Transcatheter closure of aortopulmonary window in children:
Case reports and literature review

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INTRODUCTION

Aortopulmonary (AP) window is an uncommon congenital anomaly, accounting for approximately 0.2% of all congenital heart abnormalities. It is usually nonrestrictive and is conventionally treated surgically at an early age to prevent the development of pulmonary vascular obstructive disease. In 10% of this lesion, it is restrictive and case reports of its percutaneous closure are mostly limited to these patients.

CASE REPORTS

From March 2002 to November 2010, three children underwent successful transcatheter closure of AP window at our institute.

Case 1

The first patient was an 11-month-old male child weighing 8 kg who presented with cardiac murmur asymptptomatically since he was 3-month-old. Grade III-IV/VI continuous murmur was present in the left 2nd and 3rd intercostal area. An AP window was found and left atrium and ventricle were dilated, there was no associated congenital cardiac anomaly, however, the size of AP window was not clear on echocardiography. Cardiac catheterization and selective aortogram were performed. Pulmonary artery pressure was 18/7 mmHg (mean 12 mmHg). Pulmonary to systemic flow ratio (Qp / Qs) was 2:1. Selective ascending aortogram in anterior view showed the presence of AP window measuring 3 mm in diameter. It was located in the left lateral wall of the ascending aorta, 12 mm far from the aorta valve (Figure 1). The AP window was crossed antegradely from aortic side with a 5F pigtail catheter. Once the catheter was in the MPA, a 0.032-in. wire was advanced through the catheter into the main pulmonary artery (MPA). The wire was snared out from the venous side thus creating an arteriovenous (AV) loop of the wire from MPA through right ventricle (RV) to right atrium (RA) and inferior vena cava (IVC). A 6F long sheath was then taken over the wire
from the venous side through the defect to the descending aorta. A 6–8 mm Amplatzer duct occluder (AGA Medical, Golden Valley, MN) was used to occlude the defect. There was no residual shunt on subsequent aortic root angiography (Figure 2) as well as on color Doppler. Post-procedure in hospital course of the patient was uneventful. At 1-year follow-up, the patient continues to be asymptomatic.

**Case 2**

A 2-year-old girl presented with a history of recurrent lower respiratory tract infection (LR-TI), which was managed medically. On examination, grade IV/VI systolic murmur was present in the left 3rd and 4th intercostal space. Hemoglobin was 127 g/L and baseline biochemical parameters were normal. Chest radiography revealed cardiomegaly and pulmonary plethora. Electrocardiographic finding was left ventricular hypertrophy. However, the first echocardiographic examination failed to identify the presence of AP window, although the AP window could be identified at the second echocardiography (Figure 3, 4). Echocardiogram showed severe pulmonary arterial hypertension. Left atrium and ventricle were dilated. Cardiac catheterization and selective aortogram were performed. A 20 % step-up in oxygen saturation was noted in the main pulmonary artery. Pulmonary artery pressure was 82/43 mmHg (mean 64 mmHg). Qp/Qs was 4:1. Selective ascending aortogram in anterior view showed the presence of APW measuring 10.39 mm in diameter (Figure 5). The AP window was crossed retrogradely from the venous side via the right femoral vein, inferior vena cava, right atrium, right ventricle and pulmonary trunk with a 6 F MPA2 Cordis catheter and an Amplatz super stiff J-tipped 0.035" wire leaving its tip in the descending aorta. A 10F long sheath and dilator were advanced over the wire and its tip positioned across the AP window into the descending aorta. An 18 mm HeartR muscular ventricular septal defect occluder (Lifetech Scientific Co, Shenzhen, China) was deployed across the AP window. A check aortogram was done to ensure complete occlusion of the defect without impinging upon left main coronary artery (LMCA). The device was delivered and the sheath was withdrawn to the inferior vena cava. An aortic root angiogram showed the device occluding the AP window completely with a moderate residual shunt (Figure 6). There was no impingement of left coronary artery or aortic or pulmonary regurgitation. The systolic murmur disappeared and the follow-up echocardiography done at 1 month showed no residual shunt.

**Case 3**

A 14-year-old male presented with cardiac murmur asymptotically. On examination, grade III/VI continuous murmur was present which was best heard in the left 2nd and 3rd intercostal space. Echocardiographic examination failed to identify the presence of AP window, only showed a giant patent ductus arteriosus (PDA) and severe mitral regurgitation. Cardiac catheterization and selective aortogram were performed. Pulmonary artery pressure was 45/20 mmHg (mean 28 mmHg). Pulmonary to
systemic flow ratio (Qp/Qs) was 5.7:1. Selective ascending aortogram in anterior view showed the presence of AP window measuring 14 mm in diameter. As described above, the defect was crossed retrogradely with a 6F MPA2 Cordis catheter and an Amplatz super stiff J-tipped 0.035" wire leaving its tip in the descending aorta. A long 9F sheath and dilator were advanced over the wire and its tip positioned across the AP window into the descending aorta. An 18–20 mm HeartR Duct Occluder (Lifetech Scientific Co., Shenzhen, China) was used to occlude the defect. A check aortogram was done to ensure complete occlusion of the defect without impinging upon LMCA and left coronary artery or aortic or pulmonary regurgitation. The continuous murmur disappeared and the follow-up echocardiography done at 6 months showed no residual shunt with moderate mitral regurgitation.

Figure 1  Angiogram in ascending aorta demonstrating aortopulmonary window connecting ascending aorta and pulmonary trunk. The arrow shows aortopulmonary window
Figure 2  Angiogram after successful deployment of Amplatzer duct occluder in aortopulmonary window shows complete occlusion of the defect with the device
Figure 3  Parasternal short axis 2D echo showing dropout in the area of the adjacent walls of the intrapericardial components of the aorta and the pulmonary trunk, suggestive of an aortopulmonary window
Figure 4  Color Doppler showing an aortopulmonary window
DISCUSSION

AP window, a defect in the aortopulmonary septum, is classified into various types by different authors 2–4. Ho, et al. 2 had classified AP window into four types according to its morphologic features, proximal defect is between ascending aorta and MPA having little inferior rim separating the AP window from semilunar valves, distal defect is between the origin of right pulmonary artery and ascending aorta having a well-formed inferior rim but little superior rim, confluent defect, a combination of the first and second types with little superior and inferior rims, and intermediate type, which has adequate superior and inferior rims. It is the intermediate type of defect that is best suited for device closure. Recently, Jacobs and the Congenital Heart Surgery Database Committee have advocated the use of hierarchical levels for AP window 5. The committee settled on five AP window subtypes; besides the above four, AP window and interrupted aortic arch as a subtype.

World literature on transcatheter closure of AP windows is limited to closure of restrictive defects that in adults or older children (3 years old to 27 years old) 6. Trehan V, et al 6 reported percutaneous closure of nonrestrictive aortopulmonary window in three infants. At our institute, Case 2 was a two-year-old girl who had a nonrestrictive AP window with near systemic PA pressures, while other two cases were restrictive. An antegrade (aortic to pulmonary artery side) approach was used to cross the defect in an infant while a retrograde approach was in the other two children. Different operators have used different types of devices for transcatheter closure of AP windows. We used two different kinds of devices-duct oc-
clude (6–8 mm, 18–20 mm), muscular VSD occluder (18 mm). Duct occluder has a shape suited for closing a duct or a tubular structure. When used for AP window, though the aortic end is well apposed to the aortic wall, the pulmonary end tends to remain away from the wall of pulmonary artery and hence protrudes into the lumen (5–8 mm depending on device size). Muscular VSD occluder having a waist length of 7 mm also suffers from the same disadvantage. So an aortogram must be done to ensure complete occlusion of the defect without impinging upon LMCA and left coronary artery or aortic or pulmonary regurgitation.

In our second patient, during the initial attempt to deploy the device, the fully open aortic disc of the device (18 mm muscular ventricular septal defect occluder) easily slipped out of the aorta across the AP window into the MPA. After careful review of the procedure, it was realized that aortic disc as it was approaching the aortic window was making an acute angle with the aortic wall and was not approaching parallel to it. So it was decided to use the same device again and with careful manipulation of the same sheath this time the aortic disc was made to approach the window parallel to aortic wall. Then the device could be easily delivered across the window.

When both the aortic and the pulmonary ends were positioned in the respective great arteries across the AP window, the following were systematically checked prior to release of the device from the delivery cable. Echocardiographic examination was done to visualize the device across the defect, look for any residual shunt or acquired gradient in any of the great arteries. Aortic root angiography was done to look for any residual shunt without impinging upon LMCA and left coronary artery.

**CONCLUSION**

Transcatheter closure of large (nonrestrictive or restrictive) AP window is safe and feasible in children provided the AP window is intermediate in type and unassociated with any other congenital cardiac anomaly. Significantly less discomfort, avoidance of CPB and surgical scar, and shorter duration of hospital stay make device closure the treatment of choice in such selected cases of AP window.

**REFERENCES**