A 21-year-old pregnant woman with congenital heart disease

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Figure 1. Admission electrocardiogram. See text for explication.

A 21-year-old woman was transferred from another hospital in her 23rd week of pregnancy. She had had an audible precordial murmur the day of her birth and had had pneumonia three times in the first few years of life. Otherwise, she was asymptomatic during childhood and adolescence. Aside from being small (4'11" tall and 85 lbs before becoming pregnant), she had developed normally. When she was transferred, she was asymptomatic and on no medication.

Pertinent physical findings were a regular pulse at 96 beats/min; a blood pressure of 116/60 mm Hg; normal neck veins; brisk, full, symmetrical arterial pulses in the arms and legs with no radial-femoral delay; a loud and palpable second heart sound along the upper and mid left sternal border; and a small apical impulse just outside the left mid-clavicular line. A harsh, grade 4/6, systolic ejection murmur, heard over the entire chest and back and in the neck, was loudest at the cardiac base and of

equal intensity on the right and left sides. A decrescendo diastolic murmur was maximal in the third left intercostal space. There was no gallop, ejection click, cyanosis, or clubbing.

The admission electrocardiogram showed normal sinus rhythm, left axis deviation of the QRS complex, and no septal Q waves in leads I, aVL, V₅, or V₆, but Q waves were present in leads II, III, aVF, and V₁ (Figure 1). The history, physical examination, chest radiograph (Figure 2), and electrocardiogram provided some, albeit incomplete, insight into the patient's congenital cardiac malformations and the consequent pathophysiology. A systolic murmur heard the day

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Figure 2. Admission posterior-anterior chest radiograph. The proximal right and left pulmonary arteries are at the same level. A hump is seen on the left cardiac border, and the ascending aorta is seen on the left side of the cardiac base rather than the right. The right border of the descending aorta can be seen to cross the spine obliquely from right to left and arrive in its usual location at the level of the diaphragm. These are features suggesting congenitally corrected transposition of the great arteries. In addition, the dilated pulmonary arteries suggest poststenotic dilation and/or increased pulmonary arterial pressure/flow.

of birth suggested right or left ventricular outflow obstruction; it would have taken several days for pulmonary vascular resistance to fall sufficiently for the murmur of a ventricular septal defect to be heard. The patient's brisk pulses excluded aortic stenosis, so the outflow tract obstruction had to be right ventricular. Frequent bouts of pneumonia in infancy suggested a sizable left-to-right shunt, a supposition supported by the large pulmonary arteries and pulmonary plethora noted on chest radiograph (Figure 2). The absence of septal Q waves in leads I, aVL, V₅, and V₆ and their presence in leads V₁, II, III, and aVF suggested ventricular inversion, an integral feature of congenitally corrected transposition of the great arteries, a diagnosis also brought to mind by the chest radiograph (Figure 2) (1). The anterior position of the aortic valve in that condition would have produced the loud and palpable second heart sound.

In congenitally corrected transposition of the great arteries, there are discordant atrioventricular and ventriculo-arterial connections. The right atrium receives blood from the two vena cavae and sends it across a mitral valve into a morphologic left ventricle that pumps it across a pulmonic valve, which is in fibrous continuity with the mitral valve, and into the pulmonary artery. Pulmonary venous blood travels through the left atrium and across a tricuspid valve into a morphologic right ventricle that pumps the blood across the infundibulum and the aortic valve into the aorta. Thus, ventriculoarterial discordance "corrects" the atrioventricular discordance, and the path of the blood through the heart is normal if there are no other malformations (2). Such patients have lived into their 70s (3). Whether the morphologic right ventricle, i.e., the systemic ventricle, in a patient with isolated congenitally corrected transposition of the great arteries functions as long and as well as a morphologic left

ventricle does in a normal person has been affirmed by some experts (4) and denied by others (5).

The vast majority of patients with congenitally corrected transposition of the great arteries, however, have associated cardiac malformations. Three of these occur frequently enough to be considered part of the anomaly: ventricular septal defect in approximately two thirds of patients (2); left ventricular (pulmonary ventricular) outflow tract obstruction, which can be valvular and/or subvalvular, in one half (1); and anatomic abnormalities of the tricuspid (systemic atrioventricular) valve in 90%, many of which are not clinically significant and the most common of which is Ebstein's anomaly (2). Other malformations seen in patients with congenitally corrected transposition of the great arteries include atrial septal defect; subaortic obstruction, which is often associated with coarctation of the aorta; aortic valve atresia with hypoplasia of the morphologic right ventricle; and pulmonary atresia with hypoplasia of the morphologic left ventricle.

Because of malalignment of the atrial septum with the inlet ventricular septum, the atrioventricular conduction system is abnormal (2), and up to 75% of patients with congenitally corrected transposition of the great arteries eventually have atrioventricular block ranging from first degree to third degree (1). Wolff-Parkinson-White type ventricular preexcitation may occur, as it does in other persons with Ebstein's anomaly of the tricuspid valve, and some, but not all, supraventricular arrhythmias in congenitally corrected transposition of the great arteries are associated with a left-sided accessory pathway (1). Because the morphologic left ventricle and the left bundle branch lie to the right of the morphologic right ventricle in congenitally corrected transposition of the great arteries, initial septal depolarization is from right to left and often inferosuperiorly as well, as seen in Figure 1.

As is often the case, especially with congenital heart disease, the echocardiogram and Doppler examination added important diagnostic information to that obtained by history, physical, chest radiograph, and electrocardiogram. In this patient, echo-Doppler confirmed congenitally corrected transposition of the great arteries, a nonrestrictive ventricular septal defect, and pulmonic valvular and subvalvular stenosis with a 75 mm Hg peak systolic pressure gradient between the pulmonary ventricle (morphologic left ventricle) and the pulmonary artery. Because her systemic arterial systolic pressure at the time was 105 mm Hg and the ventricular septal defect was nonrestrictive, her pulmonary arterial systolic pressure was approximately 30 mm Hg. The echo-Doppler also revealed a restrictive patent ductus arteriosus, which explained the full, brisk pulses and the decrescendo diastolic murmur. The systolic component of the continuous murmur of the patent ductus was obscured by the louder murmur of pulmonic stenosis. Both ventricles had normal systolic function, and both atrioventricular valves were completely competent.

The patient stayed on the obstetrical service throughout the remainder of her pregnancy. She entered active labor at 34 weeks of gestation and under epidural anesthesia delivered a 2425 g daughter with Apgar scores of 8 and 9. Bilateral tubal ligation

was then performed. Mother and daughter were doing well at discharge on postoperative day 4.

Two questions remain. First, why, with such a complex congenital cardiac malformation, was this woman asymptomatic after the first years of life and able to have a successful pregnancy? It was because the severe malformations were balanced in such a way that the circulatory system was quite adequate. Pulmonic stenosis prevented early severe heart failure or subsequent Eisenmenger reaction from the nonrestrictive ventricular septal defect. At the same time, there was sufficient blood flow through the pulmonic valve and the restrictive ductus to prevent cyanosis and allow normal activity and a successful pregnancy. Thus far she has avoided two common accompaniments of congenitally corrected transposition of the great arteries, i.e., tricuspid (systemic atrioventricular) valvular regurgitation and atrioventricular block.

Second, should the patient undergo operative repair? An article from the Mayo Clinic points out that most persons with congenitally corrected transposition of the great arteries eventually undergo cardiac surgery and that for many, the operation comes too late for optimal results (6). Those statements are difficult to refute. On the other hand, in this patient, so-called complete repair would require extensive complicated surgery,

which is difficult to recommend to an asymptomatic patient. In addition, few institutions have a Gordon Danielson, one of the paper's authors and one of the foremost congenital cardiac surgeons of his or any other day. Most importantly, the patient prefers not to undergo an operation as long as she feels well.

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