CASE REPORT

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Navigating aortopulmonary window repair beyond infancy: a case report on perioperative management

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ABSTRACT

Background: Aortopulmonary window (APW) represents one of the most uncommon congenital heart anomalies. Prompt surgical correction is essential once the diagnosis is established, independent of patient age. Without intervention, there is a high risk of developing severe pulmonary hypertension (PH). Delayed diagnosis and treatment remain a major obstacle, particularly in regions with limited healthcare resources. This study aims to highlight the clinical presentation, surgical management, and outcomes of a late-diagnosed APW case, emphasizing the feasibility of successful intervention beyond infancy.

Case Presentation: A two-year-old boy who was diagnosed late with type 3 APW, presenting with repeated episodes of breathlessness and poor growth. Echocardiography revealed a significant defect in the aortopulmonary septum. Surgical closure was performed successfully using a patch through a transaortic approach. Intensive postoperative care—including administration of pulmonary vasodilators, inotropic support, and regular monitoring—was provided, and the patient did not experience any PH crises. Before discharge, echocardiography confirmed complete closure of the APW with no remaining defect.

Conclusion: APW is a rare congenital heart defect that carries a notable risk of pulmonary hypertension. Although timely intervention is ideal, this case demonstrates that surgical repair can still be effective in selected patients with late-presenting APW beyond infancy. Careful perioperative management is crucial to minimize the risk of PH crises and to achieve the best possible outcomes.

Keywords: Aortopulmonary Window, Case Report, Congenital Heart Disease, Perioperative Management, Pulmonary Hypertension.
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INTRODUCTION

Aortopulmonary window (APW) is a rare congenital cardiac anomaly, defined by an abnormal communication between the ascending aorta (AA) and the main pulmonary artery (MPA), while the two semilunar valves remain anatomically separate.[1-5] This defect creates a significant left-to-right shunt, increasing pulmonary blood flow and leading to early development of pulmonary hypertension (PH), which substantially elevates the risk of mortality. If left untreated, particularly in cases with significant defects, APW can result in a mortality rate of up to 40% within the first year of life.[4,5] For this reason, early diagnosis and immediate surgical intervention are essential to prevent progression toward irreversible pulmonary vascular disease and, ultimately, Eisenmenger syndrome.

Although APW is globally considered a rare anomaly, delayed diagnosis in developing countries is not uncommon due to limited access to healthcare facilities and diagnostic modalities.[2,3] Many patients are diagnosed beyond infancy, when symptoms have already progressed, posing additional challenges for surgical repair and perioperative management. The clinical presentation may vary significantly, ranging from mild respiratory symptoms to profound failure to thrive, cyanosis, or signs of heart failure. These manifestations often overlap with other congenital heart defects, which may lead to misdiagnosis or underrecognition of APW. In resource-limited settings, the lack of advanced imaging tools, such as echocardiography or cardiac catheterization, further hinders early identification and timely intervention.

In addition to variability in the timing of diagnosis, the anatomical diversity of APW also contributes to the complexity of clinical management. Based on the widely accepted Mori classification, APW is divided into three morphological types. Type 1 (proximal defect) is the most common and is located just above the semilunar valves. Type 2 (distal defect) occurs at the uppermost portion of the ascending aorta. Type 3, the rarest form, represents a total defect involving most of

the aortopulmonary septum.[3] Type 3 APW often includes direct communication between the aorta and the right pulmonary artery, making surgical access and cardioplegia delivery more challenging. Larger and more complex defects, particularly in Type 3, are associated with a higher risk of cardiopulmonary bypass-related complications and postoperative PH crisis.

Based on those mentioned above, this case study presents a case of a two-year-old male with a late-diagnosed Type 3 APW involving a large defect and potential cardioplegia leakage. The patient was referred to a tertiary hospital in the Special Region of Yogyakarta, Indonesia, and underwent successful surgical repair. This case highlights the critical importance of comprehensive preoperative evaluation, tailored intraoperative techniques, and meticulous postoperative care, including the use of pulmonary vasodilators and inotropic agents, to prevent complications such as the PH crisis. It demonstrates that with careful perioperative planning and monitoring, surgical repair can still achieve favorable outcomes in patients presenting beyond infancy, even in settings with limited resources.

CASE PRESENTATION

A two-year-old male presented with a history of recurrent shortness of breath and failure to thrive for the past year. The patient was suspected of having acyanotic congenital heart disease at another facility, but the exact cause remained unknown due to limited medical resources. Upon examination, the patient exhibited tachycardia, tachypnea, and reduced oxygen saturation. A continuous grade 3/6 murmur was auscultated over the left midclavicular line. Cyanosis was observed, triggered by crying and physical activity.

Initial chest X-ray showed cardiomegaly and plethoric lung fields (Figure 1). Transthoracic echocardiography revealed a significant 14-18 mm defect connecting MPA and AA, showing a predominant left-to-right shunt. Patent foramen ovale (PFO) was also seen with a left-to-right shunt. The left atrium (LA) and left ventricle (LV) were dilated, with LA to Aorta (LA/Ao) ratio of 1.2 and LV internal diameter end diastole (LVIDd) of 42 mm. The LV ejection fraction was 63%. Subsequent computed tomography (CT) images confirmed a significant aortopulmonary septal defect of 14.5 mm, suggesting type 3 APW (Figure 1). Cardiac catheterization was performed in this patient to evaluate the pulmonary and systemic haemodynamics better. The pulmonary vascular resistance index (PVRI) in room air was at 0.22 WU/m2, whereas post-oxygen administration was at 0.21 WU/m^2.

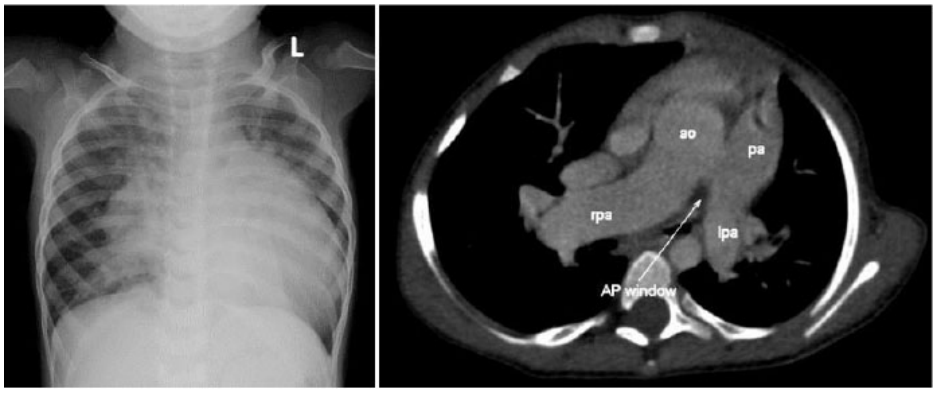

Figure 1. (Left) Anteroposterior view of chest X-ray showing cardiomegaly of all heart chambers and lung congestion; (Right) Cardiac computed tomography images showing a large aortopulmonary window connecting the aorta, MPA, and RPA.

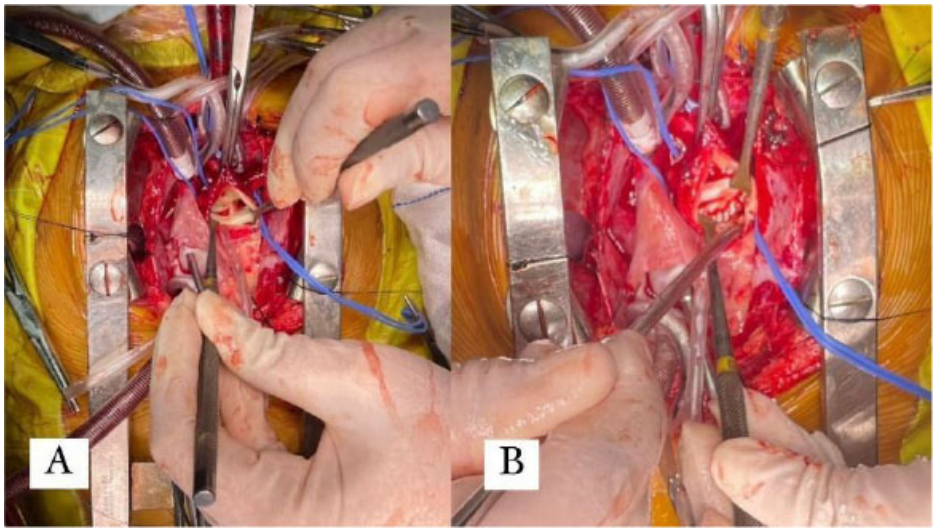

Figure 2. Operative picture showing pre- and post-closure of the aortopulmonary window. (A) Proximal and distal lumen of the right pulmonary artery were seen; (B) patch closure of the APW.

The patient was scheduled for an elective surgical repair via median sternotomy under moderately hypothermic cardiopulmonary bypass (CPB). During surgery, after administration of heparin, cannulas were inserted into the distal ascending aorta and the bicaval venous. Aortic root cannula was placed for cardioplegic solution (Custodiol) administration. The pulmonary arteries were snared to prevent the solution from entering the lungs. The patient was then cooled down to 30°C. Aortotomy was performed to access the APW. The RPA was seen as directly connected with the AA. The defect was closed using a polytetrafluoroethylene (PTFE) patch to create a new wall for the RPA (Figure 2). The PFO was intentionally left uncorrected to help reduce the right heart pressure due to the risk of developing a PH crisis. The CPB time was 87 minutes, with an aortic cross-clamp time of 54 minutes and an ischemic time of 50 minutes. Postoperative transesophageal echocardiography confirmed the successful closure of the APW without residual defects.

The patient received dobutamine (5 µg/

kg/min) and milrinone (0.5 µg/kg/min) to improve cardiac function intraoperatively. The patient was then closely monitored in the intensive care unit (ICU) for seven days. In the ICU, appropriate care was taken to prevent a PH crisis. The inotropic agent was gradually tapered off over 24 hours. Milrinone was steadily tapered off within seven days and continued with oral Sildenafil 0.5 mg/kg/8 hours. The patient did not develop a PH crisis in the postoperative period. Prior to discharge, detailed echocardiography showed a fully closed APW with no residual shunt and an intact PFO.

DISCUSSION

APW represents 0.1-0.2% of all congenital cardiac anomalies. In approximately 50% of cases, APW is associated with other cardiac defects, such as coarctation of the aorta, interrupted aortic arch, tetralogy of Fallot, or, as in the case of our patient, patent foramen ovale.[3,4] According to Mori and associates, APW can be classified into three morphological types. Type 1, the most common, is a proximal defect located a few millimeters above the semilunar valves. Type 2 is a distal defect located in the uppermost portion of the ascending aorta. Type 3, the rarest type, is a total defect involving the majority of the AA. Both types 2 and 3 are commonly associated with the connection between the RPA and the aorta.[3,6] This case represents the first encounter of APW at our center. The patient presented with a type 3 APW, which we found to have a large defect involving the aorta, pulmonary trunk, and RPA.

Symptoms typically develop very early, within 1 to 2 weeks, and often include poor feeding, recurrent respiratory infections, and failure to thrive.[3–5] In general, late presentation of APW beyond 1 year of age is uncommon. Detailed epidemiological data on this population are often not well-documented in large cohorts. However, in the developing world, this scenario is relatively common and is associated with poor access to care and misdiagnosis.[1,3,7] The challenge in late-presenting APW is ensuring comprehensive perioperative care to prevent PH crisis, beginning with assessment of operability. Clinical examination, chest radiology findings, echocardiographic evidence, and, if needed, cardiac catheterization conducted by surgeons, cardiologists, perinatal providers, and pediatricians can be used for operability assessment.[2,3,6] Transthoracic echocardiographic screening is recommended for patients with clinically confirmed congenital heart disease to assess the presence of PH early.[8] Echocardiographic parameters include left heart volume overload (LVIDd greater than +2 Z-score), left atrium dilatation (LA/Ao ratio > 1.4), flow acceleration across the mitral valve, and direction of shunt (left-to-right shunt indicated operability).[3] Cardiac catheterization is considered the gold standard for diagnosing PH with or without pulmonary vascular disease and is crucial for determining a patient's operability. A baseline PVRI of < 6 WU/m^2 and a resistance ratio < 0.3 are indicators of a favorable outcome following surgical repair, with no need for vasoreactivity testing.[3,8] Most of these operability criteria have been met by our patient. Although the patient has shown cyanosis on exertion and echocardiography indicated left heart volume overload, with LVIDd of the patient greater than +3 Z-score, there is no evidence of Eisenmenger syndrome, and the likelihood of a favorable outcome is still great at this stage. Once irreversible pulmonary hypertensive vascular disease develops, notably when the shunt reverses and becomes right-to-left, corrective surgery is no longer an option.[3,8]

Patients with a large APW often do not survive infancy due to irreversible PH and the potential development of Eisenmenger syndrome. Early intervention, ideally performed within the first few months of life, through a surgical or transcatheter approach, is crucial to prevent the onset of pulmonary vascular disease.[1,2,6,9] However, studies have shown that surgical repair in patients beyond 1 year of age can still yield satisfactory results.[2,3,5,7]

Surgical techniques include simple ligation, division and suturing, and patch repair, performed through various approaches, such as transpulmonary, transwindow, or transaortic. In addition, percutaneous transcatheter device closure for infants has also been proven to have short-term safety as well as efficacy. However, some evidence reported that the transpulmonary approach has been associated with higher re-intervention rates for pulmonary artery stenosis, making transaortic or transapical a more preferable approach.[1,10] Percutaneous transcatheter device closure is also related to device embolization, impaired coronary blood flow, aortic or pulmonary valvular defects, as well as residual shunt, encouraging reintervention.[1,9] Nevertheless, the definitive choice of technique is guided by the surgeon's judgement and is based on the patient's clinical condition. For example, ligation and division may be suited for a case of long, small-diameter APW without concurrent lesions that need to be repaired.[1]

In our case, repair of the APW was done using patch material via transaortic approach, chosen as it provides safer and easier access to both main arteries, considering the APW tends to be located posteriorly, while also offering better visualization of the coronary orifices. Several studies have supported the safety of this technique due to its low mortality and minimal aortic or pulmonary artery stenosis incidence within many years after surgery.[9] The transaortic approach is also suitable for avoiding coronary artery and aortic valve injury.[10] A significant challenge we encountered in this procedure was the risk of cardioplegic solution leaking into the pulmonary artery (PA) due to the large defect. However, no complications were experienced following the procedure.

In cases with immediate diagnosis and/or without other prominent abnormalities, there is usually low intraoperative mortality and satisfactory long-term prognosis.[11] However, late-presenting APW with pulmonary hypertensive disease can continue having high PA pressure postoperatively and even Eisenmenger syndrome. In late-repaired APW, cardiac failure or irreversible pulmonary hypertension may develop. Meanwhile, if uncorrected, APW median survival is 33 years based on the literature.[3,5,11]

Due to the possible occurrence of PH crisis periods in the postoperative period, it should be kept in mind that specified management and long-term follow-up for the patient are required to examine the PA pressure change, especially for complex AP windows, in order to examine residual

lesions and provide further management if needed. However, there has not been an ideal treatment guideline for APW early detection or long-term follow-up until today, considering the case and relevant research numbers are scarce. Nevertheless, nowadays late presentations decrease due to diagnostic imaging development and more profound knowledge of APW natural course, thus the quicker referral to surgical management before the patients develop pulmonary vascular obstructive disease. If the pulmonary reactivity remains preserved, late-presenting cases can be surgically repaired. Improvement in the perioperative and late management of those with increased pulmonary vascular resistance also supports the late presenters' survival.[3,5,11–13] In our case, the patient was operable with satisfactory outcomes despite being 2 years old, while some studies proved that surgical repair conducted early in infancy or below 1 year of age has more excellent outcomes.[3] This may be attributed to the absence of significant co-existing pathologies (e.g., Eisenmenger syndrome, increased pulmonary vascular resistance) as well as optimal perioperative management.

For the treatment of PH, administration of anti-pulmonary hypertensive agents, such as prostanoids, endothelin receptor antagonists, or phosphodiesterase inhibitors, is considered mandatory.[4,14] Milrinone and sildenafil, a phosphodiesterase inhibitor, are both potent pulmonary vasodilators and are extensively used as treatment for PH secondary to congenital heart disease. Milrinone also has inotropic properties that help enhance myocardial contractility.[14]

In addition, for patients with Eisenmenger syndrome who do not have the opportunity for surgical procedures, medical therapy is administered to relieve their symptoms, which include pulmonary hypertension, heart failure, as well as respiratory tract infections. The medical therapy recommended includes vasodilators and anti-pulmonary hypertensive agents, with heart-lung transplantation as another final treatment for APW with Eisenmenger syndrome. However, further research about the ideal treatment plan still needs to be established.[11]

Other management strategies, including inotropic support, mechanical ventilation, and parenteral nutrition, are tailored to clinical presentation.[14] Gowda et al. also highlight the importance of adequate pain control, nitroglycerine infusion, early initiation of sildenafil, diuretics, a low-salt diet, and early mobilization.[5] These measures have shown promising outcomes in late APW closure beyond 2 years, with no reported cases of postoperative PH crisis.[5]

CONCLUSION

APW is a rare congenital cardiac anomaly with a significant risk of PH. While early intervention is crucial, our findings suggest that surgical correction can still be successfully carried out in selected patients presenting with late APW beyond infancy. However, meticulous perioperative management is essential to reduce the risk of PH crisis and ensure optimal results.

ETHICAL APPROVAL

This study has been approved by the Medical and Health Research Ethics Committee of the Faculty of Medicine, Public Health and Nursing, Universitas Gadjah Mada, with the approval number of KE/FK/1417/EC/2024.

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CONFLICT OF INTEREST

The authors have fulfilled the ICMJE uniform disclosure form. The authors have no conflict of interest to declare.

AUTHOR CONTRIBUTION

MA, YK, S, HA, YAA, IA, and T were involved in conceptualizing, designing, providing supplementary data, conducting, and supervising the manuscript writing. In addition, AR contributed to editing and submitting the manuscript. All authors prepared the manuscript and agreed to this final version of the manuscript to be submitted to this journal.

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