

A Rare Case of Adult Aortopulmonary Window Combined with Anomalous Origin of the Right Pulmonary Artery from the Aorta Leading to Eisenmenger Syndrome

Journal of International Medical Research

49(1) 1–6

© The Author(s) 2021

Article reuse guidelines:

sagepub.com/journals-permissions

DOI: 10.1177/0300060520984656

journals.sagepub.com/home/imr



Mingyue Cui¹ , Binfeng Xia¹, Heru Wang¹,
Haihui Liu² and Xia Yin¹

Abstract

Aortopulmonary window is a rare congenital heart disease that can increase pulmonary vascular resistance, exacerbate left-to-right shunt and lead to heart failure and respiratory tract infections. Most patients die during childhood. We report a 53-year-old male patient with a large aortopulmonary window combined with anomalous origin of the right pulmonary artery from the aorta, with Eisenmenger syndrome and without surgery.

Keywords

Congenital heart disease, aortopulmonary window, Eisenmenger syndrome, anomalous origin of the right pulmonary artery from the aorta, pulmonary vascular resistance, surgery

Date received: 14 May 2020; accepted: 2 December 2020

Introduction

Aortopulmonary window (APW) is a rare congenital heart disease, accounting for 0.2% to 0.6% of all congenital heart diseases.¹ APW is an abnormal communication between the ascending aorta and the main pulmonary artery caused by embryonic hypoplasia. Most patients with APW

¹Center for Cardiovascular Medicine, The First Hospital of Jilin University, Changchun, China

²Department of Cardiac Ultrasound, The First Hospital of Jilin University, Changchun, China

Corresponding author:

Xia Yin, Center for Cardiovascular Medicine, The First Hospital of Jilin University, 71 Xinmin Street, Changchun, Jilin province 130021, China.

Email: xiayintong@aliyun.com



develop congestive heart failure in infancy owing to left-to-right shunt.² The survival rate of patients with large untreated APW is very low, with a mortality rate of 40% in the first year;³ few patients with APW survive to adulthood. We report a 53-year-old male patient with a large APW combined with anomalous origin of the right pulmonary artery from the aorta (AORPA) and Eisenmenger syndrome without surgery.

Case report

A 53-year-old male patient was admitted to our hospital because of chest tightness and shortness of breath for more than 40 years and intermittent haemoptysis for 10 years. Haemoptysis occurred frequently in autumn and winter, with a volume of approximately 20 to 30 mL per episode. The patient underwent medical evaluation when he was approximately 11 years old. Congenital heart disease was considered, but the exact cause of his chest symptoms was unknown because of limited medical resources at that time. Therefore, during the course of the disease, he had no systematic diagnosis and received no treatment. He had no history of hypertension, diabetes, coronary heart disease, smoking or related family diseases.

Physical examination revealed the following: blood pressure: 133/78 mmHg, heart rate: 82 bpm, respiratory rate: 16 bpm and oxygen saturation: 94%. The cardiac borders were normal in chest radiographs, and no dry or moist rales were heard on auscultation. The P2 heart sound was loud, and a grade 3/6 systolic murmur was ausculted over the left parasternal area. Routine blood test results were as follows: white blood cell count: $7.31 \times 10^9/L$, red blood cell count: $6.21 \times 10^{12}/L$, haemoglobin 190 g/L, haematocrit 0.562 L/L and B-type natriuretic peptide concentration: 43.70 ng/L (reference range, 0–100 ng/L). Liver and renal function, urinalysis and

other laboratory test results were normal. Electrocardiography suggested sinus rhythm and bilateral atrial enlargement. Transthoracic echocardiography showed right ventricular enlargement and dysfunction with an estimated systolic pulmonary artery pressure of 111 mmHg. The size of the left ventricular end-diameter was normal, and the left ventricular ejection fraction was 58%. Echocardiography also showed an interruption between the ascending aorta and the main pulmonary artery bifurcation measuring approximately 38 mm in width on the parasternal long-axis view (Figure 1). The right pulmonary artery was widened and shifted to the right, overriding the APW. The left pulmonary artery originated from the main pulmonary artery, and colour Doppler flow imaging showed bidirectional shunting. The echocardiographic diagnosis was congenital heart disease, APW, right pulmonary artery originating from the beginning of the APW and pulmonary hypertension. Lung computed tomography (CT) and second-phase enhancement with contrast showed visible communication between the aorta and pulmonary artery. The left pulmonary artery originated from the pulmonary trunk, and the right pulmonary artery originated from the aorta (Figure 2). The final diagnosis was APW combined with AORPA, and Eisenmenger syndrome was confirmed, which precluded surgery. We recommended that the patient receive medicine to reduce pulmonary arterial pressure.

Discussion

APW, also known as aortopulmonary septal defect, is a rare congenital heart disease. APW is defined as direct communication between the ascending aorta and the main pulmonary artery owing to incomplete separation of the arterial trunk into the aorta and pulmonary artery during embryogenesis.⁴ APW can appear as an

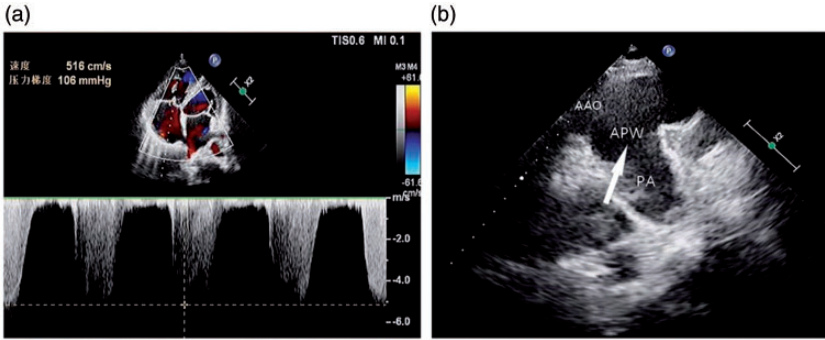


Figure 1. Transthoracic echocardiography. (a) Tricuspid regurgitation spectrum with Doppler ultrasonography revealed the following: tricuspid regurgitation velocity: 5.16 m and pressure difference: 106 mmHg (1 mmHg = 0.133 kpa). Using these values to estimate pulmonary artery pressure, owing to right-sided heart enlargement, 15 mmHg (right atrial pressure) should be added in accordance with the pressure difference of 106 mmHg, and the systolic pulmonary artery pressure should be 111 mmHg. (b) Parasternal long axis view demonstrating a defect (white arrow) between the ascending aorta and main pulmonary artery. AAO = ascending aorta, PA = pulmonary artery.

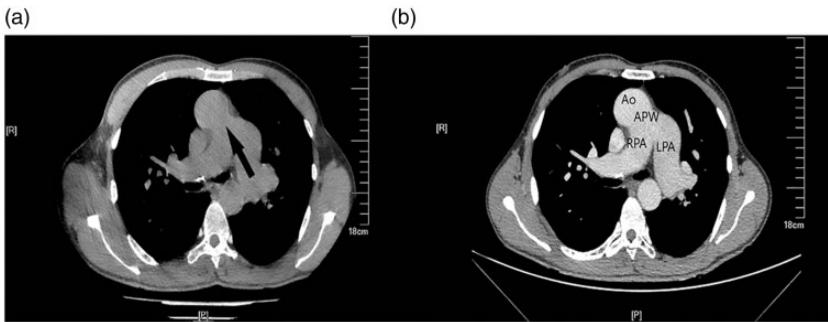


Figure 2. Lung computed tomography (CT) images. (a) Lung CT image showing an aortopulmonary septal defect (black arrow). (b) Second-phase enhancement with contrast during lung CT showing visible communication between the aorta and pulmonary artery, and that the right pulmonary artery originates from the ascending aorta.

Ao = Aorta, RPA = right pulmonary artery, LPA = left pulmonary artery.

isolated lesion, or may be combined with other cardiac abnormalities, such as ventricular septal defect, atrial septal defect, patent ductus arteriosus and tetralogy of Fallot.⁵ Our patient was diagnosed with APW combined with AORPA. According to the classification proposed by Mori et al., APW is divided into three types. Type I indicates an APW located in the pulmonary artery bifurcation before the

proximal defect, and the left pulmonary artery and main pulmonary artery maintain a normal relationship. Type II indicates an APW located at the bifurcation of the left and right pulmonary arteries, with a distal defect in an oblique direction. Type II APW accounts for only 10% of all APW cases. Berry et al.⁶ modified the Mori classification according to the origin of the right pulmonary artery. In the modified

classification, Type II is further divided into Type IIA and Type IIB. Type IIA indicates that the right pulmonary artery originates from the main pulmonary artery. Type IIB represents AORPA, often combined with aortic arch hypoplasia and patent ductus arteriosus; our patient had type IIB. Type III is a compound type with proximal and distal defects and complete absence of the aortopulmonary septum (Figure 3).

The pathophysiological changes in APW are complicated. Through the defect, a large amount of high-pressure aortic blood ejects directly into the proximal end of the pulmonary artery, which first leads to pulmonary hypertension and pulmonary congestion. Second, the

continuous impact of high-pressure blood flow can induce pulmonary arteriolar spasm, intimal thickening and luminal thinning, which increases resistance. These pathophysiological changes can lead to pulmonary hypertension. Excessive pulmonary artery pressure can increase right heart load, which can induce right ventricular enlargement and dilatation and lead to right heart failure. Large APW defects and left ventricular volume overload cause left ventricular enlargement and dilatation, which can induce left heart failure. Severe cases can quickly develop complete heart failure after birth.

Echocardiography is considered the first-line imaging tool to diagnose simple and complex congenital heart diseases; however,

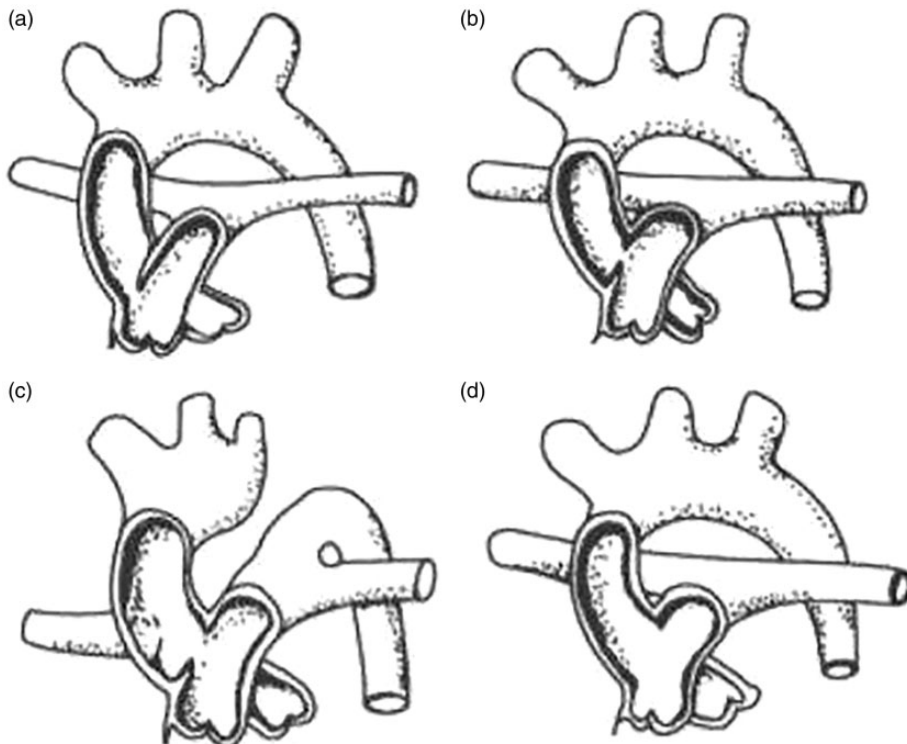


Figure 3. APW classification diagram. (A) Type I, proximal defect. (B) Type IIA, distal defect with the right pulmonary originating from the main pulmonary artery. (C) Type IIB, distal defect, anomalous origin of the right pulmonary from the aorta. (D) Type III, total defect (these images originate from: https://ssl.bdstatic.com/70cFuXSh_QIYnxGkpoWkIHF6hhy/it/u=1611928785,3978765144&fm=26&gp=0.jpg. Accessed: 15 August 2020).

with severe pulmonary hypertension, a diagnosis of APW may be missed.⁴ To some extent, an ultrasonographic diagnosis may be difficult and should be based on careful evaluation of the two-dimensional images projected on the parasternum and sternum. Our patient was underdiagnosed for a long time because of limited medical resources. His right ventricle was hypertrophic, and the left ventricle was normal; ejection fraction was in the normal range with bidirectional shunting. The reason for our patient's survival may be related to the normal left ventricular structure and function.

APW can induce early severe pulmonary hypertension, congestive heart failure, respiratory tract infections and mortality. Thus, once the diagnosis is confirmed, surgery should be performed immediately, regardless of the patient's age. Irreversible pulmonary hypertension with right-to-left shunting is the only contraindication for surgery.⁷ In cases with early diagnosis and no other abnormalities, the intraoperative mortality is low, and the long-term prognosis is satisfactory; however, surgery is not an option for patients with Eisenmenger syndrome.^{7,8} For patients who have lost the opportunity for surgery, medical treatment is the only option to relieve symptoms, such as pulmonary hypertension, heart failure and respiratory tract infections. The treatment strategies for patients with Eisenmenger syndrome are stated in major clinical guidelines.⁹ Studies have shown that vasodilators may improve symptoms, and the main medications are endothelin receptor antagonists (e.g., bosentan), phosphodiesterase inhibitors (e.g., sildenafil) and prostacyclins (e.g., iloprost). Bosentan can improve patients' exercise endurance and haemodynamics. Studies have shown that sildenafil is also beneficial regarding mortality.^{10,11} Heart–lung transplantation may be another treatment, according to a case reported by Bobylev et al.,¹² which was a patient who

underwent heart–lung transplantation for APW combined with Eisenmenger syndrome as the final treatment. The hope is that the ideal treatment plan can be determined with future research. Until more is known, early detection and diagnosis of APW is critical.

The literature suggests that the median survival for uncorrected APW is 33 years.¹³ The longest reported survival for untreated APW with Eisenmenger syndrome is 60 years,¹⁴ which our patient has exceeded, and who continues to be followed-up.

Ethics statement

All procedures followed were in accordance with the CARE guidelines.¹⁵ Verbal consent for participation in the study or use of medical data was obtained from the patient. We have de-identified the details such that the identity of the patient may not be ascertained in any way.

Ethics approval is not required in our institution for case reports.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Funding

This work was supported by the National Natural Science Foundation of China (81670353).

ORCID iD

Mingyue Cui  <https://orcid.org/0000-0002-3261-2437>

References

1. Kutsche LM and Van Mierop LH. Anatomy and pathogenesis of aorticopulmonary septal defect. *Am J Cardiol* 1987; 59: 443–447.
2. Aggarwal SK, Mishra J, Sai V, et al. Aortopulmonary window in adults: diagnosis and treatment of late-presenting patients. *Congenit Heart Dis* 2008; 3: 341–346.

3. El Dick J, El-Rassi I, Tayeh C, et al. Aortopulmonary window in adults: a rare entity leading to Eisenmenger syndrome. *Echocardiography* 2019; 36: 1173–1178.
4. Barnes ME, Mitchell ME and Tweddell JS. Aortopulmonary window. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 2011; 14: 67–74.
5. Mori K, Ando M, Takao A, et al. Distal type of aortopulmonary window. Report of 4 cases. *Br Heart J* 1978; 40: 681–689.
6. Berry TE, Bharati S, Muster AJ, et al. Distal aortopulmonary septal defect, aortic origin of the right pulmonary artery, intact ventricular septum, patent ductus arteriosus and hypoplasia of the aortic isthmus: a newly recognized syndrome. *Am J Cardiol* 1982; 49: 108–116.
7. Gowda D, Gajjar T, Rao JN, et al. Surgical management of aortopulmonary window: 24 years of experience and lessons learned. *Interact Cardiovasc Thorac Surg* 2017; 25: 302–309.
8. Talwar S, Agarwal P, Choudhary SK, et al. Aortopulmonary window: morphology, diagnosis, and long-term results. *J Card Surg* 2017; 32: 138–144.
9. Galiè N, Humbert M, Vachiery JL, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS): Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT). *Eur Heart J* 2016; 37: 67–119.
10. Basit H, Wallen TJ and Sergent BN. *Eisenmenger Syndrome. StatPearls [Internet]*. Treasure Island (FL): StatPearls Publishing, 2020.
11. Hartwig BJ and Schultze B. Review of evidence for bosentan therapy for treatment of Eisenmenger syndrome. *J Am Assoc Nurse Pract* 2019; 31: 72–77.
12. Bobylev D, Sommer W, Avsar M, et al. Aortopulmonary window: a rare untreated adult case. *Heart Lung Circ* 2014; 23: e235–e236.
13. Kose M, Ucar S, Emet S, et al. A case of aortopulmonary window: asymptomatic until the first pregnancy. *Case Rep Cardiol* 2015; 2015: 935253.
14. Su-Mei AK and Ju-Le T. Large unrepaired aortopulmonary window—survival into the seventh decade. *Echocardiography* 2007; 24: 71–73.
15. Gagnier JJ, Kienle G, Altman DG, et al. The CARE Guidelines: Consensus-based Clinical Case Reporting Guideline Development. *Glob Adv Health Med* 2013; 2: 38–43.