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Case Report

Unrecognized major aortopulmonary collateral arteries in double outlet right ventricle-ventricular septal defect: A rare case report

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ABSTRACT

Background: Major aortopulmonary collateral arteries (MAPCAs) are aberrant vessels that typically form to compensate for cyanotic congenital heart disease (CHD) with restricted pulmonary blood flow. This often obscures clinical suspicion of MAPCAs in CHDs associ-ated with pulmonary overcirculation, making it potentially unrecognized.
Case Illustration: We report the case of a 5-month-old girl presenting with dyspnea, recurrent respiratory infections, and failure to thrive. Echocardiography revealed double outlet right ventricle (DORV) with ventricular septal defect (VSD) and patent ductus arteriosus (PDA). After initial treatment for heart failure and pneumonia, she underwent pul-monary artery (PA) banding and PDA ligation. Ongoing poor postoperative status prompted a second surgery, where intraventricular tunnel repair failed due to inability to wean from cardiopulmonary bypass. An atrial septectomy and PA band retightening achieved hemodynamic improvement, but the patient remained ventilator-dependent with recurrent pneumonia. Further evaluation with multislice CT and catheterisation uncovered previously undiagnosed MAPCAs supplying the right lung and a hypoplastic right pulmonary artery (RPA), precluding embolisation. Since the MAPCAs and hypoplastic RPA were initially unrecognized, the PA banding procedure inadvertently exacerbated the right lung.
Conclusions: This case highlights the importance of considering MAPCAs in various CHD morphologies and emphasizes the value of pre-procedural imaging selection to optimize surgical planning and outcomes.

1. Introduction

Major aortopulmonary collateral arteries (MAPCAs) are aberrant vasculatures that arise from the aorta or its branches to the pulmonary vasculatures. MAPCAs typically form to compensate for cyanotic congenital heart diseases (CHDs) characterised by restricted pulmonary blood flow. This often obscures clinical suspicion of MAPCAs in CHDs associated with pulmonary overcirculation. Consequently, diagnosis can be missed or delayed, and complications such as pulmonary hypertension may have already developed, impeding the already intricate management of MAPCAs, especially with complex CHD.[1,2]

In this report, we present a case of a 5-month-old girl with double outlet right ventricle (DORV) and ventricular septal defect (VSD), in whom MAPCAs were only identified after persistent postoperative symptoms prompted further investigation.

2. Case Illustration

A 5-month-old girl was referred to our hospital with a chief complaint of shortness of breath 5 days prior to admission. She had experienced recurrent respiratory tract infections and failure to thrive since the age of 3 months, but her parents were unsure about the presence of cyanosis. On physical examination, she weighed 3.5 kg, room air oxygen saturation was 94%, auscultation of heart sounds was regular but a grade 3/6 pansystolic murmur was heard on the lower left sternal border. Echocardiography revealed both the aorta and pulmonary artery arising from the right ventricle, confirming a diagnosis of DORV accompanied by a 3 mm inlet VSD and a patent ductus arteriosus (PDA). Pressure gradients across the right ventricle–aorta (10 mmHg) and right ventricle–pulmonary artery (6 mmHg) were low, with no stenosis or regurgitation of the aorta or pulmonary artery. Increased pulmonary vascularity was noted on chest radiography. We assessed the patient with congestive heart failure and pneumonia and treated her with furosemide, captopril, and spironolactone. No significant improvement occurred, so we planned for the patient to undergo surgery immediately.

The initial surgical intervention included pulmonary artery (PA) banding and PDA ligation. The patient was stable during the first few days post-operation. However, she was unable to be weaned off ventilator support. By postoperative day 15, the patient started showing signs of pneumonia and a hyperdynamic state. The patient was decided to undergo re-tightening of the PA banding and DORV repair to improve haemodynamics and cardiopulmonary conditions. The procedure was done 21 days after the first PA banding procedure. Initially, we did an intraventricular tunnel repair. When the patient was weaned off the cardiopulmonary bypass, systolic pressure dropped to 30 mmHg and peripheral saturation to 20%, giving the impression of a pulmonary hypertension crisis. The cardiopulmonary bypass was then restarted and an atrial septectomy was done, followed by the ventricular patch removal and retightening of the PA banding. Peripheral saturation improved to 70% and systolic pressure to 70 mmHg, cardiopulmonary bypass was successfully weaned off.

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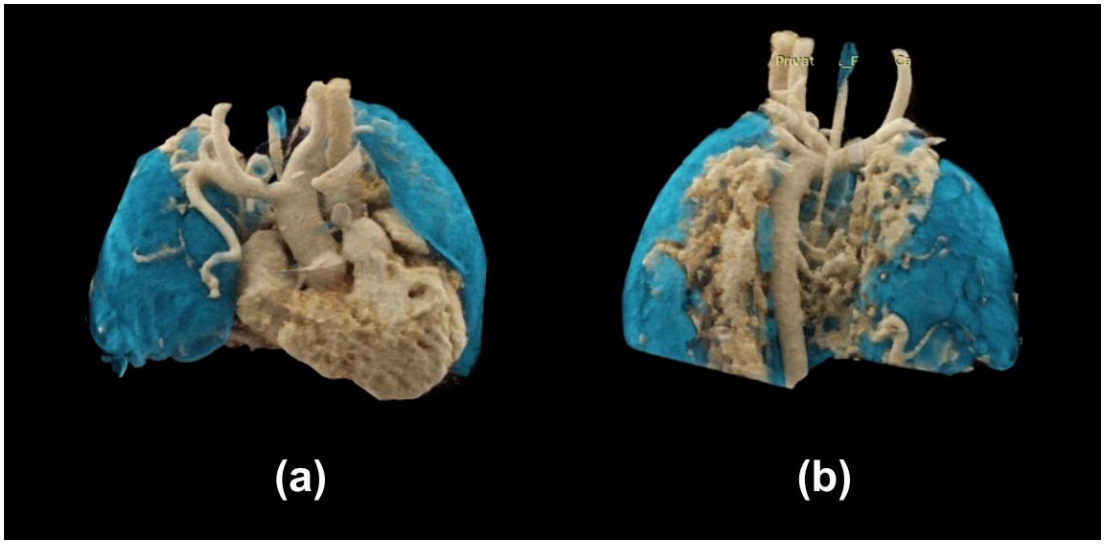

Figure 1. Three-dimensional reconstruction of computed tomography (CT) scan of the thorax demonstrating multiple major aortopulmonary collateral arteries (MAPCAs) supplying the right lung. (a) Anterior view shows multiple MAPCAs arising from the right subclavian artery (b) Posterior view further delineates the extensive collateral network entering the right pulmonary parenchyma.

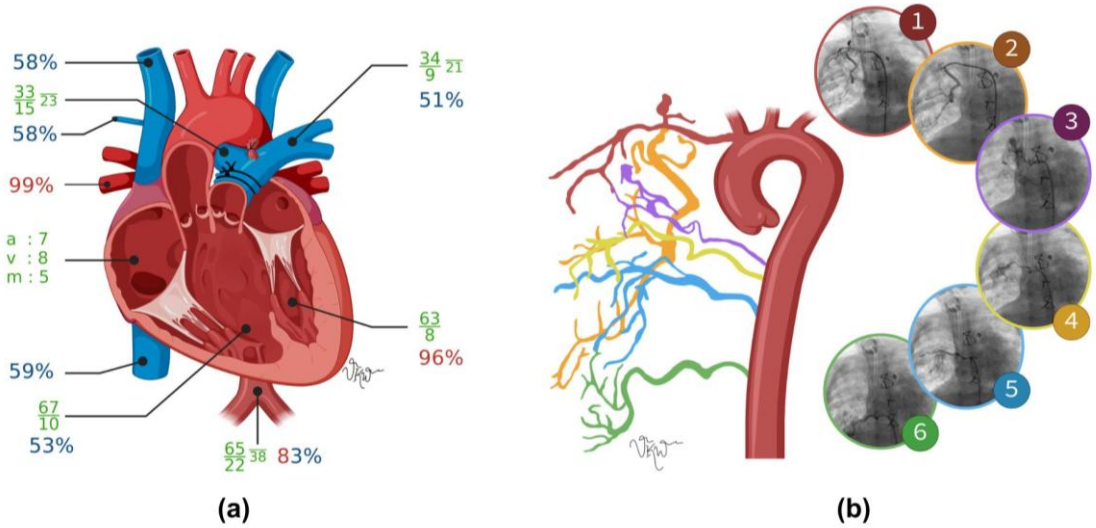

Figure 2. (a) Schematic illustration of the patient's cardiac anatomy, oxygen satura-tions, systolic and diastolic pressures along with mean arterial pressures based on the cardiac catheterization results. The diagram demonstrates double outlet right ventricle (DORV) with inlet extending to subaortic ventricular septal defect (VSD), post-ligated patent ductus arteriosus (PDA), bilateral superior vena cava (SVCs), pulmonary hypertension following re-tightening of the main pulmonary artery banding, confluent pulmonary arteries, hypoplastic right pulmonary artery, and multiple essential MAPCAs. Oxygen saturation values (percentages) and pressures (systolic/diastolic with mean in parentheses) are shown within the cardiac chambers and great vessels (indicated by black arrows), reflecting significant systemic-pulmonary mixing. (b) Pulmonary angiography demonstrating multiple MAPCAs, each color-coded to indicate a distinct essential collateral artery supplying the right lung.

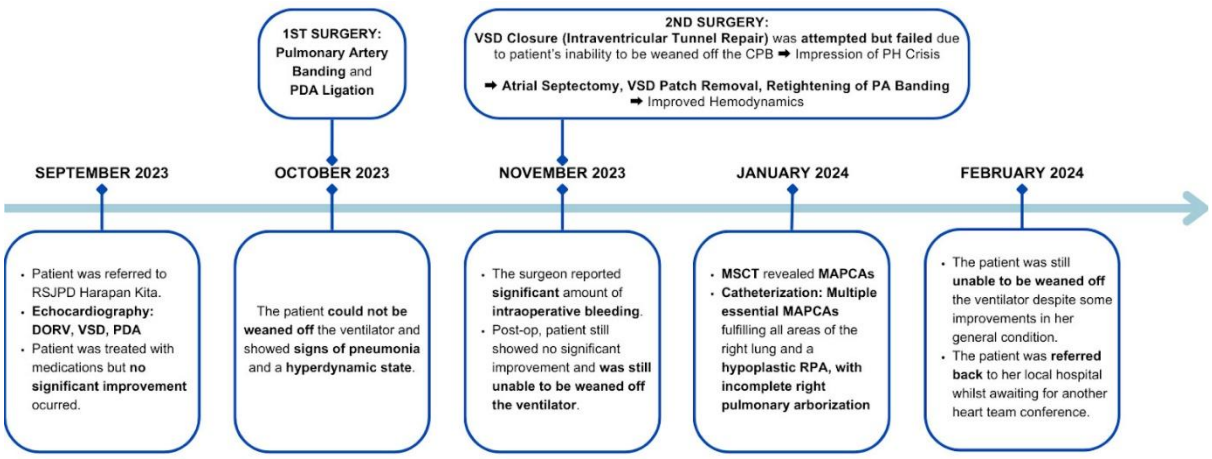

Figure 3. Timeline of the patient's clinical course.

After the second surgery, the patient did not show a significant improvement. The patient was still unable to be weaned off the ventilator and experienced recurrent bouts of pneumonia. We did echocardiography again but did not get any new findings. Due to a persistent lack of progress following multiple interventions, we proceeded with cardiac multislice computerised tomography (MSCT) to evaluate the patient's heart condition in greater detail. The MSCT revealed the presence of some previously undetected MAPCAs (Figure 1). We decided to do a cardiac catheterisation to embolise these MAPCAs. Catheterisation confirmed the presence of a DORV, an inlet VSD that expands to the subaortic, pulmonary hypertension, and several MAPCAs fulfilling all areas of the right lung. We also detected a hypoplastic right pulmonary artery (RPA) with a diameter of 1.98 mm and incomplete right pulmonary arborization (Figure 2). The hemodynamics measurements obtained are presented in Table 1.

Table 1. Hemodynamics measurement from cardiac catheterization

	PVR (WU)	PVRi (WU/m²)	PVR/SVR
LPA	5.46	0.71	0.71
RPA	5.79	0.75	0.76

Abbreviations: PVR, pulmonary vascular resistance; WU, Woods Unit; PVRi, pulmonary vascular resistance index; PVR/SVR, ratio of pulmonary vascular resistance to systemic vascular resistance.

However, this diagnosis of MAPCAs was already delayed, the pulmonary pressure and pulmonary arteries' resistance were too high, so embolisation was not feasible. Following discharge with home oxygen, the patient was monitored at her local hospital. At two-month follow-up, she remained ventilator-dependent with frequent lower respiratory infections, reflecting the limited options for definitive repair given her high pulmonary vascular resistance and hypoplastic RPA. Figure 3 summarizes the timeline of the patient's care.

3. Discussion

MAPCAs typically develop to compensate for reduced pulmonary blood flow in cyanotic congenital heart defects. In our patient, diagnosis was delayed due to a low index of suspicion, as the primary condition—DORV with VSD—usually causes pulmonary overcirculation. However, MAPCAs have been reported in various CHDs, including DORV, total anomalous pulmonary venous connection, tricuspid atresia, complex single ventricle, and acyanotic CHDs.[3,4] Rare cases of isolated MAPCAs have also been documented.[5,6] These examples, along with our case, serve as a reminder to consider MAPCAs as one of the differential diagnosis for persistent pulmonary overcirculation even in atypical presentations. In our patient, the hypoplastic RPA was ultimately identified as the factor contributing to the development of MAPCAs, all of which were located in the right lung. This finding was made only later, even after the initial detection of MAPCAs.

This case underscores the importance of selecting appropriate imaging modalities, ideally before planning interventions. For diagnosing MAPCAs, echocardiography is often the initial imaging examination; however, while it has high specificity (100%), it has low sensitivity (53%).[1,2,7] In our patient, none of the six MAPCA branches were detected on repeated echocardiographic examinations. The hypoplastic RPA was also not identified, most likely because the distal narrowing lay beyond the acoustic window. In addition, no significant pressure gradient across the pulmonary artery was observed, a finding that typically suggests obstruction. The echocardiographic findings appeared to be consistent with pulmonary overcirculation secondary to DORV-VSD; therefore, no additional abnormalities were suspected in the beginning, leading to an incomplete initial diagnosis.

Beyond echocardiography, CT scan is generally a reliable and less invasive modality for diagnosing MAPCAs. Pulmonary circulation adequacy can also be assessed using indices such as the McGoon ratio and Nakata index, which are particularly useful in cases like a hypoplastic pulmonary artery. In this case, however, the hypoplastic RPA was not detected through CT scan. Recent advancements in CT technology, such as multiphase dual-energy CT, enable differentiation of lung segment perfusion from MAPCAs and native pulmonary arteries, while 3D printing allows precise scaling useful for procedural planning and patient education. However, CT scan may still have limited sensitivity in complex vascular anomalies, often necessitating follow-up with cardiac catheterisation for definitive assessment. Such was the case in our patient, where MSCT first revealed MAPCAs but did not provide a complete picture of their distribution.[1,2,7] Whilst clinically sound, this sequential approach showed the economic implications of initial diagnostic choices. Given the complexity of MAPCAs, early catheterisation may offer a more cost-effective strategy by streamlining diagnosis.

Cardiac catheterisation remains the gold standard for diagnosis.[1,7] It revealed the true culprits of our patient's condition: a hypoplastic RPA and multiple essential MAPCAs. These two defects contributed to a perplexing clinical presentation; although the patient exhibited pulmonary overflow, the excess flow to the right lung was not through the native RPA—as initially assumed based on the DORV–VSD physiology—but instead was supplied predominantly by the MAPCAs. Because CT and catheterisation are not routinely performed at our center to assess pulmonary blood flow prior to intervention—unless severe pulmonary hypertension or pulmonary artery obstruction is suspected—we decided to perform PA banding based on an incomplete initial diagnosis. This intervention failed to alleviate right-sided overcirculation and instead further compromised perfusion through the already hypoplastic RPA.

Upon establishing the diagnosis, our management strategy shifted, and the primary goals became to unifocalise the essential MAPCAs (integrating them into a unified vascular pathway); to salvage the hypoplastic RPA (redirecting the pulmonary perfusion to the native RPA); and to re-catheterise to assess the suitability of VSD closure for optimal haemodynamic outcomes.[1,3,8,9] We evaluated several surgical options to salvage the hypoplastic RPA without increasing flow to the left lung, such as Right Blalock-Taussig shunt, RV-PA conduit or Sano shunt, and central Waterston shunt (Figure 4).[1,8-11] However, management in this patient became particularly difficult because, despite the MAPCAs in the right lung being mostly essential, pulmonary hypertension had also already developed. This created a delicate balance in deciding how to optimise right lung perfusion while mitigating hypertension progression and hypertensive crisis.

4. Conclusion

MAPCA is a relatively uncommon cardiac anomaly but can occur in various CHD morphologies and should be considered among the differential diagnoses for persistent pulmonary overflow symptoms. Tailored pre-procedural imaging selection is paramount in managing complex CHD. When the likelihood of MAPCAs is high, early catheterisation may provide a more comprehensive and efficient diagnostic pathway, ultimately guiding patient-centered management options and timely interventions before further complications deteriorate.

5. Declaration

5.1 Ethics Approval and Consent to participate
Not applicable.

5.2. Consent for publication
Written informed consent has been obtained from the patient(s) to publish this paper.

5.3 Availibility of data and materials
Data used in our study were presented in the main text.

5.4 Competing interests
Not applicable.

5.5 Funding Source
This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

5.6 Authors contributions
Idea/concept: SNS. Design: SNS. Control/supervision: SNS. Data collection/processing: SNS, EHP, VKW. Extraction/Analysis/interpretation: SNS, EHP, VKW. Literature review: EHP, VKW. Writing the article: SNS, EHP, VKW. Critical review: SNS, EHP, VKW. All authors have critically reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

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