Multiple coronary arterial fistulas to left ventricle associated with atrial septal defect

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Fig. 1 Still frames: subcostal modified bicaval view showed large ASD secundum (between arrows) with left to right shunt.



Coronary artery fistulas (CAF) rarely drain to the left ventricle (LV) leading to diastolic overload. We report a three-year-old girl in whom an atrial septal defect (ASD) on echocardiography with left to right shunt (figure 1) and possible CAF were diagnosed. At catheterization,

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Received 30 November 2014; revision accepted for publication 27 January 2015. coronary angiography identified multiple CAF to the left ventricle (figure 2). As the CAF were not amenable for closure and LV end diastolic pressure was normal, we closed the large ASD with a left to right shunt ratio (Qp/ Qs) of 2.4/1 by device to reduce the burden on the RV.

We were aware that with closure of ASD, there will be an additional overload to the LV, next to the shunt through CAF. Theoretically, this increase in diastolic volume might lead to LV function deterioration. Probably in an adult patient we would have considered leaving the ASD open.

After 6 months of follow-up, our patient remains asymptomatic, but her LV size increased within two weeks after ASD closure by 26.5% (still within normal range, z-score 0.01); figure 3. Accordingly, the right ventricle decreased in size and the paradoxical septal motion normalized (figure 2 C, D). This could also be observed in patients after ASD closure without any additional lesion.

A long-term regular follow-up by echocardiography might show signs of potential LV dysfunction. Evaluation by CT angiography, magnetic resonance imaging and myocardial stress tests might be necessary. In summary: CAF to the LV are extremely rare. In combination with ASD, the LV preload is decreased but after ASD closure it will increase in addition to the systemic-to-systemic shunt through the CAF. The result of long-standing LV overload and the initial management approach of this combination is not well defined. Further studies are needed to address this issue.



Fig. 2 Still frames: **A** and **B panel**: selective left coronary artery angiography. **A panel**: right antero-oblique (25°) and caudal (20°) view. **B panel**: left anterior oblique (90°) view. Both panels show dilated and tortuous left anterior descending artery and diagonal branch with contrast flow into the left ventricle. LAD: left anterior descending, Cx: circumflex artery, LV: left ventricle, LA: left atrium, RV: right ventricle, AAo: ascending aorta.



Fig. 3 A and B panels: parasternal long-axis echocardiography views. Left ventricle dimension increased after ASD closure from 2.2 cm to 3.0 cm (z-score -2.5 to 0.01).
C panel: M-mode before ASD closure shows paradoxical septal motion and dilated right ventricle.
D panel: M-mode after ASD closure shows normalization of septal motion with decrease in RV size.

Abbreviations, see figure 1.