Twin pregnancy, dyspnea, cyanosis, and a heart murmur

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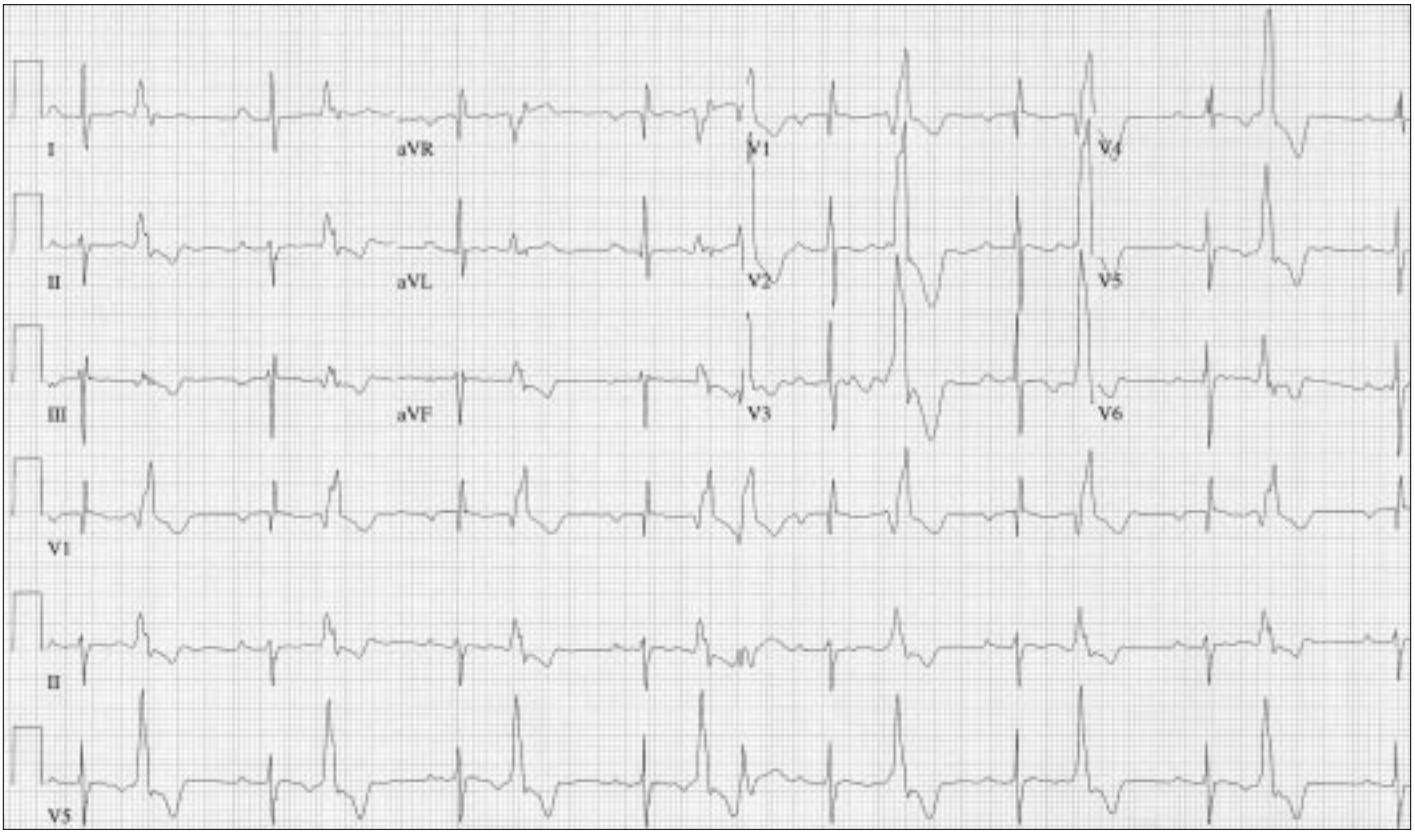

Figure 1. Electrocardiogram shows frequent ventricular premature complexes, long P-R interval, left atrial enlargement with left axis deviation of the P waves, left anterior fascicular block, and right ventricular enlargement.

A 32-year-old woman presented at 14 weeks gestation with a twin pregnancy. Her chief complaints were exertional dyspnea and orthopnea. She had a history of 2 spontaneous abortions followed by delivery of a live term infant 14 years previously. Two years previously she had suffered a cerebrovascular accident with eventual complete recovery.

Cardiovascular examination revealed mild cyanosis, 2+/4+ symmetrical clubbing of fingers and toes, jugular venous distention with large CV waves with an estimated mean right atrial pressure of 15 mm Hg, and a prominent left parasternal lift. The second heart sound was moderately and fixedly split with a loud second component. A grade 2/6 systolic ejection murmur was heard in the pulmonic area, and a grade 3/6 blowing pansystolic murmur was heard at the apex.

Electrocardiographic findings were a long P-R interval, left anterior fascicular block, and right ventricular enlargement, all typical of an atrioventricular septal defect *(Figure 1)*. In addition, there were frequent ventricular premature complexes and P waves indicating left atrial enlargement and having a frontal plane axis of –5°. This much left axis deviation of the P wave is unusual in atrioventricular septal defect limited to the lowermost portion of the atrial septum, so-called ostium primum atrial septal defect (1, 2), but often is found when the entire atrial septum is missing (3, 4).

Echocardiography confirmed the presence of a common (single) atrium *(Figure 2)* with bidirectional shunting. The ven-

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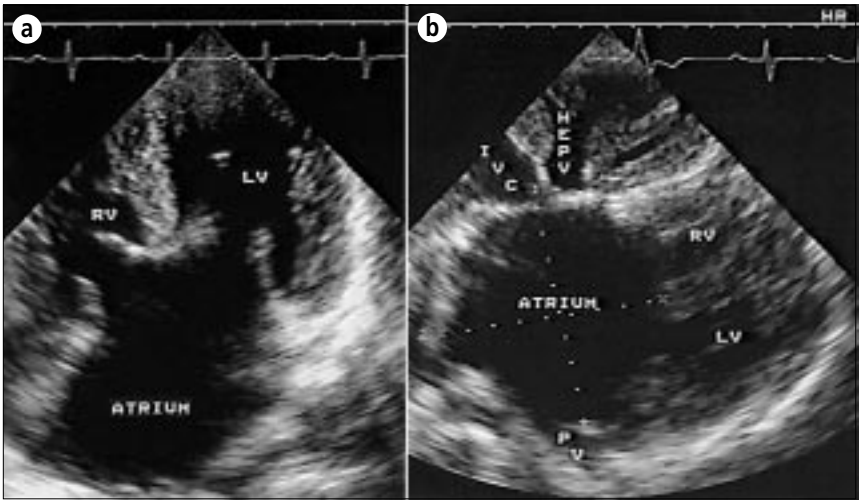

Figure 2. Two-dimensional echocardiographic frames in the **(a)** apical 4-chamber and **(b)** subcostal views. The atrial septum is completely absent, and the common atrium is large. There is a common atrioventricular valve with separate left and right orifices. Markers are spaced at 1-cm intervals. HEPV indicates hepatic vein; IVC, inferior vena cava; LV, left ventricle; PV, pulmonary vein; RV, right ventricle.

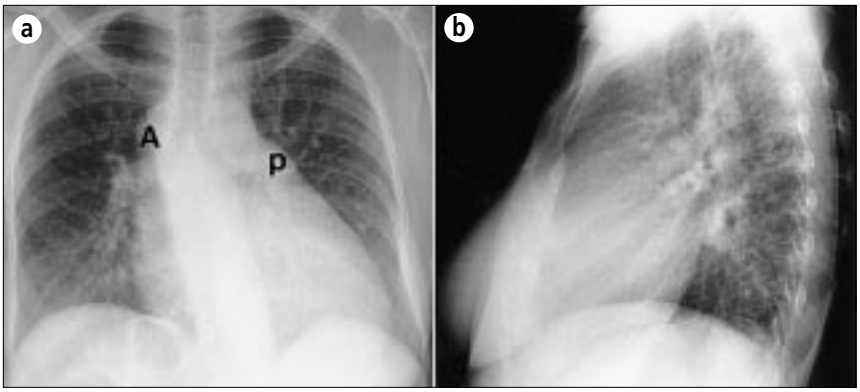

Figure 3. (a) Posteroanterior and **(b)** left lateral chest roentgenograms; A indicates dilated azygos vein; p, dilated pulmonary trunk.

Figure 4. An angiogram: the catheter passes up the azygos continuation of the inferior vena cava to the superior vena. From there its course is through the common atrium and the left-sided orifice of the common atrioventricular valve, out the long and narrow (so-called goose neck) left ventricular outflow tract into a slightly dilated ascending aorta.

tricular portion of the defect was closed by the attachments of the common atrioventricular valve. This valve had left and right orifices; both were mildly to moderately regurgitant. The velocity of the regurgitant jet, as assessed by Doppler ultrasound, and the height of the neck veins indicated that pulmonary arterial systolic pressure was approximately 75 mm Hg when systemic arterial systolic pressure, measured by cuff, was 100 mm Hg.

A posteroanterior chest roentgenogram showed scoliosis, left-sided aortic arch, left-sided cardiac apex, dilated azygos vein, generalized cardiomegaly, prominent pulmonary trunk, and plethoric lungs *(Figure 3a)*. The colon filled the right upper quadrant, interposing between the liver and diaphragm (Chilaiditi's syndrome). No stomach bubble was identified, and thus abdominal situs could not be determined. On the lateral chest roentgenogram, the retrosternal space was filled by a large right ventricle *(Figure 3b)*. An earlier cardiac catheterization at another institution documented intrahepatic interruption of the inferior vena cava with azygos continuation as the reason for the enlarged azygos vein *(Figure 4)*.

Because of the maternal risk posed by pulmonary hypertension, congestive heart failure, and the extra volume load of a twin pregnancy, therapeutic abortion was recommended, readily accepted by the patient, and performed uneventfully. Nearly 3 years later, however, she returned in the third trimester of a single pregnancy. She remained in the hospital until delivery, when care was taken to avoid hypoxia, hypotension, and air embolism.

Common atrium, a rare congenital malformation, has many similarities with the more usual atrioventricular septal defect limited to the lowermost portion of the atrial septum: a left-to-right shunt; an identical type of common atrioventricular valve that may have only one orifice (5) but usually has right and left orifices (3, 5, 6), either or both of which may be incompetent; and a rather characteristic electrocardiogram showing right ventricular enlargement, left anterior fascicular block and, in 50% of patients, a long P-R interval (7, 8). The right ventricular enlargement is due to the volume load imposed on the right ventricle by the left-to-right shunt, which often is accompanied by some degree of pulmonary arterial hypertension. In atrioventricular septal defect the atrioventricular node is in the posterior atrial wall, and the nonbranching portion of the His bundle is unusually long (9). Such an arrangement could lead to prolonged internodal and/or A-V nodal-His-Purkinje conduction. Waldo et al have shown that prolonged internodal conduction is the culprit in most patients with atrioventricular septal defect and a long P-R interval (10). A left anterior fascicle that is thinner and longer than usual has been blamed for the characteristic left axis deviation of the QRS complex in the frontal plane (8).

In several respects, however, common atrium differs from atrioventricular septal defect limited to the lowermost portion of the atrial septum. The greater likelihood of left axis deviation of the P wave has been mentioned. Patients with common atrium

rarely have Down syndrome but not uncommonly have the Ellis-van Creveld syndrome (8). Common atrium apparently also is more likely to be associated with cardiac malposition, especially isolated levocardia (4). Anomalies of the systemic veins seem to occur considerably more frequently in those with common atrium. This patient and at least 2 others have had intrahepatic interruption of the inferior vena cava with azygos continuation (6), and persistence of the left hemiazygos vein has been described (3). Persistent left superior vena cava, occasionally without a right superior vena cava, occurs in nearly half the patients (3–5). Finally, common atrium is associated with pulmonary and systemic venous admixture at the atrial level. The right-to-left component of this admixture inevitably produces some decrease in systemic arterial oxygen saturation. This cyanosis is not necessarily a sign of Eisenmenger physiology.

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