosis (Fig. 2).

The operation was finished with sternal closure. However, the chest was opened due to low cardiac output at postoperative 6 hours, and delayed sternal closure was performed on postoperative day 3. Follow-up echocardiography 2 weeks after the operation showed a patent LPA without anastomotic stenosis. Follow-up CT scanning also verified the presence of normal vascularization without anastomotic stenosis (Fig. 1C, D). The patient recovered smoothly but had to remain in the hospital for 25 additional days after surgery for treatment of other anomalies. At the follow-up visit 10 months later, she was doing well, and her echocardiogram showed no evidence of stenosis at the LPA re-implantation site. However, to carefully evaluate LPA growth, we plan to perform a lung perfusion scan, CT scan, and catheter examination to quantify the aortopulmonary collateral vessels in the near future.

The patient's parents provided written informed consent for the publication of the patient's clinical details and images.

## Discussion

Hemitruncus arteriosus is a rare congenital cardiovascular malformation in which 1 pulmonary artery anomalously originates from the aorta. Left hemitruncus is less common than right hemitruncus [1].

The anomaly was first described by Fraentzel [2] in 1868. Since then, several case reports and series of hemitruncus have been published, but reports of left hemitruncus are scarce. Early diagnosis is essential for prompt surgical repair to prevent death following congestive heart failure and the development of irreversible pulmonary vascular obstructive disease [3]. Nathan et al. [4] reported that early hemitruncus repair resulted in excellent hemodynamic and anatomic outcomes.

The anatomic correction of hemitruncus by translocating the anomalous pulmonary artery to the pulmonary trunk was first described by Kirkpatrick et al. [5] in 1967. Since then, direct re-implantation has become the treatment of choice. We also attempted direct reimplantation; however, due to the lack of tissue, we used an autologous pericardial patch hood to prevent anastomotic site stenosis.

With regard to the timing of surgery, we believe that surgery during the neonatal period is ideal for favorable results. If possible, we recommend performing the operation within 2 weeks of birth.

After a CT examination on postnatal day 7 (Fig. 1A), we initially thought that re-implantation of the LPA would be simple. However, by more than 2 weeks after birth, decreased pulmonary vascular resistance together with the increased PDA and VSD shunt had led to enlargement of the left and main pulmonary arteries (Fig. 1B); thus, dissecting and encircling the aorta, PDA, and LPA before starting cardiopulmonary bypass were difficult. Thus, if this rare disease can be detected early, it is important not to delay surgery.

With regard to postoperative myocardial swelling resulting in low cardiac output syndrome, staged repair with

LPA reimplantation and external pulmonary artery banding is a possible option to avoid needing to open the sternum in the neonatal period.

Overall, early preoperative detection of this rare congenital cardiovascular malformation and postoperative detection of restenosis requiring re-intervention are important. Three-dimensional reconstruction CT images can be useful diagnostic tools [6].

## Conflict of interest

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

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## References

1. Kutsche LM, van Mierop LH. *Anomalous origin of a pulmonary artery from the ascending aorta: associated anomalies and pathogenesis*. Am J Cardiol 1988;61:850-6.
2. Fraentzel O. *Ein fall von abnormer communication der aorta mit der arteria pulmonalis [A case of abnormal communication of the aorta with the pulmonary artery]*. Virchows Arch Pathol Anat Physiol Klin Med 1868;43:420-6.
3. Penkoske PA, Castaneda AR, Fyler DC, van Praagh R. *Origin of pulmonary artery branch from ascending aorta: primary surgical repair in infancy*. J Thorac Cardiovasc Surg 1983;85:537-45.
4. Nathan M, Rimmer D, Piercey G, et al. *Early repair of hemitruncus: excellent early and late outcomes*. J Thorac Cardiovasc Surg 2007; 133:1329-35.
5. Kirkpatrick SE, Girod DA, King H. *Aortic origin of the right pulmonary artery: surgical repair without a graft*. Circulation 1967;36:777-82.
6. Kim TH, Kim YM, Suh CH, et al. *Helical CT angiography and three-dimensional reconstruction of total anomalous pulmonary venous connections in neonates and infants*. AJR Am J Roentgenol 2000; 175:1381-6.