Echocardiographic Features of Congenital Left Ventricular Inflow Obstruction

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SUMMARY The echocardiographic features of congenital left ventricular inflow obstruction are described in six patients. The echocardiograms in two patients with cor triatriatum were distinguished by normal mitral valve motion and an abnormal echo within the left atrium. In two patients with supravalvar mitral ring, in addition to abnormal mitral valve motion, an abnormal echo, presumably originating from the obstructive membrane, was located between the anterior and posterior mitral leaflets. In two cases of parachute mitral valve, mitral valve motion was abnormal. In one of these cases there were multiple mitral valve echoes similar to those found in supravalvar mitral ring.

The echocardiographic identification of an obstructive membrane within the left atrium is difficult because of the occurrence of artifacts. However, membranes may be identified if careful scanning techniques are employed in patients in whom left ventricular inflow obstruction is suspected. The echocardiogram is useful in detecting mitral valve abnormalities in these patients and is valuable in cases where mitral valve replacement is contemplated.

COR TRIATRIATUM, supravalvar mitral ring, and congenital mitral stenosis (most commonly parachute mitral valve) are congenital lesions which obstruct filling of the left ventricle. These malformations can cause pulmonary venous and pulmonary arterial hypertension with very similar clinical manifestations. A few reports have dealt with the echocardiographic findings in cases of congenital mitral stenosis, cor triatriatum, and supravalvar mitral ring, but a comparative study of the echocardiographic findings in these three entities has not appeared previously in the literature.

Materials and Methods

The six patients in this study ranged in age from 22 days to 15 years (table 1). They represent all the patients who presented to the Children's Hospital Medical Center from May 1974 to December 1975 with surgically proven left ventricular inflow obstruction and with technically satisfactory echocardiographic studies. Patients with left atrioventricular (A-V) valve atresia were not included in this study. The diagnosis of left ventricular inflow obstruction was documented at cardiac catheterization by the presence of an end-diastolic pressure gradient across the mitral valve and by angiography. In addition to surgical confirmation of obstruction in all cases, postmortem confirmation was obtained in case 3. Three types of obstruction were found in the six patients (table 1): cor triatriatum (2), supravalvar mitral ring (2), and isolated parachute mitral valve (2). One patient with supravalvar ring had an associated parachute mitral valve, the other an abnormal mitral valve with commissural fusion.

Echocardiographic examination was performed with a Hoffr few model 101B ultrasonoscope, utilizing either a 5.0 or a 3.25 MHz transducer. The echocardiographic signal was recorded by a Cambridge Fiberoptic multichannel recorder. Standard scanning techniques were performed. Total amplitude and diastolic velocity (E-F slope) of the anterior mitral valve leaflet were measured and compared to previously published data. All patients except case 5 had echocardiograms pre and postoperatively.

Results

Cor Triatriatum (Patients 1 and 2)

The amplitude and diastolic closure rate of the anterior leaflet of the mitral valve were normal (table 2). The motion

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of the posterior mitral leaflet was normal as well. In both cases, an independent, abnormal echo was observed in the left atrium behind the posterior wall of the aortic root and behind the mitral annulus. The pattern of movement of the abnormal echo consisted of a slow anterior motion in systole and a slow posterior motion in diastole. This motion was parallel to the echoes from the posterior wall of the aortic root (fig. 1). Repeated scans were performed and careful gain adjustments were made and therefore these abnormal echoes were not believed to be artifact. The abnormal echoes were no longer present in the postoperative echocardiograms of these patients (fig. 2).

Supravalvar Mitral Ring (Patients 3 and 4)

In both cases, the amplitude of anterior mitral leaflet excursion was normal (table 2) but the posterior leaflet moved anteriorly in diastole. In one patient, the diastolic closure rate (E-F slope) was decreased and in the other it was at the lower limits of normal. In both patients, an abnormal echo was noted between the anterior and posterior mitral valve leaflets. The motion of this echo, which possibly originated from the supravalvar ring, was similar to those of the valve leaflets but clearly separated from them (fig. 3). In patient 3 this abnormal echo could no longer be demonstrated after surgical removal of a supravalvar mitral ring; no mitral valve surgery was performed (fig. 4). In patient 4, mitral valve replacement was required so that a comparative postoperative study was not possible.

Parachute Mitral Valve (Patients 5 and 6)

In both patients with isolated parachute mitral valve, the mitral valve motion was abnormal (table 2, figs. 5 and 6). In patient 5, multiple mitral valve echoes were recorded moving anteriorly in diastole (fig. 5). The total valve excursion was decreased and the E-F slope was decreased (20 mm/sec). In addition the motion of the interventricular septum closely paralleled that of the mitral valve and the two structures were difficult to distinguish (fig. 5). At postmortem examination, there was obliteration of the interchordal spaces, essentially a single papillary muscle, and the anterior and posterior leaflets were completely fused. The anterior leaflet was thickened and redundant and the only ostium was a cleft-like slit in the anterior leaflet. The multiple mitral valve echoes probably originated from the thickened, redundant anterior mitral leaflet tissue.

Mitral valve motion in patient 6 with parachute mitral valve was less abnormal than patient 5: the valve excursion was mildly decreased, the E-F slope was 30 mm/sec and the posterior leaflet moved posteriorly in early diastole. Hemodynamically, the obstruction in this patient was also not as severe as in the previous patient and he has done well following fenestration of the mitral valve.

Discussion

Cor triatriatum and supravalvar mitral ring are lesions in which a membrane within the left atrium obstructs inflow to the left ventricle. In cor triatriatum the membrane which divides the left atrium into pulmonary venous and left atrial chambers attaches to the mid-portion of the left atrium and the pulmonary venous chamber lies obliquely behind the left atrial chamber. In supravalvar mitral ring a fibrous shelf-like membrane is present at the inlet of the mitral valve.1

Nimura et al.6 reported the echocardiographic findings in two patients with cor triatriatum; in both patients, an abnor-

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**Table 1. Clinical Data**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age at diagnosis (yr)</th>
<th>LA-LVEDP gradient</th>
<th>Associated lesions</th>
<th>Diagnosed by</th>
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<tr>
<td>Cor Triatriatum</td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>1</td>
<td>14</td>
<td>—</td>
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<tr>
<td>2</td>
<td>13</td>
<td>20</td>
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<tr>
<td>3</td>
<td>13</td>
<td>16</td>
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<tr>
<td>4</td>
<td>12</td>
<td>14</td>
<td>Parachute mitral valve, subaortic stenosis, ventricular septal defect, coarctation</td>
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<td>21</td>
<td>Tetralogy of Fallot</td>
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<td>6</td>
<td>15</td>
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<td>Bicuspid aortic valve</td>
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</table>

Abbreviations: CA = cardiac catheterization and angiogram, LA = left atrium, LVEDP = left ventricular end-diastolic pressure, PM = postmortem examination, PCW = pulmonary capillary wedge, S = surgery.

**Table 2. Echocardiographic Data**

<table>
<thead>
<tr>
<th>Patient</th>
<th>AML excursion (mm)</th>
<th>Mitral E-F slope (mm/sec)</th>
<th>PML motion in diastole</th>
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<tr>
<td></td>
<td>Preop Postop</td>
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<td>30</td>
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<td></td>
<td></td>
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<td>—</td>
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<td>—</td>
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<tr>
<td>6</td>
<td>15, 18</td>
<td>30</td>
<td>34</td>
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Abbreviations: AML = anterior mitral leaflet; LA = left atrium; MVR = mitral valve replacement; PML = posterior mitral leaflet.
The pattern of mitral valve motion is normal. Two cases of supravalvar mitral ring have been reported in which abnormal echoes were located posterior to the aortic root. In our experience, echoes originating from the membrane in cor triatriatum are more likely to be found behind the aortic root and mitral annulus; in supravalvar mitral ring the membrane is likely to be found related to the mitral valve leaflets. However, location of the membrane by echocardiography will depend largely on anatomic position and motion of the membrane as well as transducer beam width and direction. The pattern of mitral valve motion can be helpful in distinguishing cor triatriatum from supravalvar mitral ring. Cor triatriatum is usually an isolated lesion with normal mitral valve apparatus, while supravalvar mitral ring is often associated with an abnormal mitral valve. Thus the presence of abnormal mitral valve motion in association with an abnormal membrane strongly suggests the presence of supravalvar mitral ring. It should be noted, however, that normal mitral valve motion has been reported in a case of supravalvar mitral ring.

In the echocardiograms of patients with total anomalous pulmonary venous return misinterpretation of the posterior wall of the horizontal pulmonary vein as the true left atrial wall will suggest the presence of a membrane within the left atrium. This is particularly likely to occur if the left atrium is small, causing the abnormal echo to be close to the aortic root. In our experience, careful scanning techniques at times have been useful in identifying the abnormal echo in total anomalous pulmonary venous return as the true left atrial wall. Contrast echocardiography has also been useful in delineating the location of the horizontal pulmonary vein behind the left atrium. Nevertheless, the echocardiographic differentiation of total anomalous pulmonary venous return from cor triatriatum and supravalvar mitral ring may be quite difficult and one must often rely on other clinical data when dealing with this problem.

Figure 1. Preoperative echocardiogram demonstrating a scan from the left ventricle to the aortic root in a patient with cor triatriatum (case 2). An abnormal echo (arrows) thought to originate from the obstructing membrane is present posterior to the aortic root and mitral annulus. Mitral valve motion is normal. Ao = aortic root, LA = left atrium, MV = anterior leaflet mitral valve, RV = right ventricle.

Figure 2. Postoperative echocardiogram of case 2. The abnormal extra echo seen preoperatively is no longer present. IVS = interventricular septum.
Identification of membranes within the left atrium is difficult. This is especially true when attempting to differentiate a true membrane from artifacts caused by wide transducer beam, abnormal transducer position, or reverberations from more anterior structures. Careful transducer position and gain control settings can often eliminate these artifacts. Nevertheless, the echocardiographic diagnosis of an obstructive membrane within the left atrium should be made with caution and only in association with clinical evidence of left ventricular inflow obstruction.

In addition to obstructing membranes, congenital left ventricular inflow obstruction can also be caused by congenital mitral valve abnormalities, most commonly a parachute mitral valve. Since parachute mitral valve is commonly associated with a supravalvar mitral ring and abnormal echoes between mitral leaflets can be found in either lesion, it is not possible to differentiate these two lesions echocardiographically. However, the important consideration in patients with left ventricular inflow obstruction is the severity of the mitral valve disease. The presence of an obstructive membrane with normal mitral valve motion is reassuring evidence that mitral valve replacement may not be necessary. Echocardiographic evidence of severe mitral valve abnormality (as indicated by decreased excursion, abnormal posterior leaflet motion, and markedly decreased EF slope), regardless of the additional presence of an obstruc-
tive membrane, will alert the surgeon to the possibility of mitral valve replacement.

Despite the difficulties encountered in diagnosing left ventricular inflow obstruction, the echocardiogram can be a useful adjunct in the management of these patients.

Acknowledgment

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