Dextrorotation of the Heart in Rheumatic Mitral Stenosis

Report of a Case Complicated by Obesity

By Nathan Pocker, M.D., and Israel Steinberg, M.D.

DEXTROTATION of the heart occurring in rheumatic heart disease is rare. Only Schreiner and Lyons and Steinberg and Ayres have reported cases. Dextrootation of the heart is a congenital anomaly in which the heart is displaced toward the right hemithorax and rotated in a counterclockwise direction on its longitudinal axis. This results in an altered spatial relation of the cardiovascular structures within the thorax. The apex of the heart lies behind the sternum, and the two ventricles are side by side. The right ventricle forms the right cardiac border, and the left ventricle is no longer the posterior ventricle but becomes part of the anterior aspect of the heart. The great vessels arise normally but the rotation to the right causes medial displacement of the pulmonary artery and opening of the loop between the ascending and descending aorta. Dextrootation may be simulated by turning a patient with a normally placed heart into the left anterior oblique position.

Recent reports have dealt with dextrootation of the heart and differentiated it from other spatial derangements. Dextrootation of the heart has been frequently associated with other congenital cardiac anomalies, but only rarely with acquired rheumatic valvular heart disease. The purpose of this paper is to report a third case of dextrootation of the heart and rheumatic mitral stenosis.

Case Report

A 40-year-old white man was admitted to the Rockefeller Hospital for treatment of life-long obesity. He denied having had sore throat, rheumatic fever, or chorea. Two years prior to admission he had had sudden onset of paralysis with numbness of the left side of the body and aphasia. He was hospitalized for 1 month because of a cerebral embolism. Findings at that time were atrial fibrillation, left hemiparesis, and obesity. The paralysis cleared rapidly during several months. Two years prior to admission he weighed 439 pounds; he later reduced to 355 pounds. He became asymptomatic until 1 year prior to admission, when following sudden severe chest pain, he was again hospitalized. Atrial fibrillation and a normal transaminase determination were found and when the chest pain subsided, he was discharged after a stay of 5 days. The electrocardiogram showed right ventricular hypertrophy and atrial fibrillation at the rate of 75 per minute. The heart sounds were distant and there were no murmurs; the blood pressure was 130/70 mm. Hg. Another episode of sudden chest pain occurred 6 months prior to admission, and again the electrocardiogram showed atrial fibrillation but without evidence of myocardial infarction. The patient has been obese for most of his life but denied having had rheumatic fever. At age 12, his weight was 181 pounds; during high school and college, it was 215 pounds; in 1947, at age 23, he weighed 300 pounds, and at age 31 he was 409 pounds. On admission to the Rockefeller Hospital on September 9, 1963, he weighed 355 pounds and measured 5 feet, 10 inches.

The chest x-ray showed a globular heart. The right cardiac border was unusually prominent in the frontal and left anterior oblique projections. There was also bulging of the pulmonary artery segment. The left atrial and ventricular contours appeared normal (fig. 1). The electrocardiogram showed atrial fibrillations, with a ventricular rate of 64. There was no deviation of the electrical axis. The T waves were flat in all standard leads, upright in leads V1 and V2, and inverted in V3 to V6. No significant ST abnormalities were noted. Because of the unusual roentgenograms, history of hemiplegia, and cardiac pain, intravenous angiography was advised. This revealed a slightly enlarged superi-
or vena cava and a posteriorly rotated and enlarged right atrium. The right ventricle, pulmonary artery, pulmonary arterial and venous branches, and left atrium were enlarged. The left atrium measured 13 by 9 cm. (The average normal measurement is 8 by 5.5 cm.\textsuperscript{5}) The left ventricle was also rotated into the left anterior oblique position and was not enlarged. There was considerable delay in emptying of the left atrium; indeed, it remained opacified for 18 seconds. Thrombi were not present although the left atrial appendage was not visualized. The ascending aorta was normal and measured 28 mm. in diameter (fig. 2). The mitral stenosis was believed to be due to rheumatic rather than to congenital heart disease.

**Discussion**

The interrelation between acquired (infectious) and congenital heart disease have frequently been recorded in the literature.\textsuperscript{6, 7} The Lutembacher's syndrome, though rare, is probably the best known example of a con-

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**Figure 1**

A, left. Frontal teleradiogram of the chest showing undue prominence of the right Cardiac silhouette and fullness of the pulmonary artery segment. B, right. Left anterior oblique x-ray showing enlargement of the right ventricle. There is no impingement on the barium-filled esophagus.

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**Figure 2**

Frontal intravenous serial angiograms. A, left. The right atrium is rotated toward the right hemithorax. The right ventricle forms the right inferior border of the cardiac silhouette. B, center. The right ventricle and pulmonary artery are dextrorotated and the interventricular septum is vertical. C, right. The left atrium is enlarged, centrally placed, and long opacified. The left ventricle is in systole, lies medially, and is nonopacified. The aorta is also dextrorotated with prominence of the aortic window.
DEXTOROTATION IN MITRAL STENOSIS

genital atrial defect complicated by rheumatic mitral stenosis.\textsuperscript{9}\textsuperscript{,}\textsuperscript{11} Rheumatic heart disease (mitral stenosis) has also been found in association with a number of other congenital cardiovascular malformations.

In mitral stenosis there is usually enlargement of the atrium, pulmonary artery, and right ventricle. The hypertrophied right ventricle casts a more prominent shadow in the right hemithorax in dextrorotation of the heart, and this with the enlarged left atrium distorts the left cardiac border (fig. 1). Angiocardiography, by delineating the cardiac chambers and revealing dextrorotation of the heart, can establish the diagnosis of mitral valvular disease and dextrorotation of the heart. In this case, this proved to be important not only for diagnosis but also for treatment. Indeed, once the patient's weight has been reduced, surgery for relief of mitral stenosis is planned; this may also prevent systemic emboli.

Recently, the patient's 1-year-old son was seen at another hospital because of an upper respiratory infection. The roentgenogram of the chest disclosed dextrorotation of the heart (fig. 3). To our knowledge, this is the first instance of the occurrence of dextrorotation of the heart in father and son.

Figure 3
Telerentgenogram of the chest showing dextrorotation of the heart in son of patient. (Courtesy of Dr. Sylvia P. Griffiths.)

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Summary and Conclusions
Diagnosis of dextrorotation of the heart and mitral stenosis was difficult in a 40-year-old man, with recurrent attacks of hemiplegia and cardiac pain, because of obesity and absence of cardiac murmurs. An unusual contour of the heart was shown by angiocardiography to be due to dextrorotation of the cardiac chambers and mitral stenosis. The enlargement of the right ventricle, pulmonary artery, and left atrium established the diagnosis of mitral stenosis and helped to explain the etiology of the chronic atrial fibrillation. Hemiplegia, aphasia, and thoracic pains were attributed to left atrial thrombi. A son of the patient, aged 1 year, also had dextrorotation of the heart, which, however, was uncomplicated.

The combination of dextrorotation of the heart and rheumatic mitral stenosis is rare; only two other cases have been previously reported. Dextrorotation of the heart should be included in the list of congenital malformations that may be complicated by rheumatic heart disease.

Acknowledgment
Drs. Jules Hirsch and Edward H. Ahrens, Jr., kindly referred and gave permission to report this case.

References
6. Dry, T. J., and Connolly, D. C.: Inflammatory Complications of Congenital Heart Disease in


Chorea.—May 7, 1812. Miss C., aged nineteen, from the time of her birth till she was fourteen years of age, enjoyed good health. . . .

Between four and five years ago the present patient began to suffer great debility, and shortness of breath, which were now and then accompanied with some degree of chorea, throughout the whole body. This complaint has increased till the present time, varying, however, at different periods, as to its force, and not obtaining any essential or permanent relief from Ether, Camphor, blisters on the head and other parts, a seton in her neck, the cold bath, sea bathing, warm bathing, &c. . . .

Miss C. now left Bath with the following instructions:—1st. Wholly to abstain from spirits and fermented liquors; to eat meat only once a-day, and always to eat less than her appetite demanded.

2dly. To walk every day as far as her strength would permit, avoiding the heat of the day.

3dly. To have four leeches applied once a week to the temples, encouraging the subsequent discharge of blood by washing the orifices with lukewarm water.

4thly. To employ a cold shower bath every other morning before breakfast.

5thly. To keep her hair constantly close cut.

6thly. To persevere for a month in the use of the pills last prescribed.—Collections from the Unpublished Medical Writings of the Late Caleb Hillier Parry, M.D.F.R.S. Vol. I., London, Underwoods, Fleet-Street, 1825, pp. 588-590.
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