Abstract: An aortopulmonary window is an uncommon congenital cardiac defect. Transcatheter closure of these defects poses a great technical challenge and are thus feasible only in selected cases. We report a successful closure of an aortopulmonary window using a 10mm Amplatzer type septal occluder, highlighting the technical challenges encountered during the procedure.

Introduction

Aortopulmonary window (APW) is a communication between ascending aorta and the pulmonary trunk, occurring above the semilunar valves. Isolated APWs account for 0.2% of cases of congenital heart diseases[1] and is associated with other cardiac anomalies in 52% of cases[2]. It has similar hemodynamic features to a patent ductus arteriosus(PDA) and even more so, to a common truncus arteriosus(CTA), the anatomical difference from the latter being the presence of well-defined aortic and pulmonary valves.

Being close to origins of great vessels, it imposes a great challenge with regards to transcatheter closure. However transcatheter closure of APW should be considered (when the anatomy is favorable in terms of location, size, and margins of the defect as well as favorable physiology) for reversible pulmonary hypertension[3].

Paucity of reports in world literature may be due to relative rarity of the defect with good margins, associated cardiac anomalies requiring cardiac surgery and technical challenges, including constraint of the sheath and dedicated device for these patients.

We report a case of a young man with a distal type of APW who presented with some clinical features of heart failure, in whom catheter occlusion was successfully achieved using a 10mm Amplatzer type septal occluder.

Case report

The patient first presented at the age of 16 years, with recurrent respiratory tract infections and a cardiac murmur. During the initial encounter he was diagnosed as having a PDA and underwent thoracotomy which had negative findings. Subsequent evaluation using cardiac catheterization had revealed an APW, but the patient was lost to follow up for next 16 years. At the age of 32, he again presented with persistent respiratory symptoms and was referred for intervention.

He had a bounding pulse, loud P2 and a continuous murmur. Chest X-ray demonstrated cardiomegaly and pulmonary plethora (Figure 1).

Transthoracic echocardiography revealed dilated left sided chambers and a moderate size (7mm) APW with left to right shunt(Figure 2, 3 and 4). It was distally located closer to right pulmonary artery (RPA) origin, well away from semilunar valves and coronary origins and was well circumscribed. There were no other associated cardiac malformations and pulmonary artery pressures remained less than half of systemic pressures.

Under general anesthesia, the femoral artery and vein were percutaneously cannulated. Ascending aortography confirmed an APW of 7mm, located in the distal MPA(Figure 5) with pulmonary artery pressures (45/31, mean 37 mmHg) remaining less than half of systemic pressures (115/57 mmHg, mean 82mmHg).
Figure 1 - Chest X-ray demonstrating cardiomegaly and pulmonary plethora

Figure 2 - Apical four chamber view showing dilated left sided chambers.

Figure 3 - Appearance of an APW on a modified 4 chamber view echo with the probe tilted anteriorly.

Figure 4 - Parasternal short axis view showing APW with left to right shunt.
The defect was retrogradely crossed using a 4F Judkins right coronary catheter and 0.035” Terumo wire combination passed from the aortic side. The Terumo wire was exchanged to a 300cm long 0.035” Terumo guide wire which was snared in the RPA with a 10mm Amplatzer gooseneck snare introduced via the femoral vein, and exteriorized to create a continuous arterio-venous loop.

Over the wire, a 7F Amplatzer PDA delivery system was advanced via the femoral vein and through the APW into descending aorta. Initial attempts to close the defect using 8mm and 10mm Amplatzer muscular VSD occluders and a 12mm x 10mm Amplatzer duct occluder I failed as they easily slipped back in to MPA. Thereafter, a 10mm Cocoon atrial septal occluder, which has a larger retention skirt, was successfully deployed across the defect. Before releasing the device appropriate device position was confirmed both by transthoracic-echocardiography and aortogram (Figure 6). There was no residual flow across the device and no obstruction to either aortic or pulmonary arterial flow including that to RPA.

Pulmonary artery pressure has dropped to near normal level(28/20, mean 23mmHg) following the device closure.

Patient was discharged the following day, with a plan of continuing an anti-platelet dose of aspirin for 6 months. Follow up echocardiogram done after 6 months revealed ideal positioning of the device without any residual flow with near normal sizes of left sided cardiac chambers (Figure 7).

**Discussion**

APW is a rare congenital cardiac anomaly, with half of the patients having associated cardiac lesions. Early surgical treatment is recommended for larger defects, to avoid the risk of progressive pulmonary vascular disease, but require the use of cardiopulmonary bypass[4].

Transthoracic echocardiography and angiography play an important role in delineation of the anatomy in patients with APW. It provides information on the exact location and size of the defect, distance of the defect from coronary artery origins and semilunar valves and associated anomalies.

Though there is paucity of data regarding transcatheter closure of APW, recent case reports...
have increasingly used duct occluders[5] for small sized defects, whilst large defects have been closed with Amplatzer septal occluder [3] and Amplatzer VSD devices[6].

**Conclusion**

Our case highlights the usefulness of Amplatzer type septal occluder when a device with a larger retention skirt for a given waist is required.

**References**