

Aortopulmonary Window in Association with Subpulmonic Ventricular Septal Defect and Non Compaction of Left Ventricle

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ABSTRACT

Aortopulmonary window (APW) is a rare congenital cardiac anomaly, which clinically mimics patent ductus arteriosus (PDA). A girl who was diagnosed to have congenital heart disease (CHD) at the age of four years presented to us at the age of 10 years with exertional dyspnoea and fatigue. She was clinically diagnosed as PDA with pulmonary artery hypertension (PAH). Two-dimensional and colour Doppler echocardiography showed a type II aortopulmonary window and a large inlet type of subpulmonic ventricular septal defect (VSD) with non-compaction of left ventricle. The diagnosis was confirmed by cardiac catheterization and angiography. This is an interesting case of type II APW with rare association that can be missed clinically, but can be diagnosed by meticulous echocardiogram. We herein report a rare case of aortopulmonary window associated with a subpulmonic ventricular septal defect and left ventricle non compaction (LVNC).

Key words: Aortopulmonary window, Patent ductus arteriosus, 2D Echocardiography, Left ventricular noncompaction, Subpulmonic ventricular septal defect.

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BACKGROUND

Aortopulmonary window (APW) is a very uncommon congenital anomaly which results from failure of septation of the aortopulmonary trunk during the fifth to eight weeks of development. APW has been classified as simple or complex types. Clinically, Aortopulmonary window mimics patent ductus arteriosus (PDA) or Ventricular Septal Defect (VSD) with pulmonary artery hypertension (PAH). The magnitude of the shunt is mainly related to the size of the defect and pulmonary vascular resistance. The left ventricle non compaction (LVNC) is usually an isolated condition. Association with subpulmonic Ventricular Septal Defect and Aortopulmonary window is extremely rare.

CASE PRESENTATION

A 10-year-old girl diagnosed to have CHD at the age of four years, was not taken to tertiary centre for surgery. Now she presented with history of exertional dyspnoea and fatigue. On examination she had BP 120/30 mm of Hg. Hyperdynamic precordium with apical impulse shifted down and out. She had a palpable loud P2. There was a continuous murmur with very short diastolic component. A chest roentgenogram showed cardiomegaly with pulmonary plethora. The electrocardiogram showed normal sinus rhythm, biventricular hypertrophy, and P pulmonale. The detailed Two-dimensional echocardiography with colour Doppler in apical four chamber view showed left atrium and left ventricle (LV) dilatation, non-compaction of LV (Figure 1A and 1B). Parasternal short axis view showed a large subpulmonic Ventricular Septal Defect (Figure 2) measuring 1.2 cms with bidirectional shunt and modified parasternal short axis view showed type II APW (Figure 3A, and 3B). The persistent left superior vena cava draining to the right atrium via an enlarged coronary sinus. Non compaction of left ventricle is seen in modified short axis. Doppler gradient across the septal defect was 25 mm Hg and tricuspid regurgitant jet gradient was 70 mmHg. The cardiac catheterization was performed to assess the PAH and know the pulmonary vascular resistance. Hemodynamic and oximetric measurements revealed a markedly elevated pulmonary arterial pressure (102/55 mmHg; mean 75 mmHg) with a

large pulmonary flow ($Q_p/Q_s=2.93$). Pulmonary vascular resistance was 0.5 (R_p/R_s). Right heart catheterization revealed left-to-right shunting at ventricular and arterial levels. Oxygen inhalation revealed reversible pulmonary hypertension. On aortography, the ascending aorta, the main pulmonary trunk, and the pulmonary arteries were simultaneously opacified, which confirmed the diagnosis of an Aortopulmonary window (Figure 4). The coronary arteries were normal. Through right femoral approach, arteriovenous loop was formed and a muscular ventricular septal occluder was negotiated from pulmonary arterial side. Device closure was attempted twice but failed. Patient was advised high risk surgical closure.

DISCUSSION

Aortopulmonary window results from failure of septation of the aortopulmonary trunk during the fifth to eight weeks of development. The more posterior the septum forms the closer the window encroaches on the pulmonary artery (type II) until it finally passes the right pulmonary artery, giving the appearance of emerging from the descending aorta (type III).¹⁻³ Aortopulmonary window has also been defined as simple and complex types. Simple Aortopulmonary window is associated with hemodynamically insignificant anomalies such as a patent ductus arteriosus, atrial septal defect, or a patent foramen ovale, whereas the complex type is associated with more complex anomalies such as an interrupted aortic arch, transposition of great arteries, Fallot's tetralogy, or anomalous coronary arteries.^{2,3}

Aortopulmonary window is a very uncommon congenital anomaly and there are very few surgical series of more than 20 patients in the literature.⁴⁻⁶ The review of literature showed only 30 cases of APW that were accompanied by a VSD, and of them, only four reports consisted of three or more cases.⁵⁻⁸ The other case was a 15-year-old boy who had a simple type I Aortopulmonary window, whose defect was successfully repaired using total circulatory arrest in 1988.

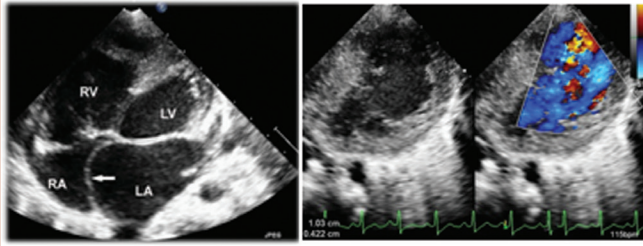


Figure 1A : Transthoracic echo in apical four chamber view shows non compaction of left ventricle with interatrial septum bulging towards right atrium. B: noncompacted myocardium is (1 cm) more than two times the compacted myocardium (0.4 cm). The color Doppler shows filling the trabeculae.

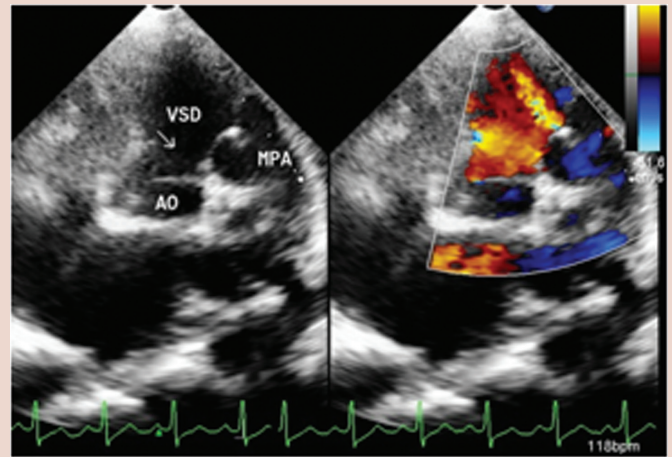


Figure 2: Transthoracic echo in short axis view with color Doppler shows large subpulmonic Ventricular Septal Defect [arrow], with bidirectional shunt and pulmonary regurgitation [arrow].

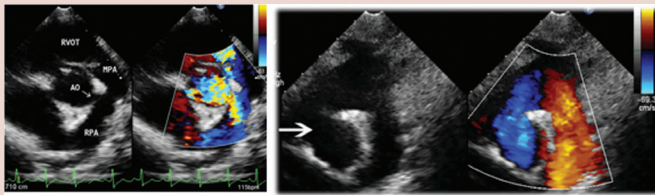


Figure 3A: Modified parasternal short axis view with color Doppler shows Aortopulmonary window (arrow) with left to right shunt. B: Suprasternal view with color Doppler shows Aortopulmonary window (arrow).

Clinically, APW presents with similar symptoms to those of a patent ductus arteriosus and a VSD with pulmonary hypertension. The magnitude of the shunt is mainly related to the size of the defect and pulmonary vascular resistance. Commonly, the defect is large and a large left-to-right shunt is present, resulting in congestive heart failure and pulmonary vascular obstructive disease. Cyanosis is usually absent unless irreversible severe pulmonary vascular disease has developed. Since elevation of the pulmonary resistance is rapid in these patients, a prompt diagnosis and treatment are mandatory. An accurate diagnosis of Aortopulmonary window based on clinical symptoms is extremely difficult when it coexists with a VSD. Although two-dimensional echocardiography is important in the diagnosis of this defect, 9 the best method of confirming Aortopulmonary window is angiogram especially when there is a coexisting VSD. The diagnosis of APW can be missed unless an angiogram is obtained.

Surgical closure is indicated in all patients with an Aortopulmonary window, except for asymptomatic patients with small defects. In most patients, closure should be undertaken at the time of diagnosis because of the risk of development of early pulmonary vascular disease.¹⁰

In the absence of associated anomalies, the late results of surgical correction are excellent; however, in patients with complex anomalies, the prognosis is largely determined by the presence of these anomalies. In older patients, the outcome will largely depend on the pulmonary

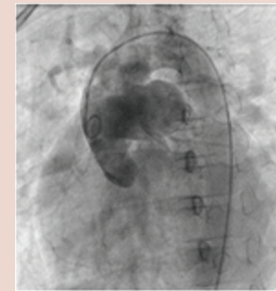


Figure 4: Angiogram of ascending aorta done in left anterior oblique shows opacification of pulmonary artery through Aortopulmonary window [arrow].

vascular resistance at the time of repair. Backer and Mavroudis reported 22 cases of APW in a 40-year period with no mortality; they used trans-aortic patch closure in the most recent six patients.⁶

CONCLUSION

Aortopulmonary window in association with subpulmonic Ventricular Septal Defect non compaction of Left Ventricle is very rare. Meticulous 2 D echocardiographic examination is required, Diagnosis and closure at early stage is imperative to prevent pulmonary artery hypertension.

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