ILLUSTRATIVE ECHOCARDIOGRAM

Echocardiographic "Pseudo-IHSS" Pattern in Atrial Septal Defect* 

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The patient is a 30-year-old asymptomatic man with a heart murmur first discovered two months prior to his referral to the Mayo Clinic for cardiac evaluation. Physical examination revealed right ventricular lift, grade 2/6 ejection systolic murmur at the pulmonic area, and persistently split second heart sound. A mid-diastolic flow murmur was heard at the left lower sternal border.

The roentgenogram of the chest revealed prominence of the main pulmonary artery segment and increased pulmonary vascular markings. An echocardiogram revealed right ventricular conduction delay (rsR' in lead V1) with a clockwise frontal plane QRS loop.

ECHOCARDIOGRAM

Figure 1 is a schematic drawing of normal anterior mitral leaflet motion. The nomenclature is as proposed by Edler.1

The echocardiogram of our patient (Fig 2 left) reveals features of right ventricular diastolic volume overload23—the right ventricular dimension (RVD) and RVD index are increased to 4.5 cm and 2.7 cm/cm², respectively (normal: RVD, 0.5 to 2.1 cm; RVD index, 0.3 to 1.1 cm/cm²). In addition, the ventricular septal motion is clearly abnormal (paradoxic) as both the septal echo and the left ventricular posterior wall echo move anteriorly during systole. The echocardiographic pattern of RV volume overload is nonspecific but, when taken into consideration along with the clinical findings in our patient, the diagnostic possibilities could be: (1) ASD, (2) partial anomalous pulmonary venous connection with intact atrial septum, and (3) combination of (1) and (2). Since echocardiographic examination would not be able to distinguish among these conditions, cardiac catheterization is required.

Of interest in this patient is the abnormal motion of the mitral valve (Fig 2 right), which reveals an exaggerated systolic anterior motion similar to that seen in patients with idiopathic hypertrophic subaortic stenosis (IHSS).4 5 This pseudo-IHSS pattern was first reported by Popp and Harrison,6 also in a patient with secundum atrial septal defect. Forker and Morgan7 recently reported the association of IHSS with secundum atrial septal defect in an adult patient, but neither the physical findings and electrocardiogram nor the postextrasystolic...

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beat response in our patients was that of IHSS. We have noticed a similar abnormality of mitral valve motion in a few additional patients with large left-to-right shunts at the atrial level. Observations in such patients have revealed a few possible differentiating features. The left ventricular internal dimension (LVID) is usually in the normal range or slightly decreased. The exaggerated systolic anterior motion of the mitral valve occurs gradually, and the diastolic closure slope of the mitral valve is normal. On the other hand, in patients with IHSS (Fig 3) the LVID usually is significantly decreased, and the anterior motion of the mitral valve occurs abruptly during midsystole and characteristically reveals a posterior motion during the isometric relaxation phase. Also, in IHSS the diastolic closure slope of the mitral valve is decreased, resembling mitral stenosis.

Cardiac catheterization data in our patient revealed an increase in oxygen saturation at the superior vena cava level and a further increase at the right atrial level. Double-sampling dye curves confirmed the presence of large left-to-right shunts at both the right atrial and superior vena cava levels, and the associated anatomic pulmonary venous connection from the right upper and middle lobes to the superior vena cava was demonstrated by angiography. The calculated pulmonary-to-systemic flow ratio (QP/Qs) was 3:4; pulmonary vascular resistance was normal.

References