Dyspnea in a pregnant 20-year-old woman with electrocardiographic right ventricular enlargement

D. Luke Glancy, MD, Sai K. Devarapalli, MD, Babu Makkena, MD, and Pramilla Subramaniam, MD

Figure. Electrocardiogram recorded when the patient came to the clinic. See text for description.

A 20-year-old woman, who was 2 months pregnant, came to the cardiology clinic complaining of exertional dyspnea of 6-months duration. Her chest was clear to auscultation, and the neck veins were modestly elevated. The second heart sound was single and loud. There was no cyanosis or clubbing. A midline sternal scar was noted and was described by the patient as the incision for a heart operation when she was 2 months old. An electrocardiogram showed sinus arrhythmia, marked right axis deviation ($+135^\circ$ as judged by equally positive QRS complexes in leads II and aVR), and right ventricular enlargement with a qR complex in lead V₁ (Figure).

Electrocardiograms with this extreme degree of right ventricular enlargement are rare in adults but may be seen in severe pulmonic stenosis with an intact ventricular septum, tetralogy of Fallot, the Eisenmenger reaction, primary pulmonary hypertension, and simple transposition of the great

arteries with surgical redirection of the pulmonary and systemic venous blood at the atrial level via a baffle, i.e., the Albert-Senning-Mustard operation (1–3). The first two diagnoses are ruled out by the absence of a precordial murmur. The second and third are excluded by the lack of cyanosis. The heart operation at 2 months of age indicates a congenital malformation, thus excluding primary pulmonary hypertension. The absence of right bundle branch block strongly suggests that the operation did not include a right ventriculotomy. Thus, the only tenable diagnosis is transposition surgically corrected at the atrial level.

From the Sections of Cardiology, Departments of Medicine, Louisiana State University Health Sciences Center and University Hospital, New Orleans, Louisiana.

Corresponding author: D. Luke Glancy, MD, 7300 Lakeshore Drive, #30, New Orleans, Louisiana 70124 (e-mail: dglanc@lsuhsc.edu).

The diagnosis was confirmed by echocardiography that revealed ventriculoarterial discordance (transposition of the great arteries), severe dilatation and hypokinesis of the systemic (anatomic right) ventricle, normal function of the pulmonary (anatomic left) ventricle, a normally functioning intraatrial baffle, mild regurgitation of both atrioventricular valves, a right atrial mean pressure of approximately 10 mm Hg, and a pulmonary arterial systolic pressure of approximately 30 mm Hg.

Without intervention, 90% of infants with simple transposition die within a year of birth (4). With the Albert-Senning-Mustard operation, the majority become adults. Thus, it is a remarkably successful operation. A residuum of the operation, however, is an anatomic right ventricle serving as the systemic ventricle, which may fail after decades of pressure overload. One solution has been to do the Idriss-Jatene arterial-switch operation (5, 6) and undo the intraatrial baffle. In this patient with a left ventricular systolic pressure <70% of systemic arterial pressure, the arterial switch operation would have to be preceded by pulmonary arterial banding to prepare the currently low-pressure left ventricle for the high-pressure systemic arterial circulation. After several months, the band would be removed, the intraatrial baffle undone, and the great arteries switched. This two-stage operation has rarely been performed successfully in patients beyond their teens (7). Thus, cardiac transplantation will probably be this patient's best option when her cardiac failure worsens despite medications.

A more immediate problem is the pregnancy. The patient is in New York Heart Association Class IIID: she has dyspnea with less than ordinary activity and has objective evidence of severe cardiovascular disease, i.e., a severely dilated and poorly

contractile systemic ventricle (8). Thus, a good case can be made for terminating the pregnancy (9, 10). She wants to continue with the pregnancy, however, and will probably do so in spite of medical advice to the contrary.

1. Albert HM. Surgical correction of transposition of the great vessels. *Surg Forum* 1955;5:74-77.
2. Senning A. Surgical correction of transposition of the great vessels. *Surgery* 1959;45(6):966-980.
3. Mustard WT. Successful two-stage correction of transposition of the great vessels. *Surgery* 1964;55:469-472.
4. Liebman J, Cullum L, Belloc NB. Natural history of transposition of the great arteries: anatomy and birth and death characteristics. *Circulation* 1969;40(2):237-262.
5. Idriss FS, Goldstein IR, Grana L, French D, Potts WJ. A new technique for complete correction of transposition of the great vessels. An experimental study with a preliminary clinical report. *Circulation* 1961;24:5-11.
6. Jatene AD, Fontes VF, Paulista PP, Souza LC, Neger F, Galantier M, Sousa JE. Anatomic correction of transposition of the great vessels. *J Thorac Cardiovasc Surg* 1976;72(3):364-370.
7. Lee C, Kuehl KS. Long-term outcomes and management of the adult with systemic right ventricle. *Am Coll Cardiol Current Journal Review* 2004;13:60-64.
8. Criteria Committee of the New York Heart Association. *Nomenclature and Criteria for Diagnosis of Diseases of the Heart and Great Vessels*, 9th ed. Boston: Little, Brown and Co, 1994:253-255.
9. Perloff JK. Pregnancy in congenital heart disease; the mother and the fetus. In Perloff JK, Child JS, eds. *Congenital Heart Disease in Adults*. Philadelphia: WB Saunders, 1991:124-140.
10. Daniels CJ, Allen HD, Fontana ME. Adolescent and young adult cardiology. In Allen HD, Gutgesell HP, Clark EB, Driscoll DJ, eds. *Moss and Adams' Heart Disease in Infants, Children, and Adolescents, Including the Fetus and Young Adult*, 6th ed. Philadelphia: Lippincott Williams & Wilkins, 2001:582-599.