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Case Reports

Surgical management of ductal origin of the right pulmonary artery in an infant

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ABSTRACT

This clinical case report describes a 1.5-month-old infant with ductal origin of the right pulmonary artery. The surgical management included reconstruction of the right ventricular outflow tract and reimplantation of the right pulmonary artery using a 6-mm ringed expanded polytetrafluoroethylene (ePTFE) graft.

Keywords: Congenital heart defect, ductal origin of the pulmonary artery, pulmonary hypertension

Introduction

We present the case of an infant initially diagnosed on transthoracic echocardiography as having an absent right pulmonary artery, which was subsequently confirmed to represent ductal origin of the right pulmonary artery. This rare congenital anomaly is associated with a high risk of pulmonary hypertension and right ventricular overload if left untreated. Early recognition and timely revascularization of the affected pulmonary artery branch are crucial to prevent irreversible pulmonary vascular changes and adverse clinical outcomes.

Case Report

The patient was a 1.5-month-old female weighing 5.6 kg who presented with dyspnea. Transthoracic echocardiography (TTE) performed one month prior had demonstrated absence of the right pulmonary artery (RPA). On admission, the patient was in good general condition. Physical examination revealed no cardiac murmurs. Vital signs were as follows: blood pressure 100/65 mmHg, heart rate 138 bpm, and respiratory rate 35 breaths per minute. Oxygen saturation (SpO_2) was 94% on room air.

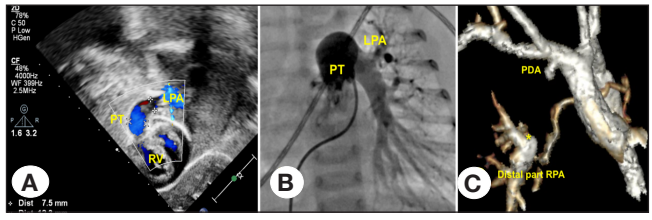

Figure 1: A) TTE showing absence of the right pulmonary artery. B) Cardiac catheterization demonstrating the PT with the left pulmonary artery. C) MSCT with 3D-reconstruction. LPA indicates left pulmonary artery; PDA, patent ductus arteriosus; PT, pulmonary trunk; RPA, right pulmonary artery; RV, right ventricle.

TTE demonstrated absence of antegrade flow into the RPA. The pulmonary trunk measured 12.3 mm (z-score +1.71), and the left pulmonary artery (LPA) measured 7.5 mm (z-score +2.28). Estimated right ventricular systolic pressure (RVSP) was 100 mmHg. No right pulmonary venous return was observed. Additional findings included right ventricular outflow tract (RVOT) muscular hypertrophy and a 6-mm atrial septal defect (ASD) with right-to-left shunting (Figure 1A).

Cardiac catheterization revealed identical pressures in the pulmonary trunk and the LPA: 124/84 mmHg (mean 98

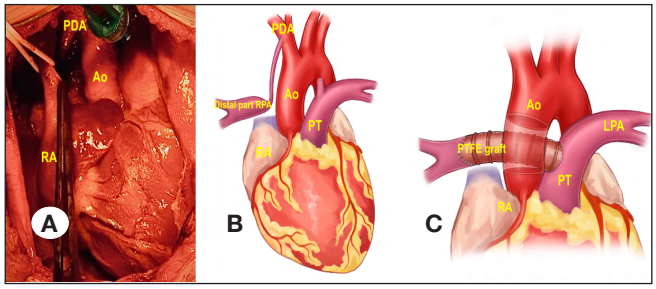

Figure 2: The operating view (A, B) and scheme of surgery (C). Ao indicates ascendens aorta; LPA indicates left pulmonary artery; PDA, patent ductus arteriosus; PT, pulmonary trunk; PTFE graft, polytetrafluoroethylene; RA, right atrium; RPA, right pulmonary artery

mmHg). The right ventricular pressure was 120/0 mmHg. Retrograde contrast injection visualized the distal RPA via collateral filling from the right pulmonary veins (Figure 1B).

Multislice spiral computed tomography (MSCT) with three-dimensional reconstruction showed the pulmonary trunk continuing solely as the LPA. A small segment of a right-sided patent ductus arteriosus (PDA) was identified near the proximal brachiocephalic trunk, and the distal RPA was also visualized (Figure 1C).

Given the imaging findings and the significant risk of pulmonary hypertension developing in the single functional lung, endovascular revascularization was attempted but was unsuccessful. Consequently, the patient was referred for surgical repair of the RPA

Via a median sternotomy, bicaval and ascending aortic cannulations were performed, cardiopulmonary bypass (CPB) was initiated, and a left ventricular vent was placed.

The ligamentum arteriosum, extending between the brachiocephalic trunk and the confluence of the distal RPA, was dissected and resected (Figure 2). To reconstruct the RPA, a 6-mm ringed polytetrafluoroethylene (PTFE) graft was anastomosed between the pulmonary trunk and the distal RPA and positioned in an anatomical course beneath the aortic arch.

Subsequently, cardioplegic arrest was induced. The RVOT was incised, and the hypertrophic muscle bundles were resected. The RVOT was reconstructed using an autologous pericardial patch. The right atrium was opened, and the ASD was closed with a Dacron patch incorporating a 3-mm fenestration.

After successful weaning from CPB, the sternum was closed, and the patient was transferred to the intensive care unit.

Early postoperative TTE demonstrated a significant reduction in right ventricular systolic pressure (RVSP) to 40–45 mmHg, along with a decrease in left ventricular end-diastolic volume. The shunt across the interatrial fenestration had converted to a left-to-right direction.

The postoperative course was complicated by bilateral pneumonia, which progressed to septic shock and necessitated a 21-day stay in the intensive care unit. The patient was discharged on postoperative day 43 in satisfactory condition on a moderate-dose sildenafil regimen.

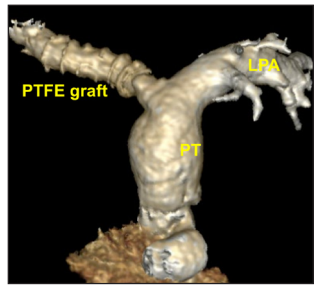

Figure 3: A 3D-MSCT performed five-months after surgery, front view. LPA indicates left pulmonary artery; PT, pulmonary trunk; PTFE graft, polytetrafluoroethylene.

At the 5-month follow-up, the patient was in good clinical condition on an unchanged medication regimen. Follow-up TTE revealed a persistent RVSP of 42 mmHg with a subnormal right ventricular volume. Hypertrophy of the interventricular septum and right ventricular walls persisted. A left-to-right shunt through the interatrial fenestration remained present. Subsequent CT angiography confirmed the graft patency and satisfactory function (Figure 3).

Discussion

Ductal origin of a pulmonary artery is a rare anomaly, with a reported incidence of approximately 0.1% (1). There are the following types: 1) PA origin from the ascendingaorta; 2) PA origin from the aortic arch; 3) PA origin from PDA; 4) PA origin from the descending aorta (2).

There is no universally accepted management strategy for patients with this anomaly. Approximately 100–120 pediatric cases detailing management approaches have been published. The two primary strategies are endovascular revascularization of the ductus arteriosus or surgical repair (3).

Endovascular intervention, while less invasive, carries a high risk of inducing unilateral pulmonary hypertension and is associated with a substantial re-intervention rate, reported to be as high as 68%.

When endovascular treatment is not feasible, surgical options include direct anastomosis, a systemic-to-pulmonary shunt, or prosthetic branch pulmonary artery grafting. A systemic-to-pulmonary shunt is associated with risks of unilateral pulmonary hypertension, shunt thrombosis, and coronary steal syndrome. Direct anastomosis between the distal pulmonary artery and the pulmonary trunk is often preferred but may be precluded by a significant distance between these structures (4). In such cases, interposition grafting remains the only viable option, although its major drawback is the need for reoperation due to patient growth (5). Alternatives to synthetic grafts include techniques utilizing autologous pericardium, xenomaterials, or homografts, and the optimal graft positioning (anatomical vs non-anatomical) remains a matter of debate.

In the present case, initial endovascular revascularization of the PDA was unsuccessful. A systemic-to-pulmonary shunt was contraindicated due to pre-existing supra-systemic pressures in the right heart chambers, which posed a significant risk of further exacerbating pulmonary hypertension in the contralateral lung. Consequently, we performed surgical reconstruction with a ringed PTFE graft,

placed in an anatomical position beneath the aortic arch to minimize the risk of graft compression and kinking.

The main limitation of this approach is the anticipated need for reoperation due to somatic growth. At the six-month follow-up, the patient remained in good clinical condition on sildenafil therapy, with an RVSP of approximately 45 mmHg. Nevertheless, long-term surveillance with periodic echocardiography and CT angiography is essential to monitor graft function and hemodynamics as the patient grows into adolescence and adulthood.

Conclusion

The optimal treatment strategy for ductal origin of a pulmonary artery remains a subject of debate. Available clinical reports describe a spectrum of approaches, ranging from conservative management with targeted pulmonary hypertension therapy to early surgical revascularization. The timing of intervention is of paramount importance, as delays of even several weeks after birth may allow the development of advanced pulmonary hypertension and heart failure, significantly complicating subsequent management.

This case underscores the importance of actively considering ductal origin of a pulmonary artery in infants with an “absent” pulmonary artery branch on echocardiography and severe pulmonary hypertension. Accumulation and careful analysis of such clinical cases will improve our understanding of this rare anomaly and help refine surgical techniques and postoperative management strategies for patients with ductal origin of the pulmonary artery.

Contribution of the authors

Study conception and design: OE, RK, FC; data collection: PE, ZSH; analysis and interpretation of results: PE, ZSH; draft manuscript preparation: ZSH, OE. All authors reviewed the results and approved the final version of the article.

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Conflict of interest

The authors declare that there is no conflict of interest.

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