Accuracy of Two-Dimensional Echocardiography in the Diagnosis of Aortic Arch Obstruction

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To evaluate the predictive accuracy of two-dimensional echocardiography in the diagnosis of aortic arch obstruction, 540 consecutive patients aged 2 days to 15 years (mean 2 months) who underwent subsequent cardiac catheterization and angiography were prospectively studied. At angiography, 51 patients had aortic arch obstruction; of these, 35 had juxtaductal coarctation, 15 isthmic hypoplasia and 1 a type B interrupted aortic arch. The presence of arch obstruction was correctly identified with two-dimensional echocardiography in 45 of 51 patients with this condition (overall sensitivity 88%).

Two-dimensional echocardiography clearly defined a juxtaductal coarctation in 33 of 35 patients and isthmic hypoplasia in 13 of 15 patients (sensitivity 94% and 73%, respectively). The form and type of interrupted aortic arch were clearly distinguished from other forms and types of arch obstruction. Among the 489 patients without aortic arch obstruction, two-dimensional echocardiography wrongly diagnosed the presence of such obstruction in 9 patients (overall specificity 98%).

Forty-six (92%) of the 51 patients had at least one associated intracardiac abnormality. Twenty-two (44%) had a ventricular septal defect, 21 (42%) a bicuspid aortic valve and 4 (18%) subaortic stenosis. Five patients had complex congenital cardiac malformations. All associated abnormalities were prospectively identified with two-dimensional echocardiography. Thus, two-dimensional echocardiography is highly specific in diagnosing aortic arch obstruction. It is less sensitive for the diagnosis of isthmic hypoplasia in the neonatal period.

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Obstruction at the level of the aortic arch may vary from a localized stenosis with bandlike constriction of the aortic lumen to tubular hypoplasia over a long segment of the aortic arch. At the extreme end of this spectrum, complete obstruction may occur (1). The correct identification of the type and site of aortic arch obstruction is mandatory when surgical repair is contemplated, because differing procedures may be appropriate to each lesion. These include simple resection with end to end anastomosis, subclavian aortic anastomosis or reconstruction necessitating a tubular synthetic graft (2,3). Furthermore, each type of aortic arch obstruction may constitute one component of the hypoplastic left heart syndrome in which a number of associated malformations involving the left side of the heart coexist (4,5). Success in the surgical correction of aortic arch obstructions is therefore also dependent on the correct identification of any associated intracardiac malformations that may be present.

Recent reports (6–9) have claimed considerable success in the description of aortic arch obstruction using two-dimensional echocardiography. This approach may be further aided by the advent of conventional and color flow Doppler modalities and, thus, potentially hazardous cardiac catheterization may be avoided. However, Doppler and color flow imaging techniques, and expertise in using them, are less widely available than two-dimensional techniques, and no studies have examined the predictive accuracy of two-dimensional echocardiography alone in a large consecutive series of patients. We therefore sought, in a prospective study of a large group of consecutive patients, to assess the predictive accuracy of two-dimensional echocardiography in defining aortic arch obstruction and to assess its role in identifying associated cardiac abnormalities.

Methods

Study patients. Over a 2 year period 540 children and neonates who were referred to the cardiac ultrasound lab-
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Laboratory for two-dimensional echocardiography underwent subsequent cardiac catheterization and full angiographic definition of the aortic arch. Within this study group, 51 consecutive patients had some form of aortic arch obstruction; their mean age was 2 months (range 1 day to 15 years); 22 were female and 29 male. All 51 patients had undergone two-dimensional echocardiographic studies but 3 had died before cardiac catheterization was performed. In these three patients the results from postmortem examination were available. Five other patients died after cardiac catheterization while awaiting surgical correction. All 43 remaining patients underwent surgical repair of the narrowed segment of the aortic arch during which complete description of the arch anatomy, precise location and type of the arch obstruction and status of the ductus arteriosus were carefully recorded. Two patients died in the early postoperative period after surgical repair.

Four hundred eighty-nine patients had a variety of other congenital intracardiac abnormalities, but a normal aortic arch anatomy as angiographically defined. In four subjects who died and who underwent postmortem examination, the normality of the aortic arch was confirmed.

**Two-dimensional echocardiography.** The study was performed using a mechanical sector scanner with a 3.5, 5 or 7.5 MHz transducer (Advanced Technology Laboratory, Mark III and Mark VI) depending on the patient’s size. All studies were recorded and stored on videotape.

A complete two-dimensional echocardiographic examination using a standardized examination (8) protocol was undertaken for each patient in accordance with usual practice in our laboratory. None of the patients had been sedated for the purpose of this study. This protocol incorporates full description of aortic arch anatomy from suprasternal and upper right parasternal positions. From these transducer locations the whole aortic arch is first imaged together with head and neck vessels in the long-axis projection. Selective images of the ascending aorta, aortic arch, aortic isthmus and descending thoracic aorta are then identified (9, 10). The origins of the left carotid, left subclavian (with its characteristic sigmoid shape) and left pulmonary arteries were carefully identified.

With the acquisition of Doppler techniques in the later period of this study, 38 patients were studied by Doppler interrogation performed using a transmitted frequency of 5 or 3 MHz with a sample volume size of 2 mm. Patients were evaluated from the suprasternal notch, with the aortic isthmus being directly visualized (11). Flow was assessed in the ascending aorta and across the aortic isthmus. Pressure gradient estimations (ΔP) were not attempted across the coarctation site because the velocity (V) proximal to the stenosis is high and therefore cannot be ignored, an assumption necessary for the application of the Bernoulli equation (ΔP = 4V^2) (11).

All 540 studies were performed in a blinded fashion without reference to the clinical diagnosis in accordance with the general practice of our laboratory during the study period. The studies were undertaken by one of two operators trained in pediatric cardiology, and a report of the cardiac anatomy including that of the great vessels was issued. Each study was then reviewed in a blinded fashion by an independent third observer who issued a second report. If there was a discrepancy at this time, a repeat study was immediately performed with the specific objective of resolving the discrepancy. Thus, the final echocardiographic report was issued before the subjects’ clinical or angiographic diagnosis was known.

**Terminology.** In accordance with previous recommendation on terminology of aortic arch obstruction, juxtaductal coarctation was defined as a localized shelflike thickening of the posterolateral aortic wall opposite the ductus arteriosus (12-14). We did not define preductal or postductal coarctation as separate entities because in our experience neither angiography nor surgery can determine the junction between the walls of the duct and aortic isthmus with sufficient accuracy so as to separate this entity from juxtaductal coarctation.

Isthmic hypoplasia was defined as a tubular hypoplasia of the aortic isthmus between the left subclavian artery and ductus arteriosus (15, 16). The term segmental hypoplasia was used to describe a hypoplastic segment of the arch before the origin of left subclavian artery. Interrupted aortic arch was defined when absence of continuity between ascending and descending aorta was identified (16).

The clinical distinction between isthmic hypoplasia and juxtaductal coarctation is not possible unless the subclavian artery is involved, because both of these forms of aortic arch obstruction are characterized by a delayed or absent femoral pulse. Thus, the use of the generic term aortic coarctation was applied when referring to either form of aortic arch obstruction localized at the aortic isthmus.

**Data analysis.** After cardiac catheterization and angiography were performed and after the surgical or autopsy descriptions were available, the individual anatomic defects were compared with the echocardiographic findings. For the purpose of the study, the cardiac catheterization data were considered as the standard for the diagnosis of intracardiac abnormalities. Surgical or autopsy findings were the standard for diagnosis of the aortic arch abnormalities.

The predictive accuracy of two-dimensional echocardiography in the diagnosis of aortic arch obstruction was evaluated by a determination of sensitivity and specificity and of positive and negative accuracy (17). Sensitivity and specificity of two-dimensional echocardiography was defined as

\[
\text{Sensitivity} = \frac{\text{true positives}}{\text{true positives} + \text{false negatives}}
\]

\[
\text{Specificity} = \frac{\text{true negatives}}{\text{true negatives} + \text{false positives}}
\]
The positive and negative accuracy of two-dimensional echocardiography was defined as:

Positive accuracy = \frac{\text{true positives}}{\text{true positives} + \text{false positives}}

Negative accuracy = \frac{\text{true negatives}}{\text{true negatives} + \text{false negatives}}

The prevalence of the associated congenital malformations in isthmic hypoplasia and juxtaductal coarctation was assessed using the chi-square test.

**Results**

**Angiographic/surgical description of aortic arch obstruction.** Of the 540 patients in our study group, 51 had aortic arch obstruction; 35 of these had juxtaductal coarctation, 15 had isthmic hypoplasia and 1 patient had interruption of the aortic arch. In 11 patients, an additional hypoplastic segment of the aortic arch was present; in 8 there was involvement of the aortic arch between the left carotid and left subclavian artery, and in 2 patients the arch between the innominate and left carotid arteries was hypoplastic. One patient had diffuse hypoplasia of the aortic arch.

**Echocardiographic visualization of the aortic arch** (Table 1). In all 540 patients, visualization of the aortic arch anatomy using two-dimensional echocardiography was possible. The two-dimensional examination correctly predicted the presence of some form of aortic arch obstruction in 45 of the 51 patients (true positives studies); thus the overall sensitivity of the method was 88%. In two patients who had anatomically proved juxtaductal coarctation and in four who had isthmic hypoplasia, however, two-dimensional echocardiography failed to demonstrate any type of obstruction (false negative studies). Juxtaductal coarctation was clearly defined in 33 of 35 patients (sensitivity 94%, specificity 99%) (Fig. 1) and isthmic hypoplasia in 11 of 15 patients (sensitivity 73%, specificity 99%) (Fig. 2). The precise type of aortic arch obstruction was ill defined in three patients with juxtaductal coarctation and in two with isthmic hypoplasia. One patient with interrupted arch and 7 of the 11 patients with an additional hypoplastic segment of the arch were correctly distinguished from patients with other forms of arch obstruction.

Among the 489 patients without any form of aortic arch obstruction, two-dimensional echocardiography wrongly suggested the presence of coarctation in 9 (false positive

<table>
<thead>
<tr>
<th></th>
<th>Total Cases</th>
<th>Sensitivity (%)</th>
<th>Specificity (%)</th>
<th>Prognostic Accuracy (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Juxtaductal coarctation</td>
<td>35</td>
<td>94</td>
<td>99</td>
<td>92 (Positive) 100 (Negative)</td>
</tr>
<tr>
<td>Isthmic hypoplasia</td>
<td>15</td>
<td>73</td>
<td>99</td>
<td>68 (Positive) 99 (Negative)</td>
</tr>
<tr>
<td>Interrupted arch</td>
<td>1</td>
<td>100</td>
<td>100</td>
<td>— (Positive) — (Negative)</td>
</tr>
<tr>
<td>Total</td>
<td>51</td>
<td>88</td>
<td>98</td>
<td>83 (Positive) 99 (Negative)</td>
</tr>
</tbody>
</table>
Figure 2. High right parasternal view from a patient with isthmic hypoplasia. Note the decreased diameter of the isthmus, commencing immediately after the onset of the left subclavian artery (LSC) (arrow) and ending with the poststenotic dilation of the descending aorta (DA). Other abbreviations as in Figure 1.

studies). Six of these infants were thought to have isthmic hypoplasia and in three a juxtaductal coarctation was falsely diagnosed. The left subclavian artery had a normal origin in all subjects with a false negative or false positive study, whereas in 8 of the 35 patients with juxtaductal coarctation the left subclavian artery had a low origin, arising at the site of coarctation. In the remaining 480 patients echocardiography correctly excluded the presence of aortic arch obstruction (true negative studies) and the left subclavian artery arose normally in all these patients.

**Doppler echocardiography.** In the later part of this study period, pulsed and continuous wave Doppler studies were performed in 38 patients, 15 with subsequently proved aortic arch obstruction and 23 without such obstruction. In all 15 patients with arch obstruction, turbulent flow was obtained across the aortic isthmus, but only in 7 was the maximal flow velocity greater than 3 m/s. Forward flow throughout diastole was also noted in the majority (87%) of these patients.

In 4 of the 23 patients without arch obstruction, a mean turbulent flow of 2.4 m/s (range 2.3 to 2.7) was recorded. In these patients, coarctation was excluded by two-dimensional echocardiography but a patent ductus arteriosus was seen on angiography. In the remaining patients, flow across the isthmus was laminar with a peak velocity below 2 m/s.

**Correlation with the clinical diagnosis.** In a retrospective analysis of the clinical findings in the 51 patients with aortic arch obstruction, the clinical diagnosis was suggestive of coarctation in 42 (82%). Of the 489 patients without any form of aortic arch obstruction, a coarctation was suspected or considered in 23 before knowledge of the echocardiographic results was available (specificity 96%).

**Associated anomalies** (Table 2). Forty-seven patients (92%) had at least one associated intracardiac or extracardiac abnormality defined at cardiac catheterization or postmortem examination. The most common associated intracardiac anomaly was a ventricular septal defect, which occurred in 44% of cases. Twenty-one patients (42%) had a bicuspid aortic valve, and four of these had a peak systolic gradient >20 mm Hg measured at cardiac catheterization. Four patients had subaortic stenosis. Patients with juxtaductal coarctation had a higher incidence of aortic valvular or subvalvular abnormalities than did patients with isthmic hypoplasia. Two patients had an atrial septal defect of the secundum type, two a mitral valve anomaly and one total anomalous pulmonary veins.

<table>
<thead>
<tr>
<th>Associated Congenital Anomalies in 50 Patients With Coarctation of the Aorta</th>
<th>Juxtaductal (n = 35)</th>
<th>Preductal (n = 15)</th>
<th>Total (n = 50)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patent ductus arteriosus</td>
<td>14</td>
<td>14 (p = 0.0004)</td>
<td>28 (56%)</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>10</td>
<td>12 (p = 0.0009)</td>
<td>22 (44%)</td>
</tr>
<tr>
<td>Bicuspid aortic valve</td>
<td>20</td>
<td>1 (p = 0.0007)</td>
<td>21 (42%)</td>
</tr>
<tr>
<td>Valvular aortic stenosis</td>
<td>4</td>
<td>4 (8%)</td>
<td>8 (16%)</td>
</tr>
<tr>
<td>Subvalvular aortic stenosis</td>
<td>4</td>
<td>4 (8%)</td>
<td>8 (16%)</td>
</tr>
<tr>
<td>Abnormal mitral valve</td>
<td>2</td>
<td>2 (4%)</td>
<td>4 (8%)</td>
</tr>
<tr>
<td>Total anomalous pulmonary veins</td>
<td>1</td>
<td>1</td>
<td>2 (4%)</td>
</tr>
<tr>
<td>Atrial septal defect (secundum)</td>
<td>2</td>
<td>2 (4%)</td>
<td>4 (8%)</td>
</tr>
<tr>
<td>Complex</td>
<td>4</td>
<td>4 (8%)</td>
<td>8 (16%)</td>
</tr>
<tr>
<td>Sequential hypoplasia</td>
<td>3</td>
<td>8</td>
<td>11 (22%)</td>
</tr>
</tbody>
</table>
pulmonary venous drainage. In all cases, these abnormalities were first diagnosed by two-dimensional echocardiography and subsequently confirmed by angiographic and surgical findings.

Fourteen (93%) of the 15 patients with isthmic hypoplasia had a patent ductus arteriosus at surgery, compared with only 14 (45%) of the 35 patients with juxtaductal coarctation (p = 0.0004). Among the 28 patients (56%) with a ductus arteriosus found to be patent at surgery or autopsy, the condition was predicted by echocardiography in 20 (71%).

Two-dimensional echocardiography correctly distinguished severe obstructive lesions from a hypoplastic segment of the transverse aortic arch in 7 (64%) of the 11 patients. Three of the four patients with undetected additional segmental hypoplasia had isthmic hypoplasia. In the fourth patient, diffuse hypoplasia of the arch in association with a juxtaductal coarctation was present.

Five patients had complex intracardiac malformations, three with single ventricle and transposition of the great arteries. Two patients had hypoplastic left heart syndrome. All diagnoses were first established with two-dimensional echocardiography and were in accordance with the subsequent angiographic and surgical findings.

**Discussion**

**Anatomic definition of the arch.** Although the echocardiographic features of coarctation have been previously described (10,18,19), no prospective study of a large unselected patient group has determined the predictive accuracy of echocardiographic diagnosis. In this study we found that two-dimensional echocardiography is a more specific than sensitive method for the prediction of aortic arch obstruction. This was particularly true for the patients with isthmic hypoplasia. Therefore, the visualization of a normal aortic arch using echocardiography strongly supports the absence of any form of obstruction. When obstruction is recognized, however, problems in precise diagnosis may arise. The junction of the ductus arteriosus, as well as some physiologic narrowing of the aortic isthmus in the neonatal period, may be the source of a false positive diagnosis of isthmic obstruction. The anatomic variability of the length of the aortic isthmus extending between the left subclavian artery and the ductus arteriosus may also be a source of interpretative errors.

The presence of a low origin of the left subclavian artery may be a helpful indirect sign, often indicating the presence of juxtaductal coarctation that could otherwise be missed because of the foreshortened isthmus. We therefore suggest that, when there is a low origin of the left subclavian artery, particular attention should be directed to interrogation of the isthmic area, because this sign appears to raise the suspicion of possible obstruction.

We support the view that further investigations are unnecessary when clinical assessment is in agreement with the echocardiographic findings. When discrepancy occurs, however, cardiac catheterization should be performed.

In none of our patients did isthmic hypoplasia and juxtaductal coarctation as defined in this study coexist. The site of entry of a ductus arteriosus into the anterior aortic wall may be confused with a juxtaductal coarctation (10). Thus, in patients with juxtaductal coarctation care in the interpretation of the ductal shelf is necessary. In contrast, both juxtaductal coarctation and isthmic hypoplasia appear to be frequently associated with an additional narrowing of the aortic segment between the left common carotid and left subclavian artery (Table 2). We therefore agree with Morrow et al. (20), who suggested that segmental hypoplasia of the arch may be useful in the echocardiographic diagnosis of coarctation.

**Potential role and limitations of Doppler echocardiography.** Because our study was designed to assess the predictive accuracy of two-dimensional echocardiography alone in the diagnosis of aortic arch obstruction, Doppler ultrasound results were not fully analyzed. From our experience, however, in the later period of this study, it does appear that an increased velocity across the aortic isthmus will not always indicate coarctation because it can also be obtained in the presence of a restrictive duct. Furthermore, although Doppler analysis may help in establishing the echocardiographic diagnosis of aortic arch obstruction, it cannot be used for quantitating pressure gradients, because the Bernoulli equation (\( \Delta P = 4V^2 \)) assumes that flow velocities proximal to the stenosis are < 1 m/s. Furthermore, flow from the collateral circulation can also interfere with the flow across the coarctation particularly when continuous wave Doppler ultrasound is used, further making the estimation of pressure gradients inaccurate. These factors potentially limit the use of Doppler ultrasound in the diagnosis of coarctation syndromes and support our view that two-dimensional imaging alone of this area will remain an invaluable diagnostic method.

**Associated abnormalities.** Associated malformations were found in 92% of patients in this series compared with the approximately 72% incidence rate reported in other studies (14,21–23). Our results are much closer to the necropsy findings of Becker et al. (24) although their figures did not include bicuspid aortic valve and patent ductus before the age of 2 months.

All the intracardiac associated anomalies observed on two-dimensional echocardiography in our study were confirmed by angiography. There were no false positive or false negative findings, although our study inevitably lacked comprehensive autopsy or surgical findings of intracardiac anatomy. Our results were similar to those of other groups (20,24) in that the most common intracardiac anomalies associated with coarctation were ventricular septal defect
(44%) and bicuspid aortic valve (42%). There was a significant difference in the prevalence of these two anomalies in relation to the type of aortic arch obstruction: ventricular septal defect was more frequently associated with isthmic hypoplasia and bicuspid aortic valve with juxtaductal coarctation. This difference may suggest a different pathogenesis of the two types of aortic arch obstruction. Of the extra-cardiac-associated abnormalities, patent ductus arteriosus was successfully detected by direct imaging in 20 (71%) of 28 patients. This detection rate is similar to that reported by Gutgesell et al. (25), who stressed the difficulties of ductus detection in the presence of aortic arch abnormalities.

Clinical implications. This study shows that two-dimensional echocardiography is a highly specific technique for diagnosing the presence of aortic arch obstruction, even when the operator has not been alerted to this diagnosis. It is also reliable in distinguishing a juxtaductal coarctation from isthmic hypoplasia. Associated intracardiac abnormalities can also be predicted with great accuracy. In contrast, the sensitivity of two-dimensional echocardiography is lower than its specificity in predicting aortic arch obstruction, particularly in neonates with isthmic hypoplasia. Doppler echocardiography with color flow mapping may increase the overall sensitivity of the technique by directly visualizing turbulent flow across the stenotic portion of the aortic isthmus.

We thank Jenny Powell for secretarial assistance.

References


