Attempting and aborting percutaneous closure of a peculiar atrial septal defect: important contribution of multiple real-time imaging modalities

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The case of a 19-year-old female with a neonatal diagnosis of Tetralogy of Fallot and complete atrioventricular (AV) septal defect (AVSD) is described. She had had a corrective surgery at the age of 6. She did well afterwards despite recent complain of fatigability with mild arterial hypoxaemia. Transoesophageal echocardiography depicted a 12 mm atrial septal defect (ASD) with a bidirectional shunt, with features of a defect of the Ostium Primum type. The reason for the bidirectional shunt was unclear, and cardiac catheterization was performed. The presence of an ASD of the Ostium Primum type was confirmed with normal pulmonary arterial pressures and resistances. The distance to the AV valves seemed compatible with the implantation of a small device and percutaneous closure of the ASD was attempted. However, it was noted that the device deployment would cause deviation of the tricuspid regurgitation jet to the left atrium. Direct coronary sinus catheterization demonstrated its direct roof opening to the left atrium. The device impaired the drainage of the coronary sinus, actually increasing the right-to-left shunt. The implantation was aborted and surgical correction was proposed. Coronary sinus septal defect is a rare form of ASD. Its diagnosis is often difficult because of non-specific features. In this case, oxygen desaturation at presentation seemed to be explained by the regurgitating jet of the right component of AV valve through the Ostium primum defect into the left atrium. Percutaneous closure of the defect was attempted. However, it became clear that the device-increased the right-to-left shunt is also allowing the diagnosis of an unroofed coronary sinus. Under these circumstances, surgery seemed to be the best option.

Case report

The case of a 19-year-old female with a residual shunting at atrial level after previous surgery is described. She had had a neonatal diagnosis of Tetralogy of Fallot and complete atrioventricular (AV) septal defect (AVSD). She had a repeat palliative balloon dilatation of the pulmonary valve during the first 2 years of life and corrective surgery at the age of 6. Re-operation was required due to a significant residual ventricular septal defect (VSD). She did well postoperatively but recently she started to complain of some fatigability with mild arterial hypoxaemia (transcutaneous oxygen saturation around 90%). Transthoracic echocardiography revealed mild tricuspid regurgitation, next to the atrial septum, allowing the estimation of a systolic right ventricular pressure of 35 mmHg. There was mild left atrial dilatation; the pulmonary venous return was normal and there was no evidence of a left superior vena cava or coronary sinus dilatation; systolic ventricular function was preserved bilaterally. Transoesophageal echocardiography depicted a 12 mm atrial septal defect (ASD) with a bidirectional shunt, located low in the septum, with features of a defect of the Ostium Primum type (Figure 1). The reason for the bidirectional shunt was unclear. In order to clarify these findings, a cardiac catheterization was performed. The presence of an ASD of the Ostium Primum type, shunting bidirectionally, was confirmed. Pulmonary arterial pressures and resistances were normal; the QP:QS ratio was 2:1. Since the patient was unwilling to have further surgeries and the distance to the...
A–V valves seemed compatible with the implantation of a small device, percutaneous closure of the ASD was attempted. Intracardiac echo guidance was used at first, but was later replaced by transoesophageal echocardiography as it seemed more favourable for balloon sizing with this defect location (Figure 2). The measured defect diameter was 11.8 mm; an 8 mm Amplatzer® device was chosen, expecting that the size of the atrial discs with be sufficient to hold it in place. During deployment, however, it was noted that the device would cause deviation of the tricuspid regurgitation jet to the left atrium (Figure 3, Supplementary data, Clip 1). The coronary sinus was then selectively catheterized and angiography was performed, demonstrating a direct roof opening to the left atrium.
Indeed, the device would impair proper drainage of the coronary sinus to the right atrium and actually increase the right-to-left shunt at atrial level (Figure 4, Supplementary data, Clip 2). The attempts at device implantation were aborted and surgical correction was proposed, although not yet implemented.

**Discussion**

Coronary sinus septal defect is a rare form of ASD resulting from a lack of formation of the atrial fold with the sinus venosus. It is also named unroofed coronary sinus syndrome, often associated with the persistence of a left superior vena cava and partial pulmonary venous return anomalies. There is a large range of defects extending from the complete absence of its walls to small fenestrations between this vessel and the left atrial cavity. Diagnosis is often difficult because of non-specific clinical and echocardiographic features. The initial diagnosis is often made by the surgeon at repair of other congenital cardiac associated anomalies such as atrial septal defect, pulmonary or tricuspid atresia.

In this case, some decrease in oxygen saturation seemed to be explained by the streaming of the regurgitating jet of the right component of AV valve into the left atrium, despite normal pulmonary arterial pressures and resistances. ASD percutaneous closure is normally not recommended for Ostium Primum type of defects. However, this patient had had previous surgeries and the edges of the patches could have made it possible to stabilize the device without interference with the left component of AV valve. However, it became clear that device implantation increased the right-to-left shunt by redirecting the regurgitation jet of the right AV valve. In addition, real-time imaging assessment of the effects of device deployment before release also uncovered the diagnosis of an unroofed coronary sinus, which had previously passed unnoticed. In fact, shunting into the left atrium and impairment of the drainage of the coronary sinus were increased by the device. Under these circumstances, the procedure was aborted, as a surgical approach seemed a better option, allowing for the simultaneous correction of the ASD and of the unroofed coronary sinus.

**Supplementary data**

Supplementary data are available at European Journal of Echocardiography online.

**References**