Echocardiography in the diagnosis and management of symptomatic aortic valve stenosis in infants

JAMES C. HUHTA, M.D., LARRY A. LATSON, M.D., HOWARD P. GUTGESELL, M.D., DENTON A. COOLEY, M.D., AND DEBRA L. KEARNEY

ABSTRACT Infants with severe aortic valve stenosis often are critically ill and require urgent surgical treatment. Currently, angiography is used at the time of cardiac catheterization to diagnose aortic valve stenosis. However, the use of this test may be hazardous in an unstable infant and may precipitate hemodynamic and clinical deterioration before surgery. Therefore, a noninvasive method of accurately making this diagnosis would be useful in that it would allow the risks of cardiac catheterization to be avoided. Between January 1982 and September 1983, 10 infants with critically severe aortic valve stenosis and intact ventricular septum were examined by echocardiography. There were no false-positive or negative results in this time period and several criteria for the noninvasive diagnosis of critical aortic valve stenosis were recognized. These included immobile aortic valve cusps and left ventricular hypertrophy with increased echo density of the left ventricular papillary muscles and mitral valve support apparatus. Patients without other aortic obstruction had poststenotic dilation of the ascending aorta, as evidenced by a ratio of the diameter of the ascending aorta to that of aortic valve anulus greater than 1.7. A disturbed Doppler velocity signal in the ascending aorta supported the presence of valvar stenosis. Nine patients underwent cardiac surgery and five survived. In five patients surgery was performed without angiographic examination. The correct diagnosis was made noninvasively in each, and four of the five patients survived surgery. Echocardiography was comparable to angiography in making the diagnosis and assessing the cardiovascular anatomic characteristics. Echocardiography could therefore replace angiography in selected infants with symptomatic aortic valve stenosis and should be routinely used in the evaluation of these patients.


SEVERE aortic valve stenosis in infancy is a highly lethal congenital condition characterized by predominant aortic valve disease (unicuspid or bicuspid aortic valve), intact ventricular septum, and absence of additional right-sided congenital cardiac lesions. Medical therapy is usually unsuccessful and urgent aortic valvotomy is indicated in the symptomatic infant. However, early operative mortality is high (30% to 80%) because of anatomic and clinical factors that result in poor preoperative condition, such as metabolic acidosis and small size of the left ventricle.

Two-dimensional echocardiography has been used to assess aortic valve stenosis in adults and children and has been useful in the definition of morphology of the left ventricular outflow tract. We recently reported the use of two-dimensional echocardiography in the selection of a group of neonates with severe left ventricular outflow tract obstruction who were most likely to survive surgical intervention. Based on this experience and using improved two-dimensional/Doppler echocardiographic equipment, we reviewed our recent experience concerning the impact of echocardiography on the diagnosis and management of symptomatic aortic valve stenosis in infancy. Two questions were addressed: (1) Can echocardiography be used to confidently diagnose aortic valve stenosis in infancy and characterize the intracardiac and extracardiac anatomy? (2) What noninvasive criteria can be used to characterize critical aortic stenosis that will allow it to be
identified independent of other causes of left ventricular dysfunction in infancy?

Materials and methods

Between January 1982 and September 1983, 500 infants underwent examination by two-dimensional echocardiography for suspected cardiac disease; in all there was angiographic or surgical confirmation of the diagnosis. Critical aortic valve stenosis was defined as valvar left ventricular outflow tract obstruction with intact ventricular septum and without significant right-sided congenital cardiac lesion plus cardiomegaly and congestive heart failure. This was the clinical picture for 10 infants (2%). Three infants (0.6%) with clinical findings of aortic stenosis were asymptomatic and therefore were excluded from the study. All 10 infants had a systolic ejection murmur plus ejection click. Chest x-ray and electrocardiographic findings are tabulated in table 1. Two-dimensional echocardiographic examinations were performed by a segmental approach to the diagnosis of congenital cardiac disease, including assessment of atrial situs, atrioventricular connection, ventriculoarterial connection, atrial and ventricular septae, atrioventricular and semilunar valves, aorta, and systemic and pulmonary venous return. This morphologic information was obtained with an Advanced Technology Laboratories 300-LX, 500 MK, or 600 MK ultrasound system and scanning from parasternal, subcostal, apical, and suprasternal approaches (figure 1).

M mode echocardiograms were available in all but two infants and were analyzed in standard fashion to quantify left ventricular end-diastolic dimension (LVEDD), systolic dimension (LVESD), and shortening fraction (SF = LVEDD − LVESD/LVEDD). These measurements were supplemented by measurements from freeze-frame two-dimensional echocardiographic images that were obtained for the quantitation of similar variables plus diameter of the aortic valve anulus, maximum dimension of the ascending aorta, and right ventricular end-diastolic dimension. The severity of poststenotic dilation of the ascending aorta was quantitated by the ratio of the diameter of the ascending aorta to that of the aortic valve anulus (figure 2). Normal values for measurements from two-dimensional echocardiographic images were obtained in 19 newborn infants without heart disease. In seven patients, pulsed Doppler echocardiography was performed with sampling in the ascending aorta to assess the pattern of blood flow velocity (figure 3). This was subjectively graded as normal, slightly disturbed, or severely disturbed. Student’s t test was used to compare nonpaired values.

FIGURE 1. Parasternal long-axis view from a neonate with critical aortic stenosis. Note the dilated left ventricle (LV), right ventricle (RV), and left atrium (LA) and the hypoplastic aortic valve anulus and poststenotic dilation of the ascending aorta (Ao). A = anterior; I = inferior; P = posterior; S = superior.

FIGURE 2. Parasternal long-axis view illustrating the site of two-dimensional echocardiographic measurements of aortic valve anulus (outside arrows) and ascending aorta at the widest site of poststenotic dilation (inside arrows). A = anterior; I = inferior; P = posterior; S = superior.

FIGURE 3. Two-dimensional echocardiographically directed Doppler sampling in the ascending aorta in critical aortic stenosis. The sampling probe was positioned as shown by the arrow in the suprasternal view (top) and the severe systolic Doppler-determined velocity disturbance (bottom) recorded was a necessary criterion for the diagnosis. Ao = aorta; Asc = ascending; other abbreviations as in figure 1.
TABLE 1
Clinical characteristics of patients

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Sex</th>
<th>Age</th>
<th>Weight (kg)</th>
<th>C/T</th>
<th>ECG</th>
<th>LVEDD (mm)</th>
<th>LVESD (mm)</th>
<th>SF</th>
<th>AVA (mm²)</th>
<th>AA</th>
<th>AA/VA</th>
<th>RVEDD (mm)</th>
<th>LVMD (mm)</th>
<th>Catheterization (LVSP:AoP)</th>
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<tr>
<td>1</td>
<td>M</td>
<td>3.5 mo</td>
<td>4.9</td>
<td>0.68</td>
<td>BVH</td>
<td>19</td>
<td>12</td>
<td>0.37</td>
<td>8</td>
<td>12</td>
<td>1.5</td>
<td>8</td>
<td>4+</td>
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<td>3.0</td>
<td>0.69</td>
<td>LAE</td>
<td>18</td>
<td>16</td>
<td>0.11</td>
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<td>9</td>
<td>1.8</td>
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<tr>
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<td>5.3</td>
<td>0.71</td>
<td>BVH + strain</td>
<td>22</td>
<td>18</td>
<td>0.18</td>
<td>7</td>
<td>13</td>
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</tr>
<tr>
<td>4</td>
<td>F</td>
<td>1 mo</td>
<td>3.5</td>
<td>0.65</td>
<td>LVH + strain</td>
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<td>21</td>
<td>0.09</td>
<td>7</td>
<td>13</td>
<td>1.9</td>
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<td>8</td>
<td>0.38</td>
<td>5</td>
<td>9</td>
<td>1.8</td>
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<tr>
<td>6</td>
<td>F</td>
<td>3 mo</td>
<td>3.0</td>
<td>0.71</td>
<td>RVH</td>
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<td>17</td>
<td>0.29</td>
<td>6</td>
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<td>2.2</td>
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<tr>
<td>7</td>
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<td>2.4</td>
<td>0.70</td>
<td>RVH + RAE + ST changes</td>
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<td>13</td>
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<td>LVH</td>
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<td>4 days</td>
<td>2.9</td>
<td>0.71</td>
<td>BVH + RAE</td>
<td>12</td>
<td>10</td>
<td>0.16</td>
<td>5</td>
<td>10</td>
<td>2.0</td>
<td>15</td>
<td>4+</td>
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AA = diameter of ascending aorta (mm); AoP = aortic pressure (mm Hg); AoV = aortic valvotomy; AVA = aortic valve anulus diameter (mm); BVH = biventricular hypertrophy; Coarc = coarctation of the aorta; C/T = cardiothoracic ratio on admission chest x-ray; LAE = left atrial enlargement; LV = left ventricle; LVEDD = LV end-diastolic dimension (mm); LVESD = LV end-systolic dimension (mm); LVH = LV hypertrophy; LVMD = LV papillary muscle-density; LVSP = LV peak systolic pressure (mm Hg); NP = not performed; PDA = patent ductus arteriosus; RAE = right atrial enlargement; RVEDD = right ventricular end-diastolic dimension (mm); RVH = right ventricular hypertrophy; SF = shortening fraction (LVEDD-LVESD/LVEDD).

*Severely disturbed ascending aortic velocity pattern.

*See text and Norwood et al.*

30 day survivor.

for aortic valve anulus, ascending aorta, and the ratio of the two to measurements in normal subjects. Papillary muscle density was graded 1 to 4, with 1 being normal and 4 being echo density comparable to that of the pericardium on short-axis parasternal scans (figure 4).

In all patients with critical aortic stenosis there was confirmation of the anatomic characteristics by angiography (in five), autopsy (in four), or surgery (in nine).

Management of infants with symptomatic aortic stenosis was individualized. In those infants in whom a complete examination of intracardiac and extracardiac anatomic characteristics was possible, including those of pulmonary and systemic venous return, the proximal coronary artery, and the aortic arch, the risks of catheterization were weighed against the possibility of an undiagnosed congenital defect being present. Five patients underwent surgery without angiographic examination. In one of these patients, who had normal left ventricular function, the left ventricular pressure was 170 mm Hg at catheterization and aortic valvotomy was done immediately after the catheterization. Four infants received infusions of prostaglandin E₁ as part of the clinical management and the echocardiographic examination was performed during this infusion in each.

Nine of the 10 infants underwent cardiac surgery, including eight who underwent aortic valvotomy and one infant who had a modified Norwood operation for hypoplastic left ventricle with anastomosis of the proximal pulmonary trunk to the ascending aorta and creation of a right subclavian artery-to-right pulmonary artery Gortex communication. Early operative mortality was four of nine (44%) overall and four of eight for aortic valvotomy (50%). Three of the four patients undergoing aortic valvotomy who did not undergo angiography survived.

**Results**

The clinical findings of aortic stenosis were present in all 10 infants and the cardiothoracic ratio on the admission chest x-ray ranged from 0.62 to 0.71 (mean = 0.69). Electrocardiographic findings varied from left or biventricular hypertrophy with strain in four to right ventricular hypertrophy alone in three. The echocardiographic and clinical results are summarized in table 1. Left ventricular function fell into two groups: those with depressed ventricular function (left ventricular shortening fraction <0.28 [five of 10]), and those with normal function (five of 10). Two-dimensional echocardiographic abnormalities were present in both groups, and included increased echo density of the mitral papillary muscles and chordae (10 of 10), right ventricular enlargement (eight of 10), immobile aortic valve cusps (10 of 10), and ascending aortic poststenotic dilation (10 of 10). The size of the aortic valve
TABLE 1  (Continued)

<table>
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<th>Surgery</th>
<th>Other lesions</th>
<th>Outcome</th>
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<tr>
<td>^</td>
<td>Yes</td>
<td>AoV</td>
<td>No</td>
<td>AliveC</td>
</tr>
<tr>
<td>NP</td>
<td>No</td>
<td>AoV</td>
<td>No</td>
<td>Died</td>
</tr>
<tr>
<td>NP</td>
<td>Yes</td>
<td>AoV</td>
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<td>Died</td>
</tr>
<tr>
<td>^</td>
<td>No</td>
<td>AoV</td>
<td>Coarc</td>
<td>AliveC</td>
</tr>
<tr>
<td>^</td>
<td>Yes</td>
<td>AoV</td>
<td>Coarc, PDA</td>
<td>Died</td>
</tr>
<tr>
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<td>Yes</td>
<td>AoV</td>
<td>No</td>
<td>Died</td>
</tr>
<tr>
<td>^</td>
<td>No</td>
<td>NorwoodB</td>
<td>No</td>
<td>AliveC</td>
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</table>

anulus (range 5 to 8 mm) was compared with that in normal subjects (6.8 ± 0.6 mm) and was not significantly different, although four infants with decreased function had anuli 5 mm in diameter. Poststenotic dilation of the aorta as assessed by the ratio of the diameter of the ascending aorta to that of the valve anulus was present in all and averaged 1.8 (range 1.4 to 2.2), which was significantly greater than the normal 1.04 (range 0.7 to 1.2) (p < .01; figure 5). Coarctation of the aorta appeared to reduce this ratio slightly in two patients. Doppler sampling in the ascending aorta was severely disturbed in seven of seven.

FIGURE 4. Short-axis parasternal view of the left ventricle in an infant with critical aortic stenosis. Note the increased density of the papillary muscles (small arrows) comparable to the density of the pericardium (open arrow). A = anterior; L = left, P = posterior; R = right; VS = ventricular septum.

CRITICAL AORTIC STENOSIS  
ASCENDING AORTA POST-STEMOTIC DILATION

FIGURE 5. Comparison of the ratios of the diameter of the ascending aorta to that of the aortic valve anulus in normal subjects and in patients with critical aortic stenosis.

A comparison of the two-dimensional echocardiographic predictions and the anatomic findings by combination of surgical, angiographic, and autopsy data showed that there were no false-positive or negative results by two-dimensional echocardiography with regard to intracardiac anatomic characteristics, including those of the atrial situs and atrial and ventricular septa and morphologic characteristics of the ventricles. The marked right ventricular enlargement that frequently accompanied severe aortic stenosis produced a typical picture at autopsy when compared with normal (figure 6). Diagnosis of extracardiac anatomic characteristics (pulmonary and systemic venous return, aortic arch branching, and the presence of proximal coronary arteries) was accurate in all but one patient with mild coarctation (see Discussion).

Discussion

The clinical diagnosis of critical aortic valve stenosis in infancy can be made more difficult in the presence of severe ventricular dysfunction. Hemodynamics at catheterization in these patients may reveal little evidence of the severity of aortic valve disease as a result of poor left ventricular function and the inability to generate elevated left ventricular end-systolic pressure. Because of this and the extreme clinical instability of infants with symptomatic aortic stenosis, an accurate noninvasive method of diagnosis would be useful. It has been our clinical impression that patients undergoing aortic valvotomy when in relatively stable condition enjoy better operative results than those whose conditions are compromised. Therefore, noninvasive echocardiographic criteria were sought that might accurately characterize these infants and per-
perhaps improve the results of surgical intervention by obviating the need for angiographic diagnosis. From noninvasive information we were able to characterize critical aortic stenosis in infancy as a combination of (1) severe immobility of the aortic valve without demonstrable systolic opening, (2) poststenotic dilation of the aorta with an ascending aorta-to-aortic anulus ratio greater than 1.7 (in the absence of coarctation), (3) severely disturbed Doppler-determined pattern of ascending aortic flow velocity, (4) left ventricular hypertrophy with increased echo density of mitral papillary muscles comparable to the density of pericardium, and (5) right ventricular enlargement. Criteria 1, 2, and 3 were diagnostic of critical aortic stenosis, and 4 and 5 were present in all patients, although most prominently when the left ventricular shortening fraction was less than 25%.

The overriding principle in the treatment of the in-

FIGURE 6. Comparison of normal and abnormal ventricular morphology in autopsy specimens cut to simulate the parasternal long-axis two-dimensional echocardiographic scan. a. Normal ventricular morphology in an infant who died of noncardiac causes. b. Severe right ventricular dilation and biventricular hypertrophy in an infant with severe aortic stenosis plus coarctation. Abbreviations are as in figure 1.
phant with critical aortic stenosis is to improve papillary muscle and endocardial left ventricular wall perfusion as early as possible. In those neonates with severe aortic valve stenosis and coarctation of the aorta it seems prudent to first treat the coarctation medically with prostaglandin infusion and then to proceed with early aortic valvotomy. They may then be weaned from prostaglandin therapy after surgery and, if this is not tolerated, then coarctation repair can be performed through a left thoracotomy. This is in contrast to the usual method of surgical treatment of multiple levels of left ventricular outflow tract obstruction in which the most distal obstruction is repaired first, but the usual approach is unsatisfactory when severe aortic valve stenosis and myocardial ischemia are present. We speculate that papillary muscle density on the two-dimensional echocardiogram may be a marker for left ventricular ischemia or fibrosis\(^\text{11}\) that can aid in this decision.

The pitfalls of noninvasive diagnosis in the critically ill hypotensive neonate are as follows:

1. Infants with coarctation of the aorta may present with ventricular dysfunction and poor cardiac output. They do not, however, manifest criteria 2 or 3 outlined above. With aortic stenosis it is necessary to exclude the possibility of severe aortic arch obstruction and this can be done with echocardiography (figure 7).\(^\text{12}\) One patient in this series (No. 5) developed coarctation of the aorta after successful aortic valvotomy. This neonate was moribund, with a pH of 6.8, on admission, and before and after aortic valvotomy there was no blood pressure gradient between the upper and lower extremities. However, at 3 months after discharge the right arm–to–right leg blood pressures differed by 40 mm Hg. This infant is now asymptomatic and underwent coarctation repair electively at 12 months of age. This appeared to be a case of coarctation developing in the first few months after birth.

2. In newborns severe cardiomyopathy may superficially resemble aortic stenosis, but these infants do not have poststenotic dilatation of the aorta or disturbed Doppler-determined ascending aortic flow velocity.

In some neonates with critical aortic valve stenoses left ventricular function is normal or hyperdynamic. In this situation hemodynamic catheterization may be needed to measure the severity of elevation in left ventricular pressure. It is likely that recent developments in the area of Doppler echocardiography will allow accurate estimation of the gradient between the left ventricle and aorta when left ventricular function is normal.\(^\text{13}\)

It is well known that left ventricular hypoplasia is an extremely poor prognostic factor in patients with critical aortic stenoses. Hypoplasia may be expressed as reduced left ventricular end-diastolic volume on the angiogram\(^\text{6}\) or by decreased cross-sectional area of the

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**FIGURE 7.** Three two-dimensional echocardiographic views of a 2.4 kg infant with critical aortic stenosis (patient No. 7) and severe coarctation of the aorta. The parasternal long-axis view (*top*) shows dilation of the left ventricle (LV) and right ventricle (RV) and mild poststenotic dilation of the ascending aorta (Ao) (less marked than in those without coarctation). A subcostal four-chamber view (*middle*) shows bulging of the atrial septum toward the right atrium (RA) (white arrow) and left atrial (LA) dilation. High parasternal scanning of the aortic isthmus (*bottom*) revealed coarctation of the aorta (black arrow) and a small patent ductus arteriosus on prostaglandin therapy. A = anterior; DAo = descending aorta; I = inferior; L = left; MPA = main pulmonary artery; P = posterior; R = right; S = superior.
left ventricle on the parasternal long-axis two-dimensional echocardiographic scan. One patient in this series had this complication of critical aortic valve stenosis and therefore was treated with a palliative operation usually only in patients with hypoplastic left heart syndrome. A neonate with a poorly functioning left ventricle with a cross-sectional long-axis area of less than 1.8 cm² is unlikely to survive valvotomy.

A complete assessment of intracardiac and extracardiac anatomic characteristics is necessary before surgery without catheterization in infants with critical aortic valve stenoses. It is not sufficient that echocardiography be accurate in the diagnosis of severe aortic valve stenosis while important associated lesions that should be corrected at the time of open heart surgery are overlooked. Particular emphasis should be placed on the noninvasive diagnosis of associated congenital defects that may influence the surgical technique, such as abnormal systemic venous return.

Compared with the usual diagnostic approach to patients suspected of having critical aortic valve stenoses in which catheterization and angiography are used, the use of echocardiography allowed expeditious surgical treatment and four of five infants managed in this way survived operation. Although it is not yet clear that echocardiography will be superior to angiography in the management of these critically ill infants, it provided comparable morphologic information and, based on this experience, clinical examination (including a chest x-ray and electrocardiographic and echocardiographic examinations) may be adequate to make a diagnosis and plan surgical therapy. This approach was designed for the markedly symptomatic infant and would not be appropriate for the asymptomatic infant with aortic stenosis and no evidence of cardiac dilation. When hemodynamic data could influence patient management, cardiac catheterization should be performed.

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References

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