CASE REPORT

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

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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.

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

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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-

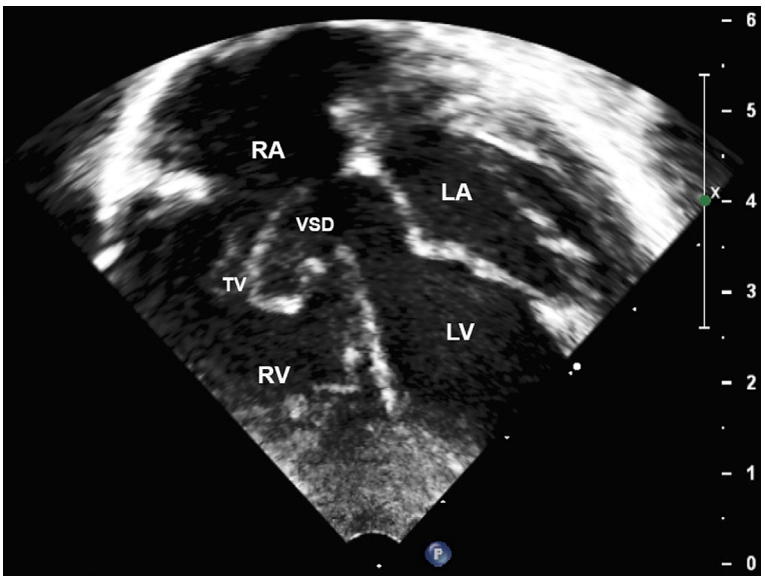

Figure 1. 2D Trans-thoracic apical view showing the ventricular septal defect (VSD) with aneurysmal tissue and multiple fenestrations from the right ventricular side. LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle, TV: tricuspid valve.