Fixed...Or Needs Fixing?

D. Brian Newman, MD; William R. Miranda, MD; Heidi M. Connolly, MD

An asymptomatic 29-year-old female was referred for evaluation of a cardiac murmur. A preappointment electrocardiogram revealed right axis deviation, right bundle-branch block, and “crochetage sign” with notched R waves in the inferior leads (Figure 1A, arrows). Physical examination revealed normal jugular venous examination and a 2+ parasternal lift. Cardiac auscultation at the left upper sternal border (Figure 1B, phonocardiogram; Audio file in the Data Supplement) revealed normal S1, grade 2/6 early-peaking systolic ejection murmur, and fixed splitting of the second heart sound (A2-P2). Chest radiography (Figure 1C) showed enlarged right heart border (arrow) and prominent pulmonary arteries (arrow head). Transthoracic echocardiogram showed moderate right ventricular enlargement and large (28×18 mm) ostium secundum atrial septal defect (Figure 1D, asterisk; Movie I in the Data Supplement) with left-to-right shunting by color Doppler (Figure 1E; Movie II in the Data Supplement), and estimated right ventricular systolic pressure was 26 mmHg.

The term “fixed” denotes absence of significant respiratory variation of A2-P2 and is a classic feature of ostium secundum atrial septal defect.1,2 Although the mechanism is incompletely understood, one hypothesis suggests that reciprocal respirophasic augmentation of systemic venous return and left-to-right shunting (ie, increased systemic venous return and decreased shunting with inspiration; vice versa with expiration) are key hemodynamic features. The cumulative effect is relatively static right ventricular stroke volume throughout the respiratory cycle.3 In the isolated right bundle-branch block, the A2-P2 interval is wide but still shows respirophasic changes. The systolic ejection murmur results from increased flow across the right ventricular outflow tract, and occasionally a soft diastolic rumble originating from the tricuspid valve is present.

The patient underwent successful percutaneous device closure with a 34-mm Amplatzer septal occluder. A preprocedure transesophageal echocardiogram with 3-dimensional imaging was performed; an oval shaped atrial septal defect was identified, with the major axis measuring 26 mm (Figure 2; Movie III in the Data Supplement). The calculated shunt fraction (Qp:Qs) was 3:1 before percutaneous device closure. There was no residual shunt postprocedure.

Disclosures

None.

References


Key Words: atrial septal defect • blood pressure • bundle-branch block • heart murmurs • stroke volume
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Figure 2. Three-dimensional transesophageal echocardiogram demonstrating the oval shaped, ostium secundum atrial septal defect measuring 26 mm across the major axis.
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