

Transcatheter Treatment of Large Aortopulmonary Window: A Case Report

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ABSTRACT

Introduction: Aortopulmonary window (APW), a large aortopulmonary septal defect (APSD), is a serious and rare defect within congenital heart diseases.

Case report: In this study, we reported an APW case with severe pulmonary arterial hypertension. This patient was successfully treated by transcatheter closure with a muscular ventricular septal defect (VSD) occluder.

Conclusion: We had a successful experience with transcatheter closure of a large APW using a muscular VSD occluder. There was no residual shunt or complications during the 6-month follow-up.

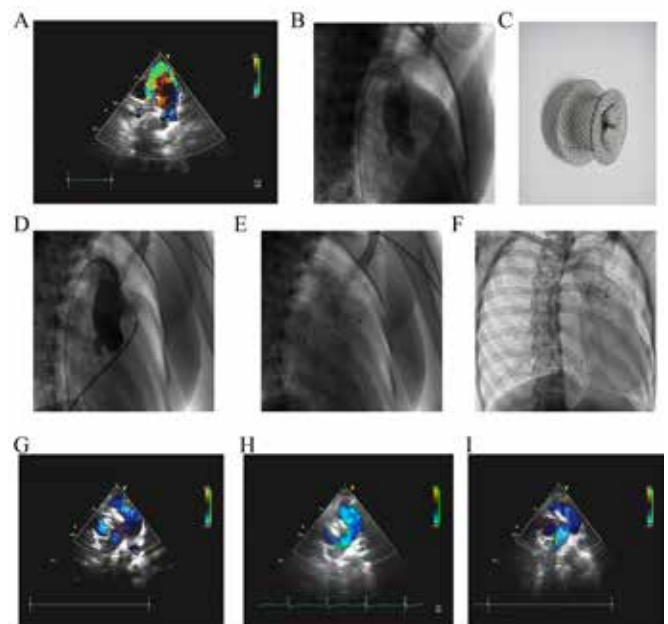
INTRODUCTION

Aortopulmonary window (APW), a kind of aortopulmonary septal defect (APSD), is a serious and rare congenital heart disease by reason of large tubular defect between the aorta and the pulmonary artery. Large APSDs can result in early heart failure, thus, treatment is needed as soon as possible to avoid the risk of progressive pulmonary vascular disease [Stamato 1995]. APSDs were treated mainly with surgery through sternotomy in the past [Erez 2004]. However, with the development of medical technology, transcatheter closure has been applied to repair APSDs [Mert 2004; Noonan 2013; Srivastava 2012]. In this study, we reported a case of successful transcatheter closure of large APW with severe pulmonary arterial hypertension using a muscular ventricular septal defect (VSD) occluder in an adult.

CASE REPORT

A year ago, a 23-year-old Chinese Han female was admitted to the hospital because of palpitation and dyspnea after activity. Physical examination revealed a continuous murmur in the left third intercostal space. Echocardiographic evaluation showed an enlargement of the left atrium

(diameter 40 mm) and left ventricle (internal diameter at end-diastole 64 mm). An expansion of the main pulmonary artery was also observed. In addition, a communication (12 mm in diameter) was found between the ascending aorta and the main pulmonary artery at 30 mm above the aortic and pulmonary valves. The continuous left-to-right shunt with a maximum velocity of 3.5 m/s was observed. The pressure gradient of the shunt was 46 mmHg and the pulmonary artery systolic pressure (PASP) was 77 mmHg



Preoperative and postoperative echocardiograms and chest X-rays of the patient. A, Preoperative echocardiogram marking the 12-mm defect (highlight); B, Preoperative chest X-ray marking the pulmonary congestion; C, The SHSMA 22-mm diameter muscular VSD occluder has a 36-mm aortic disc diameter, 32-mm pulmonary disc diameter, and 10-mm device length; D, Aortogram showing good placement of the device and little residual shunt; E, The muscular VSD occluder is released completely; F, At 1-month follow-up, a chest X-ray shows a good-shaped occlude; G, At 1-week follow-up, an echocardiogram shows good positioning of the occluder and successful closure; H, At 3-month follow-up, an echocardiogram shows left atrium and ventricle, and pulmonary artery pressure recovers gradually; I, At 6-month follow-up, an echocardiogram shows successful closure with no residual leak.

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(Figure, A). Chest x-ray showed pulmonary congestion and an enlarged pulmonary artery segment.

An ascending aortogram showed a continuous tubular shunt measuring 13 mm in diameter and 15.3 mm in length between the aorta and the pulmonary artery. The shunt was about 30 mm away from the left coronary artery orifices (Figure, B). Cardiac catheterization showed the ascending aorta pressure (118/57 mmHg) and the pulmonary artery pressure (87/45 mmHg). Therefore, this patient was diagnosed with type II distal APSD.

Because cardiopulmonary bypass (CPB) in surgical operation can affect cardiac function and pulmonary artery pressure, transcatheter closure was attempted in this patient. The surgical process was performed as previously described [Srivastava 2012; Naik 2003]. Briefly, the right femoral artery and veins were accessed by a 6 F JR4.0 and 6 F MPA2 sheath, respectively. An Amplatzer 0.035" extra stiff 260 cm wire (Terumo Corporation, Japan) was passed up to the pulmonary artery. An arteriovenous loop of guide wire by snare technique was performed for loading a 12 F long sheath (Shanghai Shape Memory Alloy Company, China) from the femoral vein to the ascending aorta. A 22 mm-diameter muscular VSD occluder (Shanghai Shape Memory Alloy Company, China) (Figure, C) was introduced into the long sheath and successfully deployed. After the operation, 47.5 mg of metoprolol succinate daily and 5 mg of amlodipine daily were given for controlling systolic pressure. Antiplatelet therapy of 5 mg/kg aspirin daily was given as well.

At 15 minutes post-operation, an ascending aortogram revealed a stable position of the occluder with abolition of the shunt (Figure, D). The aorta pressure was 134/70 mmHg and the pulmonary artery pressure was 48/19 mmHg. The continuous murmur in the left third intercostal space disappeared after the procedure (Figure, E). The patient was discharged on the following day without any complications. Chest X-ray after 1 month showed a well-shaped occluder (Figure, F). Echocardiograms at 1 week (Figure, G), 3 month (Figure, H) and 6 month (Figure, I) follow-up showed no residual shunt. The enlargement of the left ventricle was reduced, and PASP gradually returned to normal level. In addition, no other complications were noted during the 6 months of follow-up. Thus, this percutaneous closure could be considered as a success according to the postoperative results.

DISCUSSION

APSD is a severe congenital heart defect commonly associated with other cardiac anomalies, and a surgery is usually needed once diagnosed in childhood [Erez 2004]. So far, there are not many reports about transcatheter closure of APSD, probably because it is a very rare condition. Meanwhile, most of the reports are limited to small defects in children. For examples, Stamato et al [Stamato 1995] reported a transcatheter closure of an APSD in a 3-year-old child using a modified double umbrella occluder system. Further, Li et al [Li 2014] described a successful transcatheter closure of a late-onset residual defect after repair of APSD in an 8-year-old boy with a muscular occluder device. Recently,

an 18-year-old adult with a small residual shunt underwent a repair using an Amplatzer septal occlude [Prem Sekar 2012]. However, in our study, the patient was 23 years old and had a 13-mm diameter shunt.

The patient in this case had abnormal precordial beat as a child, but no obvious chest distress and suffocation were observed. When she was 23 years old, she sought medical advice for palpitation and dyspnea in our hospital for the first time. Adult presentation of APSD is not a common condition, especially for those with a large tubular shunt [Jureidini 1998]. Surgical operation and catheter-based intervention were considered for this patient. We discussed this with our cardiac surgeon. Surgery is considered as the traditional treatment option for APSD, but is associated with a risk of CPB, bleeding, and adhesion [Li 2014]. Catheter-based intervention can protect cardiac function and likely reduce pulmonary artery pressure [Michel-Behnke 2003]. Ultimately, transcatheter closure was performed because the patient was an adult with severe pulmonary arterial hypertension.

Transcatheter closure is considered in APSD when the defect is relatively small and the space is adequate for the device [Naik 2003; Atiq 2003]. The transcatheter closure by a muscular VSD occluder was selected for treating the APW in this patient because (1) she was diagnosed with APSD with evidence of a large left-to-right shunt, and the defect was away from the left coronary artery orifices; (2) the defect had an unusual shape of a long tube-like fistula; (3) the continuous tubular shunt was 13 mm in diameter. Thus, the previously reported Amplatzer occluders could not be used because of their small distal disc. The SHSMA muscular VSD occluder with a large distal disc and a wasp waist corresponding to the shape of the defect was selected [Srivastava 2012]. Finally, we had a successful experience of transcatheter closure of a large APW using a muscular VSD occluder.

Some things should be noted regarding transcatheter closure. First, the feasibility of transcatheter closure has to be evaluated by defining the location, shape, and size of the defect. Transcatheter closure should be considered only when the distance between the defect and the valves is sufficient, without other related cardiovascular abnormality. Second, the selected candidate VSD occluder should match the shape and margin of the defect. In our experience, preoperative image evaluation is important. Echocardiogram can be used to confirm its existence as well as to assess the volume and direction of the shunt [Backer 2002]. Chest X-ray and aortogram can help to evaluate the feasibility of transcatheter closure and to choose suitable devices [Naik 2003; Elhoury 2008].

Conclusion

We recommend using the muscular VSD occluder for closure of large APSD in an adult with high PASP. Further studies are required to verify its safety, efficacy, and long-term results in more patients.

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