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A Rare Anatomical Variant of Adolescent Persistent Truncus Arteriosus: Multimodal Imaging-Guided Surgical Strategy and Outcome

A 15-year-old girl was admitted with dyspnea on exertion and minimal hemoptysis lasting 1 week. On auscultation, a grade III systolic murmur was heard in the aortic valve area with localized transmission to the neck. Her oxygen saturation was 89% in room air. Subsequent transthoracic echocardiography (TTE) revealed a single widened arterial trunk (50 mm in diameter) overriding the ventricular septum by about 60%. Two separate pulmonary artery branches originated directly from the posterior left wall of the arterial trunk (Figure 1A). The common arterial valve was trileaflet with moderate-to-severe regurgitation, and a large non-restrictive ventricular septal defect (VSD) was noted beneath the valve (Figure 1B, 33 mm in diameter). Computed tomography angiography (CTA) showed a single arterial trunk supplying the coronary arteries, pulmonary arteries, and aorta. The left and right pulmonary arteries arose directly from the dilated posterior wall of the arterial trunk (Figure 1C and D; Video 1). Right heart catheterization indicated severe pulmonary hypertension: pulmonary vascular resistance (PVR) was 362 dyn·s/cm^5 (4.5 Wood units), pulmonary artery pressure (PAP) was 109/42/64 mm Hg, and Qp/Qs was 2.63. According to these findings, the patient received preoperative pulmonary vasodilator therapy, including inhaled nitric oxide and oral sildenafil at a dose of 2 mg/kg 3 times daily, aimed at reducing PVR and optimizing cardiac function. After multidisciplinary discussion, the patient underwent surgical correction of the common arterial trunk, ascending aorta and aortic valve repair, and VSD closure. Intraoperative findings confirmed the imaging diagnosis. The postoperative course was uneventful, and she was discharged 1 week later. Postoperatively, pharmacological management with sildenafil was continued to control PAP. The oral dose remained at 2 mg/kg 3 times daily to prevent complications and promote recovery.

Six months after surgery, outpatient TTE showed no residual shunt at the ventricular level, mild aortic valve regurgitation, and no stenosis in the right ventricular outflow tract, pulmonary artery, or its branches. The forward flow velocity in the right ventricular outflow tract was 2.0 m/s. Computed tomography angiography demonstrated patency of the right ventricular outflow tract and artificial graft (Figure 1E and F; Video 2), with reduction in right ventricular size and improvement in ascending aorta dilation (Figure E compared to Figure C). The patient will continue to undergo regular medical evaluations and follow-up visits to allow timely adjustment of anti-pulmonary arterial hypertension (PAH) treatment, prevent complications, and ensure optimal long-term outcomes.

Persistent truncus arteriosus (PTA) is a rare congenital heart defect, accounting for 1%-4% of all congenital cardiac anomalies.[1,2] It arises from the failure of embryonic septation between the aorta and pulmonary trunk, resulting in a single arterial trunk that supplies both systemic and pulmonary circulations. Persistent truncus arteriosus exhibits significant anatomical heterogeneity, necessitating classification systems such as Collett-Edwards (based on pulmonary artery origin) and Van Praagh (based on ventricular geometry) to guide surgical planning.[3,4] In recent years, increasing recognition of late-presenting PTA has been observed, particularly among adolescents and adults, often due to mild initial symptoms or

E-PAGE ORIGINAL IMAGE

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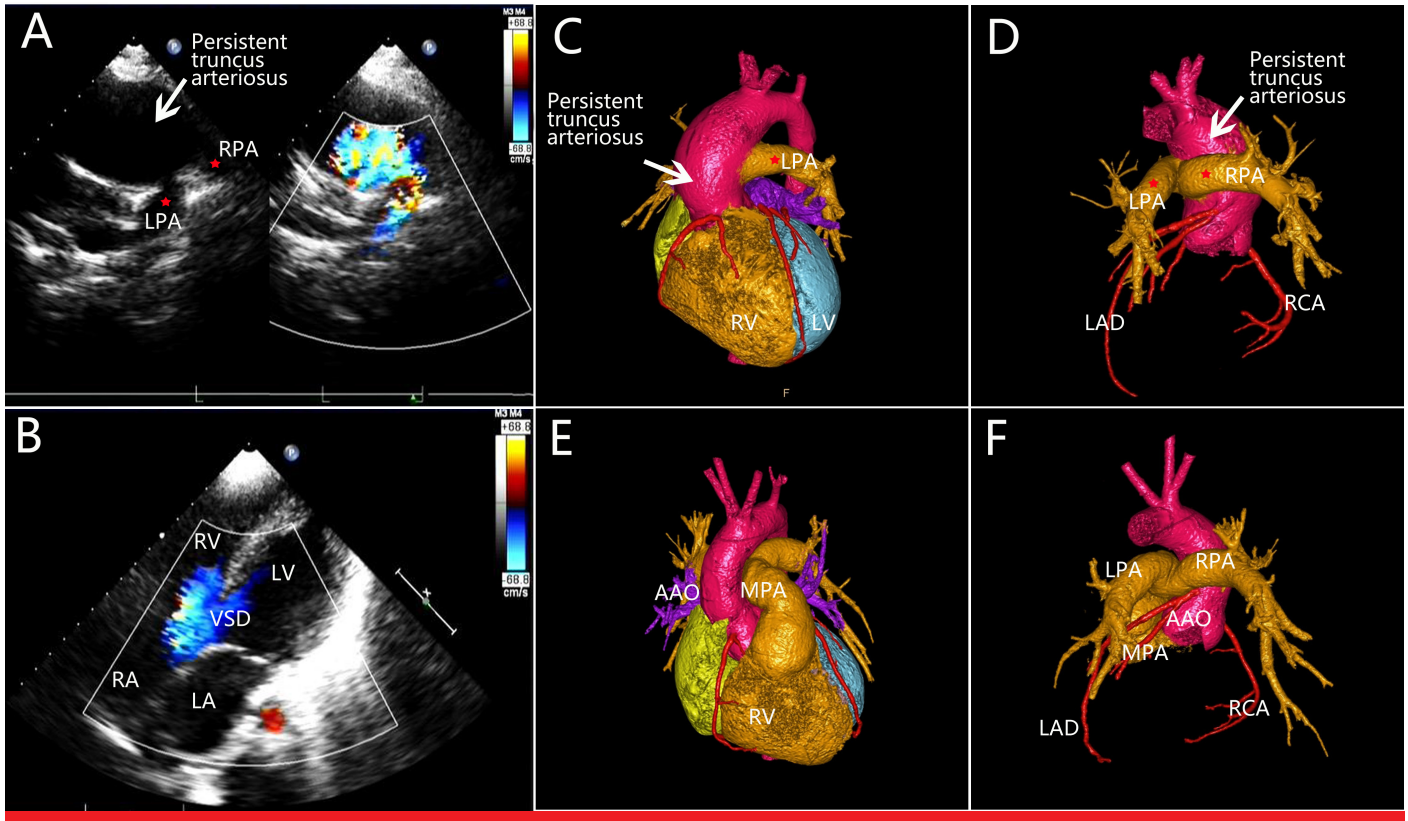

Figure 1. Multimodal images of a 15-year-old girl diagnosed with type I persistent truncus arteriosus with ventricular septal defect who underwent surgical correction. (A-B) Transthoracic echocardiography shows a single widened arterial trunk with 2 separate pulmonary artery branches originating from the posterolateral wall of the arterial trunk and a large ventricular septal defect beneath the common arterial valve. (C-D) Preoperative computed tomography angiography volume rendering shows the left and right pulmonary arteries directly arising from the dilated posterolateral wall of the arterial trunk. (E-F) Postoperative computed tomography angiography volume rendering shows patency of the right ventricular outflow tract and artificial graft, with marked reduction in right ventricular size and significant improvement in ascending aorta dilation compared to preoperative images. AAO, aorta; LAD, left anterior descending coronary artery; LPA, left pulmonary artery; LV, left ventricle; MPA, main pulmonary artery; RCA, right coronary artery; RPA, right pulmonary artery; RV, right ventricle.

limited access to medical care.[5] These patients commonly present with long-standing volume overload and progressive pulmonary vascular disease, which significantly elevate surgical risk. Despite these challenges, tailored surgical strategies guided by multimodal imaging and comprehensive hemodynamic assessment have demonstrated favorable outcomes.[6] Studies indicate that even in late-presenting cases, complete repair can be performed with relatively low perioperative mortality and satisfactory short- to mid-term results.[6] However, significant preoperative truncal valve regurgitation remains an independent risk factor for overall mortality, highlighting the importance of optimizing valve management to improve long-term survival.[7] Other operative risk factors include aortic arch abnormalities and prolonged cardiopulmonary bypass time, which are associated with increased hospital mortality.[7] Postoperative follow-up reveals substantial improvement in cardiac function, with most patients transitioning from NYHA class III/IV preoperatively to class I/II postoperatively,[7] underscoring the life-saving and quality-of-life benefits of surgical intervention. Nevertheless, mild reductions in exercise tolerance remain common among long-term survivors, emphasizing the need for structured rehabilitation and lifelong monitoring.[8] It is also critical to recognize that PTA is frequently complicated by PAH, a condition often underdiagnosed due to its nonspecific clinical presentation.[5] Delayed diagnosis and referral to specialized PH centers contribute to poor outcomes. In this context, pharmacological support—particularly with sildenafil—plays a vital role in maintaining pulmonary vascular patency, preventing pressure rebound, and supporting myocardial recovery.[9] The STARTS-2 trial further confirms that long-term use of sildenafil significantly improves survival and quality of life.[9]

This case illustrates that individualized surgical repair—including reconstruction of the arterial trunk, valve preservation or repair, and closure of the VSD—can effectively correct anatomical defects, restore hemodynamics, and achieve durable outcomes, even in adolescent patients with severe pulmonary hypertension. Given the high risk of complications such as PAH and early mortality, precise preoperative evaluation using multimodal imaging, and hemodynamic assessment is essential to optimize surgical strategy and outcomes, particularly in high-risk populations.

This approach not only enhances immediate success but also provides a valuable framework for managing similarly complex congenital heart defects.

Data Availability Statement: The data that support the findings of this study are available from the corresponding author.

Informed Consent: This report has obtained the patient's informed consent for the publication of their anonymized clinical data.

Declaration of Interests: All authors have read and approved submission of the manuscript and have no conflict of interest to disclose.

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Video 1: Preoperative CT volume rendering demonstrates bilateral pulmonary arteries directly arising from the dilated posterior wall of the common arterial trunk.

Video 2: Postoperative CT volume rendering illustrates patency of the right ventricular outflow tract and the artificial conduit.

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