Congenital Left Ventricular Inflow Obstruction Evaluated by Two-dimensional Echocardiography

A. REBECCA SNIDER, M.D., CLAUDE L. ROGE, M.D., NELSON B. SCHILLER, M.D., and NORMAN H. SILVERMAN, M.D.

SUMMARY Several forms of congenital heart disease that cause left ventricular inflow obstruction have similar M-mode findings, and frequently the exact anatomic diagnosis cannot be made by M-mode echocardiography alone. We examined five children with various forms of left ventricular inflow obstruction using two-dimensional echocardiography. The diagnosis was confirmed by cardiac catheterization and surgery in all five patients. In one patient with congenital mitral valve stenosis, a thick mitral valve with two papillary muscles was imaged. This patient was easily distinguished from a second child with parachute deformity of the mitral valve in whom a single papillary muscle arising from the left ventricular apex was seen. These two patients with mitral valve stenosis were easily differentiated from the three patients in whom the left ventricular inflow obstruction was caused by a membrane within the left atrium. The membrane could be seen in several spatial planes; however, we could not distinguish by two-dimensional echocardiography one child who had cor triatriatum from the other two patients who had a supravalvar mitral ring. Because of its spatial anatomic display, the two-dimensional echocardiogram provides information for a more detailed anatomic diagnosis in children with congenital left ventricular inflow obstruction.

CONGENITAL FORMS of left ventricular inflow obstruction include cor triatriatum, supravalvar mitral ring, congenital mitral valve stenosis and parachute mitral valve deformity. There have been several reports of the M-mode echocardiographic findings in these lesions and, in many instances, similar M-mode echocardiographic findings have been reported for very different anatomic defects. The difficulty in distinguishing between the anatomic forms of left ventricular inflow obstruction by M-mode echocardiography was noted by LaCorte, Harada and Williams in 1976 and more recently by Driscoll, Gutgesell and McNamara. We have studied a group of children with various forms of congenital left ventricular inflow obstruction to determine if two-dimensional echocardiography can provide a more precise anatomic diagnosis.

Methods

All patients with congenital left ventricular inflow obstruction who underwent M-mode and two-dimensional echocardiography, cardiac catheterization and surgery at the University of California at San Francisco from September 1976 to the present were included in this report. Five children had congenital forms of left ventricular inflow obstruction diagnosed prospectively by M-mode and two-dimensional echocardiography and then underwent cardiac catheterization and surgery. During this period, no other patients who underwent echocardiographic examination were found to have mitral stenosis at subsequent cardiac catheterization. Patients with forms of hypoplastic left heart were excluded from the study. Also, because of lack of surgical confirmation, we excluded one child with a left atrial membrane demonstrated by two-dimensional echocardiography and cineangiography. This child had a low pulmonary artery wedge pressure and has not undergone surgery.

The patients were 9 months to 16 years of age. Complete cardiac catheterization data, including oximetric shunt determinations and biplane cineangiograms, were obtained in all patients. The cardiac catheterization findings were confirmed at surgery in all patients.

All patients were studied by M-mode and two-dimensional echocardiography before cardiac catheterization. The M-mode echocardiograms were performed with an Ekoline 20A Ultrasonoscope interfaced with a Smith Kline strip-chart recorder. A variety of transducers with appropriate focal length and frequency were used. Recordings were obtained with patients in the left lateral decubitus position with the transducer in the third or fourth intercostal space at the left sternal border. The M-mode echocardiograms were evaluated to determine the direction of movement of the posterior mitral valve leaflet in diastole, the presence of mitral valve diastolic flutter, and the presence of mitral valve reopening with atrial contraction. Anomalous echoes in the left atrium or the mitral valve funnel in diastole were noted. The maximum excursion of the anterior mitral valve leaflet in diastole (D-E excursion) and the rate of early diastolic closure of the anterior mitral valve leaflet (E-F slope) were measured by standard techniques.

Two-dimensional echocardiograms were recorded with a Varian V-3000, 80° phased-array sector scanner. A 2.25- or a 3.5-MHz transducer was used. The patients were examined in the left lateral decubitus position with the transducer in the second or third intercostal space. The long-axis view of the heart was obtained by directing the ultrasonic plane between the apex and base of the heart. From this position the
transducer was rotated 90° clockwise to obtain the short-axis image of the heart. The apical four-chamber view was imaged with the technique previously described. The transducer was placed over the cardiac apex and angled so that all four cardiac chambers were shown simultaneously. The two-dimensional echocardiograms were recorded on videotape for later analysis at real time or slow speeds. The illustrations presented here were obtained from Polaroid photographs of stop-action, single-frame, scan images from the videotape recordings. This photographic process markedly reduces image quality and causes a loss of the visual integration of motion normally present in the real-time format.

Results

All five patients with congenital forms of left ventricular inflow obstruction had technically adequate M-mode and two-dimensional echocardiograms. Table 1 summarizes the pertinent clinical and catheterization findings. Two patients (nos. 1 and 2) had mitral valve stenosis diagnosed at cardiac catheterization. Patient 1 had a 16-mm Hg gradient between the pulmonary artery wedge pressure and the left ventricular end-diastolic pressure and markedly elevated pulmonary vascular resistance. At cardiac catheterization patient 2 had parachute mitral valve deformity, ventricular septal defect, coarctation of the aorta and patent ductus arteriosus. At cardiac catheterization a left atrial membrane was found in patients 3–5. In patients 3 and 4 the membrane was situated close to the mitral valve and a diagnosis of supravalvar mitral ring was made. This diagnosis was confirmed at surgery. In patient 5, a left atrial membrane was seen but a definite diagnosis of cor triatriatum or supravalvar mitral ring could not be made at cardiac catheterization. At surgery, this patient was found to have cor triatriatum.

M-mode Echocardiograms

Table 2 summarizes the M-mode echocardiographic findings, which were very similar among the

| Table 1. Clinical, Catheterization, and Surgical Data in Five Patients with Left Ventricular Inflow Obstruction |
|---|---|---|---|---|
| Pt no. | Clinical data | Catheterization data | Diagnosis | Surgical findings and procedure |
| | Age | Sex | PAP (mm Hg) | PAWP (mm Hg) | LVP (mm Hg) |
| 1 | 13 yrs | F | 96/61,74 | 28 | 156/12 |
| 2 | 9 mos | F | 110/60,90 | 110/6 | Parachute MV with stenosis, PAH, coarct of Ao, VSD, PDA |
| 3 | 14 yrs | M | 38/18,24 | 18 | 105/10 |
| 4 | 9 mos | F | 60/20,40 | 18 | 170/8 |
| 5 | 16 yrs | M | 53/28,36 | 30 | 116/9 |
| **Total** | | | | | **SVMR versus CT, PAH** |

Aortic pressure.

Abbreviations: Ao = aorta; AS = aortic stenosis; CS = coronary sinus; CT = cor triatriatum; LSVC = left superior vena cava; LVP = left ventricular pressure; MV = mitral valve; PAP = pulmonary artery pressure; PAH = pulmonary artery hypertension; PAPVR = partial anomalous pulmonary venous return; PAWP = pulmonary artery wedge pressure; PDA = patent ductus arteriosus; R SVC = right superior vena cava; SAH = systemic hypertension; SubAS = subaortic stenosis; SVMR = supravalvar mitral ring; VSD = ventricular septal defect.

| Table 2. M-mode Echocardiographic Findings in Five Patients with Left Ventricular Inflow Obstruction |
|---|---|---|---|---|---|---|---|
| Pt no. | Diagnosis | MV diastolic flutter | PMVL movement in diastole | MV "a" wave | D-E excursion (MM) | E-F slope (mm/sec) | Anomalous echoes in MV funnel | Anomalous echoes in LA |
| 1 | MV stenosis | Yes | Anterior | Absent | 12 | 16 | Yes | No |
| 2 | Parachute MV | Yes | Anterior | Absent | 5 | 18 | Yes | No |
| 3 | SVMR | Yes | Anterior | Absent | 12 | 54 | Yes | No |
| 4 | SVMR | Yes | Anterior | Absent | 8 | 20 | Yes | No |
| 5 | CT | Yes | Posterior | Present | 27 | 94 | Yes | No |

Abbreviations: CT = cor triatriatum; LA = left atrium; MV = mitral valve; PMVL = posterior mitral valve leaflet; SVMR = supravalvar mitral ring.
five patients. All of the M-mode studies exhibited diastolic flutter of the mitral valve and multiple echoes in the mitral valve funnel in diastole (fig. 1). All but patient 5 had a paradoxically moving posterior mitral valve leaflet, lack of an "a" wave on the mitral valve echogram, decreased D-E excursion, and a decreased E-F slope (fig. 1). An anomalous left atrial echo was not found in any of the three patients with left atrial membranes.

Two-dimensional Echocardiograms

Table 3 is a summary of the pertinent two-dimensional echocardiographic findings.

Mitral Valve Stenosis

Findings at two-dimensional echocardiography in patients 1 and 2 suggested mitral valve stenosis. In patient 1, the mitral valve leaflets appeared thickened (densely reflective) in all planes of examination. The valve leaflets moved stiffly and had a diminished diastolic excursion. Coarse vibrations of the mitral valve leaflets were present during diastole. Figures 2 and 3 are representative examples of the thickened mitral valve and large left atrial chamber in the long-axis and apical views. No membranous structures were imaged in the body of the left atrium or near the mitral valve annulus. In the short-axis view of the left ventricle below the plane of the mitral valve leaflets, two papillary muscles of equal size were imaged (fig. 4). The right-sided cardiac chambers appeared enlarged and the tricuspid valve appeared thickened in the apex image of this patient (fig. 3). These findings were thought to result from long-standing pulmonary artery hypertension. At cardiac catheterization, this patient had a severely stenotic mitral valve with elevated pulmonary vascular resistance. Mild aortic stenosis and partial anomalous pulmonary venous return to the superior vena cava were also found. At surgery, a thickened, deformed mitral valve with two papillary muscles and a 5-mm central orifice was replaced with a Hancock prosthesis.

The two-dimensional echocardiogram in patient 2 suggested mitral valve stenosis. As with patient 1, a thickened, stiffly-moving mitral valve was imaged in all cardiac planes. A large left atrial chamber was noted in the long-axis and apical views; and there was no evidence of a membrane in the left atrium. However, the short-axis view of the left ventricle was distinctly different from that of patient 1 with mitral

<table>
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<tr>
<th>Pt no.</th>
<th>Diagnosis</th>
<th>Long-axis view</th>
<th>Short-axis view</th>
<th>Apex view</th>
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<tr>
<td></td>
<td></td>
<td>LA size</td>
<td>RV size</td>
<td>Membrane</td>
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<td>1</td>
<td>MV Stenosis</td>
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<tr>
<td>2</td>
<td>Parachute MV</td>
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<td>Enlarged</td>
<td>No</td>
</tr>
<tr>
<td>3</td>
<td>SVMR</td>
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</tr>
<tr>
<td>4</td>
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<tr>
<td>5</td>
<td>CT</td>
<td>Normal</td>
<td>Normal</td>
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</table>

Cavity sizes were evaluated subjectively and confirmed by M-mode echocardiographic measurements.

Abbreviations: CT = cor triatriatum; LA = left atrium; MV = mitral valve; RV = right ventricle; SVMR = supravalvar mitral ring.
**Figure 2.** Long-axis image from patient 1, a 13-year-old with mitral valve (MV) stenosis. The MV leaflets are thickened and the left atrial cavity is enlarged. There are no membranous structures near the MV or in the left atrium (LA). The right ventricle (RV) is large because of long-standing pulmonary hypertension. Ao = aorta; LV = left ventricle.

**Figure 3.** Apex image from patient 1, who had mitral valve (MV) stenosis and severe pulmonary artery hypertension. The MV is thickened and the left atrial cavity is enlarged. The right atrium (RA) and right ventricle (RV) are also enlarged, probably a result of long-standing pulmonary hypertension. This frame is taken at the time of maximum diastolic opening of the MV, which is severely limited (compare with tricuspid valve opening). LA = left atrium; LV = left ventricle; PV = pulmonary veins; TV = tricuspid valve.

**Figure 4.** Short-axis image of the left ventricle below the level of the mitral valve leaflets in patient 1, with mitral valve stenosis. The presence of two distinct papillary muscles (PM) excludes the possibility of parachute mitral valve deformity. RV = right ventricle; Sept = septum.
valve stenosis. On a scan from the aortic root to the apex in the short-axis plane, the mitral valve leaflets were thickened and the mitral orifice was small. Below the level of the mitral valve leaflets, a single, large papillary muscle was seen arising from the posterior left ventricular wall (fig. 5). The echocardiogram was interpreted as showing a parachute mitral valve deformity with a single papillary muscle. Cardiac catheterization showed a parachute mitral valve deformity with a single papillary muscle arising from the left ventricular wall. Additional findings at cardiac catheterization included a small ventricular septal defect, patent ductus arteriosus and moderate coarctation of the aorta. These diagnoses were confirmed at surgery and autopsy.

**Left Atrial Membranes**

In patients 3-5, an anomalous echo stretching across the left atrium was imaged in the long-axis and apical views. This echo, which was believed to represent a left atrial membrane, was similar in all three cases in its position within the left atrium and its phasic movement. Figure 6 demonstrates the proximity of the thin membrane to the mitral valve ring. With real-time recording, the echoes from the mem-

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**Figure 5.** Short-axis image of the left ventricle below the level of the mitral valve leaflets from patient 2, a 9-month-old child with parachute mitral valve deformity. A single, large papillary muscle (PM) is seen arising from the center of the ventricular cavity. RV = right ventricle; S = septum.

**Figure 6.** Long-axis image from patient 3, a 14-year-old male who had a supravalvar mitral ring. A membrane is seen stretching obliquely across the left atrial cavity just above the mitral valve. The mitral valve (MV) is thickened and the left atrial cavity is enlarged. Because of associated dextroversion, the septum and left ventricular apex are not imaged clearly in this long-axis view. Ao = aorta; LA = left atrium; LV = left ventricle; RV = right ventricle.
brane moved toward the mitral valve funnel in diastole and away from the mitral valve leaflets in systole. The exact attachments of the membrane were extremely difficult to image; however, in the long-axis plane (fig. 6), the membrane stretched obliquely across the left atrial chamber with extensions anterosuperiorly to the posterior aortic root and posteroinferiorly to the posterior left atrial wall. In the apical four-chamber view (fig. 7), the membrane extended in a nearly horizontal plane adjacent to the mitral valve ring. It could be imaged extending to the right in the region of the primum atrial septum and to the left to the lateral left atrial wall. Patients 3 and 4 had enlargement of the left atrium in the long-axis and apex views and a slightly thickened mitral valve with a decreased diastolic excursion in all planes. Patient 5 had a normal-appearing mitral valve apparatus and normal left atrial size.

At cardiac catheterization in all three patients, a left atrial membrane was detected on the levophase of a pulmonary artery angiogram. All patients had a moderately severe gradient between the pulmonary artery wedge pressure and left ventricular end-diastolic pressure and underwent surgery. In patient 5 at surgery, a membrane of cor triatriatum was found dividing the left atrium into a dorsal, superior chamber receiving the pulmonary veins and a ventral, inferior chamber communicating with the left atrial appendage and mitral valve funnel. The mitral valve apparatus appeared normal. In patients 3 and 4 at surgery, the membrane of a supravalvar mitral ring was found closely attached above the mitral valve. In both cases, the mitral valve leaflets appeared mildly thickened. Patient 4 also underwent closure of a ventricular septal defect and resection of severe subaortic stenosis. At surgery, this patient had two papillary muscles. Two-dimensional echocardiographic examinations performed in the immediate postoperative period in all three patients showed that the anomalous left atrial echoes had disappeared. The postoperative mitral valve echograms were unchanged in appearance from the preoperative examinations.

Discussion

Congenital lesions that cause left ventricular inflow obstruction may lead to pulmonary venous and pulmonary arterial hypertension; therefore, early detection of these defects is important. The clinical manifestations of the various forms of left ventricular inflow obstruction are vague and do not aid in determining a precise anatomic diagnosis. The exact anatomy is often not clear after a complete cardiac catheterization and cineangiography. Several reports have attempted to relate the anatomy of left ventricular inflow obstruction to the M-mode echocardiographic findings. Lundstrom1 described several cases of congenital mitral valve stenosis with M-mode echocardiographic findings of decreased D-E excursion, decreased E-F slope, diastolic flutter, and absent mitral valve “a” wave. In the same report, Lundstrom described the echocardiographic findings in a patient with supravalvar mitral ring (anomalous left atrial echo) and in a patient with cor triatriatum (anomalous echo behind the anterior mitral valve leaflet).

Likewise, Chung et al.2 reported a case of supravalvar mitral ring with an anomalous left atrial echo and a normal mitral valve echogram. Several other reports of patients with cor triatriatum have described anomalous left atrial echoes associated in some cases with normal mitral valve echograms5,6 and in other cases with abnormal mitral valve echograms.7 Nimura and colleagues4 described anomalous left atrial echoes on M-mode echocardiographic examination in two cases of cor triatriatum and demonstrated the membrane by ultrasonocardiogram. The first comparative study of the M-mode findings in various forms of left ventricular inflow obstruction was con-
ducted by LaCorte and colleagues. Their report described difficulty in determining the origin of either anomalous left atrial echoes or anomalous echoes within the mitral valve funnel. Causes of anomalous left atrial echoes included supravalvar mitral ring, cor triatriatum, total anomalous pulmonary venous return or beam width artifacts. In addition, multiple echoes between the mitral valve leaflets were seen in association with supravalvar mitral ring or parachute mitral valve deformity. Recently, Driscoll and colleagues reported five cases of supravalvar mitral ring with no abnormal echoes either behind the mitral valve or in the left atrium. Also, no specific M-mode pattern was found to be characteristic of either mitral valve stenosis or parachute mitral valve deformity. In summary, the M-mode echocardiogram has thus far been important in detecting the presence of left ventricular inflow obstruction but has not proved useful in determining its exact anatomy.

Because of its ability to image the heart in several spatial planes, two-dimensional echocardiography offers a unique opportunity to define more clearly the underlying anatomy in cases of left ventricular inflow obstruction. The effectiveness of cineangiography in delineating the precise anatomy in various forms of left ventricular inflow obstruction may be limited by superimposition of shadows — a factor that is eliminated by two-dimensional echocardiography. All five of our patients with left ventricular inflow obstruction were detected on two-dimensional echocardiographic examination by identifying either an abnormally thickened, stenotic mitral valve or a membrane in the left atrium. Demonstrating the papillary muscles in the short-axis plane enabled us to distinguish a patient with mitral valve stenosis with two papillary muscles from another patient with parachute mitral valve deformity and a single papillary muscle. The two-dimensional echocardiographic examination clearly distinguished the two patients with valvar forms of mitral stenosis from the three patients with left ventricular inflow obstruction due to an abnormal left atrial membrane. However, we could not differentiate the patient with a cor triatriatum from the two patients with supravalvar mitral ring. On two-dimensional echocardiographic examination, the position and motion of the left atrial membrane in the long-axis and apical views were nearly identical in all three patients. LaCorte and colleagues suggested that cor triatriatum might be distinguished on M-mode echocardiographic examination from supravalvar mitral ring by the presence of normal mitral valve motion in cases of cor triatriatum. The only patient in our series with a normal mitral valve echogram had a cor triatriatum (table 2). With further refinement in image processing and further experience with these lesions, perhaps the exact attachment of the membrane relative to the pulmonary veins and left atrial appendage may be imaged. This would allow the accurate differentiation of cor triatriatum from supravalvar mitral ring by two-dimensional echocardiographic examination.

Congenital left ventricular inflow obstruction is an extremely rare form of congenital heart disease. Previous echocardiographic reports concerning left ventricular inflow obstruction have involved small numbers of patients. In over 2000 pediatric patients studied by two-dimensional echocardiography at the University of California at San Francisco, only five cases of congenital left ventricular inflow obstruction were found. Although our number of patients detected with left ventricular inflow obstruction is small, we believe that the five patients presented in this report are representative examples of the various forms of congenital left ventricular inflow obstruction that occur in the general pediatric population.

Patient age and size did not appear to be a limiting factor in diagnosing left ventricular inflow obstruction by two-dimensional echocardiography. Also, in over 2000 two-dimensional echocardiographic studies, we have not seen a left atrial structure that could be confused with the membrane of cor triatriatum or supravalvar mitral ring. In two patients we have studied by echocardiography with total anomalous pulmonary venous return to the coronary sinus and one patient with total anomalous pulmonary venous return to a left vertical vein, the echo arising from the pulmonary venous confluence stretched obliquely behind the left atrium from a cranial to a caudal position in a manner quite distinct from the position of the membrane in supravalvar mitral ring or cor triatriatum. Care must be taken not to confuse echoes arising in diastole from a thickened mitral valve annulus with echoes arising from a membrane in the mitral valve funnel. This mistake might occur especially in small infants in whom the resolution of the study may not be optimal; however, by directing careful attention to gain and reject settings and to the motion of the echo in question, this misinterpretation can be avoided.

We have found the M-mode echocardiogram extremely useful in detecting left ventricular inflow obstruction but of limited usefulness in defining the exact anatomy of the obstruction. Two-dimensional echocardiography allows a more precise anatomic diagnosis to be made.

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Two-dimensional Echocardiographic Findings in Right-sided Infective Endocarditis

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SUMMARY M-mode and two-dimensional echocardiograms were recorded in 12 narcotic addicts who had right-sided infective endocarditis. The two-dimensional echocardiogram showed vegetations in 10 patients (nine tricuspid and one pulmonic), while the M-mode echocardiogram was positive in six (five tricuspid and one pulmonic). The use of multiple transducer positions resulted in better visualization of the valves and appeared to be an important reason for the large number of positive two-dimensional echocardiograms. Echocardiographic findings were also available after completion of antibiotic therapy in seven of 10 patients. Of these seven patients, the vegetation appeared unchanged in three, diminished in size in three and was no longer visualized in one. No patient required valve replacement. Two-dimensional echocardiography using a wide-angle sector scanner appears to offer distinct advantages over the standard M-mode technique in evaluating patients with right-sided infective endocarditis.

THE INCIDENCE of right-sided infective endocarditis has increased markedly in recent years, particularly in persons with a history of intravenous drug abuse.1-4 Echocardiographic findings in right-sided endocarditis have been described,5-9 but have received less attention than those in the left-sided endocarditis.10 This is in part due to the relative difficulty in recording the tricuspid and pulmonic valves by M-mode echocardiography, especially in the absence of right ventricular dilatation.11

Recently, the availability of two-dimensional echocardiography has permitted more complete visualization of the tricuspid leaflets. Using a realtime, two-dimensional, phased-array, wide-angle sector scanner, we attempted to examine the tricuspid and pulmonic valves in a series of narcotic addicts with clinically diagnosed right-sided endocarditis. In this communication we describe the two-dimensional echocardiographic findings in 10 patients with right-sided infective endocarditis. The importance of multiple transducer positions, comparisons with M-mode findings and changes in morphology of the vegetations after therapy are all considered in the present study.

Materials and Methods

From June through December 1978, right-sided infective endocarditis was diagnosed clinically in 12 narcotic addicts hospitalized at the Morris J. Bernstein Institute of Beth Israel Medical Center. The diagnosis of right-sided endocarditis depended upon the presence of each of the following: a history of recent intravenous drug abuse, fever greater than 101°F and three or more positive blood cultures.12, 13 In addition, it was required that the patient have a murmur consistent with tricuspid insufficiency (a blowing systolic murmur along the lower left sternal border that became more intense with inspiration) or radiographic findings compatible with pulmonary embolization. Ten of these patients had right-sided valvular vegetations (nine tricuspid and one pulmonic) visualized on two-dimensional echocardiography and constitute the patient population for the present study. There were eight males and two females, ages 23-59 years (average 33 years). Pertinent clinical data are summarized in table 1.
Congenital left ventricular inflow obstruction evaluated by two-dimensional echocardiography.

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