Heart Surgery Forum 2024; 27(6): E568–E571
doi: 10.59958/hsf.7017

Case Report

Anomalous Origin of the Right Pulmonary Artery from the Ascending Aorta in a 10-Day-Old Boy: A Case Report

Lin Luo[1], Yulan Luo[1], Qi An[2], Mei Feng[1,*]

[1]West China Hospital Critical Care Medicine Department, Sichuan University/West China School of Nursing, Sichuan University, 610041 Chengdu, Sichuan, China
[2]Department of Cardiac Surgery, West China Hospital, Sichuan University, 610041 Chengdu, Sichuan, China
*Correspondence: 15328078733@163.com (Mei Feng)

Submitted: 19 November 2023 Revised: 28 December 2023 Accepted: 3 January 2024 Published: 6 June 2024

Abstract

Anomalous origin of the right pulmonary artery from the ascending aorta is a rare anomaly, comprising approximately 0.1% of all congenital heart diseases. Patients suffered congestive heart failure in infancy, and some patients will experience progressive pulmonary vascular disease without surgical repair. In patients of this disease, early surgical intervention is generally advised and has demonstrated a high level of safety and efficacy, yielding excellent outcomes. We report a unique case involving a 10-day-old boy, characterized by the rare anomalous origination of the right pulmonary artery from the ascending aorta. This case is further complicated by the presence of a patent ductus arteriosus (PDA) and a right descending aorta. The ligation of the PDA and reimplantation of the right pulmonary artery (RPA) were successfully performed. The patient exhibited a favorable recovery trajectory postoperatively.

Keywords

anomalous origin of the RPA from the ascending aorta; patent ductus arteriosus; reimplantation

Introduction

Anomalous origin of the pulmonary artery from the ascending aorta (AAO), also known as hemitruncus arteriosus, represents a particularly rare congenital heart anomaly, comprising approximately 0.1% of all congenital heart diseases [1]. Anomalies of the right pulmonary artery (RPA) from AAO are observed to be six times more prevalent than those of the left pulmonary artery (LPA). 60% of hemitruncus arteriosus cases present as an isolated condition. The most frequently observed accompanying anomaly is patent ductus arteriosus (PDA), but other coexisting abnormalities such as Tetralogy of Fallot, coarctation of the aorta, and atrial septal defect have also been reported [2,3].

In patients of hemitruncus arteriosus, the significant left-to-right shunt lead to a scenario where normal lung receives the entirety volume of the pulmonary circulation, while the other abnormal lung is perfused by high-speed and fully oxygenated blood from the AAO at a higher systemic pressure. This condition not only precipitates pressure and/or volume overload in the pulmonary circuit, but also initiates neurovascular reflexes and stimulate humoral vasoactive mediators. This condition accelerate progression of vascular disease in the lung circulation. Manifesting symptoms early in infancy, hemitruncus arteriosus commonly presents with recurrent respiratory tract infections, pulmonary hypertension and respiratory distress. Surgical repair is recommended as soon as possible post-diagnosis [4,5].

Case Presentation

A 10-day-old boy was admitted to our emergency department because of shortness of breath after birth. The patient has no familial history of congenital heart disease. The patient's Apgar scores were recorded at 6 and 5 points at 1 minute and 5 minutes post-delivery, respectively. His height measured 46.2 cm, falling within the 10th percentile, while his head circumference was 34 cm, placing it between the 70th and 90th percentiles. Upon examination, the patient exhibited a blood pressure reading of 72/40 mm Hg, accompanied by a heart rate of 152 beats per minute, with no detectable murmurs. The respiratory rate was noted at 38 breaths per minute, and oxygen saturation was measured at 96% in room air. Additionally, the chest X-ray revealed bilateral pulmonary congestion along with cardiomegaly. Transthoracic echocardiography (TTE) revealed that a mild dilated main pulmonary artery (MPA) gives rise the left pulmonary artery (LPA), a PDA and RPA arise from the AAO, there was no evidence indicating stenosis in the branch arteries (Fig. 1A,B). A contrast-enhanced and computed tomographic angiography (CTA) confirmed the diagnosis and revealed the anomalous origin of the RPA and a PDA arising from the AAO, and a large left branch artery arising from the MPA to supply the left lung blood with right-side aortic arch (Fig. 2A–C). The patient exhibited no additional intracardiac or extracardiac malformations.

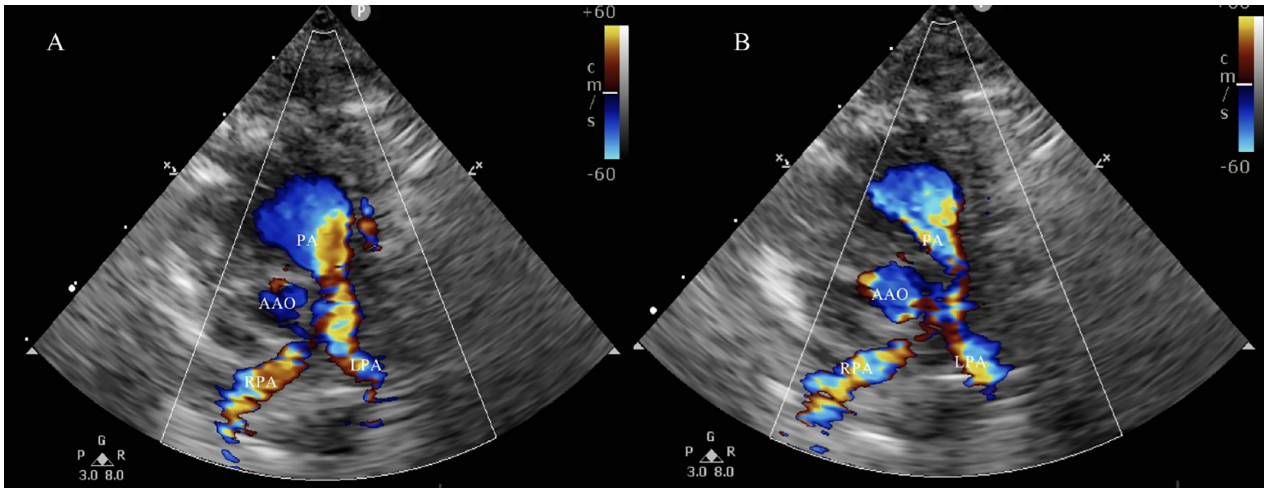

Fig. 1. Echocardiography image. (A,B) Transesophageal echocardiography (TEE) revealed mild dilated MPA gives rise the LPA, a PDA and RPA arise from the AAO. MPA, main pulmonary artery; LPA, left pulmonary artery; RPA, right pulmonary artery; AAO, ascending aorta; PDA, patent ductus arteriosus.

We performed a surgical correction for this patient, after ligation of PDA, the RPA was adequately mobilized and freed up to the point of pulmonary bifurcation, ensuring sufficient length of the RPA was obtained for the procedure, then the RPA is disconnected from AAO.

An opening was created on the right side of the MPA, with a diameter matching that of the RPA orifice. Subsequently, the distance between the RPA and the right side of the MPA was meticulously measured, if the RPA is not subjected to significant tension, direct implantation the RPA to the right side of the MPA with patch augmentation of the defect in the ascending aorta with the cardiopulmonary bypass was acceptable. Subsequent postoperative evaluations, including TTE and computed CTA, confirmed a widely patent right RPA (right pulmonary artery)-MPA (main pulmonary artery) connection. Additionally, a postoperative chest-X-ray showed no significant abnormalities in the lung fields. The patient's recovery was uneventful and discharge 10 days after the surgery. The baby has remained asymptomatic for 13 months during the follow-up period and the CTA revealed the absence of stenosis in both the LPA and right RPA.

Discussion

Patients with hemitruncus arteriosus generally exhibit atypical clinical symptoms, especially when combined with other congenital heart diseases such as atrial septal defect, PDA, Tetralogy of Fallot [6,7]. Some researches showed infant patients and adult patients with different symptoms such as cyanosis, tachypnea, respiratory distress, pulmonary hypertension [6–8]. Our case was the youngest case reported in the literature and exhibits atypical symptoms shortness of breath. Once the diagnosis of hemitruncus arteriosus is confirmed, prompt surgical repair is recommended. Because The mortality rates of the disease at 3 months and 6 months post-diagnosis are approximately 30% and 70%, respectively [3,6].

Various surgical approaches have been applied to this patient based on different anatomical types. Primary direct implantation technique means RPA was disconnected and then directly implanted into the side of the MPA. This procedure may be performed with or without patch augmentation for repairing the defect in the ascending aorta. Primary repair involving direct implantation is typically reserved for patients where the RPA originates from the posterior aspect of the aorta, in close proximity to the MPA and the RPA can be harvested without tension, because no synthetic materials and patches can keep the growth potential of the pulmonary artery (PA) and the need to prevent anastomotic obstruction [7]. Some other surgical approaches such as use of interposition graft conduit, use of aortic ring to elongate the RPA, double flap technique, suitable for patients with RPA far from the main PA, the significant drawback of this approaches are that the patient will eventually outgrow the branch pulmonary artery and reintervention rate was high.

The LeCompte maneuver is generally not recommended during initial surgical procedures. However, following the translocation of the right pulmonary artery, if there is a significant increase in right ventricular pressure, exceeding 60% of systemic circulation pressure, we advocate for the utilization of the LeCompte maneuver. This technique has been observed to substantially reduce right ventricular pressure, thereby facilitating a more stable postoperative recovery for the patient [8,9].

Cardiopulmonary bypass (CPB) was necessary for this kind of patients, however, circulatory arrest or not should be determined based on the surgeon's preference and the patient's anatomical considerations. The early surgical re-

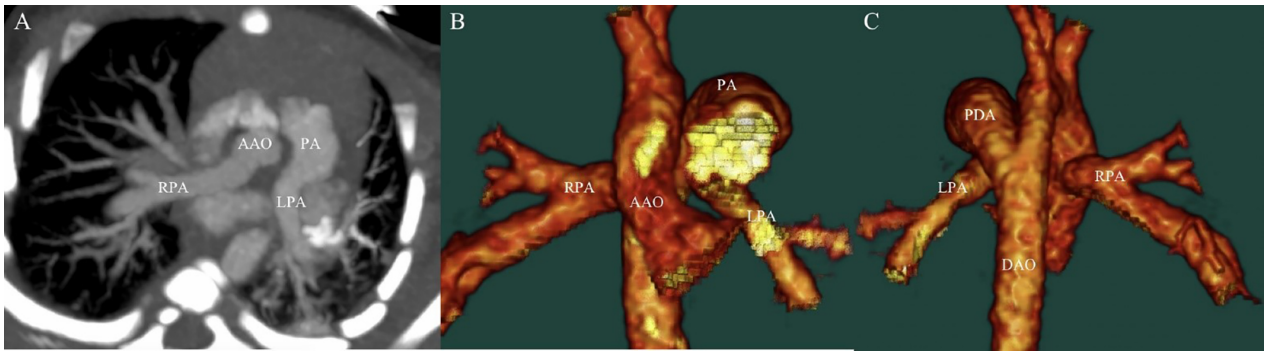

Fig. 2. CTA images. (A–C) Cinematic three-dimensional reconstruction from the CTA images showed the anomalous origin of the RPA from the AAO in the setting of a right-side aortic arch and a PDA. MPA, main pulmonary artery; LPA, left pulmonary artery; PDA, patent ductus arteriosus; RPA, right pulmonary artery; AAO, ascending aorta; CTA, computed tomographic angiography.

sult of anomalous origin of the right pulmonary artery from the ascending aorta was acceptable. The incidence of requiring reintervention was found to be 12.5%–36% during long-term follow-up [9–11].

To our knowledge, this represents the youngest documented case of an anomalous origin of the right pulmonary artery from the ascending aorta. We anticipate that this report will provide a substantial educational resource for medical practitioners. It is our earnest hope that this case will foster prompt diagnosis and proactive management in similar future cases, enhancing patient outcomes. The CARE checklist was used when writing this case report (**Supplementary Table 1**).

Availability of Data and Materials

All data generated or analyzed during this study are included in this published article.

Author Contributions

LL, YL and QA designed the research study. QA performed the research. MF and QA analyzed the data. All authors contributed to editorial changes in the manuscript. All authors read and approved the final manuscript. All authors have participated sufficiently in the work and agreed to be accountable for all aspects of the work.

Ethics Approval and Consent to Participate

This study is in compliance with the declaration of Helsinki. Written informed consent was obtained from the patient for publication of this case report in accordance with the journal's patient consent policy. This case report was approved by the Institutional Review Board of West China Hospital. Approval Number: 32209.

Acknowledgment

We would like to thank Dr. Xiao Li for his help in polishing our paper.

Funding

This research received no external funding.

Conflict of Interest

The authors declare no conflict of interest.

Supplementary Material

Supplementary material associated with this article can be found, in the online version, at https://doi.org/10.59958/hsf.7017.

References

[1] Alhawri K, Alakhfash A, Alqwaee A, HassabElnabi M, Ahmed F, Alhawri M, *et al*. Anomalous right pulmonary artery from aorta, surgical approach case report and literature review. Journal of Cardiac Surgery. 2021; 36: 2890–2900.

[2] Hadeed K, Ohanessian G, Acar P. Anomalous origin of the pulmonary artery from the ascending aorta in a neonate, assessed by two-dimensional echocardiography. Archives of Cardiovascular Diseases. 2010; 103: 493–495.

[3] Prifti E, Bonacchi M, Murzi B, Crucean A, Bernabei M, Luisi VS, *et al*. Anomalous origin of the left pulmonary artery from the aorta. Our experience and literature review. Heart and Vessels. 2003; 18: 79–84.

[4] Prifti E, Crucean A, Bonacchi M, Bernabei M, Leacche M, Murzi B, *et al*. Postoperative outcome in patients with anomalous origin of one pulmonary artery branch from the aorta. European Journal of Cardio-thoracic Surgery: Official Journal of

the European Association for Cardio-thoracic Surgery. 2003; 24: 21–27.

[5] Cho S, Kim WH, Choi ES, Lee JR, Kim YJ. Surgical Results of Anomalous Origin of One Pulmonary Artery Branch from the Ascending Aorta. Pediatric Cardiology. 2015; 36: 1532–1538.

[6] Nathan M, Rimmer D, Piercey G, del Nido PJ, Mayer JE, Bacha EA, *et al*. Early repair of hemitruncus: excellent early and late outcomes. The Journal of Thoracic and Cardiovascular Surgery. 2007; 133: 1329–1335.

[7] Kirkpatrick SE, Girod DAKH. Aortic origin of the right pulmonary artery. The Annals of Thoracic Surgery. 1968; 5: 165–170.

[8] Sha JM, Cao Y, Xu SS. Repair of Hemitruncus With Irreversible Pulmonary Hypertension. The Annals of Thoracic Surgery. 2019; 108: e35–e36.

[9] Tsoutsinos A, Germanakis I, Kanakis M, Samanidis G, Despotopoulos S, Kousi T, *et al*. Abnormal origin of right pulmonary artery from the ascending aorta in an infant ("*Hemitruncus*"). Clinical Case Reports. 2023; 11: e8103.

[10] Kopparapu SC, Saravana Babu MS, Sukesan S, Menon S. Anomalous origin of right pulmonary artery from the ascending aorta-Intraoperative transesophageal echocardiography and surgical images. Annals of Cardiac Anaesthesia. 2023; 26: 431–432.

[11] Mahajan S, Bansal V, Aggarwal P, Naganur SH. Main pulmonary artery continuing as right pulmonary artery with trifurcation at its origin and aberrant origin of left pulmonary artery from ascending aorta in a case of tetralogy of Fallot. Indian Journal of Thoracic and Cardiovascular Surgery. 2023; 39: 89–92.