Absent Pulmonary Valve with L-Transposition of Great Arteries: A Case report

Wataru Sakai[1], Hidetsugu Asai[2], Takafumi Oyasu[1], Yosuke Arai[2], Noriyoshi Ebuoka[2], and Jun Oba[2]

[1]Hokkaido Medical Center for Child Health and Rehabilitation
[2]Hokkaido Daigaku

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Abstract

Absent pulmonary valve syndrome and L-transposition of the great arteries are rare congenital anomalies. To our knowledge, patients with both have not been previously reported. We present a case of surgical success in the patient with absent pulmonary valve, L-transposition of the great arteries, dextrocardia, double outlet left ventricle, hypoplastic anatomical right ventricle, valvular aortic stenosis and tracheomalacia. We performed anatomical left ventricle to pulmonary artery shunt using an 8-mm expanded polytetrafluoroethylene bicuspid-valved conduit, pulmonary artery plication, and tracheostomy. The patient was discharged in a stable condition while waiting for the Glenn procedure until getting her parent's permission.

Introduction

Absent pulmonary valve syndrome (APVS) is a rare congenital cardiac malformation that has absent, dysplastic, or rudimentary pulmonary valve leaflets. The characteristic feature of APVS is dilation of the pulmonary artery, which leads to compression of the tracheobronchial tree and respiratory distress. Most cases of APVS are associated with tetralogy of Fallot (ToF). L-transposition of the great arteries (L-TGA) is also a rare congenital anomaly that is associated with ventricular septal defect, pulmonary stenosis, and abnormalities of the tricuspid valve. There are no previous reports on patients with both APVS and L-TGA.

Case presentation

A female infant with a birth weight of 2,231 g was delivered vaginally at a gestational age of 37 weeks and 6 days. A fetal echocardiogram revealed a heart abnormality with severe aortic and pulmonary stenosis. Tracheobronchial tree and lung abnormality were not reported in fetal diagnosis. After birth, she was tachydyspneic and cyanotic with a loud "to-and-fro" murmur over the left sternal border. Hepatomegaly was observed. Chest radiography revealed a cardiothoracic ratio of 80%. Echocardiography showed the absence of pulmonary valve with pulmonary insufficiency and stenosis, L-TGA, dextrocardia, double outlet left ventricle, hypoplastic anatomical right ventricle, valvular aortic stenosis, atrial septal defect, and patent ductus arteriosus with a bidirectional shunt. Computed tomography (CT) showed a severe hypertrophic anatomical left ventricle connected to the left displaced and dilated main pulmonary artery, with dilation of both branches (Figure 1). The compression of the left main bronchus by the dilated pulmonary artery was diagnosed as tracheomalacia (Figure 2A). The left lung was compressed by the enlarged heart and was suspected to be hypoplastic. It had a less vascular shadow than the right lung.

A tracheal tube was placed immediately after birth because the spontaneous breathing effort was severe, and left lung sounds could not be heard on auscultation. A Ventricular arrhythmia requiring adrenalin infusion immediately after birth indicated severe heart failure. Six hours after delivery, the patent ductus arteriosus

was ligated because of increased pulmonary insufficiency and exacerbation of heart failure. On postnatal day 5, percutaneous balloon aortic valvuloplasty was performed for critical aortic valve stenosis. This reduced the peak pressure gradient from 25 mmHg to 20 mmHg. At 4 months age, anatomical left ventricle to pulmonary artery shunt using an 8-mm expanded polytetrafluoroethylene bicuspid-valved conduit with bulging sinuses (Stretch Vascular Graft, W.L. Gore & Associates, Inc., Flagstaff, AZ, USA) and pulmonary artery plication were performed. Although pulmonary insufficiency could be controlled completely with the operation, tracheomalacia of the left main bronchus remained (Figure 2B), and a high positive ventilation setting was still required after the operation. Tracheostomy was performed at the age of five months. There was poor postoperative weight gain and repeated necrotizing enterocolitis. Cartelization showed that her Qp:Qs was 1.6:1, and aortopulmonary collaterals were observed from the right intrathoracic artery. Embolization of the aortopulmonary collateral was performed to decrease the pulmonary blood flow at the age of nine months. Conduit was clipped at the age of ten months to decrease pulmonary blood flow. There was good weight gain, and positive ventilation was not required after the surgery, although tracheomalacia of the left main bronchus persisted on CT. Despite recurrent respiratory tract infections, the patient was discharged at the age of 14 months in a stable condition while waiting for the Glenn procedure until getting her parent's permission.

Discussion

APVS includes rudimentary, dysplastic, or absent pulmonary valve leaflets, dilated main pulmonary artery with or without dilation of its branches, to-and-fro flow at the site of the absent pulmonary valve, and systolic pressure gradient across the narrowed pulmonary valve[1]. APVS is a rare variant of ToF, which accounts for approximately 3–6% of patients with ToF [2].

L-TGA is also rare, with a published incidence ranging from 0.02 to 0.07 per 1000 births [3]. The most common anatomic cardiac abnormalities with L-TGA include ventricular septal defect, pulmonary stenosis, and abnormalities of the tricuspid valve [3].

A dilated pulmonary artery with APVS can compress the tracheobronchial tree and the esophagus, which is a significant indicator of poor postnatal prognosis [4]. One study showed that complete repair with a valved conduit and reduction pulmonary arterioplasty can help improve tracheomalacia [5]. Although most patients might require pulmonary arterioplasty, it is still controversial whether a pulmonary valve or a valved/unvalved conduit should be inserted and how the degree of size reduction of the pulmonary artery should be determined. The standard medical therapy for tracheomalacia is positive-pressure ventilation with or without a tracheostomy[5]. We performed anatomical left ventricle to pulmonary artery shunt with a bicuspid conduit made of polytetrafluoroethylene, which has excellent biocompatibility and low antigenicity[6]. Our patient required tracheostomy and positive pressure ventilation from birth till the age of one year. The anatomical left ventricle instead of the right ejected blood to the pulmonary artery directly because of cTGA. This, along with aortic valve stenosis, could lead to pulmonary over circulation and dilate the pulmonary artery easily. Therefore, the complex of APVS, cTGA, and aortic stenosis might cause pulmonary artery dilation and tracheomalacia more easily than APVS with TOF.

Conclusion

To our knowledge, this is the first reported case of a patient with APVS and L-TGA who underwent anatomical left ventricle to pulmonary artery shunt, pulmonary artery plication, and tracheostomy. The patient was discharged in a stable condition while waiting for the Glenn procedure.

Acknowledgments: None

Abbreviations

APVS: Absent Pulmonary Valve Syndrome

TOF: tetralogy of Fallot

L-TGA: L-transposition of great arteries

CT: Computed tomography

Disclosures

Ethics approval and consent to participate: The Ethics Committee at Hokkaido Medical Center for Child Health and Rehabilitation approved this study.

Conflict of Interest Disclosure: None

Availability of data and material: Not applicable.

Author contribution:

Wataru Sakai and Hidetsugu Asai: Concept, Interpretation, Drafting article, Critical revision of article, Approval of article

Takafumi Oyasu, Yosuke Arai, Noriyoshi Ebuoka and Junichi Oba: Interpretation, Drafting article, Approval of article

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Figure 1. Preoperative CT. The left displaced and dilated main pulmonary artery with dilation of both branches.

aLV, anatomical left ventricle; Ao, aorta; MPA, main pulmonary artery; RPA, right pulmonary artery; LV, left pulmonary artery.

Figure 2. The arrow (-) shows the compression of the tracheobronchial tree by the dilated pulmonary artery at the same level of left bronchial stenosis (A) Preoperative and (B) postoperative CT at the same level of left bronchial stenosis. The arrow (-) shows the left main bronchus that is compressed by the dilated pulmonary artery. The arrowhead (¿[?]) shows anatomical left ventricle to pulmonary artery shunt.

MPA, main pulmonary artery; RPA, right pulmonary artery; LPB, left pulmonary bronchus; conduit, anatomical left ventricle to pulmonary artery shunt

Figures: 2