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Case Report **OPEN ACCESS**

Role of echocardiography-derived pulmonary to systemic flow ratio in diagnosing aortopulmonary window: A case report

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ABSTRACT

Background: Aortopulmonary window (APW) is a rare congenital cardiac anomaly characterized by abnormal communication between the ascending aorta and the main pulmonary artery. Without early surgical correction, this condition has a poor prognosis. However, few case reports have described adult survival of patients with untreated APW.

Case presentation: We report the case of a 15-year-old girl who developed irreversible pulmonary hypertension due to untreated APW. Initially, we suspected the presence of an extracardiac shunt using a simple calculation of the echocardiography-derived pulmonary-to-systemic flow (Qp/Qs) ratio during a routine echocardiography study, which provided us with a clue to proceed with further multimodal diagnostic evaluation. In this report, we describe a comprehensive diagnostic workup, including right heart catheterization and computed tomography imaging, which confirmed the diagnosis of severe irreversible pulmonary hypertension secondary to a large untreated APW.

Conclusion: This case report highlights the clinical utility of the echocardiography-derived Qp/Qs ratio as a valuable, noninvasive tool for diagnosing APW, which can lead to severe irreversible pulmonary hypertension. The multidisciplinary approach demonstrated in this case serves as a valuable example for clinicians evaluating similar cases. Therefore, APW should be considered in the differential diagnosis of severe pulmonary hypertension, even in adult patients.

Keywords: aortopulmonary window, computed tomography, echocardiography, right heart catheterization, case report

INTRODUCTION

Aortopulmonary window (APW) is a rare congenital cardiac anomaly, accounting for less than 1% of all congenital heart diseases [1]. This condition is characterized by an abnormal communication between the ascending aorta and the main pulmonary artery, resulting from a deficient septum separating the two vessels. APW can be classified into four distinct types based on its location: type I (proximal), located just above the sinus of valsalva; type II (distal), located in the upper portion of the ascending aorta; type III (total), involving the majority of the ascending aorta; and type IV (intermediate), which has adequate superior and inferior rims, making them most suitable for possible device closure [2]. While this anomaly can be seen in isolation, APW may be associated with other congenital cardiac anomalies, including patent ductus arteriosus, interrupted aortic arch, atrial septal defect, ventricular septal defect, and coronary artery anomalies, occurring in approximately half the patients [3-6].

This condition typically manifests soon after birth with a gradual decline in pulmonary vascular resistance over the first few days of life, resulting in the development of a left-to-right shunt. This leads to pulmonary edema and symptoms of congestive heart failure, as observed in other congenital left-to-right cardiac shunts. If left untreated, the prognosis of APW without prompt surgical intervention is extremely poor. Timely diagnosis and repair of the defect are essential to prevent the devastating consequences of uncontrolled left-to-right shunting and the development of irreversible pulmonary vascular disease [7, 8].

This report presents a case of APW incidentally identified on echocardiography due to a suspected dilated right-sided heart. Due to the increased echocardiography-derived pulmonary-to-systemic flow (Qp/Qs) ratio, the presence of an extracardiac shunt in the absence of echocardiographic evidence of intracardiac cardiac shunts by color Doppler was suspected. Using agitated saline contrast, saline bubbles were observed in the descending aorta, with no visible contrast seen in the left-side chambers of the heart, raising the suspicion of a pulmonary-to-aorta shunt. The diagnosis was further confirmed by right heart catheterization and computed tomography (CT) imaging.

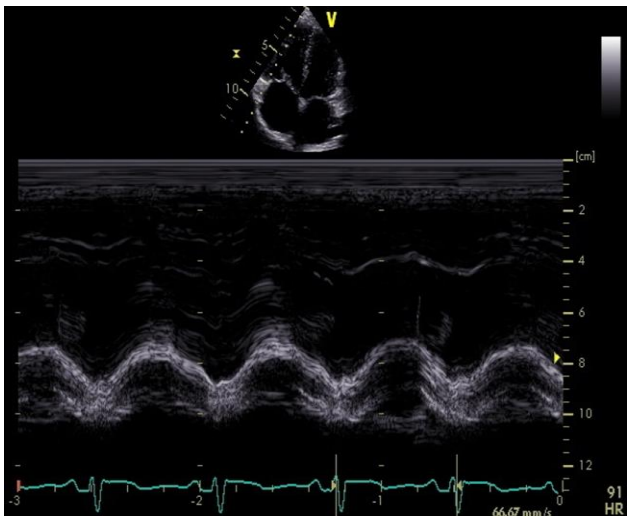

Figure 1. Trans-thoracic echocardiography image showing dilated right ventricle with normal systolic function assessed by TAPSE (TAPSE: tricuspid annular plane systolic excursion) (reprinted with permission of the patient)

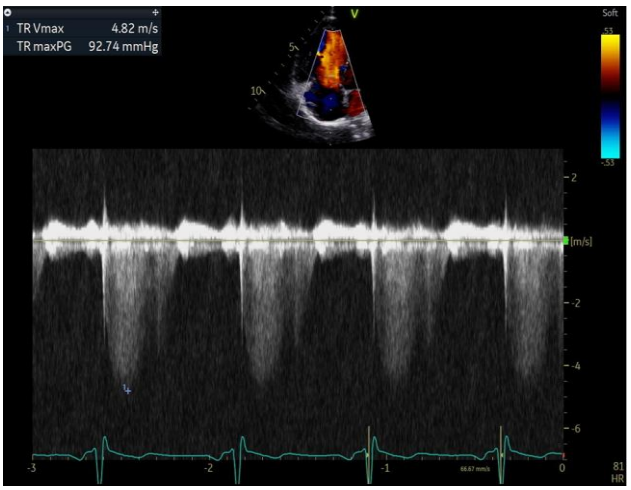

Figure 2. Trans-thoracic echocardiography image with CW Doppler, showing elevated right ventricle systolic pressure measured at 108 mmHg (estimated from peak TR velocity jet + RAp) (Rap: right atrial pressure & TR: tricuspid regurgitation) (reprinted with permission of the patient)

CASE PRESENTATION

A 15-year-old girl presented to the pulmonology clinic at our institution with a history of dyspnea on exertion (NYHA functional class II-III) and two episodes of hemoptysis. She was referred to the cardiology clinic for further evaluation, including echocardiography, to assess the presence of pulmonary hypertension and rule out the possibility of intracardiac shunts or valvular heart disease. A comprehensive cardiac workup was performed to elucidate the underlying etiology of the patient's respiratory symptoms and hemoptysis, with a particular focus on identifying potential structural or hemodynamic abnormalities within the cardiovascular system.

The patient had recurrent respiratory infections since childhood, which required repeated hospital admissions, and was subsequently diagnosed with an underlying cardiac pathology for which surgery had been advised. However, the patient's parents refused surgery. The exact diagnosis could not be definitively established as the patient's prior medical records were not available for review. However, the patient's symptoms improved over time and ultimately subsided, coinciding with normal growth and development.

Physical examination revealed pan-digital clubbing. Auscultation revealed a loud S2 with a short-ejection systolic murmur over the left lower sternal border in the second intercostal space. The oxygen saturation (SaO2) was 80-85% at rest, which showed further desaturation with exercise. Laboratory examinations were unremarkable, with no significant abnormalities identified in the blood, which helped to further confirm the absence of an underlying systemic disease process contributing to the patient's respiratory symptoms. Chest radiography revealed mild bilateral hilar prominence with no other remarkable findings.

Transthoracic echocardiography showed normal left ventricle cavity size and function (ejection fraction = 56%) with no regional wall motion abnormality (**Video 1A** & **Video 1B** - Transthoracic echocardiography showed normal left ventricular cavity size and function), moderately dilated right ventricle (RV) with normal systolic function (**Figure 1**) and flattened ventricular septum in systole and diastole, consistent with RV pressure and volume overload (**Video 2** - Parasternal short-axis view showed a dilated right-side heart with septal flattening during systole and diastole, consistent with right ventricular pressure and volume overload).

The RV systolic pressure was severely elevated at 108 mmHg (**Figure 2**) from a moderate degree of tricuspid regurgitation (**Video 3** - Trans-thoracic echocardiography with color Doppler showed moderate tricuspid regurgitation). There was no evidence of an intracardiac shunt on color Doppler (**Video 4A** & **Video 4B** - Parasternal short-axis pulmonary artery-focused view (A) and suprasternal aortic arch view (B) with color Doppler showed no evidence of shunts). However, when agitated saline contrast was injected, saline bubbles were observed in the descending aorta, with no visible saline contrast seen in the left side chambers of the heart, which raised the suspicion of a pulmonary-to-aorta shunt (**Video 5** - Apical-4 chamber was viewed with injected agitated saline contrast and showed suspected saline bubbles in the descending aorta (arrow) with no visible saline contrast seen in the left chambers of the heart). Therefore, we performed echocardiography-derived Qp/Qs calculations to determine the possibility of intra- or extra-unseen cardiac shunts. The calculated Qp/Qs was equal to 1.8 (**Figure 3**), indicating the possibility of a left-to-right shunt. Therefore, additional cardiac imaging modalities were recommended.

The patient underwent right heart catheterization via the right femoral venous access. The catheter advanced from the right atrium across the tricuspid valve into the RV and then further progressed through the right ventricular outflow tract into the pulmonary artery. Unexpectedly, the catheter was observed to have crossed over and entered the descending aorta, suggesting the presence of a connection between the pulmonary artery and aorta, consistent with an APW (**Video 6** - Right heart catheterization showed passage of the catheter from the pulmonary artery to the aorta through the aortopulmonary window). An additional arterial catheter was inserted into the right femoral artery. Contrast injection confirmed the presence of a pulmonary-to-aorta shunt, which was consistent with Eisenmenger physiology (**Video 7** - Pigtail catheter in the aorta showed the reversal of contrast from the

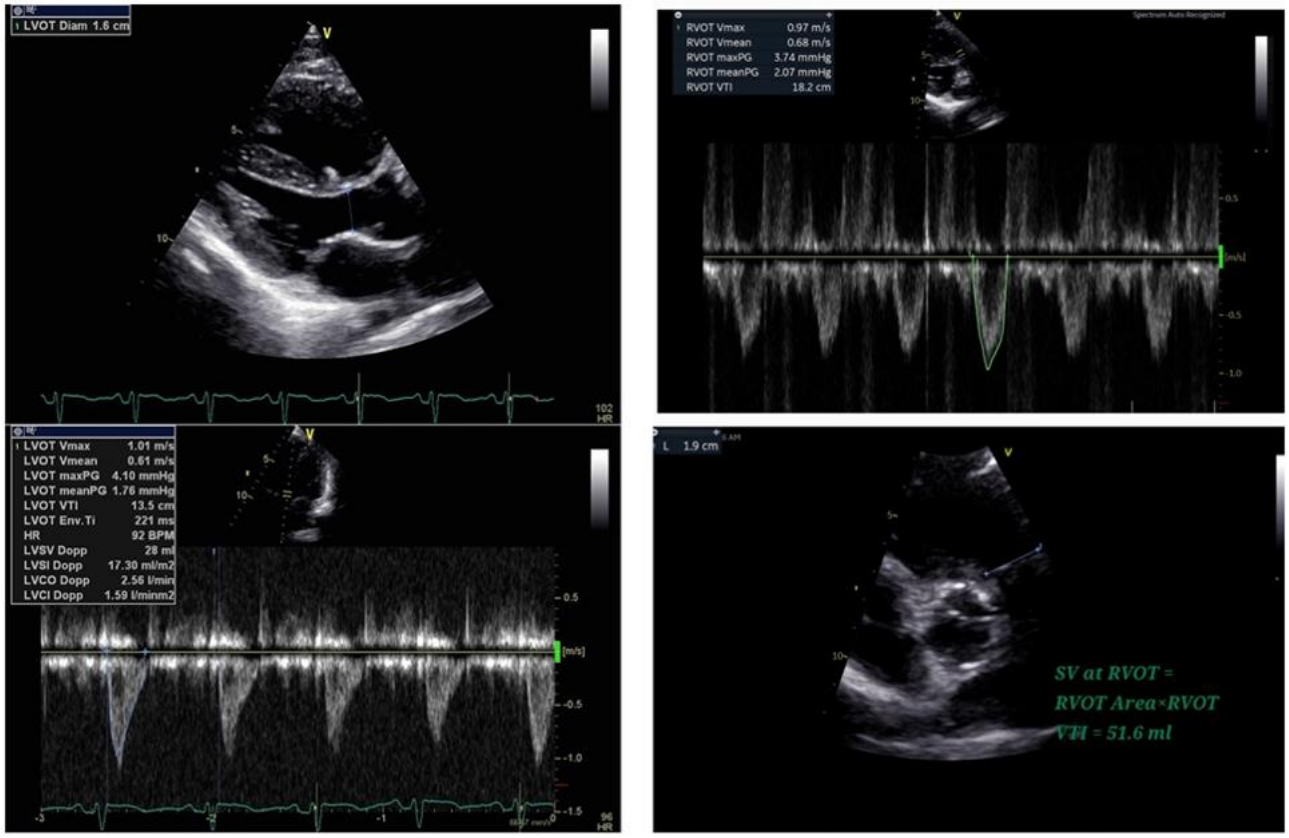

Figure 3. Echocardiography-derived pulmonary-to-systemic flow ratio calculation. The calculation was done from obtained measurements, as follows: (A) pulmonary blood flow (Qp)–measure cross-sectional area of right ventricular outflow tract (RVOT), measure velocity of blood flow across RVOT using Doppler echocardiography, & calculate (Qp) using Qp = CSA (RVOT) × VTI (RVOT); (B) systemic blood flow (Qs)–measure cross-sectional area of the left ventricular outflow tract (LVOT), measure the velocity of blood flow across the LVOT using Doppler echocardiography, calculate (Qs) using Qs = CSA (LVOT) × VTI (LVOT); (C) calculate the ratio Qp/Qs–once you have Qp and Qs values, calculate Qp/Qs = Pulmonary blood flow/systemic blood flow; & (D) interpret the results–A Qp/Qs ratio greater than 1 indicates increased pulmonary flow, while a ratio of less than 1 suggests decreased pulmonary flow relative to systemic flow (this approach provides a non-invasive approach to assess the pulmonary and systemic blood flow dynamics in patients) (CSA: Cross-sectional area in cm^2 & VTI: Velocity time interval in cm) (Calculate LVOT SV = LVOT area × LVOT VTI = 28 ml & qp/Qs = 51.6/28 = 1.84) (reprinted with permission of the patient)

pulmonary artery to the aorta, consistent with Eisenmenger physiology).

The right heart catheterization revealed a saturation of 73% in the descending aorta, while the upper limb saturation was 98%. Pulmonary artery systolic pressure was 120 mmHg, with a mean pressure of approximately 95 mmHg. The pulmonary capillary wedge pressure was 3 mmHg. The systemic systolic blood pressure was 120 mmHg. A nitric oxide challenge was used to assess responsiveness to pulmonary hypertension. It involves administering nitric oxide, a potent vasodilator, to determine whether it can improve the patient's pulmonary pressure. In this case, the nitric oxide challenge was "negative for reversibility." This means that the patient's pulmonary hypertension did not improve with nitric oxide, indicating that the condition was severe and irreversible. A right-sided saturation run was used to measure the oxygen saturation levels on the right side of the heart.

Normally, there is a step-up in oxygen saturation levels between the right atrium and the pulmonary arteries. This step-up was absent in this case, suggesting a right-to-left shunt, where deoxygenated blood bypassed the lungs and traveled directly from the right side of the heart to the left. The patient had severe and irreversible pulmonary hypertension with a large patent ductus arteriosus and a right-to-left shunt.

The patient was referred to the radiology department for CT angiography to evaluate the size of the defect and the presence of other cardiac and extracardiac anomalies. The CT angiography confirmed the presence of a patent ductus arteriosus measuring approximately 13 mm (**Video 8** - Computed tomography angiography 3D cine view showed the connection between the aorta and the pulmonary artery, **Figure 4**). The ascending aorta had a normal diameter (23 mm). The heart appeared mildly enlarged with a thickened RV wall suggestive of RV hypertrophy (**Figure 5**). The main pulmonary artery and right and left pulmonary arteries were mildly dilated, with peripheral pruning of the pulmonary vasculature. No other cardiac anomalies were observed. The aortic arch was left-sided with a normal branching pattern, origin, and course of the coronary arteries. The lung parenchyma appeared normal. A final diagnosis of APW with Eisenmenger syndrome was established.

The case was discussed with consultants in pulmonology, pediatric cardiology, and interventional cardiology. Surgical closure of APW is contraindicated because of irreversible pulmonary hypertension (Eisenmenger syndrome). Therefore, the patient is currently being treated with sildenafil. The importance of fluid and salt restrictions was discussed with the patient. She was instructed to limit fluid intake to less than 2

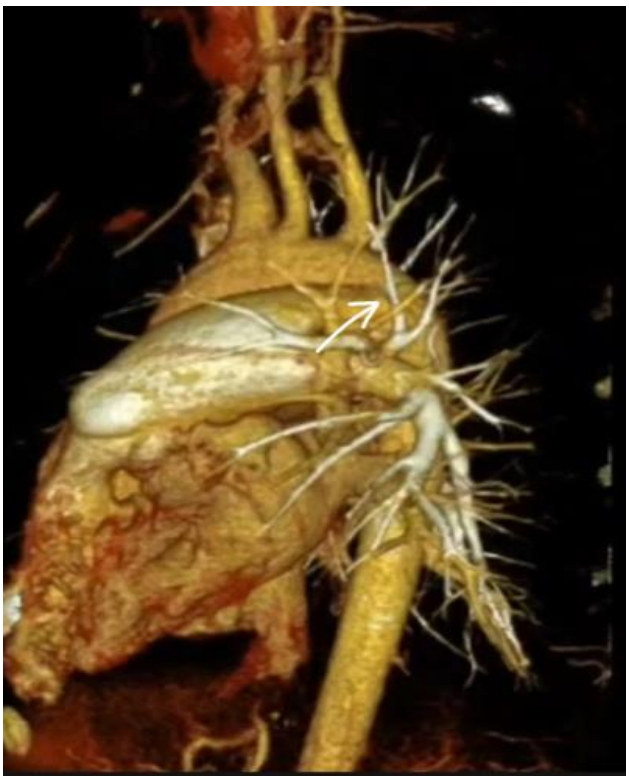

Figure 4. CT angiography 3D cine view showing the connection between the aorta and the pulmonary artery (reprinted with permission of the patient)

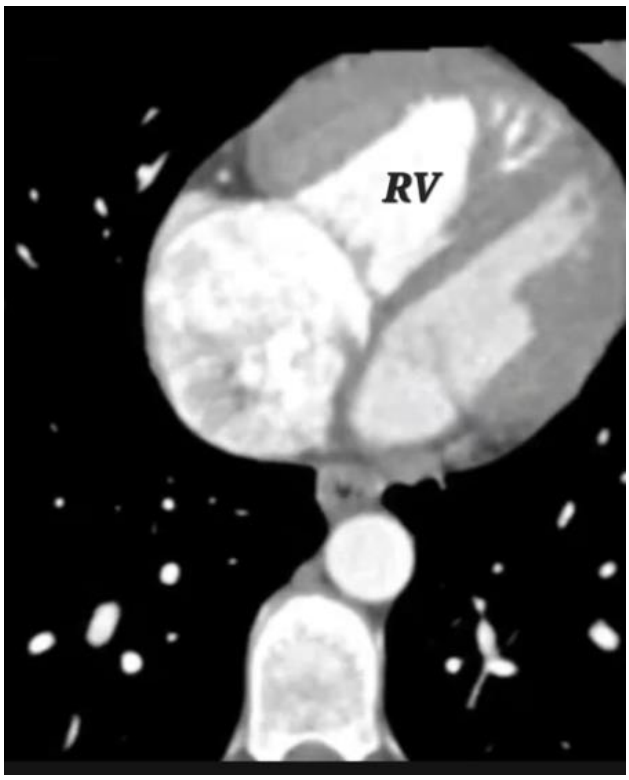

Figure 5. CT angiography showing mildly enlarged RV with thickened wall, suggestive of RV hypertrophy (reprinted with permission of the patient)

liters per day and salt intake to less than 1.5 grams per day. The patient will be asked to regularly measure her weight. This will allow the clinical team to titrate the patient's diuretic regimen accordingly, to help manage her fluid status and symptoms.

DISCUSSION

In this case report, we presented a patient with suspected APW, a rare congenital cardiac anomaly characterized by an abnormal communication between the aorta and pulmonary artery [1]. The diagnosis of this condition can be challenging due to its rarity and varied clinical presentations [2]. However, by employing a multimodal diagnostic approach, including echocardiography, right heart catheterization, and CT angiography, we were able to reach a definitive diagnosis. This condition has a poor prognosis if left untreated. However, few case reports have described adult survival in patients with untreated APW [9-13].

Echocardiography plays a crucial role in the initial evaluation of patients by providing valuable information about structural abnormalities and flow dynamics [13]. Although not commonly used, the Qp/Qs equation has proven to be a helpful tool for assessing the presence of extracardiac shunting. By calculating the ratio between pulmonary blood flow (Qp) and systemic blood flow (Qs), we were able to estimate the magnitude of the shunt and support the suspicion of APW, despite there being no clear evidence of shunts on either color flow Doppler or agitated saline contrast. Furthermore, right heart catheterization provided additional hemodynamic data, confirming the presence of a right-to-left shunt, and helping evaluate the severity of the defect. CT angiography complemented the findings by providing detailed anatomical visualization, ruling out any associated anomalies, and aiding in surgical planning.

The combination of the aforementioned diagnostic modalities allowed for comprehensive evaluation and accurate diagnosis of APW in our patient, highlighting the importance of a multidisciplinary approach in the assessment of complex congenital cardiac anomalies. The echocardiography-based Qp/Qs ratio proved to be a valuable diagnostic tool in this case, providing a noninvasive, cost-effective, and radiation-free method for assessing the presence of an extracardiac shunt and quantifying its severity. The ability of echocardiography to accurately determine the Qp/Qs ratio and thereby differentiate APW from other congenital heart defects with left-to-right shunts highlights its potential as a first-line diagnostic modality for this condition. In contrast, right heart catheterization, considered the gold standard for shunt quantification, is an invasive procedure with associated risks. Furthermore, CT, which is useful for anatomical delineation, does not provide hemodynamic data.

CONCLUSION

Here, we present a case of APW, which is a rare congenital cardiac defect. Using echocardiography, right heart catheterization, and CT angiography, we confirmed the diagnosis and assessed the severity of the anomaly. The echocardiography-derived Qp/Qs ratio, although not commonly used in clinical practice, is a valuable yet simple tool for evaluating extracardiac shunting. This case report underscores the utility of echocardiography-based Qp/Qs ratio assessment as a reliable and non-invasive tool for diagnosing APW, which can guide further management and treatment

decisions. The multidisciplinary approach demonstrated in this case serves as a valuable example for clinicians involved in evaluating and managing similar cases. Therefore, APW should be considered in the differential diagnosis of severe pulmonary hypertension, even in adult patients.

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Ethical statement: The authors stated that the study was approved by the Local Research Ethics Committee at Imam Abdulrahman Bin Faisal University on 15 July 2024 with approval code IRB-2024-03-529. Written informed consent was obtained from the patient.

Declaration of interest: No conflict of interest is declared by the authors.

Data sharing statement: Data supporting the findings and conclusions are available upon request from the corresponding author.

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