ILLUSTRATIVE ECHOCARDIOGRAM

Long Segment (Tunnel) Subaortic Stenosis*

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The echocardiogram was obtained from an 18-year-old man who was known to have had a cardiac murmur since birth.

CASE REPORT
At the age of eight years, the patient underwent cardiac catheterization to evaluate his limited exercise capacity and because of the progression of the left ventricular hypertrophy on his electrocardiogram. The clinical diagnosis of fixed subvalvular aortic stenosis was confirmed, with a mean gradient of 50 mm Hg measured across the left ventricular outflow tract. On the anteroposterior angiogram, a distinct subvalvular membrane was seen. At surgery a fibromuscular diaphragm was excised. The reports do not mention other abnormalities in the subvalvular area. During the period of follow-up prior to this echocardiogram, poor exercise tolerance was the only complaint.

Physical Examination
The patient's pulse rate was 78 beats per minute and regular. The blood pressure was bilaterally equal and measured 120/80 mm Hg. No increased jugular venous distention was present. A shudder was palpated over both carotid arteries. The point of maximal cardiac impulse was in the left fifth intercostal space just outside the midclavicular line. At the left sternal border a systolic ejection murmur of grade 4/6 was heard, with radiation to the cardiac base and carotid arteries. No ejection click was appreciated. At the lower sternal border, a diastolic decrescendo murmur of grade 2/6 was present.

The ECG showed marked left ventricular hypertrophy with early QRS-T divergence. Prominence of the left ventricle was present on the chest x-ray film.

LONG SEGMENT SUBAORTIC STENOSIS

FIGURE 1. M-mode scan from aorta (Ao) to apex of left ventricle (LV) performed with uniform speed of transducer. Note length of narrowed left ventricular outflow tract (OT). IVS, interventricular septum; c, cusps of aortic valve; F, fluttering of mitral valve; LA, left atrium; aML and pML, anterior and posterior leaflets of mitral valve; IVS, interventricular septum; ppm, posteromedial papillary muscle; and PW, posterior wall of left ventricle.
Echocardiogram

Figure 1 presents the patient's M-mode scan from the aorta to the apex of the left ventricle. The scanning speed of the transducer was as uniform as possible. The diameter of the aorta is normal, but there is a striking abnormality of the cusps of the aortic valve. The initial rapid opening is followed by an abrupt partial closure throughout the remainder of systole, and there is fluttering of the cusps. The left atrium is dilated, with a ratio of the left atrium to the aorta of 1.6 (normal, less than 1.3). The left ventricular outflow tract (that is, the area between the level of the aortic valve and that of the mitral valve) is long and diffusely narrowed (early-systolic and end-systolic dimensions of 15 and 10 mm, respectively). The anterior and posterior leaflets of the mitral valve demonstrate a tiny high-frequency flutter. There is concentric hypertrophy of the left ventricle, as the thickness of the septum and the thickness of the posterior wall are equal and measure 16 mm. At the level of the posterior papillary muscles, only a virtual space of the left ventricle is present, indicating ventricular emptying that is greater than normal (angiographic equivalent of apical obliteration). Real-time cross-sectional analysis of the sagittal plane through the left ventricle using a multicyrystal scanning device (ECHOcardioVISOR 02) clearly demonstrated a distinct narrowing of the outflow tract persisting throughout both phases of the cardiac cycle (Fig 2).

DISCUSSION

The correct diagnosis of the type of subvalvular aortic stenosis may be difficult despite careful clinical, hemodynamic, and angiographic studies. Distinctive echocardiographic findings have been reported recently. Davis et al. suggested that closure of the aortic valve in early systole that persists throughout the remainder of systole and also coarse fluttering of these leaflets, both in the absence of asymmetric septal hypertrophy, are suggestive, if not specific, for the disease. Popp et al. extended the echocardiographic criteria, including a linear echo in the left ventricular outflow tract of the thin subvalvular membrane as representing the discrete form of the disease and specifying the generalized narrowing of the left ventricular outflow tract as representing the long-segment obstructive form. Thus, these features further permit differentiation between the two basic forms of subaortic stenosis.

This patient apparently has the long-segment or tubular subvalvular type of aortic stenosis. The absence of aortic insufficiency is generally considered as an argument against the diagnosis of subaortic stenosis. The presence of aortic insufficiency may be seen on the echocardiogram as a high-frequency diastolic fluttering of the mitral valve and is thought to be the result of the regurgitant jet striking the leaflets or the result of turbulence in the left ventricle. Such fluttering is present on the echocardiogram of this patient and thus further supports the diagnosis. Fixed subvalvular aortic stenosis most commonly produces symmetric left ventricular hypertrophy (in this case, 18 mm). The differential diagnosis for idiopathic hypertrophic subaortic...
stenosis is thus easily made echocardiographically by the absence of an asymmetric thickened septum, as well as by the absence of the typical systolic anterior motion of the anterior leaflet of the mitral valve, causing the outflow obstruction in idiopathic hypertrophic subaortic stenosis; however, both of the latter abnormalities may exceptionally be present in patients with tunnel subaortic stenosis.4

Although angiocardiographic studies play a paramount role in the diagnosis of subvalvular aortic stenosis,5 confusion may prevail in the interpretation, or the diagnosis may be missed. In some cases of fixed subaortic stenosis, a membrane may co-exist with a long-segment narrowing. This case report is an example where only the membrane was diagnosed from the anteroposterior angiogram. We have had experience with two other cases of long-segment subvalvular stenosis mistakenly thought to represent valvular aortic stenosis, despite hemodynamic and angiographic evaluation. The anteroposterior end-systolic left ventriculogram of one of these patients is shown in Figure 3. A catheter was inserted into the left ventricle via the transseptal route, as it was impossible to get into the left ventricle via the retrograde method. A pressure gradient of 110 mm Hg was measured between the left ventricle and aorta, and the diagnosis of valvular aortic stenosis was accepted because no subvalvular narrowing was seen on the anteroposterior angiogram. The cusps of the aortic valve demonstrated thickening and reduced motion. However, a distinctly narrowed outflow tract was seen on the M-mode scan and ultrasonic cross-sectional images (Fig 4).

The cases discussed here do suggest that the narrowed left ventricular outflow tract may be thought of as a flattened space in some cases. It appears that the echocardiographic beam most likely passes through the small dimension of the narrowed asymmetric tunnel, whereas the x-ray beams in the anteroposterior projection are perpendicular to the tunnel, resulting in visualization of the large dimension of the tunnel on the angiogram. Thus, for an accurate analysis of the outflow tract, biplane studies are mandatory. This information can also be obtained by combining single-plane angiocardiographic and echocardiographic studies.

We concluded that examination with ultrasound may either make one suspicious or definitely determine the diagnosis of long-segment subvalvular aortic stenosis when the previously mentioned diagnostic features are present.

References