Journal of the Ceylon College of Physicians

Case Report

Aortopulmonary window presenting with severe pulmonary hypertension during pregnancy: A case report

Perera S L, Jeyapraniya A, Dissanayake D M C B, Perera H A A C N, De Silva A E, Abeysinghe S K, Pathirana W D A

Colombo South Teaching Hospital, Colombo, Sri Lanka

ARTICLE INFO

ABSTRACT

Aortopulmonary window defect is an extremely rare congenital cardiac defect characterised by a communication between the ascending aorta and the pulmonary trunk. A 37-year-old woman at 36 weeks of gestation of her second pregnancy presented with pre-term, pre-labour rupture of membranes. She was noted to have hypoxia on room air, and a bedside transthoracic echocardiogram revealed pulmonary hypertension. Urgent CT pulmonary angiogram showed no evidence of pulmonary embolism, but her HRCT Chest revealed consolidation in the upper zone of the left lung. She underwent emergency lower segment caesarean section under spinal anaesthesia. Following administration of the spinal anaesthetic, she developed hypotension and oxygen desaturation leading to emergency intubation and mechanical ventilation. A formal 2D transthoracic echocardiogram done afterwards showed an aortopulmonary window with a 3 cm defect complicated by severe pulmonary hypertension, together with right-to-left shunting, demonstrating Eisenmenger physiology. The hypoxia and hypotension were postulated to be due to spinal anaesthesia causing reduction of systemic vascular resistance and worsening of right-to-left shunting. She was managed with pulmonary vasodilators, diuretics and intravenous antibiotics. She was discharged after a lengthy ICU stay, with long-term home oxygen therapy.

*Corresponding Author: Perera S L
Email address: senuralp@gmail.com
https://orcid.org/0009-0002-8795-9769

DOI: https://doi.org/10.4038/jccp.v57i1.8194
Received: 15.04.2026; Accepted: 09.05.2026

Introduction

An aortopulmonary window (APW) is a rare cardiac malformation that accounts for 0.1–0.2% of all congenital heart defects. It is characterised by a communication between the ascending aorta and the pulmonary trunk in the presence of two separate semilunar valves.[1] The prognosis of APW without surgical correction is poor, with a mortality rate of 40% in the first year of life due to the rapid development of Eisenmenger syndrome and pulmonary artery hypertension (PAH).[2]

According to current clinical guidelines on the management of cardiovascular diseases during pregnancy, the World Health Organisation (WHO) and the European Society of Cardiology (ESC) classify PAH as a modified WHO Class IV risk.[3] Although this category carries a 40%–100% risk of major maternal complications, there are reported cases of such patients presenting during pregnancy and requiring a multidisciplinary approach.[4,5]

This case describes a pregnant woman presenting after pre-term rupture of membranes, who was incidentally found to have persistently low SpO_2 despite oxygen therapy. Subsequent evaluation led to the diagnosis of an APW defect and Eisenmenger syndrome. This case is notable because APW window is rarely diagnosed for the first time in adulthood, and even rarer in pregnancy.

Case presentation

A 37-year-old woman in her second singleton pregnancy with one previous normal vaginal delivery presented at 36 weeks of gestation with dribbling of clear fluid per vagina, suggestive of rupture of membranes. She did not have abdominal pain, contractions or per vaginal bleeding. She complained of a productive cough over the past 3 days but had no fever or shortness of breath. On further enquiry, she reported exertional dyspnoea (MRC grade 3) and poor exercise tolerance (NYHA class 3) for more than 2 years, for which she had not undergone formal evaluation. Her current pregnancy was uncomplicated except for foetal growth restriction.

She was investigated for a systolic murmur at age 5, but a transthoracic echocardiogram did not reveal any abnormality. Her first pregnancy 3 years ago, was uneventful, with delivery of an underweight baby (2.0 kg) via normal vaginal delivery. Her oxygen saturation had never been documented during antenatal check-ups in either pregnancy.

On examination, blood pressure was 130/80 mmHg and pulse rate was 76 bpm, a loud second heart sound was noted in the pulmonary area, and bilateral lung fields were clear. Grade 2 finger clubbing was also seen together with central cyanosis.

Her oxygen saturation was 76% on room air, which improved to 92% with oxygen at 15L/minute via a non-rebreather mask. An arterial blood gas analysis (ABG) performed on room air showed a PaO_2 of 55 mmHg and a $PaCO_2$ of 26.6 mmHg. ECG showed sinus rhythm, with no S1Q3T3 pattern. Bedside transthoracic echocardiography revealed a dilated right atrium and right ventricle with moderate tricuspid regurgitation (TRPG- 56 mmHg) and severe pulmonary hypertension.

Foetal status was assessed using cardiotocography (CTG) and ultrasound imaging, and the foetus was deemed to be in good health.

Urgent computed tomography pulmonary angiogram (CTPA) excluded pulmonary embolism. High-resolution computed tomography (HRCT) of the chest showed patchy consolidation predominantly in the upper lobes, suggestive of active infection. Full blood count, renal and liver function tests were normal. C- reactive protein (CRP) was elevated at 29.8 mg/dL. She had no proteinuria.

The patient was admitted to the intensive care unit (ICU) due to persistent maternal hypoxaemia in the setting of preterm prelabour rupture of membranes and concern for maternal deterioration. The patient underwent an emergency lower segment caesarean section under spinal anaesthesia. Approximately 5 minutes after administration of spinal anaesthesia, the patient developed profound hypoxia, with saturation falling from baseline to 58% despite high-flow oxygen supplementation. Her blood pressure also fell to 96/58 mmHg. The patient was electively intubated, which normalised oxygen saturation. A healthy, albeit underweight (1.8 kg), female baby was delivered and admitted to the neonatal ICU. The patient was mechanically ventilated overnight and extubated the following morning.

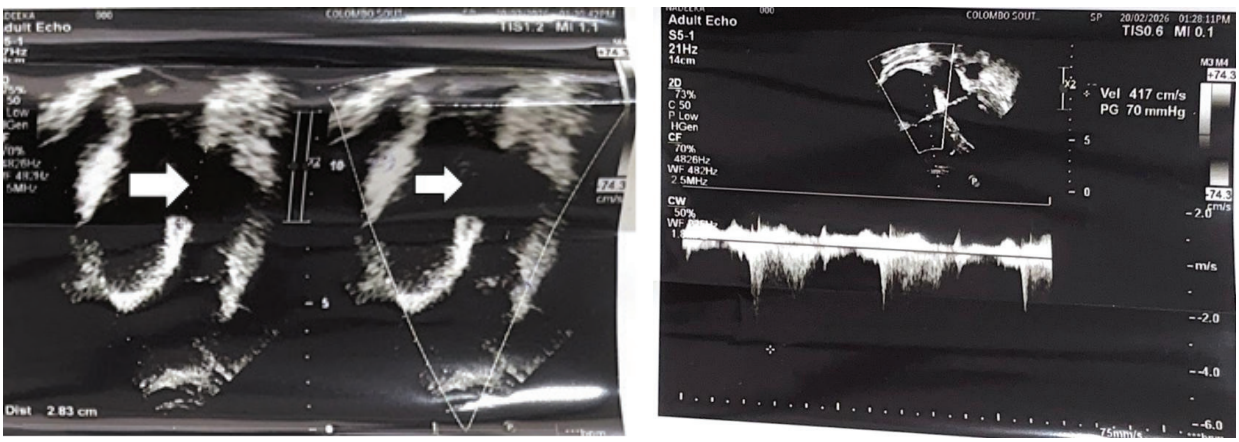

Figure 1. Trans-thoracic echocardiogram showing an aortopulmonary window 2 cm distal to the pulmonary valve annulus (arrow).

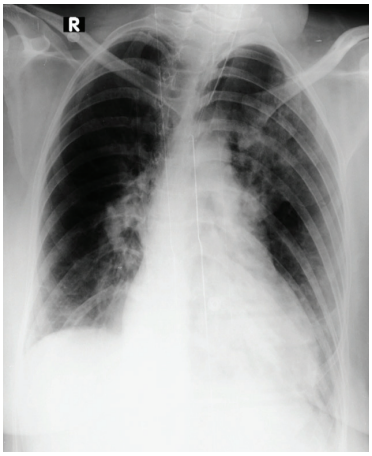

Figure B. Chest X-ray showing patchy consolidation of left upper and lower zones.

She required high-flow nasal oxygen at 60 L/minute to maintain her oxygen saturation at 88-92%.

Transthoracic 2D echo was done, which revealed a large aortopulmonary window (3 cm defect), located 2 cm distal to the pulmonary annulus, extending towards the right pulmonary artery, with right-to-left shunting – consistent with Eisenmenger physiology, together with severe pulmonary hypertension and severe tricuspid regurgitation (TRPG 80-85 mmHg). Formal right ventricular function assessment and cardiac magnetic resonance imaging (MRI) were not performed due to unavailability.

The patient was started on sildenafil 25 mg 8-hourly, oral furosemide 40 mg twice daily, and oral spironolactone 25 mg once daily. She was also treated with intravenous teicoplanin for 9 days and intravenous levofloxacin for 8 days as directed by sputum culture and antibiotic susceptibility testing. She improved steadily, and we were able to wean her oxygen over a period of 10 days to 2L/min. She was discharged after long-term home oxygen therapy was arranged, but her functional status remained NYHA class 3. Cardiac MRI was not available at the time of admission but was planned for the future.

Discussion

Congenital heart diseases occur in about 0.8% of all births. APW accounts for approximately 0.1-0.2% of all congenital heart disease cases, making it an exceedingly rare condition. The majority of patients with congenital heart disease survive into adulthood with advances in management.[6] Those missed in childhood may be unmasked by multiple physiological adaptations during pregnancy.

To support the developing foetus and meet increased maternal metabolic demands, the maternal cardiovascular system adapts by increasing cardiac output, elevating heart rate, expanding blood volume and decreasing systemic vascular resistance and blood pressure.[7] Due to these adaptations, congenital cardiac lesions with left-to-right shunting may worsen during pregnancy and may even lead to right-to-left shunting with Eisenmenger syndrome or congestive cardiac failure. Our patient, who was found to have an APW, had developed severe pulmonary hypertension and Eisenmenger syndrome by the time of diagnosis.

APWs are commonly classified according to the Mori classification into proximal (Type I), distal (Type II), and total (Type III) defects. The defect in our patient appeared consistent with a distal or extensive defect extending toward the right pulmonary artery. APW may also be associated with other cardiac disorders, such as atrial septal defects, ventricular septal defects, patent ductus arteriosus, and Tetralogy of Fallot, in approximately 50% of patients.[8]

In our patient, though a systolic murmur had been noted in childhood, the follow-up 2D echocardiogram was normal, and the APW had gone unnoticed. This shows how congenital cardiac defects can be missed in early life, especially when symptoms are minimal. The majority of APW cases present during infancy, and surgical correction is indicated for all, without which the prognosis would be poor.[9] Therefore, survival into adulthood without surgery is rare and is usually associated with severe complications.

This case is particularly unusual because the diagnosis remained unrecognised until late in the patient's second pregnancy, despite longstanding symptoms suggestive of pulmonary hypertension, including poor exercise tolerance and a loud pulmonary second heart sound, while low oxygen saturation and digital clubbing pointed towards a right-to-left shunt. Low birth weight babies in both pregnancies may reflect the presence of chronic maternal hypoxaemia leading to impaired placental perfusion.

Sui Mei et al described a clinical case of a patient with APW who successfully went through three pregnancies with vaginal delivery and remained relatively asymptomatic until 6th decade of life.[10] There are also a few other case reports of patients diagnosed during pregnancy or after delivery.[2]

Sympathetic blockage during anaesthesia causes an abrupt reduction in systemic vascular resistance. In patients with severe pulmonary hypertension, this exacerbates right-to-left shunting leading to sudden hypoxia and hypotension. This explains the rapid haemodynamic collapse and worsening hypoxaemia in our patient after spinal anaesthesia. The 2022 ESC guidelines for the diagnosis and treatment of pulmonary hypertension state that there is a significantly increased risk of peri-operative mortality in patients with pulmonary hypertension. It also affirms that general recommendations cannot be made regarding the mode and method of anaesthesia for these patients,

and that a multidisciplinary effort is required to determine the preferred management in the pre- and peri-operative periods.

Diagnosis of APW in adults can be challenging and may be misdiagnosed as more common causes of pulmonary hypertension, such as pulmonary embolism. In this case, the initial echocardiography demonstrated right heart dilatation with severe pulmonary hypertension, without clear identification of the underlying lesion. The diagnosis became evident only after detailed reassessment following perioperative deterioration. This case emphasises why adults with unexplained pulmonary hypertension should be thoroughly evaluated for uncommon congenital cardiac lesions.

When performed before the onset of permanent pulmonary vascular remodelling, surgical closure remains the mainstay of management and yields positive long-term outcomes. However, established pulmonary hypertension poses serious treatment challenges. Management of such cases focuses on pulmonary vasodilator therapy and long-term oxygen therapy. This patient was initiated on long-term home oxygen therapy due to persistent hypoxia.

Pulmonary hypertension during pregnancy poses an increased risk to the mother, with high morbidity and mortality rates; up to 6% in pulmonary hypertension and up to 15% in Eisenmenger syndrome, as shown in some studies.[11] Recognising subtle signs and symptoms is crucial in early diagnosis and better risk assessment.

Conclusion

APW is an extremely rare congenital heart disease that is seldom seen in pregnancy. This case emphasises the importance of early congenital heart disease detection to prevent its progression to Eisenmenger syndrome. This case also highlights the importance of maintaining a high index of suspicion in pregnant patients presenting with hypoxia and features of pulmonary hypertension. Early diagnosis and a multidisciplinary approach are essential to optimise maternal and foetal outcomes. Echocardiography remains the cornerstone for the diagnosis of pulmonary hypertension and congenital heart disease. However, in our relatively resource-limited setting, the absence of comprehensive right ventricular assessment facilities and cardiac magnetic resonance imaging (MRI) hindered the thorough evaluation of this patient. The case also highlighted the grave consequences that anaesthesia can have for these patients, and careful planning of the method and timing of anaesthesia is paramount when proceeding with surgery on patients with Eisenmenger physiology.

Author Declarations

Consent

Written informed consent for the publication of patient information and images was provided by the patient.

Competing interests

No potential conflicts of interest

Criteria for authorship

All the authors contributed to the diagnosis and management of this patient, and to the drafting, critical revision and final approval of the manuscript.

Funding

This study did not receive any specific grants.

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