Two-dimensional echocardiographic assessment of the aorta in infants and children with congenital heart disease

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ABSTRACT To determine the accuracy of two-dimensional echocardiography in the identification of congenital anomalies of the aorta, we compared two-dimensional echocardiographic with angiographic results in 261 consecutive infants and children with congenital heart disease (age 1 day to 20 years, mean 3.3 years). Two-dimensional echocardiography was performed and interpreted without knowledge of angiographic results. Complete visualization of the ascending and descending aorta and aortic arch branches was possible by two-dimensional echocardiographic examination in suprasternal, parasternal, and subcostal views of 255 patients (98%). Identification of the esophagus during swallowing aided the diagnosis of anatomic characteristics of aortic arch. One or more significant aortic arch anomalies were present on angiograms of 116 of 255 patients (46%) and were detected by two-dimensional echocardiography in 110 (sensitivity 95%, 99% specificity). Anomalies detected by two-dimensional echocardiography angiography were ascending aorta hypoplasia in four/four, truncus arteriosus three/three, right aortic arch 31/31, anomalous subclavian artery 11/16, coarctation 27/29, and patent ductus arteriosus 53/57. We conclude that two-dimensional echocardiography can be used to determine the anatomy of the aorta in most infants and children. In selected patients, two-dimensional echocardiography may eliminate the need for angiographic examination before surgery for congenital heart disease.


NONINVASIVE IMAGING of intracardiac anatomy and various forms of congenital heart disease has been possible for several years. However, diagnosis of extracardiac abnormalities has only recently been attempted. Advances in imaging technology make feasible a detailed evaluation of the aorta in newborn infants and children. We therefore examined 261 consecutive children with congenital heart disease by two-dimensional echocardiography. A segmental approach to the aorta was necessary so that it could be visualized completely and this approach was made possible by a combination of parasternal, subcostal, and suprasternal echocardiographic scans. Findings of two-dimensional echocardiography were compared with those of angiography or at autopsy in an attempt to answer the two following questions: (1) What is the sensitivity of two-dimensional echocardiography in detecting an abnormality of the aorta in infants and children with congenital heart disease? (2) What is the accuracy of two-dimensional echocardiography in diagnosing specific congenital abnormalities of the aorta?

Materials and methods

Between July 1982 and February 1983, each of 261 neonates, infants, and children underwent prospective two-dimensional echocardiographic examination of the aorta in the pediatric echocardiographic laboratory of Texas Children's Hospital, Houston. Complete examination of the ascending aorta, aortic arch branches, aortic isthms, and descending aorta was possible in 255 (98%) and in each the anatomic characteristics of the aorta were confirmed by angiography and/or at autopsy. Of the six patients who were excluded due to incomplete examination results, five were adolescents. Patients ranged in age from 1 day to 20 years (mean 3.3 years) and there were 157 male and 98 female patients. Ninety-eight patients were less than 1 year of age (42%) and 33 were less than 2 weeks (13%) old. Eighty-two
(32%) patients had undergone previous surgery, 40 intracardiac operations and 42 palliative operations not requiring cardiopulmonary bypass. Examinations were performed with Advanced Technology Laboratories equipment and 3.5, 5.0, and 7.5 MHz frequency transducers. The two-dimensional echocardiographic examinations were performed and interpreted without knowledge of the aortic anatomy.

Minor anomalies of the aorta such as mild dilatation of the ascending aorta, anomalous origin of a vertebral artery, or origin of the left carotid from the right innominate artery were not included in the analysis, although they often could be detected by two-dimensional echocardiography. Patients with postoperative defects such as recurrent or residual coarctation were evaluated without knowledge of previous surgery and were included in the analysis.

Imaging technique. Because the aorta is a complex structure, no single tomographic view images it completely. Therefore, a segmental approach was necessary. Figure 1 shows schematically the four approaches to the aorta that were part of each examination. The ascending aorta was imaged from parasternal, subcostal, and suprasternal views. The examination of the ascending aorta required scanning to the left and the right from the standard suprasternal view (figure 2, a). In this scan the descending aorta could sometimes be visualized as well. In the first 25 patients, the “sidedness” (left or right) of the aortic arch was determined by identification of the esophagus during swallowing of formula or other liquid by scanning from the left to the right side of the esophagus. We observed that the esophagus could be identified during normal swallowing without contrast and the upper thoracic vertebral bodies were also useful as markers of the midline. In the subsequent 230 patients, esophageal contrast was performed only in those patients in whom normal brachiocephalic branching could not be detected. Aortic arch branching was evaluated from suprasternal scans. With orientation of the transducer toward the right shoulder of the patient, the normally positioned right innominate artery with branching of the right subclavian artery and right carotid arteries could be seen (compare figure 1 and figure 2, b). This was also an aid in identifying the aortic arch as right or left sided. Care was taken to avoid imaging two carotid arteries and mistaking this for the innominate artery in patients in whom the origin of both carotid arteries was from the same orifice. Anomalous subclavian artery was detected by tracing the arch branching after determining the side of the arch (figure 3). The aortic isthmus was visualized from suprasternal and high parasternal scans (figure 2, c). It was necessary to obtain a view in which the main pulmonary artery, left pulmonary artery, and descending aorta were visualized simultaneously. This view was entirely out of the plane of the ascending aorta and aortic arch branches, although occasionally the left subclavian artery was visualized. From this position the region of the ductus arteriosus could be evaluated. Care was necessary to avoid confusing the normal crossing of the left pulmonary artery and descending aorta with the ductus arteriosus. The descending aorta was visualized from parasternal and subcostal views (figure 2, d) and its position was localized with respect to the spine to allow detection of a right-sided descending aorta. In patients with a right aortic arch the usual transition from a right-sided upper descending aorta to a left-sided lower descending aorta takes place at the region of the diaphragm and this could be traced in all. Note was made of the pulsatility of the descending aorta in patients suspected of having aortic obstruction. Once an abnormality of the aorta was detected it was necessary to continue to pursue a segmental approach to exclude the possibility of multiple aortic abnormalities.

Confirmation of aortic anatomy was obtained by angiography in 253 (99%) and at autopsy in eight patients (3%). The final angiographic results were compared with the echocardiographic anatomic diagnoses.

Results

Detection of aortic abnormalities. One or more abnormalities of the aorta were present in 116 of the 255 patients (46%) and were detected by two-dimensional echocardiography in 110 (95% sensitivity). Of the 139 patients with normal aortae by angiography or at autopsy, 137 were correctly recognized as normal by two-dimensional echocardiography (specificity 99%) (figure 4). There were two false-positive results, and both were cases of patent ductus arteriosus that appeared on the two-dimensional echocardiograms but which were not present on the angiograms. In addition, there were six false-negative results: two patients in whom small ductus arteriosus was missed, one patient with a small patent ductus and mild coarctation, one patient with a small patent ductus and an anomalous right subclavian artery, one patient with coarctation of the aorta, and a final patient with an isolated anomalous right subclavian artery that was not detected.

Diagnosis of specific aortic abnormalities. There were 154 abnormalities of the aorta observed in the 116 patients (table 1). There were 13 abnormalities of the
descending aorta, including four with severe hypoplasia and three with truncus arteriosus (figure 5). Aortic arch abnormalities were common and were found in 31 patients with right aortic arches (figure 3). Abnormalities of the aortic isthmus were the most common and included patent ductus arteriosus (in 57; figure 6) and coarctation (in 29; figure 7). Abnormalities of the descending aorta were rare. Those found included obstruction as the result of an aneurysm in one patient and right descending aortae below the diaphragm in four.

Two-dimensional echocardiography was highly accurate in diagnosing these defects, whether there were multiple abnormalities or they occurred in isolation, including patent ductus arteriosus in 53 of 57 (93%), right aortic arch in 31 of 31 (100%), coarctation of the aorta in 27 of 29 (92%), and anomalous subclavian artery in 11 of 16 (69%). Of the remaining 22 abnormalities, two-dimensional echocardiography correctly diagnosed all life-threatening abnormalities in newborn infants, including three cases of interrupted aortic arch (two type B and one type A) and one of aortopulmonary window. Visualization of the esophagus with or without swallowing of formula or other liquid allowed confident diagnosis of right- or left-sided aortic arch in all. This technique aided in the identification of retroesophageal aortic segments, including anomalous subclavian artery; in one patient a double aortic arch was correctly diagnosed in this way.

Discussion

Noninvasive imaging of extracardiac cardiovascular anatomy such as the aorta is more difficult than the evaluation of intracardiac anatomy. Early reports of experience with two-dimensional echocardiography showed that portions of the ascending and descending aorta could be visualized from parasternal scans. Two suprasternal views have been described that allow visualization of the aorta and results of several studies have shown that the entire aorta can be evaluated in adults. The suprasternal approach has been more
FIGURE 3. Two-dimensional echocardiographic diagnosis of right aortic arch by visualization of a left innominate artery (LIA) (a) and of a right arch with anomalous left subclavian artery (LSA) (b through d). a, Suprasternal scan obtained with transducer aimed toward the left shoulder of the patient showing mirror-image branching of a right aortic arch with visualization of the left carotid (LCA) and left subclavian arteries (LSA). b and c, Suprasternal scan of the right-sided descending aorta (DAo) and the anomalous origin of the LSA (arrow). d, Suprasternal sagittal scan of the esophagus (E) during swallowing (black arrowheads) showing the retroesophageal course of the anomalous left subclavian artery. Ao = aorta, other abbreviations as in figure 2.

Detection of Abnormal Aorta
2-D Echo Versus Angiography

<table>
<thead>
<tr>
<th>2-D ECHO</th>
<th>ANGIO</th>
</tr>
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Sensitivity: 95%
Specificity: 99%

FIGURE 4. Sensitivity and specificity of two-dimensional echocardiography for the detection of at least one abnormality of the aorta. Of the 116 patients with an abnormality of the aorta, 110 were detected (95% sensitivity). Of the 139 patients without an abnormality, 137 were recognized (99% specificity).

successful in children because of the shorter distances involved and because of improved echocardiographic penetration. Recent reports of success in imaging some of the extracardiac congenital abnormalities in children with higher frequency transducers make feasible visualization of the entire aorta in every examination of a neonate, child, or adolescent suspected of having congenital heart disease. In this study complete diagnostic examination of the aorta was possible in 98% of the newborns, infants, and children with congenital heart disease. The suprasternal examination was limited in several older patients but in only one infant. This is in contrast to reported results in adults.

It is useful to divide the aorta into four parts as follows: ascending aorta, aortic arch, aortic isthmus, and descending aorta, the latter of which consists of the upper and lower portions. The examination of the aorta should be structure oriented and not view orient-
TABLE 1
Diagnosis of congenital abnormalities of the aorta

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Sensitivity</th>
</tr>
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<tbody>
<tr>
<td>Ascending aorta</td>
<td>10/10</td>
</tr>
<tr>
<td>Hypoplasia</td>
<td>4/4</td>
</tr>
<tr>
<td>Truncus</td>
<td>3/3</td>
</tr>
<tr>
<td>Aortopulmonary window</td>
<td>1/1</td>
</tr>
<tr>
<td>Aneurysmal dilation (Marfan’s)</td>
<td>1/1</td>
</tr>
<tr>
<td>Anomalous origin of left pulmonary artery</td>
<td>1/1</td>
</tr>
<tr>
<td>Aortic arch</td>
<td>43/48</td>
</tr>
<tr>
<td>Right arch</td>
<td>31/31</td>
</tr>
<tr>
<td>Anomalous right subclavian artery</td>
<td>7/11</td>
</tr>
<tr>
<td>Anomalous left subclavian artery</td>
<td>4/5</td>
</tr>
<tr>
<td>Double arch</td>
<td>1/1</td>
</tr>
<tr>
<td>Aortic isthmus</td>
<td>83/89</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>53/57</td>
</tr>
<tr>
<td>Interruption</td>
<td>3/3</td>
</tr>
<tr>
<td>Coarctation</td>
<td>27/29</td>
</tr>
<tr>
<td>Descending aorta</td>
<td>7/7</td>
</tr>
<tr>
<td>Obstruction</td>
<td>1/1</td>
</tr>
<tr>
<td>Right descending aorta (three with situs inversus)</td>
<td>6/6</td>
</tr>
</tbody>
</table>

Ed. In other words, the examiner should trace the aorta and examine it from all possible approaches and not be limited to obtaining two or three "views" of this structure. This is true because two-dimensional echocardiography is a tomographic modality and multiple scans are necessary to examine completely such a complex structure. The plane of the ascending aorta, for example, is normally very different from that of the aortic branches and the aortic isthmus. These normal differences can be markedly exaggerated in association with congenital heart disease and therefore no single view can be used to adequately evaluate all four segments. This was well illustrated in a patient with dextrocardia and pulmonary atresia with a right aortic arch. The ascending aorta coursed to the left and the arch then passed to the right with mirror-image branching. The ascending aorta was in an entirely different plane from the descending aorta.

Ascending aorta. Abnormalities of the ascending aorta may be missed with the standard parasternal approach and we observed that parasternal and suprasternal views were complementary in visualizing this region. The major congenital aortopulmonary connections, including truncus arteriosus, aortopulmonary window, anomalous origin of pulmonary artery, and coronary fistulas may each coexist with patent ductus arteriosus. Therefore, evaluation of only one portion of the aorta or diagnosis with Doppler echocardiography alone cannot be sufficient in diagnosing congenital aortic abnormalities.

Dilation of the ascending aorta was present in many infants and children with right-to-left shunting and the size of the ascending aorta with respect to the pulmonary arteries correlated qualitatively with the severity of cyanosis. Isolated dilation of the sinuses of Valsalva was typical of Marfan’s syndrome and may be the sine qua non of this condition.

Aortic arch. Evaluation of the aortic arch by twodimensional echocardiography requires a standardized, segmental approach evaluating (1) the sidedness of the aortic arch, (2) origin of right subclavian, right
lesions such as interrupted aortic arch and hypoplasia of the aortic branches, transducer position alone may be of limited value in determining the location of the arch. Smallhorn et al.11 and Riggs et al.12 have shown that interrupted aortic arch, particularly that occurring between the left carotid and left subclavian arteries, can be accurately diagnosed. Since the conclusion of this study, we have examined a premature neonate with right aortic arch, interrupted arch, and right ductus arteriosus and the side on which the arch was located could not be confidently determined by transducer position alone.

It has been reported that suprasternal examination is hazardous in the newborn infant.6,13 Our experience has been to the contrary and a careful technique can allow suprasternal examination even in the very small premature infant.

Aortic isthmus. Correlations of suprasternal two-dimensional echocardiographic/pathologic data have

and left carotid, and left subclavian arteries, and (3) size and course of the upper descending aorta. George et al.6 performed two-dimensional echo-angiographic correlation with data from 29 neonates and infants including three with a right aortic arch, and Snyder and Silverman2 showed that correct positioning of the transducer during examination of the descending aorta should provide a clue to a right aortic arch. Because the ascending aorta may course from the left or right chest toward the left or right before passing adjacent to the trachea, an intrathoracic marker (the trachea, the esophagus, or the thoracic spine) is necessary to locate the aortic arch with confidence. Combining information concerning the pattern of the aortic arch branching (left or right innominate artery) with scans of the esophagus with and without swallowing and transducer positions, we were able to determine the sidedness of the aortic arch in all 255 patients. However, in

FIGURE 6. Patent ductus arteriosus (PDA) from suprasternal scans. Top, Wide patency in a newborn infant. Bottom, Narrow pulmonary end of the ductus arteriosus in a neonate with transposition of the great arteries. Abbreviations as in Figure 2.

FIGURE 7. Suprasternal scans in a 1-month-old infant with coarctation of the aorta and large patent ductus arteriosus who presented with severe congestive heart failure and equal pulses. The coarctation ledge (black arrow) and the widely patent ductus arteriosus (white arrows) were best visualized in the aortic isthmus scan (bottom). Abbreviations as in figure 2.
shown that the region of the aortic isthmus is almost entirely out of the plane of the ascending aorta and aortic arch. Suprasternal and high parasternal scanning in a sagittal plane to examine the aortic isthmus allowed visualization of this region in each patient in this study. Our results predicting ductus arteriosus patency demonstrated that current imaging equipment is limited in resolving ductus patency under 2 mm in diameter. A combination of 7.5 MHz imaging and Doppler echocardiography with sampling in the pulmonary end of the ductus arteriosus may be superior to imaging alone.

Careful assessment of the ductus arteriosus is important in an organized clinical approach to the noninvasive diagnosis of coarctation of the aorta. Because patency of the ductus arteriosus may mask proximal aortic isthmus obstruction (figure 7), information concerning morphologic characteristics of the isthmus, ductus patency, and size of the transverse aorta is necessary in critically ill newborn infants. The most readily recognized feature of coarctation of the aorta was the posterior ledge, which often could be visualized extending to the ductus. Coarctation in children and adolescents was more difficult to image completely due to the marked medial deviation of the aortic isthmus in these patients. The ascending aorta and aortic branches were prominent in older patients with coarctation, with a marked "hyperpulsality" of the brachiocephalic branches. High parasternal scanning in patients in the left lateral decubitus and sitting positions allowed visualization of the descending aorta in this anomaly and demonstrated an abrupt cessation of normal arterial pulsation at the isthmus in all these patients.

Descending aorta. Subcostal two-dimensional echocardiographic scans of the descending aorta have been used to detect position of the umbilical artery and abnormal situs. Abnormalities of the descending aorta were those of position or obstruction. The descending aorta should be described in two segments: the upper thoracic descending aorta and the abdominal descending aorta. It is known that a right-sided upper aorta usually occurs with a right aortic arch, but it may be present with a left aortic arch and a retroesophageal aortic course after aortic branching. Three patients in this study had upper descending aortae contralateral to the side of the arch and diagnosis of this anomaly required careful scanning and esophageal visualization. The usual right-sided upper aorta courses gradually to the left at or slightly above the level of the diaphragm so that the descending aorta in the abdomen is on the left. However, we encountered six patients with a right lower descending aorta: three with situs inversus totalis and two with situs ambiguous (right isomerism). In these latter two, we observed the phenomenon described by Elliot et al. in patients with asplenic syndrome in which the descending aorta "seeks" the inferior vena cava in the abdomen, with the aorta abruptly changing direction in the region of T12 to pass to the contralateral side. There were five patients with situs ambiguous and total anomalous hepatic venous connection (left isomerism) and in each the upper and lower aorta were positioned in the midline of the chest and abdomen. Obstruction of the descending aorta occurred in one patient with Takayasu's disease. One example of the use of two-dimensional echocardiography in patients with abdominal coarctation of the aorta has been described.

This study was designed to evaluate the capabilities of two-dimensional echocardiography in patients with congenital heart disease. For this reason, we included all patients in whom there was confirmation of the aortic anatomy by angiography or at autopsy, including those with previous palliative or corrective surgery. Based on these data, we conclude that two-dimensional echocardiography may be useful (1) in evaluation of residual ascending aortic abnormalities such as mild supravalve or aortic narrowing at the site of aortotomy and (2) in the detection of residual coarctation of the aorta. Doppler echocardiography may complement two-dimensional echocardiography in evaluating residual aortic left-to-right shunting (incompletely ligated patent ductus arteriosus or aortopulmonary window).

Two-dimensional echocardiography can be used to detect abnormalities of the aorta in children with congenital heart disease. The diagnosis of specific congenital abnormalities of the aorta can be made with a noninvasive, segmental approach. Visualization of the esophagus combined with transducer positions and arch branching allows confident diagnosis of the side of the aortic arch. Based on this experience, surgery for abnormalities of the aorta may be performed without prior catheterization in selected patients.

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Circulation. 1984;70:417-424
doi: 10.1161/01.CIR.70.3.417

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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