Echocardiography in Coexisting Hypertrophic Subaortic Stenosis and Fixed Left Ventricular Outflow Obstruction

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SUMMARY

Echocardiographic study was done on four pediatric patients with coexisting hypertrophic subaortic stenosis (HSS) and fixed left ventricular outflow (LVO) obstruction. One had valvar aortic stenosis, two had discrete subaortic stenosis and one had coarctation of the aorta. Preoperatively, HSS was not suspected in any of these patients because of the classical findings of fixed LVO obstruction. The echocardiographic examination revealed an abnormal systolic anterior movement (SAM) of the anterior mitral leaflet with ventricular septal hypertrophy in all, but these findings were overlooked or thought to be related to a subvalvar diaphragm. One or two years after the surgical removal of their fixed obstruction, all showed clinical pictures of HSS. Postoperative echocardiographic examination showed an abnormal SAM with asymmetrical septal hypertrophy, and cardiac catheterization revealed mild to severe degrees of resting pressure gradients across LVO tracts. Amyl nitrite inhalation produced a more prominent SAM with increasing pressure gradient, and angiotensin infusion abolished the SAM and reduced the pressure gradient.

From our present study, echocardiography has the potential for identification of HSS coexisting with a fixed LVO obstruction. We recommend that all patients with LVO obstruction have an echocardiographic examination along with pharmacological testing. The demonstration of an unusually thickened ventricular septum and especially, abnormal SAM of the mitral valve would require careful evaluation of left heart hemodynamics and the determination of the level of the major obstruction during cardiac catheterization for pre and postoperative management.

Additional Indexing Words:
Ultrasound cardiography
Idiopathic hypertrophic subaortic stenosis
Hypertrophic obstructive cardiomyopathy

VALVAR AND FIBROUS SUPRAVALVAR or subvalvar forms of aortic stenosis cause a fixed and nonvariable obstruction to left ventricular outflow. On the other hand, when the obstruction is due to hypertrophic subaortic stenosis, it may vary in the same subject from one moment to the next and even within the systolic segment of one cardiac cycle. Because of its labile and variable nature, there has been no single satisfactory method for diagnosing hypertrophic subaortic stenosis. When the obstruction is only of mild degree or coexists with fixed left ventricular outflow obstruction, the precise clinical or laboratory diagnosis is more difficult and requires a provocative test to increase the left ventricular outflow tract obstruction.1, 15

Several investigators, using cineangiography2, 3 and ultrasound techniques,4-6 have suggested that the pathogenesis of the obstruction in hypertrophic subaortic stenosis is the narrowing of the left ventricular outflow tract produced by septal hypertrophy in association with systolic apposition of the anterior leaflet of the mitral valve to the ventricular septum. Echocardiographic demonstration of this characteristic abnormal systolic anterior movement has recently been shown to be highly specific in diagnosing hypertrophic subaortic stenosis as well as capable of separating patients with persistent resting obstructions from those with latent and
labile obstructions.6,7 The echocardiographic demonstration of asymmetrical septal hypertrophy represents an additional valuable diagnostic sign in hypertrophic subaortic stenosis.8

The purpose of this study is to demonstrate the value of the ultrasound technique in the preoperative recognition of hypertrophic subaortic stenosis when it coexists with lesions which produce fixed left ventricular outflow tract obstruction.

Materials and Methods

This study consists of four pediatric patients with ages ranging from six to fifteen years. All were studied by cardiac catheterization, angiography, and echocardiography but without employing any provocative maneuvers. All exhibited fixed left ventricular outflow tract obstruction (2 with discrete subaortic stenosis, 1 with valvular aortic stenosis, 1 with coarctation of the aorta). All underwent cardiac surgery where the fixed obstruction was identified and surgically corrected. Each patient was restudied one or two years postoperatively by the methods described because of lack of any significant improvement in their clinical status after surgery. Retrograde catheterization was used to place catheters in the ascending aorta and body of the left ventricle. Patients were studied at rest, following amyl nitrite inhalation and during intravenous infusion of angiotensin in doses ranging from 0.02-0.08 μg/kg/min. In two patients, continuous pressure and ultrasonic recordings were carried out simultaneously during cardiac catheterization. In the other two patients, ultrasound was employed independently of the cardiac catheterization study, and brachial artery blood pressure was used to monitor the effect of angiotensin infusion.

The echocardiographic examination was carried out using a commercially available ultrasonoscope (Picker Model 103) and a 2.0 Mhz transducer. A Tektronix 565 Dual Beam oscilloscope was operated as a slave and displayed the ultrasonic data in B mode on the upper beam. The lower beam was used in the multi-trace operation to record physiological data. Continuous recordings were made on 35 mm film using an oscilloscope record camera. Film transport speed was selected to equal 125 mm/second as obtained by ordinary strip chart recorders. Each examination included multiple beat recordings of mitral valve motion and evaluation of the width of the left ventricular outflow tract, thickness of the ventricular septum and of the posterior wall of the left ventricle. During provocative testing, attention was centered on the mitral valve and left ventricular outflow tract. The entire amyl nitrite response was recorded and serial studies were made during the angiotensin infusion.

Results

In preoperative presentation, all patients showed clinical, cardiac catheterization and angiographic evidence of their fixed left ventricular outflow obstruction only. But retrospectively, two patients had ventricular septal hypertrophy by left ventricular angiogram. Preoperative echocardiographic examination revealed an abnormal systolic anterior movement of the anterior leaflet of the mitral valve, either as a typical (fig. 1) or atypical (fig. 2) form with ventricular septal hypertrophy in each case. All patients met the criteria for surgical relief of their fixed left ventricular outflow obstruction. During surgical procedure, subaortic muscular hypertrophy was noted in three patients which did not appear to be sufficiently massive to require a ventriculomyotomy. There was no improvement in clinical symptoms or electrocardiogram after surgery. After removal of the afterload provided by the fixed obstruction which was masking the muscular component of left ventricular outflow obstruction, these patients showed clinical pictures of hypertrophic subaortic stenosis. Their immediate postop-

Figure 1
Comparison of a normal mitral valve (MV) recording (lower panel) and one obtained from a patient with coarctation of the aorta and hypertrophic subaortic stenosis (upper panel). Note an abnormal systolic anterior motion of the mitral valve and a ventricular septum (VS) which is twice as thick as the posterior wall (PW) in the latter. The arrows identify the systolic segments of the record. ECG = electrocardiogram; PHONO = phonocardiogram.

Figure 2
Atypical presentation of the systolic abnormality of mitral valve (MV) motion in a patient with a discrete subaortic stenosis and hypertrophic subaortic stenosis. The onset of the abnormal motion occurs late in systole and the echo pattern is fragmentary. Also note the poor gain control which failed to bring out complete picture of abnormal systolic anterior movement of the mitral valve. These resulted in preoperative oversights of the functional component of obstruction. ECG = electrocardiogram; PHONO = phonocardiogram.
ERATIVE and subsequent serial echocardiographic studies showed a classical abnormal systolic anterior movement of the mitral valve with asymmetrical septal hypertrophy in all. The width of the left ventricular outflow tract was measured and it was noted that its size was reduced to two-thirds or less than that seen in patients of similar age who did not have left heart outflow tract obstruction (fig. 3.)

Postoperative cardiac catheterization studies revealed mild to severe resting pressure gradients between the left ventricle and the ascending aorta. Inhalation of amyl nitrite was regularly associated with a marked increase in the pressure gradient\(^9\) and the simultaneously recorded echocardiograms showed an increase in the amplitude and duration of the systolic anterior motion of the mitral valve. Administration of angiotensin produced a marked reduction or elimination of the pressure gradient\(^9\) and abolished the systolic anterior movement in the echocardiogram (fig. 4). When the echocardiograms were obtained independently of the cardiac catheterization procedure, the same response to amyl nitrite and angiotensin could be observed. Three of the four patients had angiocardiographic evidence of ventricular septal hypertrophy and two showed an abnormal systolic motion of the anterior mitral leaflet. In the fourth patient, these observations could not be made.

**Discussion**

Hypertrophic subaortic stenosis may coexist with valvar aortic stenosis, discrete subaortic stenosis or systemic hypertension and usually presents with the clinical features of fixed obstruction predominating.\(^{11-13}\) The hypertrophic subaortic stenosis component of obstruction has been separated from the fixed portion by observing the response to provocative measures such as amyl nitrite inhalation and isoproterenol infusion as well as the character of the arterial pulse pressure in post-extrasystolic beats.\(^{14-18}\) In the patients presented in this study, the possibility of the coexistence of hypertrophic subaortic stenosis and fixed left ventricular outflow was not raised at the time of the initial studies. The clinical, cardiac catheterization and angiocardiographic presentations were regarded as indicative of a single fixed obstructing lesion. However, there are some findings that suggest that hypertrophic subaortic stenosis was probably present in the preoperative period. Analysis of the preoperative echocardiogram showed evidence of an abnormal echo pattern which appeared in the left ventricular outflow tract during systole and which resembled that seen in hypertrophic obstructive cardiomyopathy. Initially, these findings were overlooked because of their atypical presentation for hypertrophic subaortic stenosis or were thought to be related to a subvalvar ridge or diaphragm. Retrospectively, it appears clear that they represented abnormal systolic anterior movement of the anterior mitral leaflet and therefore represented reliable...
indicators of coexisting hypertrophic subaortic stenosis. The typical or incomplete finding which was shown in figure 2 cannot be disregarded and requires re-examination with better technique and especially with the use of provocative tests to establish the relationship of left ventricular outflow tract echo to hypertrophic subaortic stenosis. Ventricular septal hypertrophy could be observed in all recordings, further supporting the presence of pre-existing hypertrophic subaortic stenosis.

We have found echocardiography to be a highly sensitive detector of hypertrophic subaortic stenosis. The abnormal systolic anterior movement of the anterior leaflet of the mitral valve is highly specific for this condition and has not been noted in any other form of left ventricular outflow obstruction or in other primary endomyocardial diseases. The examination technique requires observation of the movement pattern of the entire mitral leaflet since false negative examinations can be obtained from the base of the mitral leaflet (fig. 5). In some instances, a provocative test may be required to demonstrate the abnormal systolic anterior movement when the obstruction is labile and latent.

Our present study documented that these children had coexisting hypertrophic subaortic stenosis and fixed left ventricular outflow obstruction. Demonstration of the association of these two types of left ventricular outflow obstructions raises the question as to their etiology. Do they exist as two entirely separate entities or is the hypertrophic subaortic stenosis secondary to the afterload imposed by the fixed obstruction? If the hypertrophic subaortic stenosis exists simply as a reactive muscular hypertrophy proximal to the fixed obstruction element20 (such as seen in right ventricular outflow hypertrophy in patients with severe valvar pulmonic stenosis),21, 22 one would expect its rather rapid disappearance or regression postoperatively. However, this was not the case in these four patients presented here. Over one to two years periods, the left ventricular outflow obstruction remained the same or actually increased in severity despite removal of the fixed obstruction. The persistence or increase in the degree of left ventricular outflow obstruction in the patients presented here, as well as those reported by others,13–16 would tend to support at least a degree of independent functioning of the two types of obstruction, although it is difficult to propose truly separate etiologies without further investigation.

Patients with combined fixed and variable left ventricular outflow obstruction present a problem in management different from those with fixed left ventricular outflow obstruction alone. Surgical correction of the fixed obstruction alone may not only fail to relieve the burden on the left ventricle, but it may also be accompanied by low cardiac output in the postoperative period as a direct result of the remaining hypertrophic subaortic stenosis. There are reports of patients dying in the postoperative period when this cause for low output was not recognized and isoproterenol therapy was initiated, effectively accentuating the very symptom prompting its use.14–16 Both surgical and medical therapy are available for consideration in such patients. When the obstruction is significantly or primarily the result of hypertrophic subaortic stenosis, those patients might well respond to a beta-adrenergic blocking agent such as propranolol.23, 24 In those children in whom the fixed obstruction itself is sufficient to require surgical relief, careful attention should be given to the possible need for ventriculomyotomy as well.13, 25, 26

Our studies indicate that echocardiography has a potential for identifying hypertrophic subaortic stenosis when it coexists with a fixed left ventricular outflow obstructing lesion. It appears necessary that all patients with left ventricular outflow obstructing lesions at any level should have echocardiographic examination including amyl nitrite provocation before cardiac catheterization and angiocardiology.

**Figure 5**

Effect of beam direction on ultrasound recording of mitral valve motion in hypertrophic subaortic stenosis. In all 3 panels recorded a few seconds apart during beam angulation, the same degree of obstruction is believed to be present as judged by the nature of the carotid pulse trace (CP) and the intensity of the systolic murmur recorded on the phonocardiogram (PCG). Recordings made near the top of the leaflet (A) show the largest systolic anterior movement. Echos obtained in more proximal portions of the mitral leaflet (B) show an attenuated systolic excursion, while those obtained near the base of the leaflet indicate no systolic abnormality (C). The small arrows show the onset of the abnormal mitral valve motion at its expected time of occurrence. AM = anterior mitral leaflet; ECG = electrocardiogram.
The demonstration of an unusually thickened ventricular septum and especially of an abnormal anterior systolic movement of the mitral valve would require careful evaluation of the left heart hemodynamics during cardiac catheterization. This should include a specific attempt to determine the level of major obstruction as well as the use of pharmacological provocative agents to detect hypertrophic subaortic stenosis masked by afterload. When the major obstruction component is hypertrophic subaortic stenosis, conservative treatment employing the use of beta-adrenergic blocking agents is probably the treatment of choice. In the absence of significant improvement, surgical treatment should then be considered. If the fixed obstruction predominates and surgical treatment is required, the surgeon should be alerted to the presence of both lesions so that consideration can be given to the use of ventriculotomy at the time of surgery. Knowledge of the coexistence of these lesions is of considerable importance in the postoperative management of patients who have undergone surgical correction of the fixed obstruction alone. The use of inotropic agents for the correction of low cardiac output in the postoperative period might be exceedingly dangerous since it will accentuate the very symptom being treated.

References
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