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Anomalous origin of a right pulmonary artery identified with echocardiography combined with CT: a case in a juvenile patient

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Abstract

An anomalous origin of the pulmonary artery (AOPA) from the ascending aorta is a relatively rare but important cardiac malformation that frequently involves the right pulmonary artery (RPA). Its clinical manifestations depend mainly on the associated significant pulmonary hypertension, with an extremely high mortality rate in the first year of life. Here, we present a rare survival case of an 11-year-old child with the disease, who was hospitalized due to intermittent abdominal pain, but without any apparent signs of chest tightness or shortness of breath. The low oxygen saturation as discovered during the physical examination. Subsequent examination with transthoracic echocardiography (TTE) and pulmonary artery computed tomography angiography (CTA) revealed this unexpected congenital malformation. Although the estimated mean pulmonary artery pressure (MAP) from the TTE was 51 mmHg, which seemed to contraindicate corrective cardiac surgery, the limitations of TTE were considered. Consequently, after multidisciplinary consultation, surgical intervention was ultimately decided upon, resulting in a favorable prognosis for the patient. This case provides a new insight for clinicians in the diagnosis and treatment of complex congenital heart diseases.

Keywords Anomalous origin of the pulmonary artery, Congenital heart malformation, Transthoracic echocardiography

Introduction

Anomalous origin of the pulmonary artery (AOPA), a congenital heart disease, is a rare anomaly that affects the right pulmonary artery (RPA) more frequently than the left pulmonary artery (LPA) and was first described by Franenttzel in 1868 [1]. This anomaly results in a substantial left-to-right shunt, with the entire cardiac output from the right ventricle traveling to one lung while the other lung receives blood at systemic pressure from the aorta [2]. These patients are at risk of developing significant pulmonary hypertension early. Therefore, early recognition and prompt surgical correction are crucial and favorable for ensuring good outcomes for AOPA patients [3]. In this report, we describe our TTE and CTA findings and deliberate upon the feasibility of operative intervention.

Case report

An 11-year-old female patient was admitted to our hospital with intermittent abdominal pain for more than 1 month. There was no previous medical history or

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family history of inherited disease in the patient's medical record. She had slight shortness of breath after activity but no other symptoms, such as chest pain, dyspnea or syncope. Upon physical examination, the temperature, respiratory rate, and heart rate were within normal limits. Furthermore, the patient presented with an arterial oxygen saturation level of 90% while breathing room air. During auscultation, An increased P2 (pulmonic) component of the second heart sound was finded. The lung sounds were unremarkable in all fields, and no abnormalities were detected during the abdominal examination. Blood analysis revealed an elevated hemoglobin level (218 g/L), red blood cell count (7.23 × 10^12/L), and hematocrit (63.1%). The features of multiview TTE were as follows (Fig. 1): (A) The parasternal aortic long axis view revealed that the RPA, which possessed an internal diameter of 16 mm, originated from the posterior wall of the ascending aorta, approximately 27 mm from the aortic valve ring. Color Doppler flow imaging (CDFI) revealed continuous flow signals from the ascending aorta into the anomalous pulmonary artery. (B-C) The pulmonary artery long-axis view revealed the existence of an arterial duct between the LPA and the descending aorta, with an internal diameter of 6.6 mm, accompanied by red and blue bidirectional shunt flow signals. (D) The flow spectrum indicated a maximum velocity (V_{max}) = 0.96 m/s and a pressure gradient (PG) = 4 mmHg; V_{max} = -0.72 m/s, PG = -2 mmHg, for an estimated pulmonary artery systolic pressure of 92 mmHg, whereas the measured brachial artery systolic pressure was approximately 94 mmHg. Further CT pulmonary artery angiography revealed the following (Fig. 2): (A) The RPA originated from the ascending aorta and had a lower density than the LPA did; (B) An arterial duct with an internal diameter of approximately 6.6 mm and a length of approximately 18 mm was observed between the starting part of the LPA and the descending aorta.

We understand that although the TTE parameters suggest the patient may not be suitable for cardiac surgery, TTE data alone cannot serve as the sole criterion for surgical correction in cases of complex congenital heart disease. Therefore, after thorough discussion by the multidisciplinary team and fully informing the family of the significant risks associated with the surgery, we decided

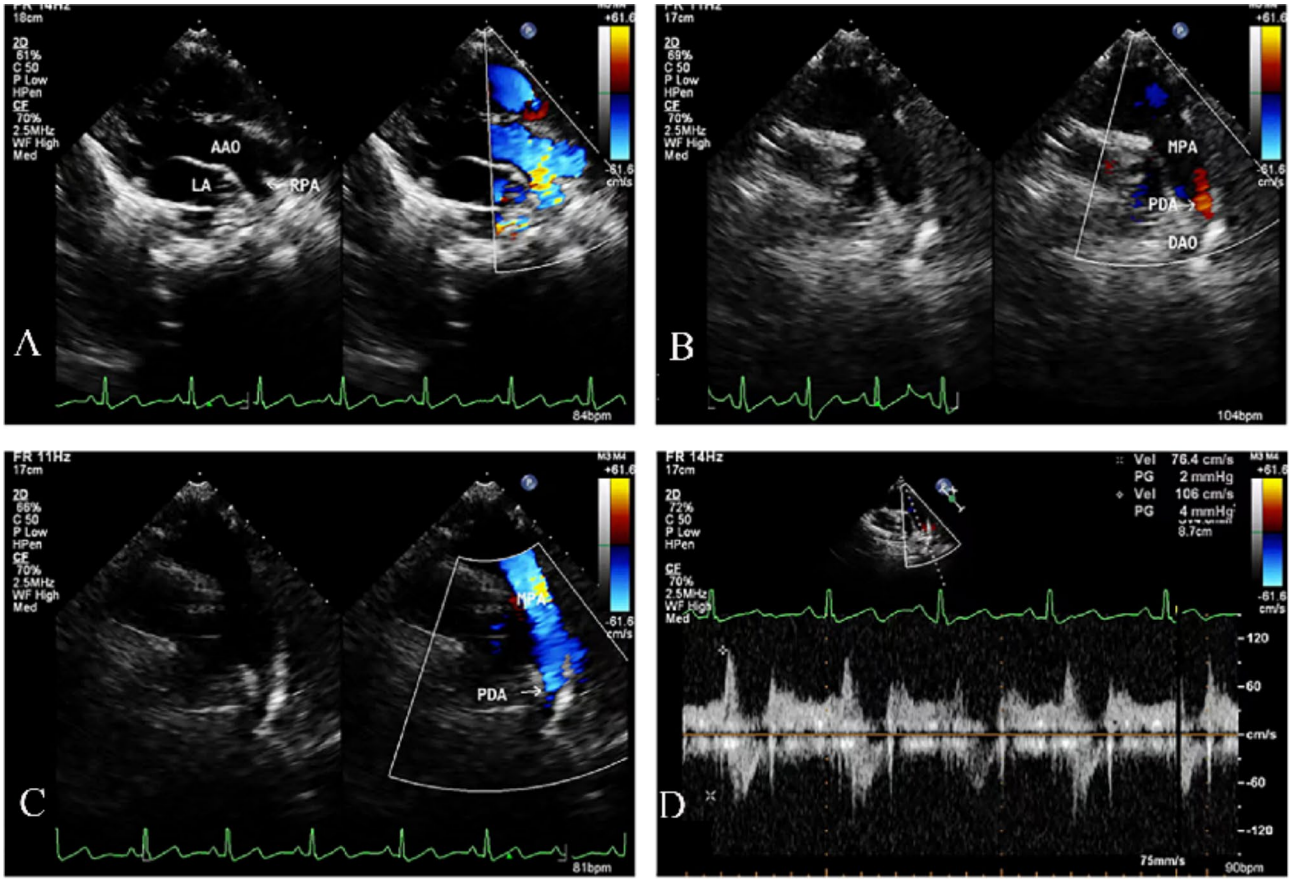

Fig. 1 Preoperative transthoracic echocardiography. (**A**) Parasternal aortic long-axis view; (**B**-**C**) parasternal short-axis view; (**D**) pulmonary artery long-axis view. LA, left atrium; MPA, mean pulmonary artery; RPA, right pulmonary artery; PDA, patent ductus arteriosus; AAO, ascending aorta; DAO, descending aorta

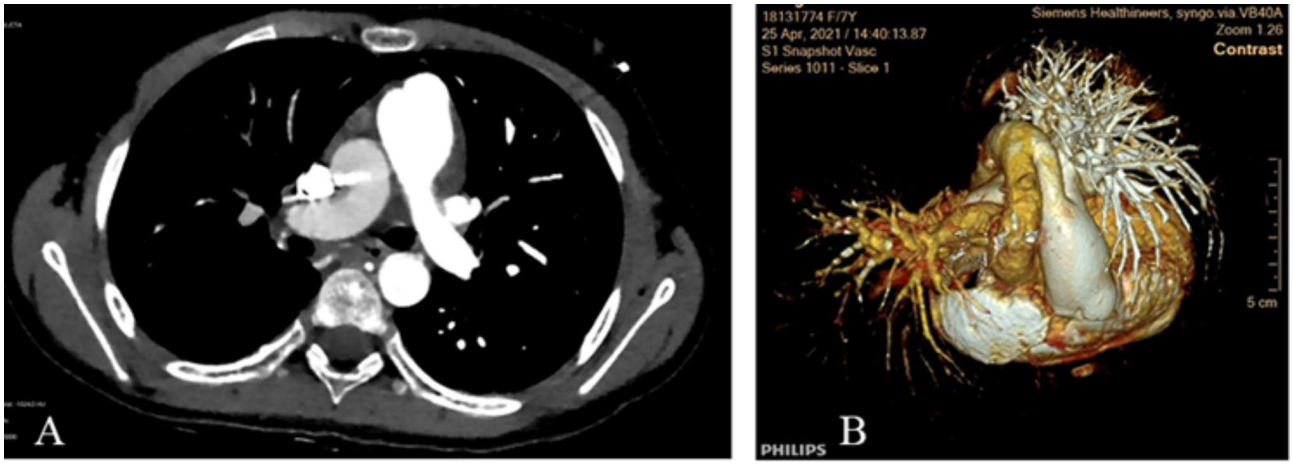

Fig. 2 CT pulmonary artery angiography. (**A**) The right pulmonary artery arises from the ascending aorta; (**B**) the arterial duct lies between the left pulmonary artery and the descending aorta. PA, pulmonary artery; RPA, right pulmonary artery; LPA: left pulmonary artery; PDA, patent ductus arteriosus; AAO, ascending aorta; DAO, descending aorta

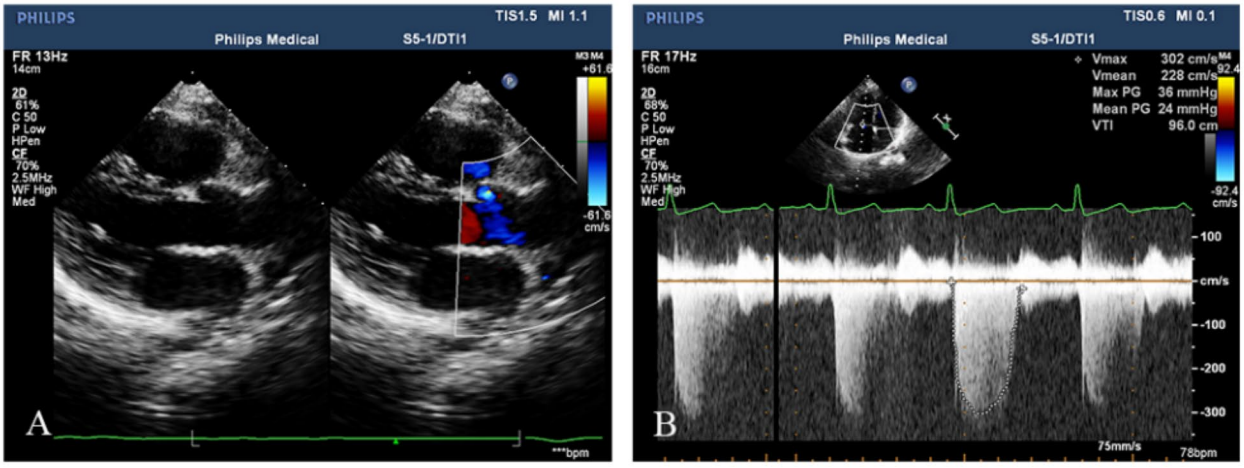

Fig. 3 Transthoracic echocardiography after the operation. (**A**) The abnormal pulmonary artery origin sign has disappeared. (**B**) Pulmonary artery pressure is estimated by tricuspid regurgitation

to attempt surgical correction to give the child a chance at survival. Under cardiopulmonary bypass and general anesthesia, the surgery was performed with a median thoracotomy. Initially, after ligation and separation of a PDA with a diameter of 8 mm, cardiac catheters showed that the pressure of the LPA dropped to 33 mm Hg, while the pressure of the RPA dropped to 55 mm Hg. This indicated that the elevated pressure in the LPA was secondary to the PDA. Consequently, a decision was mede to proceed with radical reconstruction of the pulmonary artery. Second, the anomalous pulmonary artery was disconnected from the ascending aorta, and the resulting opening in the RPA was reconnected to the right side of the pulmonary trunk. Finally, the artificial blood vessels were anastomosed with the proximal and distal parts of the ascending aorta. A significant decrease in pulmonary artery mean pressure to 45 mmHg was observed after the operation. The patient had an uncomplicated postoperative recovery and was discharged home one week after the operation. At the 6-month follow-up after the surgical repair, the heart sounds were normal, and no murmurs were heard. Echocardiography indicated (Fig. 3) that the pulmonary artery abnormal origin sign had disappeared; furthermore, according to tricuspid regurgitation, the pulmonary artery systolic pressure was approximately 36 mmHg. These findings suggested a significant decrease in pulmonary artery pressure compared with the presurgical values.

Discussion

Anomalous Origin of the Pulmonary Artery (AOPA) is a rare congenital heart disease, accounting for only 0.12% of all congenital defects [4]. It is characterized by one pulmonary artery originating from the ascending aorta, while the other is connected to the main pulmonary artery. This condition leads to a significant left-to-right shunt, subjecting one lung to systemic circulation pressure [5]. More seriously, this disease is often accompanied by other congenital heart malformations, with Patent Ductus Arteriosus (PDA) being the most common [6], leading to more complex pathophysiological and hemodynamic changes. Ultimately, within the first year of life, it can lead to pulmonary hypertension and congestive heart failure, resulting in death [7]. Currently, Transthoracic Echocardiography (TTE) and Computed Tomography (CT) are clinically applied to assist in the diagnosis of complex congenital heart diseases [8, 9]. However, it is important to note that the parameters provided by these examinations cannot simply determine whether cardiac surgery can be performed for radical correction, as evidenced in our case.

At present, pulmonary angiography or cardiac catheterization is the ultimately reliable method for diagnosing AOPA, but they are invasive and expose patients to high doses of radiation [5]. Due to family considerations regarding the lack of severe clinical symptoms in the child and the potential adverse factors mentioned above, cardiac catheterization was not acceptable. Studies have shown that TTE has a significant contribution to the non-invasive and accurate diagnosis of AOPA [10]. TTE can clearly display the exact location and origin of AOPA, as well as other related malformations and hemodynamic changes. Furthermore, according to Mansi Verma's research [11], Computed Tomography Angiography (CTA) helps provide precise anatomical depiction and identification of related anomalies, thereby assisting in preoperative planning for surgical intervention. In this report, we used a combination of TTE and CTA to clearly diagnose AOPA with PDA, but the pulmonary artery pressure provided by TTE seemed not to be suitable for further cardiac surgical correction. Surgical intervention is the only way to improve the long-term prognosis of the disease, and the therapeutic effect is closely related to age, category of malformation, and complexity. There have been reports of death after surgical treatment in patients with complex congenital heart malformations, making thorough preoperative assessment crucial [12]. As Dr. Saurabh K demonstrated in a detailed multimodal hemodynamic assessment for a 15-year-old boy, and successfully managed the surgery based on the assessment results [13]. Complex cardiac malformations with special hemodynamic changes, such multimodal hemodynamic assessment is well worth our reference to ensure good postoperative surgical outcomes for patients. However, in this specific case, it is difficult to make a choice between conservative treatment and surgical correction, as both carry significant risks. Although the TTE data seems to reflect a severe condition, the child has no severe clinical symptoms, and TTE indeed has limitations in examining complex congenital heart malformations. Therefore, after multidisciplinary discussion, it was decided to attempt surgical intervention, with a well-planned preoperative scheme, intraoperative attempts to close PDA and hemodynamic assessment by cardiac catheterization, and successful surgery after full assessment. Postoperatively, Bosentan medication was used as per the standard regimen, and the follow-up results were satisfactory.

Consequently, in this particular case, owing to the patient's inability to undergo invasive examinations, we opted for the diagnosis of the congenital heart malformation through TTE and CTA scans. After multidisciplinary consultation, surgical intervention was ultimately decided upon, resulting in a favorable prognosis for the patient.

Conclusion

AOPA is a rare congenital cardiovascular disease with a poor prognosis that requires timely identification and prompt surgical correction. The suggested diagnostic approach for AOPA requires the use of TTE in conjunction with CTA. This is a simple, noninvasive and safe method. However, data from TTE alone cannot serve as the standard for surgical procedures. Therefore, a stepwise multimodality hemodynamic evaluation was instrumental in facilitating successful surgical repair in an adolescent male patient.

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Author contributions
Yan-ling Li performed the main tasks and participated in the writing of the manuscript. Ping Xie provided suggestions for and revised the manuscript during its writing. Jia Wei collected and organized the data. Zhao-xia Guo revised the manuscript.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval
Not applicable.

Consent to participate
Not applicable.

Consent for publication
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Competing interests
The authors declare no competing interests.

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