

## CASE REPORT

# LARGE AORTO-PULMONARY WINDOW ASSOCIATED WITH SEVERE PULMONARY HYPERTENSION: A CASE REPORT ✦

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## BACKGROUND

Aortopulmonary window (APW) is a rare congenital cardiac abnormality, accounting for 0.2% to 0.6% of patients [1]. Hence the reported surgical experience is minimal. It is a communication between pulmonary artery (PA) and the ascending aorta (Ao), in the presence of two separate semilunar valves. Heart failure and irreversible pulmonary hypertension are the inevitable outcomes of an untreated aortopulmonary window. Therefore, surgical closure is the first-line curative choice. We present a case of a young boy in his second decade of life with severe signs and symptoms.

## CASE PRESENTATION

A 15-year-old male patient, laborer, from Baluchistan, (a high-altitude province of Pakistan), presented in National Institute of Cardio Vascular Diseases (N. I. C. V. D), Karachi, Pakistan with complaints of high-grade fever for 1-week and palpitations for 12 d. He had a past history of shortness of breath (SOB) since he was six years old; which has now progressed to New York Heart Association (NYHA) II.

On examination, the patient was anemic, cyanosed with raised jugular venous pressure. However, there were no signs of jaundice, clubbing or edema. On precordial examination, apex beat was localized at the 6th intercostal space shifted to mid-axillary line. S1 was normal but S2 was loud with a prominent pulmonary component. Grade IV/VI continuous murmur was heard in the left upper sternal border. Electrocardiography was compatible with right ventricular pressure overload. Chest X-ray showed cardiomegaly with prominent pulmonary artery and bilateral lung congestion. Echocardiography showed enlarged and hypertrophied right ventricle associated with dysfunction dilated pulmonary artery (size = 44 mm). A large aortopulmonary window was seen (size = 26 mm),

with severe pulmonary hypertension of 102 mmHg. Diagnosis of an aortopulmonary window was confirmed on cardiac catheterization. To further determine the type, cardiac CT was performed. This showed type I APW with severe pulmonary hypertension and large patent ductus arteriosus. Irreversible pulmonary hypertension develops quickly if the lesion is left untreated; hence early surgical repair was advised.

The patient was operated upon by a median sternotomy and pericardiotomy was performed. Patent ductus arteriosus was identified but couldn't be approached due to an aneurysmally dilated main pulmonary artery. Aorta and vena cavae were canalized and the patient was put on partial extracorporeal circulation. After achieving a temperature of 34 °C, flow was reduced to one-fourth and aorta was cross-clamped. The main pulmonary artery was opened through longitudinal aortotomy. Patent ductus arteriosus was occluded with fringes, then ligated and transfixed. The aortopulmonary window was closed using autologous pericardium. With the closure of aorta and MPA, the patient was re-warmed and cross-clamping was removed.

Patient symptoms remarkably improved with a reduction in pulmonary hypertension (Pulmonary artery pressure=40 mmHg) after surgery and at 6 w follow-up. Follow-up echocardiography showed complete closure of the aortopulmonary window. Pharmacological therapy was no longer required in the patient.

A verbal consent from the patient and a written informed consent was obtained from patients' guardian (patient was underage) for publication of this case report and any accompanying images.

## DISCUSSION

The incomplete fusion of conotruncal ridges of the heart during fifth to eight weeks of gestation results in APW.

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Consequently, there is a failure of septation of truncus arteriosus, causing abnormal communication between the right and left region [2]

APW is classified by two means. Considering Mori's classification [3] a simple defect between ascending Ao and the PA is type I (70%). Type II (25%) comprises of distal involvement of Ao with any of the PA branch. Type III (5%) is simply the abnormal origin of PA from Ao [4]. The second classification divides the defect into being either simple (isolated or in association with an ASD or VSD) or complex (association with IAA, TGA or TOF) [5]. When there are associated malformations, the clinical manifestations can be atypical [6]. The incidence of associated lesions ranges between 47% and 77% [4,7,8]

The most commonly reported associated defects are interrupted aortic arch (15-20%), PDA (11%), coarctation, coronary artery anomalies (8%) and ventricular septal defect (8%). Other associated malformations include right aortic arch, tetralogy of Fallot, subvalvular aortic stenosis, bicuspid aortic valve, ventriculoarterial discordance, double aortic arch and tricuspid atresia.

Presently echocardiography is being considered for confirmation of APW. Cardiac catheterization is important in complicated or doubtful cases. It also provides useful information concerning complexly associated lesions, as well

as revealing elevated pulmonary vascular resistance its reversibility. Therefore, it is essential to be performed in all patients with a late diagnosis or with suspected/severe PH. Our patient had type I defect which was confirmed on CT.

Surgical correction of APW is indicated, as soon as it is diagnosed, because spontaneous closure does not occur [7] The first reported surgical closure of the defect was by Robert E Gross in 1946, which involved direct ligation without CPB. However, innovation and evolution in the field of congenital cardiac surgery presently offer trans-aortic single patch technique.

In-hospital mortality from this pathology is higher in cases associated cardiac malformations and pulmonary hypertension. Postoperative pulmonary hypertensive crises have been reported, so patients should be carefully monitored [8]

In our patient, APW window was closed using trans-aortic single patch technique. The patient recovered successfully, despite having a severe PH. Recently, cases of percutaneous closure of APW have been reported. These should be limited to specific circumstances; the lesion should be type I (proximal) and small (3-4 mm), and there should be no anomalous origin of the coronary arteries. In our patient, trans-catheter was not feasible due to its size and the monetary constraint.



Fig. 1: Pre-op parasternal short axis view showing large ap window (12CM) between aorta and MPA

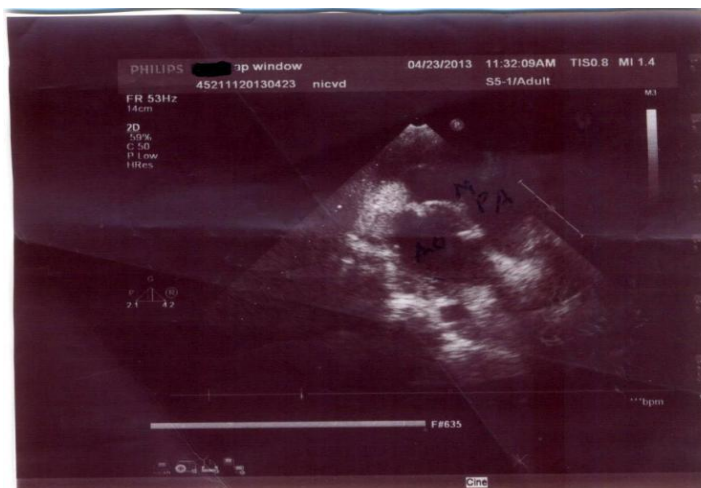
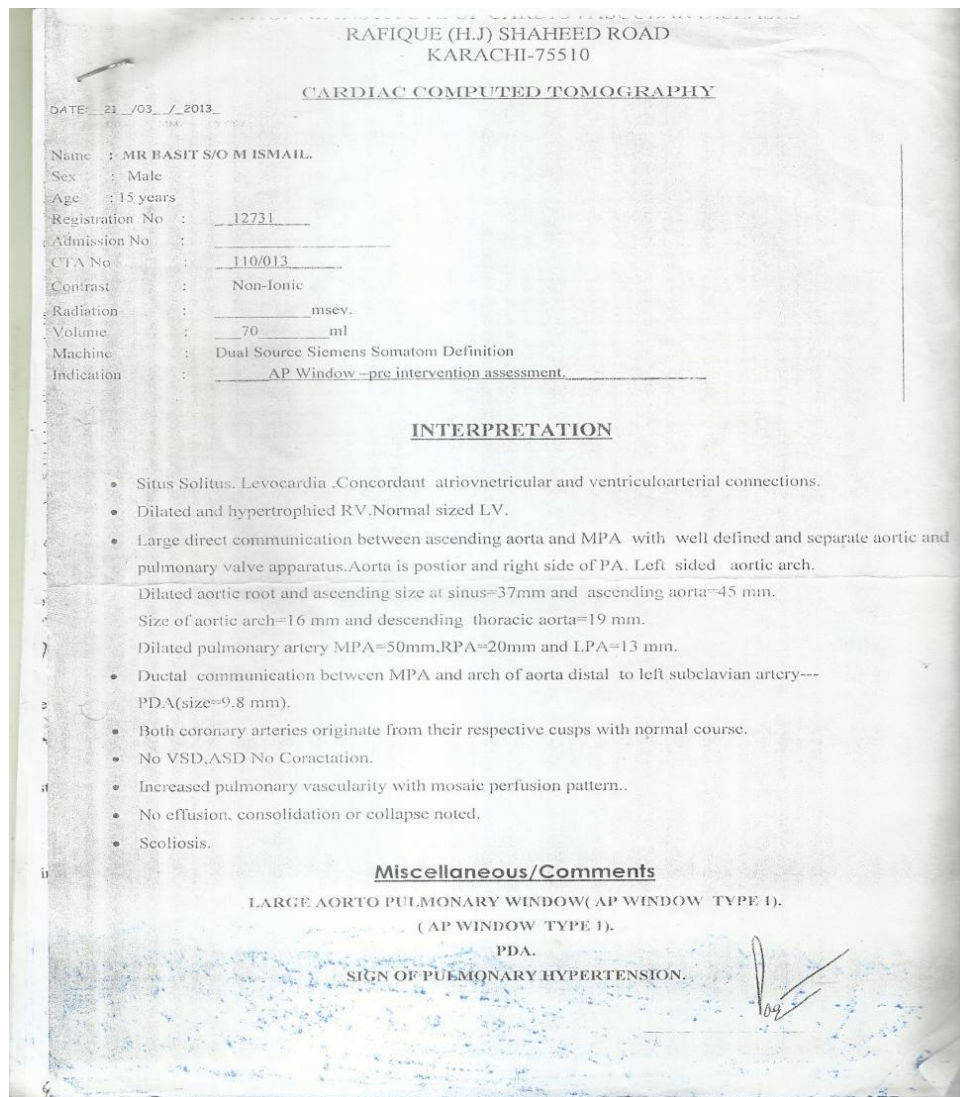


Fig. 2: Post-op parasternal short axis view shows a pericardial patch



**Fig. 3: Chest X-ray showing cardiomegaly and bilateral lung congestion**



**Fig. 4: Showing cardiac CT**



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Requested: **ECHOCARDIOGRAPHIC REPORT**

Reg. No. 1099000 Echo No. 1166/23 Tape No. 329

21/1/13 Age: \_\_\_\_\_ Sex: \_\_\_\_\_ Height: \_\_\_\_\_ Weight: \_\_\_\_\_

cal Diagnosis: 1) Sullama Habits

ode/2 D

Measurements in mm Figures in bracket indicate normal values in mm

Left Ventricle	Systolic <u>22</u>	EPSS (<7)
	Diastolic (<55) <u>42</u>	E.F. (>55%) <u>76%</u>
	Septal Thickness (11) <u>10</u>	Post. Wall Thickness (<11) <u>9</u>
Atrium (<40)	<u>21</u>	Aorta (<40) <u>28</u>
Aortic Valve		Aortic Valve
Right Ventricle (<25)	<u>25</u>	Pulmonary Artery

Structural Interpretation:

enlarged and hypertrophied RV is dysfunction  
 large gap seen b/w pulmonary artery and aorta (size = 26mm)  
 Dilated PA (size = 44mm)

Color Doppler:

Normal tricuspid valve  
 Normal size LV is normal function  
 Normal mitral and aortic valves

Pressure Gradient (mm Hg)

EDP = 32 mmHg

Estimated RV/MPA Pressure (mm Hg)

85 mmHg

Final Echo Dx:

- Large AP window is bidirectional shunt
- Enlarged and hypertrophied RV is dysfunction
- Severe PAH

H.H. 2

Fig. 5: Showing echocardiography

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 KARACHI-75510

**CARDIAC CATHETERIZATION**

DATE: 13-02-13 HOSPITAL NO.: 1099000 CATH NO.: 808/13  
 NAME: MR. BASIT AGE: 15Y HEIGHT (CM.): 158cm WEIGHT (KG.): 32kg  
 SEDATION: \_\_\_\_\_ TIME: \_\_\_\_\_ BSA (M<sup>2</sup>): 1.18 m<sup>2</sup>  
 VESSEL: \_\_\_\_\_ METHOD: RHC  
 HEMOGLOBIN G%: 10.5 HEMATOCRIT: \_\_\_\_\_ O<sub>2</sub> CAPACITY (VOL%): 14.28

PO <sub>2</sub> TIME	SITE	SATURATIONS		PRESSURES (mm Hg)				BPM	REMARKS
		SAT%	VOL%	SYSTOLIC	DIASTOLIC	EDP	MEAN		
				Pre O <sub>2</sub>					
	RA	67		a = 20, v = 3, m = 11					
	SVC	66.3		<del>120/13</del>					
	RV	86.7		120/13					
	LPA	85.2		110/60				76	
	LV	97.7		120/12				80	
	Ao	97.1		120/60				80	
				Post O <sub>2</sub>					
	SVC	55.9							
	LPA	91.8		110/57				74	
	PA	66.6		a = 17, v = 2, m = 10					
	Ao	99.7		110/60				76	
	RV	92.7		110/60					

O<sub>2</sub> CONSUMPTION (ML/MIN.): 180 SYSTEMIC FLOW (L): 4.09 (2.55)  
 PVR 7.77 (4.19) SVR 16.87 (22.9) SYSTEMIC INDEX \_\_\_\_\_  
 QP/QS 2.07 (5.29) SHUNT - (L-R) 4.76 (12.4) PULM FLOW (L): 8.49 (15.25)  
 SHUNT - (R-L) 0.36 (0.06) PULM INDEX \_\_\_\_\_

FINAL DIAGNOSIS  
 1. Large AP WINDOW  
 2. Severe PAH  
 3. Show Reversibility for AP WINDOW CLOSURE

RECOMMENDATION  
 CLOSURE OF AP WINDOW

PHYSICIAN: \_\_\_\_\_

Fig. 6: Showing cardiac catheterization report

**CONCLUSION**

APW is a congenital anomaly. It should be surgically corrected earlier, to prevent the development of obstructive pulmonary vascular disease. In our experience, short term surgical results are excellent.

**CONSENT**

Consent has been taken from the guardian since the patient was under-age. Hence, patient has given their consent for the case report to be published.

**ABBREVIATIONS**

- APW: aorto=pulmonary window
- PH: Pulmonary hypertension
- PA: pulmonary artery
- PDA: Patent Ductus arteriosus
- MPA: main pulmonary artery
- CPB: cardio-pulmonary by-pass

**CONFLICT OF INTERESTS**

The authors declare that they have no competing interests.

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**AUTHORS CONTRIBUTIONS**

SH reported the case. AWK and FM drafted the manuscript. LM and SH proof read the manuscript. LM and AWK collaborated in reporting and constructing the structure of study. All authors read and approved the final manuscript.

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