Echocardiographic Findings in Discrete Subvalvular Aortic Stenosis

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SUMMARY
Echographic features of three patients with discrete subvalvular aortic stenosis are presented. These include a narrowed area of the left ventricular outflow tract just below the aortic valve cusps in all cases. A high-intensity but thin echo was seen in the high left ventricular outflow tract at the level of the mitral annulus in one patient with a subaortic diaphragm. In one of the three patients abnormality of aortic cusp motion was suggested as well.

Additional Indexing Words:
Ultrasound Angiography Idiopathic hypertrophic subaortic stenosis Diagnostic methods Congenital heart disease

THE DIAGNOSIS of discrete subvalvular aortic stenosis may be quite difficult despite careful clinical, hemodynamic, and angiographic studies.1, 2 Highly unusual echocardiographic findings were noted in three cases with subvalvular aortic stenosis recently studied at Stanford Medical Center. In the first case, the diagnosis was made prospectively, while in the second and third cases, with previously known subaortic stenosis, echocardiography was done to look for abnormalities.

Methods
Echocardiography was performed using a Smith Kline Ekoline 20A ultrasonoscope and 2.25 MHz transducer of 0.5 inch diameter, having an acoustic lens providing beam collimation to 5 cm tissue depth. The echocardiograms were recorded on trip chart records, using the Honeywell #1856 Fiberoptics recorder or the Electronics for Medicine DR8 recorder. In addition, records of the oscilloscope face were made, using a Polaroid camera.

The patients were studied in the supine position, with the transducer placed in the intercostal space which would allow recording of the free edge of the mitral valve with the transducer oriented perpendicular to the chest wall. Phonocardiograms were obtained on the DR8 recorder using a Maico contact microphone and a band-pass filter for frequencies between 120 and 500 Hz.

Case Reports
Case No. 1
This 54-year-old man had a heart murmur since birth. Three months prior to admission he had a febrile flu-like syndrome and was given antibiotic therapy for several days. Fatigue, sweats, and intermittent fever as high as 102°F persisted for three months. Two days prior to admission he developed blurred vision in the right eye, with accompanying headache. The night of admission he developed total loss of vision in the right eye and sought medical attention.

Physical examination showed a blood pressure of 110/60 mm Hg, pulse 92 and regular, temperature 101°F. There was a right homonomous hemianopsia with normal optic fundi. The carotid pulse was felt to have an anacrotic shoulder and shudder. A precordial thrill was palpable at the cardiac base, and a grade V/VI harsh ejection systolic murmur was heard throughout the precordium, with maximum intensity at the base and radiation to the carotid vessels. No ejection click could be appreciated, and the aortic second sound was soft. There were no symptoms of congestive heart failure.

Electrocardiogram showed left bundle branch block, and chest X-ray showed a prominent ascending aorta without evidence of calcium within the cardiac silhouette. The initial four blood cultures were positive for alpha hemolytic streptococcus, and the patient was started on penicillin therapy. Echocardiography was interpreted as showing an abnormality of the left ventricular outflow tract just below the level of the aortic valves. A fine, but high-intensity echo was recorded below the level of the aortic valve and near

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the annular insertion of the mitral valve (fig. 1). This echo remained within the left ventricular outflow tract throughout the cardiac cycle. The structure apparently made contact with the anterior mitral valve leaflet during diastole, but separated from the anterior mitral leaflet during systole (fig. 2). It approached the interventricular septum during systole and moved back towards the mitral valve during diastole. This echo was lost as the sound beam was directed closer to the free edge of the mitral valve, but was present on each sweep of the left ventricular outflow tract from the body of the ventricle to the aorta (fig. 1). On such sweeps a short, narrow area just below the aortic valve was noted (fig. 2). High-frequency vibrations of the aortic valve cusps were recorded during systole, and in addition, less than normal separation of the valve cusps was seen (fig. 1). Echographic studies during provocative maneuvers for idiopathic hypertrophic subaortic stenosis were negative for the pattern associated with this condition (fig. 3). After the course of antibiotic therapy, cardiac catheterization showed a subaortic pressure gradient of modest degree (fig. 4) and no evidence for idiopathic hypertrophic subaortic stenosis.

A radiolucent line just below the aortic valve and a mild degree of aortic regurgitation were shown by angiography. Because of the lack of hemodynamic impairment and the patient’s good clinical state, surgery was not performed.

Case No. 2
This 16-year-old boy was known to have had a murmur since birth. He had a ventricular septal defect repaired at age nine, and at surgery a broad, “fibromuscular subaortic diaphragm” was excised “as completely as possible.” At age 12 he was again catheterized to evaluate a persistent murmur, and a gradient of 75 mm Hg was documented within the left ventricular outflow tract. Over the year prior to admission, the patient noted exertional chest pain and was re-evaluated for this symptom.

Physical examination showed a blood pressure of 120/70; pulse 80 and regular. Carotid pulse was felt to be within normal limits, but precordial examination showed a systolic thrill in the second, third, and fourth left intercostal spaces along the left sternal border. The first and second heart sounds were within normal limits with no ejection click, and there was a grade IV/V ejection systolic murmur at the base, with poor

Figure 1
Echocardiogram of M-mode arc-scan of the left ventricular outflow tract from the aorta (left) to the left ventricle (right) from case 1. The speed of transducer motion was not uniform during this scan. Note the area with multiple echoes just below the aortic valve leaflet and thin echo (arrows) in the left ventricular outflow tract showing systolic anterior motion and diastolic motion to the level of the anterior mitral valve leaflet. This is not the echographic pattern of idiopathic hypertrophic subaortic stenosis, since the systolic anterior motion is seen high in the outflow tract (center) but is not seen near the free edge of the valve (right). This is the reverse sequence of the findings in HHSS (see fig. 3). Aortic valve motion shows systolic vibrations in the center of the aorta (AO) at the left of the panel. PHONO = phonocardiogram; 2 RSB = second interspace right sternal border; ECG = electrocardiogram; LA = left atrium; IVS = interventricular septum; LV PW = left ventricular posterior wall.

Figure 2
Echograms from case 1 made with Polaroid camera. A. Photograph of the multiple echoes in the narrowed area just below the aortic level. B. Slightly below the area shown in A, note the fine echo between those of the interventricular septum and the anterior mitral leaflet. This is thought to represent the subaortic diaphragm (SAD). LVOT = left ventricular outflow tract; MV = mitral valve. 1 cm between vertical calibration dots.

Figure 3
Echocardiogram of M-mode arc-scan of the heart from the aorta (left) to the left ventricle (right) in a patient with idiopathic hypertrophic subaortic stenosis. The speed of the transducer motion was uniform during this scan. Note the area of outflow tract narrowing seen during systole (double arrows). This narrowing is most striking near the free edge of the valve (right of mitral valve [MV] label), and is not well seen at the level of the mitral annulus (left of MV label). Note the thickened interventricular septum (IVS) compared with the posterior wall (PW) of the left ventricle (LV). A mid-systolic closure motion and vibrations are recorded in the aortic valve (single arrow, left). ECG = electrocardiogram; PHONO = apex phonocardiogram; LA = left atrium.
radiation to the carotid vessels. A soft early diastolic blowing murmur was heard along the left sternal border.

In this case, the thin echo described near the mitral valve in case 1 was not found. However, on scanning of the left ventricular outflow tract (by sweeping the transducer from the area of the aorta, above the aortic valve, down to the body of the left ventricle below the mitral valve) a long area of narrowing was appreciated just below the aortic root (fig. 5). Care was taken to sweep the transducer along the left ventricular outflow tract at constant speed in order to achieve a rough estimation of the length of the narrowing. In contrast to the usual thin echoes of the mitral valve near the annular insertion, which make a smooth transition to the rather bright but thin aortic root echoes, this patient showed multiple thick echoes in the area of the left ventricular outflow tract near the mitral annulus and high intraventricular septum. No abnormalities of the aortic valve could be appreciated, despite the technical adequacy of their recording (fig. 5).

Electrocardiogram showed evidence of left ventricular hypertrophy. Minimal dilation of the ascending aorta and evidence of left ventricular enlargement were seen in the chest X-ray. At cardiac catheterization there was no evidence of residual intracardiac shunt, but a 56 mm Hg pressure gradient was present on pullback recording from the body of the left ventricle to the subvalvular area, with no further pressure drop across the aortic valve. Maneuvers to demonstrate idiopathic hypertrophic subaortic stenosis were negative. Left ventriculogram showed a long, narrow subaortic chamber (fig. 7). The patient underwent re-operation, and remnants of the fibrous membrane and hypertrophic muscle bands were widely excised. The aortic valve was congenitally deformed, with abnormally small right and left valve cusps resulting in aortic regurgitation.

The preoperative echocardiogram showed a minimal end-systolic outflow tract dimension less than 8 mm, due to thick septal echoes at the level of the mitral annulus; septal thickness at this level was 10 mm and equal to that in the body of the ventricle (fig. 5). The postoperative echogram was performed in a fashion nearly identical to the preoperative study (fig. 6). The minimal end-systolic outflow tract dimension was 12 mm. While the thickness of the septal echoes in the body of the ventricle remained 10 mm, the septum at the outflow level was only 5 mm.

Case No. 3

This five-year-old girl had a heart murmur detected at birth, and at ten days of age the diagnosis of aortic coarctation was made. Digoxin was given at that time because of mild congestive heart failure. At age three, the patient was asymptomatic, and digoxin was discontinued. She was admitted for cardiac evaluation...
Anteroposterior and lateral biplane left ventriculograms exposed in systole in case 2 (top panel) and case 3 (bottom panel). Arrows indicate subaortic narrowing in each patient. A linear lucency can be seen on each of the lateral views. Note the dilated ascending aorta and the distance between the lucency and the aortic valve in case 2 (top panel). Narrowing and lucency are immediately below the sinuses of Valsalva in case 3 (bottom panel).

because of increasing evidence of left ventricular hypertrophy on serial electrocardiograms.

Physical examination showed a blood pressure of 140/90 mm Hg in the upper extremities, and 50/20 mm Hg in the lower extremities. Femoral pulses were not palpable, and the carotid pulse was normal. A systolic thrill was felt at the cardiac base, and there was a grade IV/VII harsh ejection murmur throughout the precordium, with maximum intensity at the base and radiation to the neck. The second heart sound was single and was followed by a short, soft diastolic murmur maximal at the left sternal border. No ejection click could be heard. There was no evidence of congestive heart failure.

Electrocardiograms showed left ventricular hypertrophy. Prominence of the left ventricle was present on chest X-ray. A peak systolic gradient of 190 mm Hg across the left ventricular outflow tract, with a left ventricular pressure of 320/12 mm Hg, was recorded at cardiac catheterization. Left ventriculography showed a discrete area of narrowing of the left ventricular outflow tract just below the aortic cusps (fig. 7). Coarctation of the aorta was also demonstrated.

On scanning the left ventricular outflow tract of this patient, a very small distance between the annular insertion of the mitral valve and the interventricular septum was noted (fig. 8). This was consistently recorded. The aorta was thought to be of normal size for a patient of this age, but the subvalvular area was narrow relative to the aorta. The subvalvular chamber of the outflow tract measured 6 mm on the echocardiogram (fig. 8). Aortic valve motion could not be recorded.

At operation the aortic valve was found to be bicuspid. Six millimeters below the aortic annulus severe stenosis of the left ventricular outflow tract due to a thick fibromuscular collar was found. This had a

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**Figure 7**
Anteroposterior and lateral biplane left ventriculograms exposed in systole in case 2 (top panel) and case 3 (bottom panel). Arrows indicate subaortic narrowing in each patient. A linear lucency can be seen on each of the lateral views. Note the dilated ascending aorta and the distance between the lucency and the aortic valve in case 2 (top panel). Narrowing and lucency are immediately below the sinuses of Valsalva in case 3 (bottom panel).

**Figure 8**
Preoperative echocardiogram of M-mode arc-scan of the heart from case 3. The transducer was swept from the body of the left ventricle (LV) (left) to the level of the aorta (AO). Note the small size of the left ventricular outflow tract (ot) in comparison with the scale and the aorta. The aortic valve was not recorded in this patient. ecg = electrocardiogram.

**Figure 9**
Postoperative echocardiogram of M-mode arc-scan of the heart from case 3. Technique and labels as in figure 8. The size of the left ventricular outflow tract is very similar to the preoperative record. (The slightly altered appearance from figure 8 is due to different recording apparatus.) The aortic valve is recorded with normal motion (arrow).
central circular orifice approximately 5 mm in diameter. After excision of this ring, the left ventricular outflow tract easily accommodated a 14 mm probe. The patient’s postoperative recovery was uneventful.

The postoperative echocardiogram is shown in figure 9. A subvalvular narrowing of the outflow tract still was recorded. No significant change in the dimensions of the outflow tract was appreciated in comparison with the preoperative record. However, it was interesting to note a reduced end-systolic measurement of the left ventricle. The aortic valve motion was normal.

Discussion

Discrete subvalvular subaortic stenosis may closely mimic the usual type of congenital valvular aortic stenosis or supravalvular aortic stenosis in all aspects of the history, physical examination, chest X-ray and electrocardiogram.\(^1\)\(^-\)\(^8\) It is stated that the distinction between subvalvular aortic stenosis and other forms must be made at cardiac catheterization and angiography.\(^1\)\(^-\)\(^2\) Obviously the accuracy of diagnosis will depend on the care exercised during these procedures resulting from a high index of suspicion, as well as on the anatomy of the subvalvular obstruction. There are at least two basic forms of subaortic stenosis of a discrete variety.\(^1\)\(^-\)\(^8\) There may be a thin subvalvular membrane or diaphragm immediately below the aortic valve cusps and attached to the mitral valve, which may make this abnormality difficult to recognize by withdrawal pressure tracings and/or angiograms. In the other common form of this lesion, subaortic obstruction is due to a rather long narrow fibromuscular chamber which is more easily recognized by hemodynamic and angiographic study. In case 1, we believe the thin subvalvular membrane to be the type of obstruction present. This is suggested by the definite intraventricular pressure gradient without a gradient from the high left ventricular outflow tract to the aorta (fig. 4), lack of a demonstrable subvalvular chamber on angiograms, as well as a radiolucent line immediately below the aortic valve. It is suggested that the fine, high-intensity echo in the left ventricular outflow tract represents the subaortic diaphragm itself. This fine echo has not been seen in any other case recorded in our laboratory, despite studying over 2000 patients with heart disease. This is certainly a distinctive finding, and its presence strongly suggests the diagnosis of fixed membranous subvalvular aortic stenosis. Echograms from two patients with subaortic diaphragm were published by Ultan, Segal and Likoff.\(^9\) However, it is difficult to appreciate differences between the published waveform and the waveform of the mitral valve near its attachment to the annulus. The waveform of the subaortic diaphragm reported here demonstrated motion towards the transducer during systole and away from the transducer during diastole. This is the reverse of the waveform demonstrated by Ultan, Segal and Likoff.\(^9\)

The pattern of the abnormal echo within the outflow tract seen in case 1 can be explained by the fact that such tissue may be attached as a ring inserted between the base of the aortic valve cusps and the anterior mitral valve annulus. In this way the anterior mitral leaflet would be at the same level and possibly strike this diaphragm when it is in its open position, while closure of the mitral valve and bowing of the diaphragm towards the aorta would lead to separation of the echo from the mitral valve echo and its motion closer to the transducer. Lundström and Edler showed an echogram from one of four cases of membranous subaortic stenosis.\(^10\) The linear echo in the “outflow tract” is apparently above the level of the annulus in this case. Davis et al.\(^11\) recently reported the finding of high-frequency vibrations of the aortic valve cusps, with failure of the valve cusps to open fully and remain open during systole in three patients with fixed subvalvular aortic stenosis. The cause of this finding is not clear, but the aortic valve motion in case 1 (fig. 1) was very similar to that demonstrated in their presentation. A similar phenomenon has been seen in angiograms.\(^8\) They did not comment upon findings within the left ventricular outflow tract in their cases. There was a definite abnormality in the subvalvular area in case 2 of this report. The multiple thick echoes from the ventricle at a level near the annular attachment of the anterior mitral valve leaflet and below the sinuses of Valsalva are distinctly abnormal. By taking care to sweep the sound beam along the outflow tract at a uniform speed and repeatedly, we were able to confirm that this was a long subvalvular area which was definitely present (fig. 5). Reduced septal thickness and increased outflow tract dimensions were recorded postoperatively (fig. 6). Dr. Raymond Gramiak (personal communication) has also encountered one patient with fixed subvalvular aortic stenosis in whom there was an apparent narrowing of the outflow tract below the aortic valve area. While the end-diastolic echocardiographic measurement of the left ventricular outflow tract was similar to the surgical measurement in case 3, the postoperative study did not show this correlation (figs. 8 and 9). It is possible that the relief of the obstruction reduced the
systolic distending pressure in the outflow tract. A reduced end-systolic size of the left ventricle in the postoperative study is compatible with this hypothesis, but in view of the persistent hypertension due to coarctation of the aorta in this patient, we must conclude that the size of the subaortic chamber was not accurately measured by the echocardiogram in this last case.

It has been recognized that fixed subvalvular aortic stenosis may be associated with aortic insufficiency in many cases.\(^1, 8, 12\) This may be related to damage of the valve tissue by a subvalvular jet,\(^8\) to infective endocarditis in some cases,\(^12\) to interference with proper closure of the aortic valve by the subvalvular tissue, or to congenital deformity of the valve. Absence of dilation of the first portion of the aorta is a sign of nonvalvular aortic obstruction, but such dilation has been reported in the presence of subaortic stenosis and in the absence of valvular aortic stenosis.\(^1\) In our cases, this did not lead to the proper diagnosis (fig 7). The presence of calcification within the aorta leads one toward the diagnosis of valvular aortic stenosis, but the absence of calcification is of little diagnostic value in trying to differentiate congenital valvular stenosis from subvalvular aortic stenosis.\(^1\) The absence of an ejection click may be the most consistently reliable clue to differentiate subvalvular from valvular aortic stenosis.\(^1\) It is suggested that screening with echocardiographic techniques may lead one to a greater suspicion of membranous or fibromuscular discrete subvalvular aortic obstruction, so that hemodynamic and angiographic studies can be carried out with special care, in order to differentiate these two conditions and separate them from cases of valvular aortic stenosis.

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