Cross-sectional Echocardiographic Assessment of the Severity of Aortic Stenosis in Children

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SUMMARY Real-time, cross-sectional echocardiographic studies of the aortic valve were performed in 28 children with congenital valvular aortic stenosis and in 22 normal subjects. The presence of a stenotic valve was indicated by increase in echo production, abnormal motion pattern, and abnormal systolic position of the valve leaflets. Comparison of the maximum aortic cusp separation (MACS) to calculated aortic valve area yielded an r of 0.91. MACS was then expressed as a percentage of aortic root diameter (AOD) to correct for patient size. In normals MACS averaged 72.7% (range 63–92%) of AOD. With mild aortic stenosis MACS averaged 53.1% of AOD (range 42–62%) (P < 0.001 vs normal). With moderate and severe aortic stenosis MACS averaged 29.9% of AOD (range 20–35%) (P < 0.001 vs mild AS). Comparing the ratio MACS/AOD to peak systolic gradient yielded an r of 0.88. Further comparing this ratio to calculated aortic valve area yielded an r of 0.80. Cross-sectional echocardiography can detect the presence of aortic stenosis in children and, by comparing the ratio MACS/AOD, can provide information concerning the severity of the stenotic lesion.

USING THE STANDARD M-MODE echocardiographic technique, it is generally not possible to detect the presence or determine the severity of valvular aortic stenosis in children. Although eccentricity of the diastolic position of the aortic leaflet echo has been described with bicuspid aortic valves, severe valvular aortic stenosis may be associated with apparently normal aortic valve echo motion on the M-mode record. This occurs because doming of the valve appears to the narrow one-dimensional M-mode view as normal opening motion. Since valvular aortic stenosis is one of the more common congenital cardiac anomalies, a noninvasive technique which permitted visualization of the abnormally moving valve along with an estimation of the severity of the stenotic lesion in children would be of clinical value.

In a recent report we described the use of cross-sectional echocardiography to quantify the severity of aortic stenosis in adults. In adults direct measurement of maximum aortic cusp separation appeared to be a accurate reflection of the aortic valve orifice size and severity of stenosis. Correction for body surface area resulted in a slight statistical improvement but generally appeared unnecessary. In children some correction for individual size is required. Initial attempts to correct cusp separation for patient size in children using body surface area, however, resulted in a consistent underestimation of the severity of the stenotic lesion. The degree of underestimation appeared to increase further as patient size decreased. Therefore, this study was undertaken 1) to demonstrate the ability of cross-sectional echocardiography to determine the presence of aortic stenosis in children, 2) to establish an appropriate correction factor to relate maximum aortic cusp separation to individual size and hence severity of stenosis, and 3) to evaluate the relationship of aortic cusp separation and aortic size to body surface area to explain the apparent failure of this correction factor to relate aortic cusp separation in children to the same measurement in adults.

Materials and Methods

Real-time cross-sectional echocardiographic studies of the aortic valve were recorded in 28 consecutive children undergoing cardiac catheterization for valvular aortic stenosis. In two other cases adequate aortic valve echograms could not be obtained due to lack of patient cooperation in one case and failure of the cross-sectional probe to fit in the narrow intercostal spaces in the second. Ages in the study group ranged from 18 months to 16 years, mean 8.9 years. There were 18 children with mild aortic stenosis (peak systolic gradient less than 50 mm Hg), and 10 with moderate or severe stenosis (peak systolic gradient greater than 50 mm Hg). The latter group contained four children with gradients between 50 and 75 mm Hg (moderate) and six with gradients greater than 75 mm Hg (severe). Cardiac catheterization was performed using standard catheterization techniques. Left ventricular pressures were recorded using a fluid-filled catheter manometer system. Peak systolic aortic valve gradients were determined by continuous recording during catheter pullback from the left ventricle to the ascending aorta. Cardiac outputs were calculated in 20 patients using either the indicator dilution or Fick oxygen consumption technique. Aortic valve areas were calculated using the modified Gorlin formula with appropriate correction factor. Trace to mild aortic insufficiency was present in 13 of these patients. No patients had more than mild aortic insufficiency. In 26 of the 28 cases cardiac catheterization and echocardiographic studies were performed within 24 hours of each other. In the other two cases cross-sectional studies were performed at the time of the first follow-up visit after discharge from the hospital, or approximately two weeks after catheterization. Cross-sectional and hemodynamic studies were performed by separate observers at different hospitals. Cross-sectional echograms were interpreted without prior knowledge of the hemodynamic data. In two cases the pattern of aortic valve motion observed on cross-sectional study was not consistent with valvular aortic stenosis. At catheterization neither of these patients had obstruction at the valvular level and therefore they are not included in this study. Patients with aortic stenosis were compared with a

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group of 22 normal subjects. Normal patients were free of heart murmurs or findings suggestive of aortic stenosis either by history or physical examination. Cardiac catheterization was not performed in the normal group.

The cross-sectional echocardiographic examination was performed using a mechanical sector scanner developed in conjunction with the Fortune-Fry Research Laboratories at Indiana University. This system consisted of a modified Ekoline 20A echograph with a pulse repetition rate of 4.5 kc/sec. The scanner probe contained a 2.25 or 3.5 MHz transducer mechanically driven through a 30° sector at a variable rate from 0 to 30 cycles per second. The system was routinely operated at a frame rate of approximately 40 frames per second (20 cycles per second) which yielded a line density of approximately 120 lines per frame. The image thus produced was recorded by a standard portable television camera (GBC-CTC-6000) on ½ inch video cassettes using a Sanyo VTC-7100 cassette recorder. Cross-sectional images could be reviewed in real-time during the examination or subsequently from videotape in a real-time, slow motion or single frame presentation. Individual frames were converted to hard copy display using a video scan converter and a modified Honeywell 1856 strip chart recorder. An R wave triggered electronic counter superimposed on the upper right hand corner of each frame permitted timing of individual frames within the cardiac cycle to 1/100 of a second. Further details of this system have been previously reported.5-10

Cross-sectional echocardiographic studies were performed with the patients in a supine or slightly left lateral position. The cross-sectional probe was initially aligned with the long axis of the transducer sweep parallel to the long axis of the aorta. The aortic valve was located and placed in the center of the thirty degree scan. Figure 1 illustrates the relationship of the transducer to the aortic valve and the area encompassed by the thirty degree sector scan. From this transducer position it was possible to record the full extent of the aortic leaflets as well as the proximal portion of the aortic root. The maximum aortic cusp separation (MACS) was obtained by locating the aortic valve orifice at the apex of the domed stenotic valve and then sweeping the probe laterally across the valve orifice. The largest separation between the anterior and posterior aortic leaflet echoes was then measured. Measurement was made from the inner margin of the anterior leaflet echo to the inner margin of the posterior leaflet echo. This measurement was felt to approximate an aortic orifice diameter.

To relate aortic cusp separation to patient size and hence the severity of stenosis, we initially attempted to correct this value for body surface area. Comparing the values determined in this matter to similar values in adults resulted in a consistent underestimation of the severity of the stenosis in children. To evaluate this apparent failure of the standard correction for body surface area to adequately relate aortic cusp separation to patient size, the relationship of aortic size to body surface area was examined in 108 normal subjects. Aortic cusp separation was first corrected for body surface area and these values were then plotted against body surface area.

Figure 2 illustrates the relationship of aortic cusp separation corrected for body surface area to body surface area. From this figure it can be seen that the aortic cusp separation increases relative to body surface area as body size decreases. This is consistent with our initial experience and explains the apparent failure of body surface area to adequately correct aortic cusp separation for body size.

We then attempted to relate maximum aortic cusp separation (MACS) to body size by comparing MACS to aortic size at the level of leaflet insertion. The aorta was chosen since the aortic leaflets are intimately related to the aorta

![Diagram illustrating the relationship of the transducer probe and 30° sector scan to the aortic valve and surrounding structures. CW = chest wall; LV = left ventricle; PPM = papillary muscle; PLV = posterior left ventricular wall; S = sternum; RV = right ventricle; IVS = interventricular septum; AO = aorta; LA = left atrium.](image)

**Figure 1.**

![This figure illustrates the relationship of aortic cusp separation corrected for body surface area to body surface area in a group of 108 normal subjects. These data suggest that aortic cusp separation increases relative to body surface area as patient size decreases.](image)

**Figure 2.**
and their amplitude of opening motion is limited by the aortic wall. The aortic root at the leaflet level should be a close approximation of anulus size and therefore would represent a potential orifice if the leaflets were removed or were to lie flush against the aortic walls during systole. Since the maximum aortic cusp separation appears to be a reflection of actual orifice size, and the aortic root represents potential orifice size, it would then be possible to express the actual orifice as a percentage of the potential orifice. Both structures lie within the same scanning area which further facilitates comparison.

In this study then, the diameter of the aorta (AOD) was utilized as a reference to which aortic cusp separation was related. To avoid difficulties created by poststenotic dilatation of the aorta, or artifactual widening in cases with prominent sinuses of Valsalva, measurement of the aorta was taken at the base of the aortic valve. In cases in which the aortic valve was attached eccentrically to the aortic wall, AOD was measured by drawing a vertical line across the aortic root intersecting the base of the cephalad leaflet. Measurement of the aortic diameter was also made from the internal margin of the echoes from the anterior and posterior aortic walls at the level of the attachment of the aortic valve leaflets.

Statistical calculations were performed using the Student’s t-test and the least squares method of regression analysis.

Results

Cross-sectional echocardiographic study of the aortic valve in normal children demonstrated that the leaflets open widely during systole and assume a position parallel and in close apposition to the walls of the aorta. Figure 3 is a cross-sectional recording illustrating the systolic relationship of normal aortic leaflets to the walls of the aorta. The maximum aortic cusp separation is indicated by the shorter vertical arrow in the line drawing to the right of the figure. The longer vertical arrow indicates the diameter of the aortic root at the base of the valve. In this group the maximum aortic cusp separation represented a high percentage of the aortic root diameter, mean 72.7%, range 63 to 92% (fig. 4).

Aortic Stenosis

In children with aortic stenosis there was an increase in the echo density produced by the thickened aortic valve leaflets. In addition during systole rather than remaining parallel to the aortic root, there was an obvious curvature of one or both of the leaflet echoes toward the center of the aorta, reflecting the systolic doming of the valve (fig. 5). This inward curvature of the anterior and posterior leaflets reduced the maximum separation of the leaflet echoes at the valve orifice. Figure 6 demonstrates the relationship between the MACS, measured from the cross-sectional echogram, and the calculated aortic valve area in 20 patients with valvular aortic stenosis (r = 0.91). This measurement fails to relate MACS or calculated aortic valve area to patient size and hence to severity of stenosis. To relate MACS to patient size max-

![Figure 3](http://circ.ahajournals.org/)

**Figure 3.** Cross-sectional echogram of a normal aortic valve. This frame is recorded during systole. The aortic leaflets lie parallel and in close apposition to the walls of the aorta. The aortic valve leaflets lie at the tips of the vertical arrow. RVOT = right ventricular outflow tract; AV = aortic valve; LAX = long axis. The line drawing to the right of the figure illustrates the positions in which the measurement of aortic cusp separation and aortic diameter were taken.

![Figure 4](http://circ.ahajournals.org/)

**Figure 4.** Table illustrating the data derived by expressing maximum aortic cusp separation (MACS) as a percentage of aortic diameter (AOD) in 22 normal subjects, 18 children with mild aortic stenosis and 10 children with moderate or severe aortic stenosis. The latter group is further subdivided into those with moderate stenosis (peak systolic gradients from 50 to 75 mm Hg) indicated by the solid dots and those with severe stenosis (peak systolic gradient greater than 75 mm Hg) indicated by the open circles.
In the group with moderate and severe aortic stenosis (peak systolic gradient > 50 mm Hg) maximum aortic cusp separation represented a much smaller percentage of aortic root diameter, mean 29.9%, range 20-35%, \((P = < 0.001 \text{ vs mild aortic stenosis})\) (fig. 4, left hand column).

Further separation of this group into those with gradients of 50 to 75 mm Hg (moderate) and those with gradients greater than 75 mm Hg (severe) yielded the following results. In those with moderate aortic stenosis, maximum aortic cusp separation averaged 33.5% (range 31-35%) of aortic root diameter (closed circles, fig. 4 left). In those with severe aortic stenosis, maximum aortic cusp separation averaged 27.5% (range 20-30%) of aortic root diameter (open circles, fig. 3 left).

The direct relationship of MACS/AOD to peak systolic gradient in the 28 patients with valvular aortic stenosis is demonstrated in figure 7 \((r = 0.88)\). All patients with a peak systolic gradient greater than 60 mm Hg had a ratio of MACS/AOD of less than 35%. In patients with gradients of 50 mm Hg or less this ratio was greater than 40%.

The relationship of aortic valve area determined at cardiac catheterization to MACS/AOD is expressed in figure 8. This relationship, which compares an uncorrected with a corrected value and introduces the greatest number of measurement variables, demonstrates the poorest correlation \((r = 0.80)\) of the parameters evaluated.

![Figure 5](image1.png)

**Figure 5.** Cross-sectional echogram of a child with congenital valvular aortic stenosis. Systolic doming of the stenotic valve, increased echo production from the thickened valve leaflets, and a decrease in the maximum aortic cusp separation are apparent. The line drawing on the right indicates the area in which the maximum aortic cusp separation (shorter vertical arrow) was measured.

![Figure 6](image2.png)

**Figure 6.** Relationship between aortic valve area \((\text{cm}^2)\) determined at cardiac catheterization and maximum aortic cusp separation (MACS) \((\text{mm})\) determined from the cross-sectional echogram.

![Figure 7](image3.png)

**Figure 7.** Relationship of peak systolic aortic valve gradient \((\text{mm Hg})\) to the ratio maximum aortic cusp separation/aortic root diameter \((\text{mm})\). MACS is expressed as a percentage of AOD.
Discussion

This study demonstrates the ability of cross-sectional echocardiography to detect the presence of valvular aortic stenosis in children. The stenotic aortic valve can be differentiated from the normal valve by the increase in echo production, abnormal motion pattern, and abnormal systolic position of the stenotic leaflets. Because of the relative ease with which the echocardiographic examination can be performed in children the alterations in the normal pattern of valve motion are generally more readily demonstrated than in adults. This was true even in the smallest subject examined in this study (18 months) in whom systolic doming of the valve, the valve orifice, and cusp separation were well visualized. The absence of calcification of the aortic valve in children further facilitates the echocardiographic examination of this area.

In addition to qualitatively determining the presence of aortic stenosis it appears that more quantitative information can be derived from the cross-sectional echocardiogram. As previously noted in adults, if one can record the aortic valve orifice and measure the maximum separation between the valve leaflets at this orifice, then a relationship between this measurement and the aortic valve area can be established. As figure 6 illustrates this relationship is also present in children. In the collapsed, nondistended configuration in which the congenitally stenotic valve is usually pictured in surgical and pathological illustrations, the orifice appears slit-like and distorted. If this were a true reflection of the in vivo systolic configuration of the distended orifice, it would be difficult to obtain a transverse diameter which would accurately reflect valve area. The data presented in this study, however, indicate that the MACS is a reflection of the aortic valve area suggesting that the distended orifice does assure a relatively geometric shape. Our limited experience in recording the aortic valve orifice in a short axis presentation with the cross-sectional echocardiographic system indicates that the in vivo, distended valve orifice assumes a relatively circular configuration with the result that the long axis diameter is a reflection of orifice size and hence valve area.

Having established a relationship between calculated aortic valve area and MACS determined by cross-sectional echocardiography, it was then necessary to relate these values to the severity of stenosis. Since a given aortic valve area may be normal in one individual and represent severe aortic stenosis in another, some method of correction is necessary to relate both aortic valve area and MACS to body size. Our original observation that standard correction for body surface area appeared to underestimate the severity of the stenotic lesion, and subsequent findings that aortic size appears to increase relative to body surface area as patient size decreases, indicated a need for some other method of relating aortic cusp separation to patient size. We felt that it would be ideal if maximum aortic cusp separation could be related to a relatively fixed intracardiac structure which would reflect the size of the individual heart. The aortic root was selected because it is present in the same scan area as the aortic leaflets, is a reflection of the potential aortic orifice (if the valve were removed), and has been shown previously to be of value as a standard to which the size of other cardiac structures could be related.11 By measuring the aortic diameter at the base of the aortic valve, it was anticipated that abnormalities in the aortic diameter arising from poststenotic dilatation of the aorta or prominent sinuses of Valsalva could be avoided.

The relationship of the ratio MACS/AOD to the severity of aortic stenosis was then evaluated by comparing this ratio to peak systolic aortic valve gradient. The peak systolic aortic valve gradient will vary directly with cardiac output at any given aortic valve area. Since the cardiac output is directly related to individual size the gradient should correct itself for patient size and hence be a useful method for determining severity of stenosis. Figure 7 demonstrates the reasonably good correlation between the ratio MACS/AOD and peak systolic aortic valve gradient. Figure 4 further demonstrates the usefulness of this ratio in separating normal subjects from patients with mild or moderate to severe aortic stenosis based on the peak systolic gradient.

We have further evaluated this ratio in a larger group of 70 subjects including both adults and children. In this group the ratio MACS/AOD again proved useful in patients with normal sized aortas. In the adult group, however, particularly in elderly patients with primary disease of the ascending aorta, marked aortic dilatation may occur and thus result in overestimation of the severity of the stenosis. We therefore concluded that direct measurement of aortic orifice diameter with or without correction for body surface area is an adequate method for estimating severity of aortic stenosis in adults, while in children use of the ratio MACS/AOD is preferred.

Although only still frames are illustrated in this report, the cross-sectional system used in this study permits real-time evaluation of valve motion. As a result, it is frequently possible to determine the presence or absence of aortic stenosis and its severity by observing the pattern of leaflet motion in real-time. This technique should provide a rapid, noninvasive method for differentiating patients with mild aortic stenosis from those with functional murmurs and for defining those patients with mild aortic stenosis in whom cardiac catheterization is probably unnecessary.
There are two apparent limitations of this technique. The first relates to the frequency with which adequate cross-sectional studies can be performed, and the second to data interpretation and measurement. In this study it was possible to record diagnostic cross-sectional studies in 28 of 30 children examined (93%). In the remaining two cases failure to obtain adequate cross-sectional studies resulted from lack of patient cooperation in one and failure to make adequate contact between the oscillating transducer head and the patient's chest in the second. The first difficulty could have been overcome by sedating the child; however, it was not felt that this was appropriate for the purposes of this study. The problem of contact between the oscillating transducer head and the patient's chest in subjects with narrow intercostal spaces has recently been alleviated by the development of a new coupling material which permits these studies to be performed without direct contact between the transducer and the patient (a plastisal compound using polyvinyl chloride and a plasticizer, DO2). The compound has a consistency of firm Jello and can be produced in any size or shape required. Overall, however, the frequency of success in this study is consistent with our experience in examining patients with other forms of congenital heart disease and appears to be a realistic figure. The second difficulty rests with data interpretation and measurement. In our experience to date, there has been no difficulty or conflict in detecting the presence of aortic stenosis. In addition, the measurements of aortic cusp separation have been remarkably constant from frame to frame and from observer to observer. The measurement of aortic root diameter, however, is more difficult since the root may vary in diameter at the level of the sinus of Valsalva and in cases with poststenotic dilatation. We have attempted to standardize this measurement by determining AOD at the base of the aortic leaflets. In certain cases the leaflets may insert eccentrically again complicating this problem. We further recognize that small changes in the measurements of MACS and AOD may produce relatively significant changes in the ratio MACS/AOD. While this has not proven to be a major problem, because of these recognized difficulties we have considered this to be a semi-quantitative method.

References

7. Eggleton RC: Ultrasonic visualization of the dynamic geometry of the heart. Presented at Second World Congress on Ultrasonics in Medicine, Rotterdam, June 1973
10. Eggleton RC, Johnston KW: Real time mechanical scanning system compared with array techniques. Institute of Electrical and Electronic Engineers. Proceedings in Sonics and Ultrasonics, November 1974
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