



An Overview of Aortopulmonary Window

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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ABSTRACT

Aortopulmonary (AP) window is exactly a rare congenital anomaly that represents approximately 0.2 and 0.5% of all congenital heart abnormalities. It consists of communication between the aorta and the pulmonary artery or its branches. Although closely related to AP window, the pulmonary artery origin from the ascending aorta (also termed "hemitruncus") is classified as a separate defect. AP window was first described by Elliotson in 1830 in an autopsy study. In 1948, Robert Gross successfully ligated an AP window in a patient undergoing a thoracotomy for closure of a patent ductus arteriosus (PDA). In 1957, Cooley and associates described the first successful repair of AP window using cardiopulmonary bypass. Aortopulmonary window occurs due to the abnormal development of the heart's major blood vessels during early fetal growth. In most cases, this heart defect occurs by chance, with no clear reason. This condition can occur on its own or with other heart defects such as: 1. Tetralogy of Fallot, 2. Pulmonary atresia, 3. Truncus arteriosus, 4. Atrial septal defect, 5. Patent ductus arteriosus, 6. Interrupted aortic arch. Treatment for aortopulmonary window involves surgery to close the hole between the aorta and the pulmonary artery with a patch or device. This surgery is normally done as soon as possible after the diagnosis is made, usually when the child is a newborn. Associated lesions are usually repaired during the same surgery. Associated lesions are usually repaired during the same surgery. More complex repairs and myocardial protection strategies are required in patients with associated lesions, increasing the morbidity and mortality associated with the operation.

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1. INTRODUCTION

The aortopulmonary window (AP window) is a rare cardiac anomaly between the main artery that is the aorta which taking blood from the heart to the body and the blood from the heart to the lung (pulmonary artery). The condition is congenital, which means it is present at birth [1].

The normal circulation of blood flows through the lung, where it collects oxygenation, through the pulmonary artery. Afterward, the blood returns back to the heart and it goes to the aorta and rest of the body [2].

Babies with a cardiac anomaly like an aortopulmonary window that having a hole between the aorta and the pulmonary artery. Because of this defect, blood from the aorta pours into the pulmonary artery, and due to the defect much of the blood goes to the lungs. This results in pulmonary hypertension (high blood pressure in the lungs) and congestive heart failure [3]. The aortopulmonary window is an uncommon anomaly caused by the breakdown of two contrasts between the aortas and the pulmonary artery that separate the truncus arteriosus from fusion. In one-third to one-half of instances, the aortopulmonary window can develop as a single lesion or in combination with other cardiac defects [4]. Arch abnormalities, especially interrupted aortic arch and aortic coarctation, are the most prevalent related lesions. In today's world, early mortality after the closure of a simple aortopulmonary window is nearly negligible and is dependent on the existence of other lesions, particularly an interrupted aortic arch. The long-term outcome should be good. Pulmonary artery stenosis and persistent aortopulmonary septal abnormalities are among the early morbidities. Long-term monitoring is recommended to check for recurring abnormalities like branch pulmonary artery stenosis and arch blockage [5].

2. CLASSIFICATION

According to the occurrence of Aortopulmonary window may be categorized into 3 types:

- Type I:- It is also called as proximal defects which occur in the proximal part of the aortopulmonary septum; This defect is more proximally located between the origin of the

main pulmonary artery and the ascending aorta immediately above the sinus of Valsalva (due to deficient separation of the aortopulmonary trunk during development) with little inferior rim separating the AP window from the semilunar valves. These defects tend to be large, round or oval shaped, and are more often seen.

- Type II:- it is also called as distal defects which occur in the distal part of the aortopulmonary septum adjacent to the right pulmonary artery; This defect is more distal, between the ascending aorta and the origin of the right pulmonary artery (due to abnormal migration of the 6th aortic arch during development). These defects are more rare and tend to be smaller in size.
- Type III:- It is a combination of types I and II. A large defect combining Types I and II. Consists of extension of the defect into the right pulmonary artery with anomalous origin of the right pulmonary artery from the ascending aorta (secondary to unequal separation of the aorto-pulmonary trunk during development) with a well-formed inferior rim but little superior rim and involves the majority of the ascending aorta. This type is more frequently linked with other cardiac anomalies [6,7].

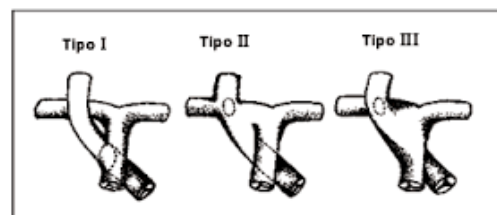


Fig. 1. Schematic drawing of the anatomical types of aortopulmonary window

3. EPIDEMIOLOGY

Aortopulmonary window is a very rare defect accounting for 0.2% to 0.6% of all congenital malformation [8,9]. There is a male preponderance. No specific genetic abnormalities have correlations with AP window, although Berry syndrome, a combination of AP window, interrupted aortic arch, and right pulmonary artery originating from the aorta is a defined entity [10].

4. ETIOLOGY

The aortopulmonary window is quite uncommon. It causes around 1% of all congenital cardiac abnormalities. The faulty/abnormal development of the heart's coronary arteries during early fetal growth causes the aortopulmonary window. The majority of the time, this cardiac abnormality arises accidentally and without any apparent cause. This condition can develop by itself or in conjunction with other cardiac conditions such as:

1. Tetralogy of Fallot
2. Pulmonary atresia
3. Truncus arteriosus
4. Atrial septal defect
5. Patent ductus arteriosus
6. Interrupted aortic arch [11]

5. CLINICAL PRESENTATION

The clinical features of APW are not specific, but majority of patients have the manifestations of a large left to right shunt. Patients with small defects may be asymptomatic. Patients with large APW usually have symptoms of pulmonary hypertension and congestive heart failure (tachypnea, diaphoresis, failure to thrive, and recurrent respiratory difficulty) in the first weeks of life. Severe pulmonary vascular hypertension can occur in the first months of life.

- Tachypnea
- Diaphoresis
- Poor feeding
- Poor growth
- Delayed growth
- Rapid heartbeat
- Respiratory infections
- Minimal cyanosis present
- Heart failure symptoms generally emerge in early childhood,
- The cardiac murmur is usually systolic with a mid-diastolic rumble as a result of increased blood flow over the mitral valve [12].

6. PATHOPHYSIOLOGY

AP window pathophysiology is equivalent to that of a ventricular septal defect, PDA, or truncus arteriosus with a left-to-right shunting. The amount of shunting from left to right is proportional to the extent of the defect and the pulmonary vascular resistance. Patients with small defects can be completely asymptomatic. With large defects, and as the pulmonary

vascular resistance decreases in the first weeks of life, symptoms of congestive heart failure develop rapidly, and irreversible pulmonary vascular disease can occur as early as during the first 12 months of life. If untreated, 40 percent of patients will die of intractable heart failure during the first year of life, and survivors will succumb to the sequelae of congestive heart failure and severe irreversible pulmonary vascular disease during childhood

Normally, blood flows from the pulmonary artery into the lungs, where it gathers oxygenated blood, and then through the pulmonary veins back to the heart.



With AP window blood from aorta flows into pulmonary artery



Large amount of blood flow to pulmonary artery results in pulmonary hypertension and cardiac failure

7. DIAGNOSIS

Some of the diagnostic procedure that reveals a heart defect are as follows:

7.1 Echocardiography

Echocardiography is the gold-standard diagnostic tool for aorto-pulmonary window. As patients with AP window are likely to present earlier in life due to symptomatic CHF from significant left-to-right shunting or cyanosis due to the higher risk of associated CHD lesions, a high-frequency probe is recommended in these neonates for evaluation.

Important views for transthoracic echocardiography include:

- Parasternal short-axis 2D and color Doppler: This view allows visualization of both MPA and aorta to evaluate for communications between these structures. Color Doppler is important to confirm that a communication truly exists and there is not just artifactual drop-out in great arterial vessel walls. This view can also help to identify extent of involvement of the RPA. Size measurements of the AP window defect can be made in this view.

- Subcostal short-axis and long-axis views: It is important to evaluate the great arteries and AP window defect in the subcostal plane. These views not only allow for evaluation of the location of the AP window defect, but also: distance from both semilunar valves, extent/presence of the inferior and superior rims, extent of involvement of the ascending aorta, and if involvement of the right pulmonary artery exists, all of which will affect surgical management. In this view, the great arteries association, ventricular septum, pulmonary valve morphology, and right ventricular outflow tract can also be evaluated since AP window can be seen with associated cardiac lesions (ie - VSD, TOF, TGA).
- Suprasternal notch views: 2D and color sweeps from this view between the aortic arch and main pulmonary artery not only allow for evaluation of shunting across the AP window between the great arteries, but allows for thorough evaluation of the arch due to incidence of association of AP window with arch anomalies (type A interrupted aortic arch or coarctation) [7].
- **Radiographic-** A radiographic image displays heart enlargement and prominence of the pulmonary artery and intrapulmonary vasculature by using x-rays, gamma rays, or other kinds of ionizing radiation to see the inside object.
- **Magnetic resonance angiography (MRA)** - The cardiac defect can also be seen with an MRI [13].
- **Cardiac catheterization-** Cardiac catheterization is a technique used to diagnose and treat cardiovascular diseases. A catheter is placed into an artery or vein in the groin, neck, or arm and threaded through your blood vessels to the heart during cardiac catheterization [14].
- **Selective aortography-** The injection of contrast medium into the ascending aorta reveals the lesion, and catheter manipulation from the major pulmonary artery straight to the ascending aorta is also a diagnostic technique for cardiac abnormality [15].

8. MANAGEMENT

8.1 Initial management

- Medical therapy is focused on preoperative stabilization.
- It will retrogress spontaneously during follow up in majority of cases.
- The only effective therapy for aortopulmonary window is surgical repair (APW).
- However there are reports of transcatheter occlusion of simple APW.

8.2 First line treatment

- Intravenous prostaglandins (e.g., alprostadil) may be required in persons with an interrupted aortic arch to maintain the ductus arteriosus open and enable blood flow to the lower half of the body.
- The increased pulmonary blood flow may be exacerbated by the accompanying pulmonary arterial vasodilation.
- Digoxin and furosemide are widely used to treat heart failure and volume overload associated by this lesion.
- Inotropic drugs (for example, dopamine and dobutamine) may also be an effective therapy for babies with severe heart failure and low cardiac output due to myocardial dysfunction.

9. SURGERY

- Aortopulmonary window is generally treated with surgery. Surgery should be done as soon as feasible after initial stabilization and correction of acidosis.
- Surgery is performed with the use of cardiopulmonary bypass. The aortopulmonary window, the major pulmonary artery, or the anterior portion of the aorta can all be incised.
- Associated lesions are generally treated at the same time as the primary lesion. In patients with associated lesions, more complicated repairs and myocardial protection methods are necessary, increasing the morbidity and mortality associated with the surgery.

9.1 Preoperative care

- Preoperative treatment focuses on correcting acidosis and stabilizing the infant. Digoxin, furosemide, and inotropes

are used to treat symptoms of congestive heart failure as needed.

- Elective intubation is also possible, and pulmonary blood flow can be controlled by adjusting the inspired proportions of oxygen and carbon dioxide.
- Echocardiography is used to characterize anatomy and evaluate ventricular function. In the case of complicated lesions or when the coronary arteries cannot be seen clearly, cardiac catheterization and/or computed tomography (CT) scanning may be necessary.
- Patients over the age of 6 months must have a cardiovascular catheterization to rule out irreversible pulmonary hypertension.

9.2 Post-operative care

- Milrinone, epinephrine, dopamine, or other medications may be used to provide inotropic support in the early postoperative period.
- Depending on the patient's preoperative health, length of time on cardiopulmonary bypass, and duration of hypothermic circulatory arrest, these can typically be weaned off over the following several hours or days.
- Individuals with postoperative pulmonary hypertension or pulmonary hypoxia crisis may necessitate therapy for Older patients.
- Inhaled oxygen at high doses is still one of the most powerful pulmonary vasodilators.
- Hypertensive crises can also be avoided by deep sedation and paralysis. Additional sedation should be given for endotracheal suctioning and other operations if paralysis is not utilized.
- Intubated patients may benefit from inhaled nitric oxide for the treatment of pulmonary hypertension.
- Patients may need to continue taking digitalis and furosemide, which are usually stopped during outpatient therapy [16].

10. CONCLUSION

Aortopulmonary Window is a rare abnormal congenital heart defect. The aortopulmonary window is produced by a failure to fuse the conotruncal ridges which divide the truncus arteriosus into the aorta and the pulmonary artery. An aortopulmonary window may appear as an isolated injury or other heart anomalies. Arch abnormalities, especially interrupted aortic

arch and coarctation of the aorta, are the most prevalent related lesions. Antenatal diagnosis is rare. In the current era, early mortality following repair of simple aortopulmonary window approaches zero and depends on the presence of associated lesions, especially interrupted aortic arch. The long-term outcome should be excellent. Early morbidity includes pulmonary artery stenosis and residual aortopulmonary septal defects. Long-term monitoring is needed to check for recurrent lesions such as development of branch pulmonary artery stenosis and arch blockage. Surgery remains the treatment of choice in adults. However, percutaneous techniques are also feasible. In our case, the Amplatzer Muscular Septal Occluder device (Abbott) allowed us to achieve a complete closure without residual shunt and vascular wall injuries during follow-up.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Barnes ME, Mitchell ME, Tweddell JS. Aortopulmonary window. In Seminars in

- Thoracic and Cardiovascular Surgery: Pediatric Cardiac Surgery Annual 2011; 14(1):67-74. WB Saunders.
2. Miller C. 14.3 Heart. Human Biology; 2020.
 3. Carini L, Cantarano MS, Lombardi M, Congiu S, de Siena P, Bizzarri F. A rare case of an unexpected aortopulmonary window.
 4. Doty JR, Doty DB. Cardiac Surgery E-Book: Operative and Evolving Technique. Elsevier Health Sciences; 2012.
 5. Naimo PS, Yong MS, d'Udekem Y, Brizard CP, Kelly A, Weintraub R, Konstantinov IE. Outcomes of aortopulmonary window repair in children: 33 years of experience. The Annals of thoracic surgery. 2014; 98(5):1674-9.
 6. Mori K, Ando M, Takao A, Ishikawa S, Imai Y. Distal type of aortopulmonary window. Report of 4 cases. Heart. 1978;40(6):681-9.
 7. Griselli M, McGuirk SP, Winlaw DS, Stümper O, De Giovanni JV, Miller P, Dhillon R, Wright JG, Barron DJ, Brawn WJ. The influence of pulmonary artery morphology on the results of operations for major aortopulmonary collateral arteries and complex congenital heart defects. The Journal of thoracic and cardiovascular surgery. 2004;127(1):251-8.
 8. Barnes ME, Mitchell ME, Tweddell JS. Aortopulmonary window. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2011;14(1):67-74. [PubMed]
 9. Kutsche LM, Van Mierop LH. Anatomy and pathogenesis of aorticopulmonary septal defect. Am J Cardiol. 1987;59(5):443-7. [PubMed]
 10. Murayama H, Watanabe T, Yasuda K, Kobayashi A. Mirror image Berry syndrome: a case report of complex aortopulmonary malformation in viscerotriangular situs inversus. Minerva Chir 2007; 62(2):129-32. [PubMed]
 11. Black J, Hawks J, Medical Surgical nursing clinical management for positive outcomes 8th edition, Missouri, Vol.II, Elsevier publishers
 12. MORROW AG, GREENFIELD LJ, Braunwald E. Congenital aortopulmonary septal defect: clinical and hemodynamic findings, surgical technic, and results of operative correction. Circulation. 1962; 25(3):463-76.
 13. Kervancioglu S, Soydinc S, Davutoglu V, Kervancioglu R, Sirikci A, Bayram M. Aortopulmonary window: a rare adult case demonstration by echocardiography, MRI, CE-MRA and angiography. Cardiovascular and interventional radiology. 2004;27(2): 175-8.
 14. Chen FL, Ge S. Aortopulmonary window demonstrated by real-time 3-dimensional echocardiography and color Doppler. Journal of the American Society of Echocardiography. 2007;20(11):1314-e7.
 15. Sondhi S, Negi PC, Sharma R, Mehta A. Case Report: Aortopulmonary window with pulmonary atresia with ventricular septal defect with D-transposition of great arteries: extremely rare anomaly. BMJ Case Reports. 2018;2018.
 16. Backer CL, Mavroudis C. Surgical management of aortopulmonary window: a 40-year experience. European journal of cardio-thoracic surgery. 2002; 21(5):773-9.

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