Simultaneous Transcatheter Valvuloplasty and Amplatzer Septal Occlusion for Pulmonary Valvar Stenosis and Secundum Atrial Septal Defect

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Abstract

Transcatheter balloon valvuloplasty had been established as the treatment of choice in patients with pulmonary valvar stenosis. Non surgical closure of secundum atrial defects by various occlusive devices is currently being evaluated. We report the first successful simultaneous transvenous Mansfield balloon dilation and Amplatzer septal occlusion in a 7-year-old girl with moderately severe pulmonary valvar stenosis and secundum atrial defect in April 1997. Satisfactory reduction of pressure gradient across pulmonary valve and complete obliteration of left to right atrial shunt were achieved.

Key words: Pulmonary valvuloplasty, Non surgical closure of atrial septal defect

Introduction

Congenital pulmonary valvar stenosis (PVS) and secundum atrial septal defect (ASD) are relatively common forms of congenital heart disease (CHD). Since the first successful clinical application of transcatheter balloon dilation of PVS in 1982,1 the latter has become the treatment of choice for children with PVS who have significant transpulmonary valvar peak to peak systolic pressure gradient greater than 50 mm Hg.2 In recent years, non surgical closure of ASD using different occlusive devices has attracted much interest. Initial enthusiasm has been dampened, at least in part, because of imperfection in design and size of the delivery system and mechanical failure like stress fracture of the metallic frame of one such occlusive device (Bard clamshell).3 However, one recently available device, the Amplatzer septal occluder,4 appears to be promising, in terms of ease of loading, delivery and deployment and satisfactory initial clinical outcome. We report the first successful concurrent transvenous balloon dilation of significant PVS and Amplatzer septal occlusion of a moderately large ASD in a 7-year-old Chinese girl.

Case Report

A 7-year-old girl was referred by a general practitioner for treatment of CHD. Cardiac murmur was first noted on the second day of life by her paediatrician. The patient, the first of a pair of non-identical twins, was born at term by elective caesarean section with a birth weight of 2500 gm. The neonatal period was uneventful. There was no history of cyanosis. Her effort tolerance was satisfactory, although she was not active in sports. There was no family history of CHD. Her developmental milestones were normal.

Clinical examination revealed satisfactory weight (19.9 kg) and height (122 cm), both at the 50th centile for local children of the same age and sex. No cyanosis or clubbing was noted. The peripheral pulses were normal. There was no radiofemoral delay. Her sitting brachial blood pressure measured 88/50 mm Hg. Slight precordial bulge and mild left parasternal heave were noted and the apex beat was palpable just outside the midclavicular line. A grade 4/6 ejection systolic murmur was present over the pulmonary area. The second heart sound was noted to be widely split and a prominent systolic ejection click was audible over the mid left sternal edge. The pulmonic component of the second heart sound was not accentuated. No definite diastolic murmur was detected. There was no sign of congestive cardiac failure. The clinical diagnosis was that of signifi-
cant pulmonary valvar stenosis, although an associated secundum atrial defect could not be excluded by clinical signs alone.

Electrocardiography showed regular sinus rhythm, right axis deviation, right atrial and right ventricular hypertrophy. Chest X-ray revealed borderline cardiomegaly with prominent pulmonary conus and relatively normal looking distal pulmonary vasculature. Transthoracic echocardiography confirmed the clinical diagnosis of moderately severe PVS with an instantaneous peak systolic pressure gradient of 64 mm Hg and post-stenotic dilation of the main pulmonary artery. In addition, a moderately large centrally located secundum atrial defect, measuring 12.9 mm with good length of upper rim (12.4 mm) and lower rim (8.2 mm) was imaged. Volume loading of the right atrium and right ventricle with paradoxical septal movement were present.

The plan of definitive management was discussed with her parents. The options of surgical correction of PVS and patch closure of ASD versus concurrent pulmonary balloon valvuloplasty and Amplatzer device occlusion were detailed. Parents elected to have transcatheter treatment of both conditions in one sitting. Informed consent was obtained. Cardiac catheterisation was carried out on 18 April 1997. Briefly, percutaneous punctures of the right femoral vein and left femoral artery were performed. 5F sheath was placed in the left femoral artery for blood pressure monitoring. Right heart oximetry and manometry were carried out, using GL 7F catheter. NIH 7F catheter was next placed in the right upper lobe pulmonary vein and single plane angiogram in four-chamber projection was performed to profile the atrial septum and delineate the size and morphology of the ASD. NIH 7F catheter was then placed in the right ventricle and biplane (lateral and posteroanterior projections) angiograms were performed to outline the right ventricular outflow tract and pulmonary valve and pulmonary venous return in the levo-phase to exclude anomalous pulmonary venous return. Pulmonary valve measured about 16 mm to 17 mm angiographically and by transoesophageal echocardiography. Doming of the pulmonary leaflets with a central jet during systole and marked post-stenotic dilation of the main pulmonary artery were clearly demonstrated (Fig. 1). Sizing of the ASD was carried out using Miller-Edwards balloon catheter under fluoroscopic and TEE guidance. Stretched ASD size was 17 mm with 3.0 ml dilute contrast inflation. ASD and atrial septal morphology were assessed using Vingmed CFM 800 multiplane paediatric TEE probe. ASD was judged to be centrally located with adequate superior and inferior margins and was away from the coronary sinus and right upper lobe pulmonary vein.

Long exchange 0.038-inch guide-wire was placed into the left pulmonary artery. A single 18 mm x 3 cm Mansfield balloon valvuloplasty catheter was advanced through the guide-wire and the balloon was placed optimally across the stenosed pulmonary valve. Balloon inflation was carried out until the “waist” was abolished (Fig. 2). Two additional inflations were made. Mansfield balloon catheter was then removed. Simultaneous right and left ventricular peak systolic pressures were 74 and 87 mm Hg, respectively. The pre-balloon dilation peak to peak systolic pressure gradient across the pulmonary valve was 53 mm Hg, which was reduced to 22 mm Hg after successful balloon dilation. Guide-wire was then placed in the left upper lobe pulmonary vein. 8F Mullins sheath was advanced through the guide-wire from right atrium to left atrium across the ASD. The distal end of the sheath was placed in the left atrial cavity. Amplatzer 17 mm septal occluder was loaded into the delivery system which was advanced through the Mullins sheath. The left atrial disc was released and positioned to engage on the left atrial side of the septum, under fluoroscopic and TEE guidance. The Mullins sheath was slowly withdrawn through ASD to release the conjoint ring or “waist” of the device which “straddled” the ASD. Subsequently, the right atrial disc was next released by further withdrawal of the Mullins sheath while the delivery system was held stationary. Stability of the position of the device was ensured by gentle “rocking” movements of the system and device. TEE colour flow imaging was performed to ensure adequate occlusion of the ASD. The device was then detached and the delivery system was withdrawn into the inferior vena cava. After a lapse of 10 minutes, repeat right ventricular angiography was carried out. This showed complete occlusion of the ASD without residual left to right shunt (Fig. 3). Catheters were removed and haemostasis was secured by digital pressure. The procedure time was 90 minutes and the screening time was 24 minutes. The patient was discharged the next morning after transthoracic echocardiography confirming complete occlusion of the ASD and satisfactory reduction of instantaneous peak systolic pressure gradient to 20 mm Hg across the pulmonary valve.

Discussion

Since 1982, transcutaneous balloon dilation has become an established treatment of choice for PVS in children as well as in adolescents and adults. It has now replaced surgical valvotomy, except in the situation of dysplastic valve which requires surgical removal to ensure adequate relief of obstruction. Both short and long term follow-up studies showed excellent results in children and adults. The place of non surgical closure of ASD by transcutaneous placement of occlusive device is less well defined, although a number of such devices have been tried with initial encouraging results since the late 1980s. Major concerns were difficulty in placement.

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because of large and clumsy delivery system with some danger of dislodgement and significant residual left to right shunt and long-term structural integrity of the device. Indeed fatigue fracture of the metallic frame of the Bard clamshell device led to its removal by the Food and Drug Administration from clinical application. In the past one year, a new occlusive device, the Amplatzer Button™ ASD occluder (AGA Medical Corporation, MN, USA) appears to overcome the technical problems due to its unique design and special materials employed. Briefly, it is a self-expandable double disc made from Nitinol wire mesh linked together by a short connecting “waist” filled with polyester fibres. To improve on the occlusive capacity, two polyester patches, one to each disc, are sewn. The device which is securely screwed onto a delivery cable can be easily folded and loaded into a 7F introducer sheath. We found that for bigger device with “waist” above 16 mm, 8F Mullins long sheath allows easier delivery of the device. Encouraged by initial excellent results in United Kingdom and Australia, we had recently in April 1997 embarked on the programme of percutaneous transluminal occlusion of ASD using the Amplatzer septal occluder.

This patient was one of the initial series of 14 cases who had participated in this programme. She had complete occlusion of the ASD by percutaneous placement of Amplatzer septal occluder. In addition, concurrent satisfactory relief of pulmonary valvar obstruction was achieved by transluminal balloon dilation in one sitting. Successful dual interventional procedures to treat co-existing congenital cardiac defects have been described in the same patient with coarctation of aorta and patent arterial duct (PAD) and with PVS and PAD (unpublished data—W C L Yip and K Y Chan). Compared with surgical treatment, the recovery period following treatment of congenital heart defect is significantly shorter. Furthermore, by performing these two procedures in one sitting would make this therapeutic approach even more attractive. Recently transcutaneous balloon dilation in adult patients with PVS and ASD had been reported. The ASD was left alone, although no increased left to right shunt was detected after the balloon dilation. In the case of our patient, however, we felt that concurrent relief of moderately severe pulmonary valvar obstruction and abolition of the significant left to right atrial shunt could be safely achieved with the combined

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Fig. 1. Right ventricular (RV) angiogram during systole in lateral (LAT) projection showing stenotic jet of contrast into the markedly dilated main pulmonary artery (MPA) before balloon dilation.

Fig. 2. Mansfield 18 mm x 3 cm valvuloplasty catheter being inflated with dilute contrast showing partially filled balloon with central constriction (marked by black arrows) due to stenotic pulmonary valve in lateral (LAT) projection.

Fig. 3. Left frame showing deployment of Amplatzer 17 mm septal occluder with delivery catheter (C) just being withdrawn in four-chamber projection (4CV) under transoesophageal guidance (P = multiplane paediatric TEE probe). Right frame showing normal pulmonary venous return into left atrium (LA) and subsequently left ventricle (LV) in postero-anterior (PA) projection without detectable residual left to right shunt across Amplatzer septal occluder (marked by 2 white arrows).
interventional procedures. As the total procedure time was only 90 minutes, the two interventional procedures could justifiably be performed concurrently rather than sequentially in two separate occasions. Indeed, complete occlusion of the ASD and adequate relief of PVS were demonstrated at the end of the interventional procedures and reconfirmed the next morning, and again one month and three months later by transthoracic echocardiography.

REFERENCES