Cross-Sectional Echocardiographic Characterization of Aortic Obstruction

1. Supravalvular Aortic Stenosis and Aortic Hypoplasia

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SUMMARY Cross-sectional echocardiographic and cineangiographic studies of the left ventricular outflow tract and ascending aorta were performed in five patients with supravalvular aortic stenosis (four hourglass and one hypoplastic). Visualization of the area of obstruction was possible in each patient using the cross-sectional system. In each case the echocardiographically determined diameter at the level of obstruction was within 3 mm of the similar angiographic value. Assessment of the extent of the lesion was possible in four of five cases. In three of these four cases the echocardiographic measurement was within 5 mm of the angiographic measurement while in the fourth the obstruction was felt to involve the total ascending aorta by both techniques. Determination of percent decrease in LVOT diameter from the aortic annulus to the level of obstruction was useful in defining obstruction and estimating severity. Cross-sectional echocardiography is a valuable noninvasive method for evaluating the ascending aorta in patients with supravalvular aortic stenosis.

SUPRAVALVULAR AORTIC STENOSIS is an obstructive congenital deformity of the aorta which originates just distal to the origin of the coronary arteries and produces either localized or diffuse narrowing of the ascending aorta. Although the designation “supravalvular aortic stenosis” encompasses a heterogeneous group of anatomic lesions three specific anatomic types have been characterized. At one end of this spectrum is the so-called membranous type, which consists of a simple fibrous diaphragm containing a single perforation. At the opposite end of the spectrum is a uniform hypoplasia of the entire ascending aorta designated the hypoplastic type. Between these two extremes is the hourglass type of deformity characterized by extreme thickening of the medial layer of the ascending aorta associated with a hourglass deformity of the external aspects of this segment of the vessel and corresponding narrowing of the aortic lumen. Over the narrowed segment there may be intimal fibrous thickening which accentuates the narrowing of the aortic lumen imparted by the medial deformity.

Historically supravalvular aortic obstruction was originally described and assigned its present designation by Mencarelli in 1930. At the time of the review of Denie and Verheugt in 1958 only 13 pathologic examples of this disorder had been reported. More recently the increased potential for surgical correction of aortic stenosis at all levels has prompted more precise characterization of the various lesions producing left ventricular outflow obstruction. This, plus the advancements in diagnostic technique, has resulted in a large number of subsequent cases of supravalvular stenosis being described.

Several reports have suggested that M-mode echocardiography may offer a relatively simple noninvasive method for detecting the supravalvular stenotic lesion. The M-mode diagnosis rests on the observation of a change in aortic diameter while scanning superiorly from the region of the sinuses of Valsalva into the ascending aorta. Attempts to characterize changes in luminal diameter of a tubular structure using the single dimensional M-mode technique, however, are difficult. In addition while it may be possible to demonstrate the decrease in aortic diameter with the M-mode recording it is not possible to characterize precisely the location of the area of obstruction or to estimate its extent. The spatial orientation provided by the cross-sectional echocardiographic technique should permit more detailed characterization of the area of obstruction in patients with supravalvular stenosis and hence augment the M-mode diagnosis. This report describes our experience in examining the ascending aorta in a group of patients with supravalvular aortic stenosis of varying types and severity.

Materials and Methods

Cross-sectional echocardiographic studies of the ascending aorta were performed in five patients with supravalvular aortic stenosis. There were four with an hourglass type of deformity and one with severe hypoplasia of the ascending aorta and aortic arch. Each of the four cases with hourglass type deformities were males while the single case with aortic hypoplasia was a female. Ages ranged from 8 to 32 years (mean 17.3 years). Specific data on the individual cases are contained in table 1. The diagnosis of supravalvular aortic stenosis was determined in each case by standard cardiac catheterization and cineangiographic techniques. Peak systolic gradient at the level of obstruction was determined by continuous recording during catheter pullback from the left ventricle into the ascending aorta. In the one patient with severe hypoplasia, apparent complete obstruction to aortic flow was present at the aortic valve level; therefore no gradient could be determined. In the other four patients peak systolic gradients ranged from 18 to 100 mm Hg (mean 67 mm Hg) (table 1).

The ascending aorta was also examined in a group of 20 normal subjects. Normals consisted of subjects with no
TABLE 1. Clinical, Cross-Sectional Echocardiographic, and Angiographic Features of Patients with Supravalvular Aortic Stenosis

<table>
<thead>
<tr>
<th>Pt/Age/Sex</th>
<th>Type</th>
<th>Gradient</th>
<th>Obst (mm)</th>
<th>Angio % decrease in diameter</th>
<th>Echo % decrease in diameter</th>
<th>Angio Length of Obst.</th>
<th>Echo Length of Obst.</th>
</tr>
</thead>
<tbody>
<tr>
<td>J.B./32/M</td>
<td>Hourglass</td>
<td>75 mm Hg</td>
<td>13 mm</td>
<td>10 mm 56.6%</td>
<td>61%</td>
<td>21 mm</td>
<td>—</td>
</tr>
<tr>
<td>J.G./13/F</td>
<td>Hourglass</td>
<td>100 mm Hg</td>
<td>14 mm</td>
<td>13 mm 36.2%</td>
<td>56.6%</td>
<td>21.5 mm</td>
<td>18 mm</td>
</tr>
<tr>
<td>A.J./9/M</td>
<td>Hourglass</td>
<td>18 mm Hg</td>
<td>13 mm</td>
<td>12 mm 13.5%</td>
<td>25%</td>
<td>15 mm</td>
<td>10 mm</td>
</tr>
<tr>
<td>J.W./16/M</td>
<td>Hourglass</td>
<td>74 mm Hg</td>
<td>17 mm</td>
<td>15 mm 50%</td>
<td>46%</td>
<td>28 mm</td>
<td>28 mm</td>
</tr>
<tr>
<td>M.B./13/F</td>
<td>Hypoplastic</td>
<td>3.4 mm</td>
<td>3 mm</td>
<td>—</td>
<td>—</td>
<td>total</td>
<td>total</td>
</tr>
</tbody>
</table>

Abbreviations: Obst. A = aortic diameter at the point of maximal obstruction by angiography; Obst. E = aortic diameter at the point of maximal obstruction by echocardiography; % decrease in diameter by angio = percent decrease in left ventricular outflow diameter from the aortic anulus to the area of peak obstruction determined from the left ventricular angiogram; % decrease in diameter by echo = percent decrease in left ventricular outflow diameter between the aortic anulus (level 1) and the areas of maximal obstruction (level 3) determined by the cross-sectional echogram; Angio length of obst. = length of the area of obstruction estimated from aortic angiogram; Echo length of obst. = echo length of obstruction estimated from the cross-sectional echogram.

Evidence of heart disease either by history or physical examination. In addition these patients had normal M-mode and cross-sectional echocardiographic examination. (In this group there were 17 males and three females, mean age 23 years, range 2-67 years).

Cross-sectional studies were performed using either a mechanical sector scanner developed in collaboration with the Fortune-Fry Research Laboratories at Indiana University Medical Center or a commercially available mechanical scanner (Smith-Kline Instruments Echo Sector I). These systems consisted of a modified Ekoline 20A echograph with a pulse repetition rate of approximately 4 kilocycles/sec and a scanner probe containing a 2.25 MHz transducer which was mechanically driven through a 30° sector at a variable rate of from 0 to 30 cycles/sec. The operating characteristics of these systems have been previously described. Cross-sectional images were recorded on videotape using a Sanyo VTC 7100 cassette recorder and were available for analysis in a real-time, slow motion, or single frame format. Individual frames were converted to hard copy using a Polaroid photographic system.

Cross-sectional studies were performed with patients in either the supine or 30° left lateral position. The cross-sectional probe was initially aligned parallel to the long axis of the aorta at the level of the aortic valve leaflets. The probe was then angled superiorly to record the distal tips of the aortic valve leaflets, the sinuses of Valsalva, and the proximal portion of the ascending aorta. Figure 1 is a diagram of the relationship of the 30° sector to the sinuses of Valsalva and proximal portion of the ascending aorta. In general, as indicated in the figure the superior border of the sinuses of Valsalva coincided roughly with the superior margin of the left atrium.

To evaluate the accuracy of the cross-sectional estimate of aortic size at the level of obstruction, the cross-sectional measurement was compared with the diameter of the aorta at the point of maximal narrowing determined from the corresponding angiographic studies. To correct for magnification angiographic measurements were compared to an aortic catheter of known size in the ascending aorta.

Detection of obstruction in patients with supravalvular stenosis depends on visualization of an obvious decrease in caliber of the vessel relative to surrounding normal areas. To determine the relationship of the aortic diameter at the level of obstruction to aortic size proximal and distal to the lesion, the diameter of the outflow tract was measured in each case at 1) aortic anulus, 2) sinuses of Valsalva, 3) the point of maximal obstruction, and 4) the aorta distal to the area of obstruction. The locations at which these measurements were taken are indicated in figure 1. In normals, where no obstruction was present, level three was measured at the superior margin of the sinuses of Valsalva and the ascending aorta (level four) at the superior limit of the scan plane. Measurements were taken from the inner aspect of the anterior root echo to the inner aspect of the posterior aortic root echo at each level. To ensure that the measurements were recorded at the mid point of the lumen, the transducer was swept from medial to lateral surface of the outflow tract and the largest diameter at each level recorded.

The echocardiographic diameter at the level of obstruction was then compared to contiguous normal areas of outflow tract to define the degree of change required to reflect
obstruction and hopefully to relate percent of change to the severity of obstruction. The wide variability in normal diameter of the sinus of Valsalva and variable decrease in diameter from the sinuses to the adjacent ascending aorta suggested that this would be an impractical level for comparison. There was a good correlation in normals between the diameter at the level of superior margin of the aortic sinuses (level 3) and the more cephalad measurement (level 4); however, the latter was the most difficult of the four areas to record and might be influenced by post-stenotic dilatation and was therefore not felt to be optimal for comparison. The aortic anulus (level 1), however, lies within or in close proximity to the scan plane, is relatively easy to record, and has a diameter similar to that of the ascending aorta and therefore appeared to be the most appropriate structure with which to compare the diameter at the level of obstruction. The diameter of the ascending aorta at the superior margin of the sinus of Valsalva or the area of greatest obstruction was therefore determined and expressed as a percent increase or percent decrease relative to the similar measure obtained at the level of the aortic anulus. Finally the length of the area of obstruction was determined from both the angiographic studies and cross-sectional echograms and the estimated extent of the lesion by these two techniques compared. Angiographic and echocardiographic measurements were determined independently by separate observers.

Results

Normal Subjects

Cross-sectional scans of the ascending aorta in each of the 20 normal subjects revealed a variable increase in diameter of the vessel just distal to its origin at the aortic leaflets. This area of dilatation corresponded to the sinuses of Valsalva and extended to a point distal to the most cephalad tips of the opened aortic leaflets. Outflow tract diameter at the anular level (level number one) averaged 21.2 mm (range 9-30 mm). This diameter increased to a mean of 26.8 mm at the level of the sinuses of Valsalva with an increase in individual diameter ranging from 2 to 10 mm. Above the sinuses of Valsalva the diameter of the vessel returned to a level similar to or slightly greater than that recorded at the aortic anulus (mean 24.4 mm, range 10-34 mm). There was a mean net increase in diameter of 3.1 mm between the anulus and the supravalvular area of the aorta (between levels one and three). In no case was there a decrease in diameter between the anulus and the proximal ascending aorta. Distal to the sinuses of Valsalva the diameter of the vessel remained relatively constant as the scan was continued cephalad. Mean aortic diameter (level four) was 24.4 mm. In only one case was there more than a 1 mm difference in aortic diameter between levels three and four. Figure 2 is a cross-sectional scan of a normal proximal ascending aorta illustrating the normal appearance of this anatomic area.

Supravalvular Aortic Stenosis

In each of the five patients with supravalvular aortic stenosis an area of obvious decrease in luminal diameter was evident at the superior border of the sinuses of Valsalva. This area of narrowing varied in both severity and extent, but in each case the appearance of the obstruction on the cross-sectional echogram was similar to its angiographic appearance. Relationship between the cross-sectional aortic diameter at the level of obstruction and the corresponding angiographic diameter is indicated in table 1. In each case the difference between these measurements was ≤3 mm, with the cross-sectional measurement being slightly less than the angiographic measurement. Estimates of the length of obstruction were possible using the cross-sectional system in three of the four patients with hourglass type lesions. In the fourth case the lesion extended beyond the plane of the cross-sectional scan. In the three cases the cross-sectional measurement was within 5 mm of the angiographic measurement (table 1). In the patient with a hypoplastic ascending aorta the obstruction was considered to involve the total ascending aorta by both techniques.

Comparison of the outflow tract diameter at the aortic anulus with the diameter at the area of obstruction in the four patients with hourglass type lesions demonstrated a mean percent decrease of 47.2% compared to a normal increase of 12.5% \( (P < 0.001) \). Although the number of patients examined is small there appeared to be a definite
relationship between percent decrease in diameter and peak systolic aortic gradient in this group of patients (table 1).

Figure 3 is a cross-sectional scan of the supravalvular portion of the aorta from a patient, A.J., demonstrating an area of aortic narrowing at the cephalad margin of the aortic sinuses followed by a gradual return of the aortic root to a normal diameter distal to the area of obstruction. Figure 4 is an angiogram from the same patient showing the similarities between the angiographic and echocardiographic appearance of this lesion. Figure 5 is a cross-sectional scan from patient, J.W., in which a more marked degree of luminal narrowing as well as more extensive involvement of the aortic root can be appreciated. Figure 6 is an angiogram from this patient again demonstrating the similarity in appearance of the areas of obstruction between these two techniques.

In the case with aortic hypoplasia the entire ascending aorta as well as the aortic anulus were of diminished size. Figure 7 is a cross-sectional scan recorded at the level of the proximal ascending aorta in which a very small, thin, strand-like aortic lumen is evident between the echoes from the anterior and posterior margins of hypoplastic vessel. The diminutive aortic size is present throughout the scan plane. As indicated in the accompanying diagram the hypoplastic aorta was associated with corresponding hypoplasia of the left ventricle and mitral valve. The left hand portion of the scan demonstrates an area of slight dilatation reflecting the sinuses of Valsalva. This is followed by an area of decreased diameter as one continues above the rudimentary sinuses with persistence of the diminutive vascular structure as the scan continues superiorly. Figure 8 is an angiogram again demonstrating the severe degree of aortic hypoplasia in this case.

Discussion

In this report we have described the cross-sectional echocardiographic features of the congenital obstructive lesion in a small group of patients with supravalvular aortic stenosis. Although supravalvular stenosis is not a common disorder it occurs with sufficient frequency to warrant serious diagnostic consideration. In studies of the types of stenosis occurring in the region of the aortic valve, supravalvular stenosis has represented between 0.6 and 6% of cases examined. Although a number of clinical features may suggest the presence of a supravalvular lesion, the precise diagnosis of supravalvular stenosis and its differentiation from valvular or subvalvular obstruction has classically required cardiac catheterization and angiography.

More recent reports have suggested that M-mode echo-
cardiography may be a useful noninvasive diagnostic method for detecting supravalvular stenosis.\textsuperscript{4,5} Unfortunately, while the M-mode technique may be valuable in this area, there are significant difficulties involved in attempting to characterize an area of narrowing of a tubular structure such as the ascending aorta using the single dimensional M-mode system. A true diameter of such a tubular vessel will be obtained only when the beam of ultrasound is perpendicular to the walls of the vessel and traverses the lumen at its mid portion. Apparent narrowing of the vessel may occur should the beam be angled eccentrically and thus fail to traverse the vessel at its largest dimension. In addition, as the beam is moved in a superior direction along the long axis of the aorta the angle at which the beam traverses the vessel will become increasingly more acute and the path of the beam from the anterior to the posterior margins of the vessel will gradually increase. This will artificially increase the vessel diameter. Thus in the study by Bolen et al.,\textsuperscript{5} the authors suggested that this phenomenon accounted for the fact that no significant difference was noted between the aortic diameter at the level of the aortic sinuses and the corresponding diameter in the more distal ascending aorta in their normal group. Since the position of the aorta in the chest may vary greatly relative to the position of the transducer, the spatial relationship of this vessel to the interrogating beam may be difficult to establish and hence the aortic diameter at a given point inaccurately measured.

The enlarged field of vision presented by the cross-sectional systems permits visualization of a greater extent of the ascending aorta. This allows both the area of obstruction and surrounding normal areas to be recorded simultaneously, hence highlighting the presence of the obstructing lesion.

In the present study the obstructing supravalvular lesion was visualized by the cross-sectional echogram in each of the five patients examined. There was a good correlation between the echocardiographically determined aortic diameter at the level of obstruction and the comparable angiographic measurement. The slight difference between these two measurements is similar to that encountered in previous studies comparing the angiographic and echocardiographic measurement of aortic root diameter\textsuperscript{15} and undoubtedly reflects the small measurement errors inherent in both of these methods.

In addition to defining the location and measuring the diameter of the lesion, it was also possible to estimate the extent of the area of obstruction in four of the five cases with the cross-sectional system. In the fifth case the superior margin of the lesion lay beneath the sternum and could not be visualized within the cross-sectional plane, preventing the full extent of the lesion from being recorded.

It was then attempted to quantitate the decrease in outflow tract size in patients with supravalvular stenosis relative to the normal variation in outflow tract diameter and to relate this value to severity of stenosis. As previously noted Bolen et al., in an M-mode study, found a significant decrease in aortic diameter from the aortic sinuses to the proximal ascending aorta only in patients with supravalvular aortic stenosis. In contrast, in our normal group, there was a consistent decrease in diameter between these levels (mean 9.8%, range 0–26%). While the mean percent

\textbf{Figure 5.} Cross-sectional echocardiographic study from patient J. W., illustrating a more marked decrease in diameter of the ascending aorta beginning just distal to the junction of the ascending aorta with the sinuses of Valsalva and producing an hourglass type of aortic narrowing. In addition to the marked decrease in aortic size the extent of the area of obstruction is also greater than that observed in figure 3.

\textbf{Figure 6.} Angiographic recording of the ascending aorta from patient J. W., illustrating the similarity in severity of obstruction and extent of the lesion (horizontal arrows) compared to the cross-sectional echogram in figure 5.
decrease of normals did not fall within their obstructive range, four of our normal cases were within this range. Although we observed a far greater percent change in diameter in the four patients with hourglass type obstruction compared to normals (44.7% vs. 9.8%), the percent decrease in obstructive patients did not appear to correlate with the severity of obstruction. This undoubtedly reflects the large individual variability in size of the aorta at the level of the sinuses. Since the direction of our data is similar to that observed by Bolen et al., the difference in absolute values undoubtedly reflects (as the authors suggested) the variation in diameter created by oblique M-mode beam angulation in this region.

Because of the variability in relationship between the sinuses of Valsalva and the ascending aorta, we examined the relationship of the aortic anular measurement (level 1) to the proximal aortic measurement (level 3). In the normal group these measurements were very similar, with the anular measurement being equal to or slightly smaller than the aortic measurement. This suggested that in the absence of anular hypoplasia, as in case M.B., the anular diameter should reflect the normal ascending aortic diameter. As anticipated in each of the other four patients with supravalvular stenosis there was a consistent decrease in diameter between the aortic anulus and the superior margin of the sinuses of Valsalva as well as apparent correlation between the percent decrease in diameter and the severity of the obstructing lesion. More data are required to define these relationships in larger groups of patients; however in this study a decrease in left ventricular outflow diameter of 25% or greater from the anulus to the proximal ascending aorta was indicative of supravalvular aortic stenosis.

All measurements in this study are based on long axis recordings of the ascending aorta. It might appear that by viewing the aorta in a short axis configuration one could directly record the cross-sectional area of the vessel both at and adjacent to the level of obstruction. Unfortunately quantitative determination of the cross-sectional area of the aorta requires the plane of the cross-sectional scan be oriented parallel to the plane of the vessel in space. Because the proximal portion of the ascending aorta generally lies superior to the position of the cross-sectional probe on the chest, the plane of the cross-sectional scan will generally be oblique to the aorta in this region. As one sweeps the transducer superiorly therefore one encounters the same problems which were previously noted with the M-mode technique, specifically that the cross-sectional area of the vessel will appear to enlarge artifactually due to increasing obliqueness of the scan plane relative to the vessel. Because the long axis permits simultaneous visualization of the vessel both proximal and distal to the area of obstruction, it is uninfluenced by these angulation artifacts and therefore appears to be a more convenient and reliable method for evaluating the ascending aorta.
Another technical concern which deserves comment is the potential for creating artifactual aortic narrowing by oblique angulation of the plane of the cross-sectional scan across the vascular lumen. This type of improper angulation may give the appearance that the aorta is "cut off," as the beam passes obliquely through the lateral margin of the vessel. This potential false-positive can be relatively easily eliminated by careful attention to aligning the probe parallel to the long axis to the vessel and adequate visualization of the lumen of the vessel both proximal and distal to the area of obstruction. It should be apparent however that should one fail to visualize the vessel distal to the area of obstruction, one could not say with absolute certainty that obstruction was, in fact, present.

In addition to visualizing supravalvular aortic obstruction and determining its extent and severity, it is also important to be able to differentiate this lesion from other types of stenosis in the area of the aortic valve as well as to detect or exclude concomitant areas of obstruction at other levels of the outflow tract. The primary lesion from which supravalvular stenosis must be differentiated is congenital valvular aortic stenosis. Previous studies from this laboratory have suggested that the enhanced spatial orientation provided by the cross-sectional method permits the domed stenotic valve to be visualized in its entirety and hence more reliable diagnosis of valvular stenosis established than was previously feasible with the M-mode systems. In addition more detailed characterization of the patterns of subvalvular obstruction has also been described using the cross-sectional systems. These studies indicate that cross-sectional echocardiography is a useful method for detecting stenosis at all levels of the left ventricular outflow tract and hence permits a more complete examination than is currently possible with other noninvasive methods.

In this group we have observed two of the three characteristic patterns of supravalvular aortic stenosis; specifically, the hourglass type and the hypoplastic type. In a review of 68 cases Patterson et al. found that 45 were of the hourglass type, while 14 were of the hypoplastic variety, and nine of the membranous type. The ratio encountered here of 4 hourglass to 1 hypoplastic type therefore is consistent with the expected distribution of these lesions. We have not as yet encountered an example of the less common membranous type of obstruction.

This report suggests that cross-sectional echocardiography offers a useful clinical method for detecting supravalvular aortic obstruction, determining the aortic diameter at the level of obstruction, and estimating the extent of the region of narrowing. In addition, by comparing the diameter of the vessel at the level of obstruction to other nonaffected areas such as the aortic anulus, it may be possible to estimate percent decrease in diameter as a reflection of severity.

References

Supravalvular aortic stenosis and aortic hypoplasia.
A E Weyman, R L Caldwell, R A Hurwitz, D A Girod, J C Dillon, H Feigenbaum and D Green

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