Aortic Stenosis in Children
Experience with Echocardiographic Prediction of Severity


SUMMARY Fifty-six children with aortic stenosis were investigated both by echocardiography and cardiac catheterization. The ratio of end-systolic wall thickness (WS) to internal diameter (LVES) across the minor axis of the LV as determined by echocardiography and multiplied by a factor of 245 predicts left ventricular pressure ($r = 0.83$). Subtracting the arm systolic blood pressure from the predicted intraventricular systolic pressure (FISP) gives a predicted systolic pressure gradient ($r = 0.91$).

The technique is found to be useful for both initial and sequential noninvasive assessment of aortic stenosis with normal LV function irrespective of the level of obstruction. It is shown to apply equally well to a wide range of LV pressures, is independent of volume load, and is useful in predicting small gradients. Some common problems encountered with the measurements are examined in detail.

NONINVASIVE PREDICTION OF LV PRESSURE has a wide application in some forms of congenital heart disease where it cannot be measured from a systemic artery, as with aortic stenosis, d-transposition of the great arteries, or coarctation of the aorta with abnormal brachial arteries due to anatomical variants or previous catheterization.

From a combination of the Laplace relationship, wall stress studies, and the concept of "relative" wall thickness, a simple formula has been developed to predict LV pressure by echocardiography. A similar study by Bennett et al. gave encouraging results for adults. This study of children with aortic stenosis was undertaken primarily to establish that the gradient that would be found at cardiac catheterization could be predicted by echocardiography.
Subjects and Methods

Fifty-six children with aortic stenosis were studied over a two year period at Hospital for Sick Children. Each child had an echocardiogram and blood pressure recorded in a resting state, prior to cardiac catheterization the following day. Excluded from the study were all infants under six months of age, those with asymmetric septal hypertrophy, and those with congestive heart failure. Thirty-six were studied in the first year by retrospective analysis of the echocardiogram without knowledge of the catheterization data. The 21 children studied in the second year all had their gradients predicted prior to cardiac catheterization.

Echocardiograms were obtained using either an Ekoline 20A or Hoffrel 101C single crystal ultrasonoscope interfaced to a Cambridge multichannel recorder. With the patient in a shallow left lateral position, a 2.25 mHz transducer was applied to the chest wall perpendicular to the mitral valve. A full sweep was obtained to display the structures of the left ventricle by angling the transducer inferolaterally to the apex, and superomedially to the outflow tract and aortic valve. Extra recordings were made at the position just below the mitral valve leaflets where epicardium, endocardium, and left septal surface were clearly seen (fig. 1). At the end of the procedure, the right brachial artery systolic blood pressure (BP) was measured with an appropriate sized cuff connected to a calibrated mercury manometer, usually with a Doppler detection system.

End-systolic wall thickness (Ws) was measured from epicardium to endocardium of the posterior wall of LV at the site inferior to the mitral valve as described. The end-systolic minor axis (LVES) was measured in the same region from endocardium to the level of the maximum posterior systolic movement of the left septal surface (fig. 2). Averages of both measurements over at least five cardiac cycles were made. The end-diastolic minor axis dimension (LVED) was measured in the same area from endocardium to left septal surface just prior to the dip of the posterior wall that precedes the systolic anterior motion (fig. 1). Systolic time intervals were taken from aortic valve leaflet opening and closure in relation to the onset of the QRS in the electrocardiogram at a paper speed of 100 mm/sec. The heart rate (HR) was measured from the accompanying ECG. Assessment of LV function by ejection fraction (EF), percentage shortening of minor axis, and velocity of circumferential fiber shortening (Vcf) was calculated from LVES, LVED, and LV ejection time (LVET). In all children the EF was greater than 60% and Vcf greater than 1.0 circumferences per second, indicating normal left ventricular function.

Cardiac catheterization was performed to assess the underlying defect. Sedation for the procedure was standardized using CM3 mixture (Demerol 25 mg, Chlorpromazine 6.25 mg, and Promethazine 6.25 mg in each 1.0 ml) in a dose of 0.1ml/kg up to a maximum of 2.0 ml. No additional sedation was required. Calibrated pressures were measured in the resting state and gradients were measured from ectopic free withdrawal traces prior to angiography. Four children had concurrent echocardiograms recorded with direct LV pressure.

The data obtained from these children are shown in table 1. The ages range from ten months to 19 years. The site of obstruction was valvular in 45%, subvalvular in six, and supravalvular in five. Nine children had moderate aortic regurgitation including one with combined aortic and mitral rheumatic valve disease. One patient with fibrous subaortic stenosis had associated moderate pulmonary valve stenosis.

Analysis was designed to explore the relationship of the ratio Ws/LVES to the LV pressure measured by cardiac catheterization, and to the prediction of the gradient by subtracting the arm systolic blood pressure. Percentage shortening and LVET were also tested against the measured gradients. The methods described by Snedecor and Cochran were used.

Results

For the 56 children, the relation of LV pressure to Ws/LVES can be correctly described by a single conversion...
factor of 245 (r = 0.83). This also remains correct for the prediction of gradients, but with a better correlation (fig. 3). Thus we derive the formula:

\[ \text{Gradient} = 245 \times (\text{LVES}/\text{LVET}) - \text{BP} \quad (r = 0.91) \]

There was no statistical difference between gradient predictions in those evaluated prospectively or retrospectively.

There is a poor correlation between gradient and percentage shortening (r = 0.31) and virtually no correlation with ejection times.

There were no discrepancies in pressure or gradient predictions in patients with valvotomy (7) or in patients with volume overload of the left ventricle (9) with moderate or severe aortic or mitral regurgitation.

Discussion

The use of the Laplace relationship enables prediction of the LV pressure during wall stress (systole) from a knowledge of the LV diameter and wall thickness at end systole.

**Figure 3. Relationship between predicted and measured gradients for aortic stenosis.**
The ratio of these components (Ws/LVES) is the same as that of an angiographic concept of "relative wall thickness," which has been previously explored and documented.7-9 Relative wall thickness is an index of the LV response to a chronic mechanical burden and readily separates volume overload from pressure overload, rises with the concentric hypertrophy of pressure overload, and falls when cardiac failure occurs. The ratio is not valid if asymmetric septal hypertrophy or cardiomyopathy is present. When pressure and volume overload co-exist, the ratio rises only in proportion to the pressure load component. This latter observation is particularly useful in clinical application, for example, when aortic stenosis is associated with aortic or mitral regurgitation.

An essential part of the technique lies in the recording and interpretation of the echocardiogram and blood pressure measurements. Perhaps the most critical part of the technique is the recording and measurement of Ws. Lack of attention to detail and standardization will introduce unwanted errors into the method and reduce its reproducibility among different observers.

It is recognized that slightly different LV dimensions can be recorded in the one patient by different transducer positions on the chest wall, and by different operators. In practice we have found that the transducer site for the most reproducible results is taken from a position perpendicular to the chest wall over the mitral valve from which an entire sweep of LV is clearly obtained. Minor axis LV dimensions are then measured over a number of cardiac cycles just inferior to the mitral valve where epicardium, endocardium, and left septal surface are clearly recorded.

From our experience with the technique, we have found it necessary to carefully define the measurement of LVES and Ws. In 22% of our patients the left septal surface echo had started to move anteriorly before the posterior LV wall had completed its maximal systolic thickening. To avoid underestimating pressures in this group it is necessary to measure LVES as the distance from the level of the maximum anterior motion of the posterior endocardium to the level of the maximum posterior motion of the left septal surface (fig. 2). This apparent asynchrony was independent of LV pressure or outflow gradient.

The most critical part of the technique is associated with the measurement of Ws. Identification of endocardium and epicardium can be difficult due to variations in appearance and artifacts in the recording. Since minor differences in the value of Ws have a marked effect in the calculation of LV pressure, careful attention must be given to the recording and interpretation of the posterior wall.

The most frequently encountered problem is the presence of strong echoes arising from chordae tendineae (fig. 4a). Their movement is similar to endocardium but is usually seen only in systole and has a reduced excursion compared to the endocardium. It is useful to follow the endocardium from mid-diastole through its early systolic posterior dip to the peak anterior motion.

Less frequently encountered is the presence of multi-layering in the posterior wall myocardium (fig. 4b). This is probably due to poor lateral resolution and provides the most difficult exercise in recognition. To resolve this problem, one must study all parts of the posterior wall recording looking for areas where there is clear endocardium with no multilayering.

Another problem is apparent separation of the two pericardial layers at end systole (fig. 4c). This may be due to a real separation or again be a problem of lateral resolution. We traditionally measure from the posterior echo when there is blurring or separation of the layers. These problems are illustrated in the echocardiogram shown in figure 5, which would be regarded as an unacceptable tracing for the prediction of LV pressure.

Measurement of Ws is of crucial importance in the calculations: a small change in the numerator of the ratio Ws/LVES can cause an important change in the ratio value derived. Minor variation in the LVES has a similar but less marked effect because of differences in their relative magnitude. Two important ramifications of this are first in the regression of the ratio Ws/LVES against LV pressure to calculate a conversion factor, and secondly in the use of this
derived factor to predict LV pressures. A simple example of this is shown in table 2 with calculations involving 0.5 mm differences. For these reasons, our own measurements were made to the nearest 0.5 mm from the echocardiographic calibration scale. While this was originally done in an attempt to improve accuracy, it should also be realized that echocardiographic resolution is no better than 1 mm at the frequencies used. Simple direct linear measurements of the two values, without references to the calibration scale, is equally valid to provide a ratio.

From the time the method was developed in our laboratory, 21 children have had their gradient estimated and recorded prior to cardiac catheterization. No difference in the reliability of the method has been found between the 36 retrospective and 21 prospective cases and attests to the validity of the method.

The measurement of blood pressure is another source of introducing error. Attention to cuff size,14 manometer calibration, and use of Doppler ultrasound are all important. Measurements in the resting state are easy to standardize by having the patient lie supine during echocardiography.

A possible source of error is the amplification of peripheral blood pressure when compared to that recorded centrally.15 This could be important where the results of predicted gradients (from echocardiography and arm blood pressure) are being compared to those measured at catheterization. Amplification varies and is greatest with shorter ejection periods, smaller pressure gradients, and in younger children. This factor could overestimate the central aortic pressure and thus underestimate the pressure gradient in aortic stenosis, particularly in the younger children with small gradients. In fact, this did not occur as there was no age effect, and an equal scatter above and below the measured gradients was present. It does not appear necessary to invoke a correction factor for this amplification effect.

Noninvasive assessment of the severity of aortic stenosis in children has been notoriously unreliable.16 Direct measurements from cardiac catheterization have remained the most reliable means of assessment and are the basis for comparison for any noninvasive method. Difficulties with catheterization reflect the effects of sedation, posture, and varying hemodynamics which are common to any method.

In relating a new method of noninvasive assessment to catheterization data, the two methods should be tested in close relationship and ideally at the same time. The comparison in this study was accomplished by performing the noninvasive assessment the day prior to cardiac catheterization with the patient at rest but not sedated. In the four cases where echocardiography was performed together with direct LV pressure measurements, the results were virtually identical to those obtained outside the catheterization laboratory.

Many noninvasive techniques have previously been applied to assess the severity of aortic stenosis in children. These include clinical examination, electrocardiography, vectorcardiography, phonocardiography, pulse contour analysis, systolic time intervals, apexcardiography18-20 and more recently a combined clinical and electrocardiographic approach.21 Published statistics with these studies show correlation coefficients no better than 0.41.

Three recent reports deserve special comment. A preliminary report from Silverman et al.22 suggested a correlation coefficient of 0.95 between gradients and ejection times corrected for heart rate. We were unable to support this with our data. Johnson et al.23 found a correlation coefficient of 0.74 comparing measured gradients to percentage shortening. This is not supported by our data, nor by the work of Dodge and Baxley24 who suggest that ejection fraction does not change significantly in compensated aortic stenosis. Glanz et al.25 have applied a similar method to ours in 13 patients, and although there are minor differences in the formula, the end results are similar.

In that study and in the work of Bennett et al.1 in adults, the conversion factors for calculating LV pressure in mmHg are quoted as 225 for both normals and volume overloaded patients. The same conversion factor has been used for patients with aortic stenosis. Our previously reported work16 describes a conversion factor of 237 for 30 children without aortic stenosis. Analysis of this current group with aortic stenosis gives a conversion factor of 245. The reason for these variations may be found in the derivation of the conversion factor and the different diagnostic groupings. The relative importance of the measurement Ws and the lesser importance of the exact conversion factor in the context of the values involved has been demonstrated in table 2.

The apparent discrepancy of correlations between LV pressures and between gradients is due to the variability of subsets of data obtained under slightly different conditions. The better correlations for gradients suggests that LV pressures are more variable than are the gradients.
is not the actual gradient. This technique of estimation is most useful for detecting the patient with a small gradient, thereby avoiding the trauma of cardiac catheterization. Our own experience shows that a predicted gradient of 35 mmHg or less was never associated with a measured gradient of more than 45 mmHg.

The overall clinical assessment of a patient with aortic stenosis should include echocardiography as an important but not the sole criterion for prediction of severity of the condition.

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References

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