

Transcatheter Closure of a Large Aortopulmonary Window with Pulmonary Arterial Hypertension: Lessons to be Learnt

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ABSTRACT

Aortopulmonary window is an uncommon congenital anomaly which behaves hemodynamically similar to patent ductus arteriosus. Being close to origin of great vessels, it poses a great technical challenge with regards to transcatheter closure and are thus, preferably closed surgically. We present a case of transcatheter closure of one of the largest aortopulmonary window with pulmonary arterial hypertension ever reported and highlighting the technical challenges encountered during the procedure.

Keywords: Aortopulmonary window, Pulmonary arterial hypertension, Amplatzer septal occluder, Device closure, Restrictive, Transthoracic echocardiography.

Key Message:

- Aortopulmonary window is a rare congenital malformation which usually presents as large non-restrictive defects manifesting early in infancy, with pulmonary hypertension.
- Rarely, these defects have suitable margins and are correctable by device closure.
- Appropriate planning and selection of hardware is essential for procedural success.

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BACKGROUND

Aortopulmonary window (APW) is a communication between the ascending aorta and the pulmonary trunk, occurring above two normally formed semi-lunar valves.^{1,2} Isolated APW accounts for 0.2% of cases of congenital heart disease^{3,4} and is associated with other cardiac anomalies in 52% of cases.⁵ It is restrictive in only 10 % cases⁶ and transcatheter closure has been tried only in this subset of population. Paucity of reports in world literature may be due to relative rarity of the defects with good margins, associated congenital cardiac anomalies requiring cardiac surgery, early development of Eisenmenger syndrome in this population or due to technical challenges including constraint of the sheath and dedicated device for these patients.

CASE REPORT

A 5-year-old male child admitted to our hospital with grade IV/VI systolic murmur, best heard at left 2nd and 3rd intercostal space near parasternal area with loud P₂. His resting oxygen saturation was 96% and there was no exercise oxygen desaturation of any limb. Transthoracic echocardiography revealed 7-10 mm large APW with predominant left to right shunt in various views. It was anteriorly located 8 - 10 mm above the sinus of valsalva with significant pulmonary arterial hypertension (estimated mean pulmonary artery pressure = 55 mmHg) (Figure 1 & 2). There were no associated congenital abnormalities. Aortic root angiogram was done which revealed a 9.5 ± 0.7mm APW which was located in mid part of aortopulmonary septum (Figure 3). Cardiac catheterization revealed very high pulmonary artery pressure (85/46 mmHg, mean=56 mmHg, more than 2/3rd of systemic pressures). Main aim of this angiogram was to look for associated coronary anomalies as well as measure the distance of coronary artery origin and pulmonary valve from the APW in two orthogonal plane for appropriate selection of type and size of device.

Despite significant pulmonary arterial hypertension, 12 mm Amplatzer™ septal occluder was taken to close the defect because of thin aortopulmonary septum and high pressure in both the great vessels and stability provided by the two rims of Amplatzer™ septal occluder. We

approached the defect from venous side with tiger catheter and 150 cm J tipped 0.035 inch Terumo guidewire to cross the defect. We exchanged the Terumo with 360 cm Amplatzer superstiff wire and passed a 6 F 80 cm Amplatzer™ Torque™ 180° delivery sheath over it. In first attempt, the device just pulled through the defect because of inadequate opening of the aortic rim due to small diameter of the arch of aorta. However, during next attempt, aortic rim was carefully allowed to open completely before it was pulled and device was successfully placed. Before releasing the device, pulmonary artery pressure was measured which was 50 / 26 (34) mmHg without fall in systemic blood pressure. Appropriate device

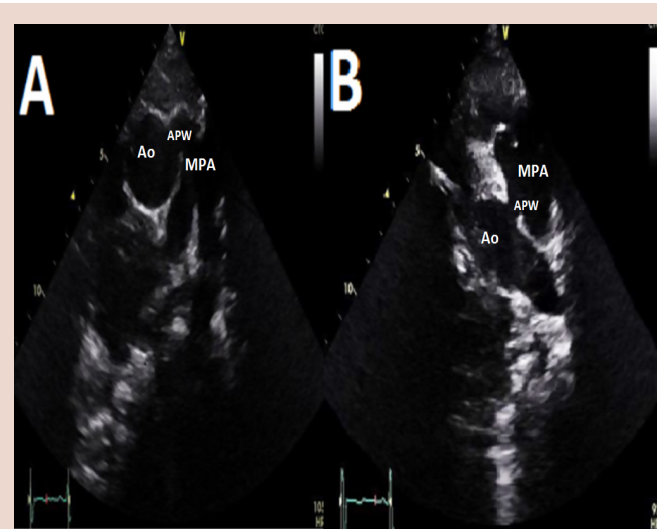


Figure 1: Modified Parasternal short axis view (A) and apical 5 chamber view(B) showing the anteriorly located large aortopulmonary window away from semilunar valves and pulmonary artery bifurcation.



Figure 2: Modified parasternal long axis view showing 6.38 mm restrictive aortopulmonary window with left to right.

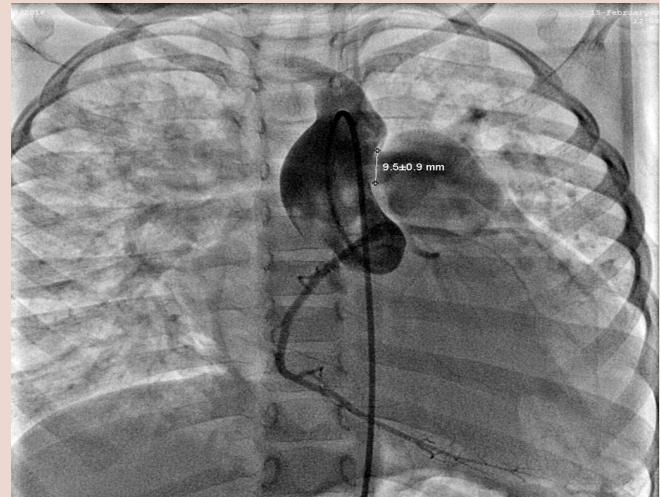


Figure 3: Ascending aorta angiogram showing 9.5 mm large aortopulmonary window.



Figure 4: Ascending aorta angiogram post device closure without residual flow.

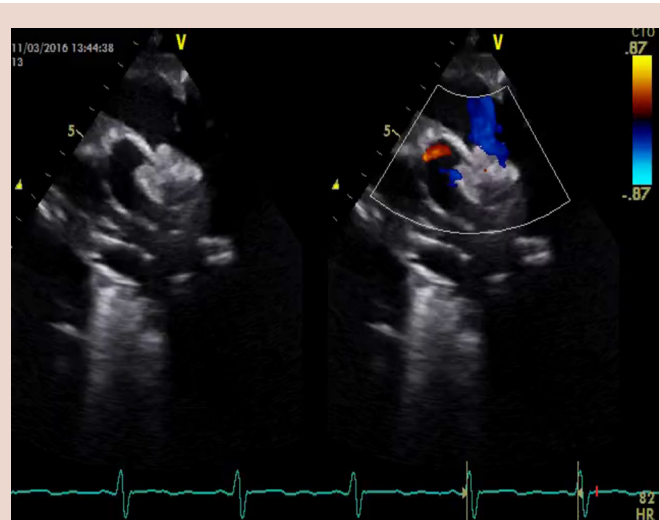


Figure 5: Modified parasternal long axis view showing device *in situ* with no residual flow.

position was confirmed both by transthoracic echocardiography and angiograms in multiple views (Figure 4 & 5). There was no residual flow across and no obstruction to either aortic or pulmonary arterial flow. Patient was put on oral aspirin for 6 months and tab. Sildenafil for Pulmonary arterial hypertension. Post device closure angiogram was done which didn't show any residual flow. Transthoracic echocardiography after 7 days revealed near normalization of mean pulmonary arterial pressure to 24 mmHg with no residual flow.

DISCUSSION

APW is a rare congenital cardiac anomaly with half of the patients having associated cardiac lesions. Mori *et al.* has proposed a classification into three types of aortopulmonary connection. Type 1 is the most common type and refers to a small defect midway between the semilunar valves and the pulmonary artery bifurcation. Type 2 is a more distal defect with the distal border formed by the pulmonary bifurcation. Type 3

is a large confluent defect involving almost the entire aortopulmonary septum and is rare.⁷ APW is most commonly confused with either a large patent ductus arteriosus or persistent truncus arteriosus.

Before proceeding with any transcatheter intervention for structural heart disease, detailed evaluation of anatomy of the defect and surrounding structures is a must for procedural success. Transthoracic echocardiography and angiogram played an important role in delineation of the anatomy in patients with aortopulmonary window. It provides information regarding the exact location of the defect, distance of the defect from coronary artery origin, pulmonary valve and pulmonary artery bifurcation, pulmonary artery pressures, and associated anomalies for selection of the suitable device for closing the defect. Double umbrella shaped devices like Amplatzer septal occluder are more suitable as they provide adequate retention skirts on both sides to prevent device dislodgement secondary to associated pulmonary arterial hypertension.

Though the procedure is technically similar to PDA device closure, but position of this defect near the origin of great vessels with acute take off and presence of large defect that require manipulation of long sheath and a large device in the great vessels make this procedure technically challenging. Perpendicular and anterior origin of defect to the origin of semilunar valves can create potential hurdles in crossing the lesion via pulmonary valve. Selection of appropriate hardware is essential to cross the lesion successfully. Another important issue is size of the device which depends on the rims surrounding the defect and pulmonary artery pressures. It is also important because large devices are difficult to manipulate in comparatively smaller great vessels. Though there is paucity of data regarding transcatheter closure of APW. Recent case reports have increasingly used duct occluders⁷⁻¹⁰ for small sized defects while large defects have been closed with Amplatzer septal occluder,¹¹ muscular VSD device,¹³ and perimembranous VSD device.⁶ Our case once again stresses the importance and usefulness of temporary device occlusion for patients with severe pulmonary arterial hypertension, closure of APW with device resulted in near normalization of pulmonary artery pressures suggesting that it was hyperdynamic and defect occlusion should be tried in case of doubt just like we do in patients with patent ductus arteriosus for reversibility.

CONCLUSION

We conclude that transcatheter device closure of APW should be considered when the anatomy is favourable in terms of location, size, and margins of the defect as well as favourable physiology in terms of reversible pulmonary hypertension even in the setting of near systemic pulmonary arterial pressures.

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None

CONFLICT OF INTEREST

None

ABBREVIATIONS USED

APW: Aortopulmonary window; VSD: Ventricular septal defect; PDA: Patent ductus arteriosus; TM: Trademark.

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