Closure of a large perimembranous ventricular septal defect in a 4.8 kg baby with Down syndrome using a duct occluder

Mohammed H. Alghamdi a,d,⇑, M.O. Galal b,c, Fahad Al-Habshan a, Mansour AL-Mutairi a

a King Abdulaziz Medical City, Riyadh
b Prince Salman Heart Center, Riyadh
c University of Essen
d King Fahad Cardiac Centre, King Saud University, Riyadh
a b d Saudi Arabia, c Germany

A 9.5-month-old boy with Down syndrome, weighing 4.8 kg, presented with history of failure to thrive. Clinically, he had symptoms and signs of congestive heart failure. His echocardiogram showed a large perimembranous ventricular septal defect (pmVSD) with some inlet extension covered by a large aneurysmal tissue with multiple right ventricular (RV) exits. Additionally, he had hypothyroidism and Hirschsprung disease. Instead of closing the VSD surgically, the VSD was successfully closed utilizing an 8 × 6 mm duct occluder. The baby remained in the intensive care unit for one night. The day after the procedure, the infant was stable and showed clinical improvement. Electrocardiogram (ECG) showed normal sinus rhythm with no evidence of heart block. Twenty-four hours later, echocardiography showed the device was in an excellent position, with a small residual leak. There was normal tricuspid valve inflow and normal aortic valve outflow with no significant valvar insufficiency. The baby was discharged after 3 days in stable condition. We believe infants with such co-morbidities which might complicate their post-operative course and prolong the intensive care unit admission, might benefit from such alternative management.

© 2014 King Saud University. Production and hosting by Elsevier B.V. All rights reserved.

Keywords: Down syndrome, Perimembranous ventricular septal defect, Transcatheter closure, Duct occluder

Introduction

Transcatheter closure of ventricular septal defects (VSD) has not yet received general acceptance. It is not well-established in the younger age group with a weight category of less than 5 kg. Occasionally, hybrid procedure has been proposed [1,2]. While surgical closure of VSD is well-established, some types and locations of VSD remain challenging [3]. Additionally, infants
with Down syndrome in the postoperative period can have a difficult course. The presence of additional co-morbidities will add more risk during the postoperative period [4]. We present an infant with Down syndrome and other associated non-cardiac conditions in whom it was possible to successfully close the VSD utilizing a duct occluder. We believe that certain types of VSD can be closed safely by transcatheter approach even if weight is less than 5 kg, especially if the trend of miniaturizing devices continues. This approach should be considered as a valid alternative to a surgical option.

Case report

Our patient is a 9.5-month-old boy with Down syndrome who presented with symptoms and signs of congestive heart failure and failure to thrive. His weight was 4.8 kg (less than the 5th percentile in the Down syndrome growth chart) and his height was 66 cm (25th percentile in Down syndrome growth chart). He was the product of a spontaneous vaginal delivery at full term with birth weight of 3.2 kg. Chromosomal analysis confirmed the diagnosis of Down syndrome. Additionally, he was found to have hypothyroidism and was initiated on L-thyroxine. Investigation by rectal biopsy due to chronic constipation confirmed Hirschsprung disease. He underwent corrective surgery by resection of the retro-sigmoid segment at the age of 6 months.

The diagnosis of a ventricular septal defect (VSD) was established at birth. He was started on furosemide and captopril. Despite maximum medical therapy, he continued to be in congestive heart failure. His weight remained below the 5th percentile throughout this course.

On examination, there were good femoral pulses, and the liver was 4 cm below the right costal margin. There was cardiac heave and grade 2/6 systolic murmur over the left precordium. The first heart sound was normal but the pulmonary component of his second heart sound was slightly loud. Chest X-ray showed cardiomegaly and increased pulmonary vascularity, consistent with significant left-to-right shunt. The electrocardiogram (ECG) showed normal sinus rhythm, normal axis deviation for age and biventricular hypertrophy. Echocardiography (echo) showed situs solitus, levocardia, intact atrial septum, mild tricuspid regurgitation (TR) and no mitral regurgitation (MR). There was a large perimembranous VSD extending into the inlet portion with left-to-right shunt and well-formed windsock-like aneurysmal tissue that had multiple fenestrations from the right ventricular side (Fig. 1). The peak systolic pressure gradient across the VSD was 50 mmHg. The VSD from the left ventricular (LV) side measured 10 mm and the largest fenestration from right ventricle (RV) measured 5 mm. There was evidence of volume loaded left atrium (LA) with left atrium to aortic ratio (LA/Ao) ratio of 1.7. LV was severely dilated with LV end-diastolic diame-
ter (LVEDD) z-score of more than +4. There was good biventricular systolic function, unobstructed right and left ventricular outflows, and unobstructed pulmonary artery branches and left-sided aortic arch. There was no patent ductus arteriosus or aortic coarctation. Drainage of pulmonary and systemic veins was normal.

Management

Cardiac surgery was considered as the first option. However, due to associated abnormalities, cardiac surgery was deemed to carry slightly higher risks than ‘straight-forward’ VSD surgery. The parents of the patient were reluctant to agree to surgical repair and opted for cardiac catheterization and possible device closure. The existing aneurysmal tissue covering the VSD encouraged us to attempt VSD closure in the catheter laboratory with the intention of using the PFM Le VSD coil (PFM company, Köln, Germany), a transcatheter approach with less risk of heart block [5].

Cardiac catheterization procedure

The procedure was performed under elective intubation and trans-esophageal echocardiography (TEE) guidance. The intervention was done as previously described for VSD closure using different devices [5]. In brief, after obtaining femoral venous access and femoral arterial access, hemodynamic assessment was first performed to obtain pressures and saturations. The pulmonary artery pressure measured 28/9 mmHg with mean of 18 mmHg while the systemic pressure was 68/24 mmHg with mean of 43 mmHg. The ratio of pulmonary to systemic flow (Qp:Qs) was about 3:1 on room air. The LV angiogram was obtained in 30° left anterior oblique (LAO) view and 20° cranial tilt. The VSD was clearly identified and measurements were obtained (Fig. 2A). The LV angiogram was repeated while the 7 French (Fr) long sheath was parked into the ascending aorta across the VSD. Judging from angiograms and TEE assessment, the defect was considered too large and not suitable for the PFM coil. Because of its vicinity to the tricuspid valve, we feared the coil might get entangled in the tricuspid valve chordae. Instead, we decided to close the VSD using an 8 × 6 mm duct occluder device (AGA, Minnesota, USA). After having established the typical arteriovenous loop, the device was inserted from the venous side, utilizing a 7 Fr delivery sheath. After device implant, angiography was performed using a 4 Fr pigtail catheter, which showed the device in good place, with only mild residual leak (Fig. 2B). The intervention was uneventful. The procedure time was about 125 min. Fluoroscopy time was 16 min.

Pre procedure TEE

Large perimembranous VSD was covered by tricuspid aneurysmal tissue with some inlet extension. The VSD from LV side measured about 10 mm while from the RV side there were many exits, and the largest defect was about 5 mm (Fig. 3A).
Post-procedure TEE

Amplatzer 8 × 6 mm duct occluder was deployed in the sac of aneurysmal tissue with small residual leak from its lower aspect and only mild tricuspid valve regurgitation (Fig. 3B).

Follow up

On follow up, no clinical nor laboratory findings were suggestive for hemolysis, and ECG showed normal sinus rhythm with no heart block. Chest X-ray showed the device in position. Post-intervention echo revealed the device in an excellent position, with only mild TR, as before the procedure. There was a tiny (2 mm) residual leak as shown by color Doppler (Fig. 4). The infant was discharged after 3 days on Aspirin 5 mg/kg/day for 6 months. Two weeks later, the infant started to gain more weight (5.3 kg). Since the VSD device closure, he remains clinically stable with no symptoms or signs of congestive heart failure. Anti-failure medications were discontinued.

Discussion

Transcatheter closure of VSD was introduced in the late 80s and early 90s [6–8]. While closure of
muscular VSD found acceptance, it was the peri-
membranous VSD, which came into discredit, 
with previous studies finding a high rate of com-
plete atrio-ventricular block associated with this 
procedure [9]. In general, transcatheter closure 
has been limited to body weight above 10 kg 
[10,11]. Our patient had Down syndrome, a body 
weight of 4.8 kg, and he belonged to the category 
of patients in which surgical intervention is 
usually indicated. Hybrid procedure as an alterna-
tive was contemplated but would have included 
not only median thoracotomy, but also longer 
intensive care unit. The facts were that the patient 
had other associated congenital abnormalities, 
failure to thrive and, importantly, his parents were 
unwilling to have him undergo surgical repair. All

Figure 4. Trans-thoracic echocardiography (2D and color) at follow up showing the duct occluder device in good device with minimal residual left-to-right shunt across it. LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle.
this encouraged us to look for a less invasive alternative. The presence of a large aneurysmal tissue covering the VSD made the risk of heart block with transcatheter approach unlikely because any device positioned in the aneurysmal tissue will be a distance from the conduction tissue [12]. We opted to perform this procedure on our patient to spare him the potential complications of surgical repair.

As the VSD had some inlet extension, we felt that the newly released PFM VSD coil device might interfere with the tricuspid valve leaflets and its chordae, and possibly lead to more TR. The muscular VSD device was excluded as it would leave a 6 mm skirt from the RV side and would potentially interfere with the normal movement of the tricuspid valve. However, the duct occluder device had the right shape and hence seemed to be the right choice for this infant.

Off-label use of Amplatzer ductal occluders in perimembranous VSD has been reported [13,14]. As the entrance to the aneurysm from the LV side measured 10 mm, we decided an 8 × 6 mm duct occluder device would be the appropriate choice. This device along with its 4 mm skirt was positioned in the aneurysmal sac completely and away from the crest of the interventricular septum. After critical exploration with TEE and by fluoroscopy, the device was successfully released.

We believe that in such an aneurysmatic VSD and in infants with such low body weight, with or without association of Down syndrome, a transcatheter approach with its miniaturized devices and delivery systems, either with the PFM Le coil or with a duct occluder device, might be a good alternative to spare such infants the potential complications of surgery.

References