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Transcatheter Closure of Aortopulmonary Window in Infants with Amplatzer Duct Occluder-I

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Aortopulmonary window (APW) is a septation defect between the ascending aorta and main pulmonary artery, and it accounts for 0.2-0.6% of all congenital heart diseases. The diagnosis is made by detecting the defect between the ascending aorta and pulmonary artery while both semilunar valves are fully developed. Based on the anatomical characteristics, APW is classified into four types: proximal (Type 1) defect, distal (Type 2) defect, total (Type 3) defect and intermediate (Type 4) defect. APW is traditionally treated by surgery, and there are a few reports about transcatheter APW closure in infancy. Only defects with adequate superior and inferior rims can be considered for device closure. We describe two cases who underwent transcatheter APW closure with the Amplatzer duct occluder-I (ADO-I). Our experience shows that the ADO-I can achieve good results in closure of APW for selected patients.

Key Words: Amplatzer duct occluder-I • Aortopulmonary window • Infant

INTRODUCTION

Aortopulmonary window (APW) is a septation defect between the ascending aorta and main pulmonary artery, and it accounts for 0.2-0.6% of all congenital heart diseases.[1] The diagnosis is made by detecting the defect between the ascending aorta and pulmonary artery while both semilunar valves are fully developed. More than half of the patients with APW have other cardiovascular anomalies, such as atrial septal defect, ventricular septal defect, patent ductus arteriosus and interrupted aortic arch.[2] Based on the anatomical characteristics, APW is classified into four types: proximal (Type 1) defect, distal (Type 2) defect, total (Type 3) defect and intermediate (Type 4) defect.[3] While proximal defects have little inferior rim in relation to the semilunar valves, distal defects have a well-formed inferior rim but little superior rim. Total defects refer to confluent defects with little superior and inferior rims. Intermediate defects, which are more suitable for transcatheter closure, have both well-formed inferior and superior rims.

The course of the disease depends mainly upon the degree of left-to-right shunting into the pulmonary trunk resulting in pulmonary vascular disease. If left untreated, irreversible obstructive changes in the pulmonary vascular bed develop, and death occurs in the first or second decade of life. Although surgical closure is the primary treatment, transcatheter APW closure can be performed as an alternative to surgery in uncomplicated patients with adequate rims. We describe two cases who underwent transcatheter APW closure with the Amplatzer duct occluder-I (ADO-I) (AGA Medical, Minn, USA).

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CASE 1

A one-month-old boy, weighing 3.5 kg, was referred

to our outpatient clinic with a diagnosis of coronary fistula. A physical examination revealed tachypnea, dyspnea and a 3/6 systolic murmur on the upper left margin of the sternum. Chest radiography showed cardiomegaly, and electrocardiography revealed normal sinus rhythm with sinus tachycardia. An echocardiographic examination demonstrated a 3.5-4 mm wide, Type 4 APW defect between the ascending aorta and main pulmonary artery (Figure 1A, B). A bicuspid aorta, right aortic arch and aberrant left subclavian artery accompanied the APW. Computed tomography revealed a tubular shaped APW with well-formed inferior and superior rims (Figure 1C). Congestive heart failure secondary to APW was considered, and decongestive treatment was administered. However, the clinical features of congestive heart failure did not regress during follow-up, and it was decided to schedule the patient for transcatheter closure of the APW after informed consent was obtained. The procedure was performed when the baby was two months old and weighed 4 kg, under general anaesthesia while ventilating with a laryngeal mask. On angiography, the APW was found to be 4.1 mm at the narrowest part with a 7-mm longitudinal length, and the Q_P/Q_S ratio was calculated as 2.5 (Figure 1D). The pulmonary artery pressure was 35/16 (mean 25) mmHg and the systemic arterial pressure was 71/40 (mean 55) mmHg. A 4F right Judkins catheter (Cordis Corporation, Miami, FL), located in the ascending aorta, was retrogradely advanced into the APW, pulmonary artery, right ventricle, right atrium and inferior vena cava through the hydrophilic guidewire. An atrioventricular loop was created by capturing the guidewire in the inferior vena cava with a 15-mm snare catheter. The 7F sheath was then inserted into the femoral vein and advanced through the wire to the descending aorta. An 8/6 mm ADO-I device was advanced through the long sheath. The device retention disc was opened in the aorta and the system was pulled back through the APW. A control angiogram was performed before the device was completely released, and the location of the device was checked by echocardiography (Figure 1E). The system was released after detecting that the position was appropriate (Figure 1E). There was no gradient indicative of obstruction at the aorta or pulmonary artery. After the procedure, the patient was admitted to the paediatric cardiology inpatient clinic for 2 days with regular follow-up, and was dis-

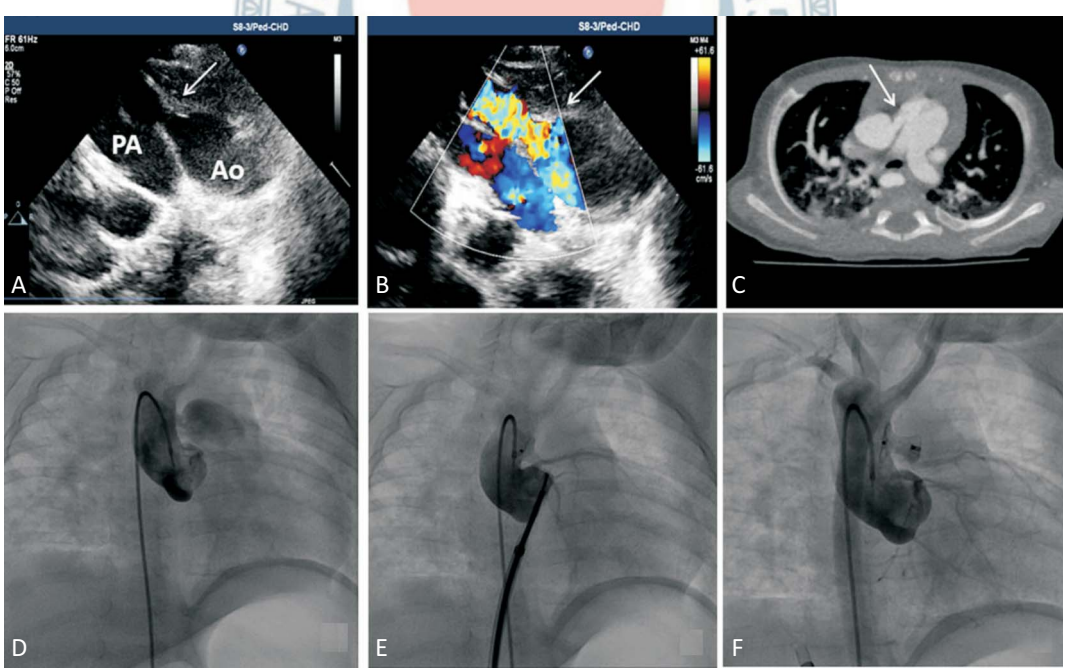

Figure 1. *(A) Echocardiographic examination demonstrated a 3.5-4 mm wide defect between the ascending aorta and the main pulmonary artery (white arrow). (B) Color Doppler shows defect between the ascending aorta and the main pulmonary artery (white arrow). (C) Computed tomography revealed a tubular shaped APW with well-formed inferior and superior rims (white arrow). (D) Angiography shows a 4.1 mm APW at the narrowest point with a 7 mm longutidinal length. (E) Control angiogram was performed before the device was completely released. (F) Angiogram after the device was completely released. Ao, aorta; APW, aortopulmonary window; PA, pulmonary artery.*

charged without any complications. Outpatient follow-up revealed no residual defects and no obstructions of the aorta-pulmonary artery after two years.

CASE 2

A six-month-old girl, weighing 5.3 kg, was admitted to our outpatient clinic due to heart murmur. An echocardiographic examination revealed enlargement of the left ventricle (left-ventricle-end-diastolic diameter: 27 mm z-score: +2.8, left-ventricle-end-systolic diameter: 19 mm z-score: +2.1), a 5-mm defect between the ascending aorta and pulmonary artery (Type 4 APW), an additional small muscular ventricular septal defect and pulmonary arterial hypertension. Cardiac catheterization was performed under general anaesthesia. Angiograms were performed to determine the anatomy of the APW, and the diameter of the defect was found to be 4.5 mm at its narrowest point with a 6-mm longitudinal length, well separated from the semilunar valves. The pulmonary artery pressure was 45/21 (mean 33) mmHg and the systemic arterial pressure was 76/29 (mean 53) mmHg, and the Q_p/Q_S ratio was calculated as 2.6. The defect was crossed from the aortic side using a 4F right Judkins catheter (Cordis Corporation, Miami, FL) over a hydrophilic wire. The hydrophilic wire was then snared in the superior vena cava to create an arterio-venous circuit. A 7F sheet was introduced from the venous side, and a 10/8 mm ADO-1 device was deployed under transthoracic echocardiography and fluoroscopy guidance. One day after the procedure, the patient was discharged in a stable general condition.

DISCUSSION

APW is a defect resulting from incomplete separation of the walls of the main pulmonary artery and aorta at the conotruncal septum during early embryogenesis. With regards to the anatomical characteristics, APW is traditionally treated by surgery, and there are numerous reviews in the literature about surgical outcomes.[5] The first treatment by transcatheter closure was described in the 1990s using a double umbrella occluder system.[6] Since then, reports of transcatheter APW closure with different devices have emerged sporadically.[7-9]

The first step for successful treatment is to determine which patients are appropriate for transcatheter APW closure. The location and size should be carefully defined in order to avoid device embolization, interference with pulmonary valve function, impairment of coronary artery flow and obstruction of the great vessels. Only defects with adequate superior and inferior rims can be considered for device closure. Therefore, defects suitable for percutaneous closure are generally proximal and intermediate-type APWs, which refer to a small defect midway between the semilunar valves and the pulmonary artery bifurcation.[10] In most cases, angiographic and echocardiographic measurements are sufficient to select the device; however, computed tomography and cardiac magnetic resonance imaging can also be performed if echocardiography or angiography is not sufficient to evaluate the diameter of the defect. Balloon sizing can be a good alternative if the defect diameter cannot be assured when necessary. In our first patient, computed tomography was performed before the decision was made for transcatheter closure of the APW.

It is also a challenge to choose the appropriate device, and there is currently no consensus on the optimal device for APW closure. While there are currently no commercially available devices that are specifically designed for the transcatheter closure of APW, cases have been reported in which atrial septal defect, patent ductus arteriosus and muscular-perimembranous VSD occluder devices have been used.[7-12] APWs do not differ in type, but only in shape. In some cases, the longitudinal length of the defect is higher, which results in a tubular shaped APW. Ductal occluders can be appropriate alternatives in patients with tubular APWs without the risk of protruding toward the main pulmonary artery. Therefore, ADO-I was preferred in both of our patients. However, in patients with short longitudinal lengths, devices with a shorter waist can be used. On the other hand, the direction of the intervention can also be determined according to the device features, the defect size and weight of the patient. The procedure can be done in an antegrade or retrograde way according to this information. If the patient's artery is suitable and the device is symmetrical, the defect can be closed without performing an atrioventricular loop.

Transcatheter APW closure has several advantages:

extracorporeal circulation is avoided during the procedure, and the postoperative hospital stay is shorter. Numerous reviews in the literature about surgical outcomes have shown excellent results.[5] Therefore, appropriate patients should be carefully selected for transcatheter APW closure to avoid complications such as stenosis in both sides of the defect and device embolization.

In conclusion, a transcatheter approach can be used as an alternative to surgery in infants with APW. Our experience shows that the ADO-I can achieve good results for the closure of APW in selected patients. Patient and device selection should be made very carefully according to the type, size, longitudinal length and shape of the APW.

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CONFLICTS OF INTEREST

All the authors declare no conflict of interest.

ETHICAL STANDARDS

The authors assert that this work complies with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008. This case was approved by the patients family.

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