Hybrid muscular ventricular septal defect closure in a 4.5 kg infant followed by sildenafil treatment and transcatheter atrial septal defect occlusion

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A 4.5 kg 3-month-old girl was admitted with heart failure symptoms. Transthoracic echocardiography (TTE) revealed 5.5 mm midmuscular ventricular septal defect (VSD) with bidirectional shunt, 6 mm typically located secundum atrial septal defect (ASD), right heart enlargement and signs of pulmonary hypertension (PH). Our heart team decided to perform a hybrid VSD closure. After sterno- and pericardiotomy and heart apex elevation the right ventricle (RV) was punctured on the beating heart under epicardial echocardiography (EE) guidance (Figure 1). Right ventricle pressure of 40/0/6 mm Hg and arterial pressure of 64/40/50 mm Hg were measured. The VSD was crossed with a J-tip guidewire, and then a 8 French (Fr) delivery sheath was advanced. Taking into consideration the delicate manual maneuvers, the 7 mm Hyperion VSD Muscular Occluder (Comed B.V., The Netherlands/Lepu MT Company, China) was successfully deployed under EE (Figure 2) – an insignificant residual leak was observed. The intervention was uneventful. On the first day after the procedure during weaning from

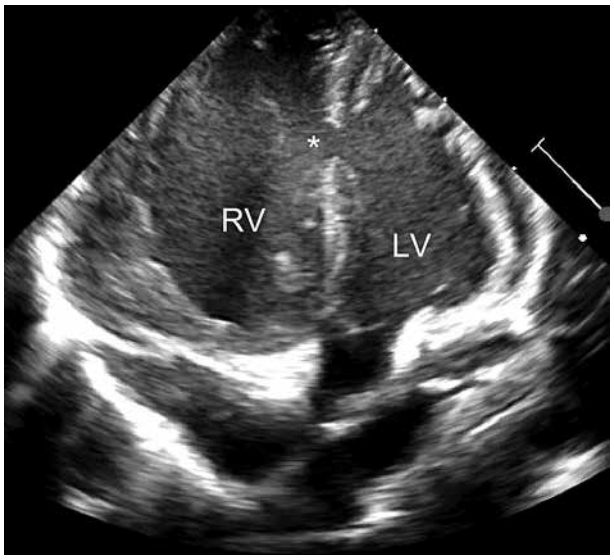

Figure 1. Epicardial echocardiography. Ventricular septal defect (VSD) – asterisk
RV – right ventricle, LV – left ventricle.

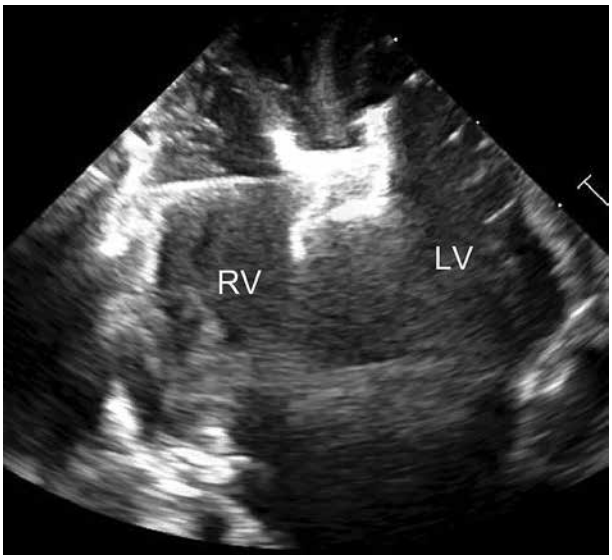

Figure 2. Epicardial echocardiography. 7 mm Hyperion VSD Muscular Occluder deployed in VSD over delivery cable
RV – right ventricle, LV – left ventricle.

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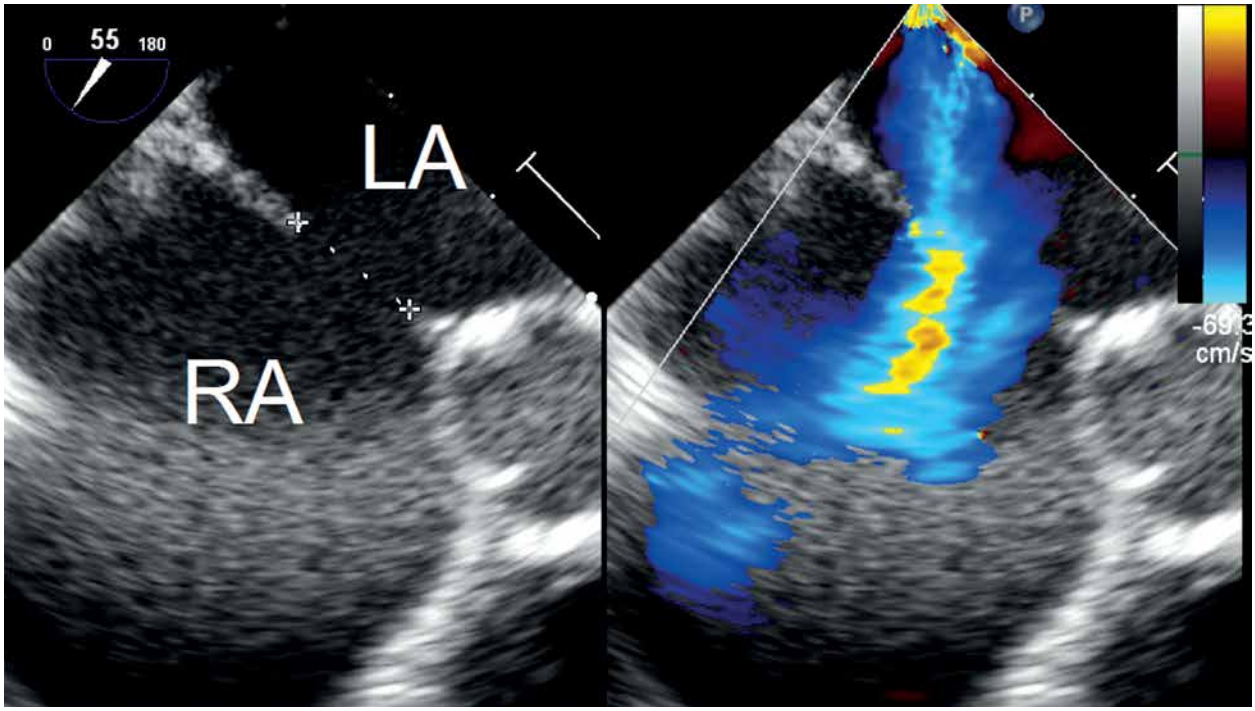

Figure 3. Transesophageal echocardiography with color Doppler. Atrial septal defect with left-to-right shunt
LA – left atrium, RA – right atrium.

mechanical ventilation a pulmonary hypertensive crisis occurred, manifested by significant bradycardia and arterial saturation fall. Therefore, NO inhalation, sildenafil (15 mg/day), milrinone and furosemide were administered. The treatment enabled successful weaning from mechanical ventilation after 1 week. The patient's 1-year follow-up with sildenafil administration at the same dose was uneventful. At the age of 15 months and weight of 10 kg, the girl was readmitted in order to perform heart catheterization with pulmonary artery (PA) pressure measurement. At that time diaphoresis during activity was noted in the anamnesis. In TTE 11 × 9 mm ASD with left-to-right shunt, two insignificant small residual muscular VSDs and right heart enlargement were observed. The PA pressure of 34/9/22 mm Hg and RV of 37/0/9 mm Hg were measured. Therefore, successful percutaneous ASD closure with a 12 mm Cocoon Septal Occluder (Vascular Innovations Co., Nonthaburi, Thailand, 8 Fr sheath) was performed under transesophageal guidance without balloon calibration (Figures 3, 4). In a 15-month follow-up the child was asymptomatic, TTE did not show residual leak through the ASD, and RV dimensions decreased, although on a decreasing dose of sildenafil.

Surgical closure of muscular VSDs in small infants is technically challenging [1]. Especially VSDs located apically are difficult to identify surgically. Hybrid VSD closure with Amplatzer occluders has become an attractive alternative option [2]. Recently published multicenter mid-term results have confirmed high efficacy and safety of such an approach and avoidance of cardiopulmonary bypass [2]. Our preliminary experience also indicates suitability of devices other than Amplatzer, such as Hyperion (China, VSD) or Cocoon (Thailand, ASD). Percutaneous access is another useful alternative, although limited by the patient's weight. Secundum ASD percutaneous closure is the method of choice even in small children [3].

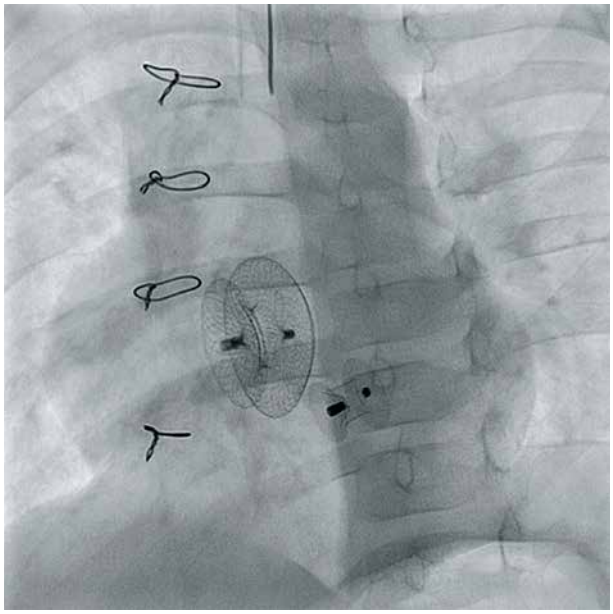

Figure 4. Fluoroscopy. 7 mm Hyperion VSD Muscular (below) and 12 mm Cocoon Septal Occluders

Pulmonary hypertensive crisis is an important cause of morbidity and mortality in patients with pulmonary

arterial hypertension secondary to congenital heart disease who require intervention [4]. Non-restrictive VSD accompanied by a large ASD may cause early development of PH. Sequentially performed interventions can counteract this process. Early hybrid VSD closure has become an attractive method of treatment, but it carries a potential risk of pulmonary hypertensive crisis in the post-procedural period.

Conflict of interest

The authors declare no conflict of interest.

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