Mitral Valvotomy in a Patient with Dextrocardia and Situs Inversus

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Mitral valvotomy was performed in a 45-year-old woman with mirror-image dextrocardia, situs inversus and acquired mitral stenosis. The literature was reviewed and a total of 15 reported cases was found, with surgical treatment in 4. Closed mitral valvotomy has been the treatment in all cases to date, and was successful in each. An open-heart approach was perfectly feasible in our case, but was reserved for the future when valve replacement might be required.

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Dextrocardia with situs inversus is an uncommon defect, and the association of this congenital lesion with acquired valvular heart disease is even rarer. A recent experience with this combination led to our review of the literature and case report.

Case Report

The patient was a 45-year-old woman with a history of acute rheumatic fever 22 years before, at which time she was found to have a heart murmur and dextrocardia. Over the subsequent nine-month period the patient was admitted to the hospital three more times for treatment of the rheumatic fever. She was then asymptomatic until one year prior to her admission for cardiac evaluation on Oct. 11, 1971. During this period she had noted a gradually progressive increase in dyspnea on exertion, easy fatigability, difficulty in doing her housework, paroxysmal nocturnal dyspnea, bilateral pedal edema, and two-pillow orthopnea. At the time of admission the patient was considered to have cardiac functional disease class 3, but had been taking no cardiac medication.

Physical examination revealed a moderately obese woman with a blood pressure of 130/90 mm Hg and normal sinus rhythm at a rate of 88/min. The physical positive findings were limited to the heart. The apex beat was in the fifth right intercostal space at the midclavicular line. There were no palpable heart sounds or thrills, and there was no evidence of ventricular hypertrophy. The first heart sound was moderately accentuated as was the pulmonary component of the second heart sound. There was no third or fourth heart sound. An opening snap was heard over the entire precordium, which the phonocardiogram demonstrated .06 sec after the aortic closure, and there was a grade 2/6 diastolic rumbling murmur at the apex, with presystolic accentuation. There was no systolic murmur. The chest x-ray film revealed dextrocardia and situs inversus, a cardiothoracic ratio of 0.4/0.8, left atrial enlargement, and increased prominence of the central and upper lobe vessels (Fig 1). The electrocardiogram was consistent with dextrocardia and also showed...
FIGURE 2. Electrocardiogram consistent with (a) dextrocardia and (b) left atrial hypertrophy.

normal sinus rhythm, a mean QRS axis of +120 degrees, and left atrial enlargement (Fig 2). Routine laboratory studies were entirely normal.

Cardiac catheterization and angiography revealed dextrocardia, situs inversus and severe mitral stenosis (Table 1). There was no mitral regurgitation, valvular calcification, aortic valvular disease, or intracardiac shunt. The end-diastolic gradient across the valve was 14 mm Hg at rest, and 40 mm Hg after supine bicycle exercise, indicating severe mitral stenosis. The pulmonary artery pressure of 50/25 mm Hg while at rest rose to 70/42 mm Hg with exercise. The cardiac index was 2.8 l/M2/min (Fick method) while at rest, and the pulmonary vascular resistance was normal.

On Dec 8, 1971, the patient underwent thoracotomy through the fourth right intercostal space. Exploration of the left atrium revealed mitral stenosis typical of rheumatic disease. The valve was thickened, the orifice admitted only one fingertip, and there was rubbery fusion of the commissures and shortening of the chordae tendinae. There was no mitral regurgitation. Finger fracture was attempted, but was not successful. The transventricular dilator was then inserted through an apex stab wound, and the valve was serially opened to 40 mm, with the production of only minimal regurgitation. The patient's recovery after operation was complicated only by mild bilateral atelectasis. She was discharged on December 19, at which time she was afebrile, the lungs were clear, and the first heart sound was slightly increased in intensity, but there was no murmur or opening snap. The patient was dismissed on therapy with digoxin 25 mg daily, furosemide 40 mg daily, and a potassium supplement.

Two years after operation, she had no evidence of heart failure or ventricular hypertrophy, the blood pressure was 105/60 mm Hg, and she had a normal sinus rhythm at 78/min. The first heart sound was only slightly louder than normal, the second heart sound was normal in intensity and closely split, and there was no opening snap. There was a faint mid-systolic click at the upper right sternal border. There was a very early short diastolic rumble at the apex and a 1/6 mid-systolic blowing murmur at the apex that radiated to the axilla and the right lower sternal border. Thus, there was no evidence of clinically significant pulmonary hypertension, mitral stenosis or mitral insufficiency. The patient had been taking no medication for 15 months and was asymptomatic.

DISCUSSION

Dextrocardia is a congenital anomaly in which the heart is displaced toward the right hemithorax, and in mirror-image dextrocardia, of which this case is an ex-

<table>
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<tr>
<th>Pressure</th>
<th>Rest, mm Hg</th>
<th>Exercise, mm Hg</th>
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<tbody>
<tr>
<td>Right atrium</td>
<td>A = 11</td>
<td>6</td>
</tr>
<tr>
<td></td>
<td>V = 6</td>
<td></td>
</tr>
<tr>
<td>Right ventricle</td>
<td>56/7</td>
<td>72/10</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>50/25</td>
<td>70/42</td>
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<tr>
<td>Pulmonary capillary wedge</td>
<td>AC = 28</td>
<td>26</td>
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<tr>
<td></td>
<td>V = 40</td>
<td></td>
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<tr>
<td>Left ventricle</td>
<td>142/12</td>
<td>148/12</td>
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ample, the left atrium and ventricle lie posteriorly and form the right cardiac border, the venae cavae are on the left side, and the pulmonary artery arises anteriorly and to the right of the aorta. A number of congenital heart defects have been reported with dextrocardia, including ventricular septal defect, patent ductus arteriosus, atrial septal defect (secundum and primum), tetralogy of Fallot, pentalogy of Fallot, infundibular pulmonary stenosis, transposition and patent ductus arteriosus, transposition and pseudotruncus, cor biloculare, atrial-ventricular canal with patent ductus arteriosus, and total anomalous pulmonary venous return. 5, 6 Multiple skeletal defects have also been reported in association with dextrocardia, including multiple deformities of the spine and skull asymmetry. 4

Total situs inversus is uncommon, its incidence being one per 10,000 births, and the association of acquired valvular lesions with dextrocardia and situs inversus is unknown but must be very rare. 6 The first case of dextrocardia and acquired valvular heart disease was reported by Owen 8 in 1911; the electrocardiogram in this case was interpreted by Dr. Thomas Lewis. A comprehensive review of acquired heart disease and dextrocardia including 1 new case and 12 additional cases in the world literature was reported in 1963 by Bopp et al. 1 A total of 15 cases has been reported previously in the literature, 9 of these being in females. 2, 4, 5, 6, 8, 10, 11 Three patients had isolated aortic lesions, 10 isolated mitral lesions, and 2 combined aortic and mitral lesions. Although the association of acquired valvular heart disease with dextrocardia is felt by some authors to be more than a chance association, there is little direct evidence to implicate dextrocardia as an etiologic factor in the development of rheumatic valve disease.

The diagnosis of dextrocardia is made from physical examination, chest x-ray film findings and electrocardiogram. When it is associated with situs inversus, the heart is usually normal except for the mirror-image rotation defect. However, when it is an isolated phenomenon, other congenital cardiac defects are the rule, frequently of a complex nature. 8 Their diagnosis must be confirmed by cardiac catheterization.

Four patients have been reported to have undergone surgical correction of associated mitral stenosis, and all underwent closed mitral commissurotomy. 5, 6, 8 In each case the immediate course has been satisfactory after operation. Unfortunately, longterm followups were not reported, except for the patient reported by Bopp et al, 3 who developed stenosis again six years later.

An open-heart approach through a left thoracotomy would have been easy to perform in the present case. However, the satisfactory results that had been reported in the literature, the mobile valve, the absence of calcification or mitral insufficiency, the presence of normal sinus rhythm, and the lack of any history of peripheral embolism led us to choose the closed approach this time, and to reserve the open approach for future valve replacement if it becomes necessary.

Closed mitral valvotomy in the presence of dextrocardia was performed through a right anterolateral fourth intercostal space incision (Fig 3). The left atrial appendage was enlarged and formed the upper right cardiac border, the left anterior descending coronary artery was present and identified the ventricular septum, the apex was formed by the left ventricle, and the anterior right ventricle gave rise to the pulmonary artery that originated in front of the aortic root. Purse-string sutures were placed in the apex of the left ventricle and the base of the left atrial appendage. The need for the surgeon to reverse the role of his hands in this situation, using the left index finger to explore the atrium and mitral valve and attempt finger fracture, while preserving the right hand to manipulate the valve dilator, presented no difficulty.

REFERENCES
9 Owen S: A case of complete transposition of the viscera, associated with mitral stenosis; including a description of the electrocardiographic tracings. Heart 3: 113-17, 1911

Figure 3. Diagram of procedure at operation showing (a) incision, (b) position of heart and great vessels, and (c) valvotomy with dilator.