Prospective Diagnosis of d-Transposition of the Great Arteries in Neonates by Subxiphoid, Two-dimensional Echocardiography

Fredrick Z. Bierman, M.D. and Roberta G. Williams, M.D.

SUMMARY  Subxiphoid, two-dimensional echocardiograms (S2DE) were performed and interpreted before diagnostic cardiac catheterization on 59 neonates who weighed 1.3–6.0 kg (median 3.3 kg) and were 1–35 days old (median 4.5 days). The echocardiographic studies were successfully performed on 56 of 59 infants. Using appropriate longitudinal and transverse projections, S2DE permitted simultaneous visualization of the branch pulmonary arteries, proximal thoracic aorta and ventriculoarterial attachments. D-transposition of the great arteries was correctly predicted in all 16 infants with levocardia, situs solitus and atrioventricular concordance. In these patients, the standard left ventricular longitudinal projection demonstrated the bifurcating main pulmonary artery attached to the posterior ventricle; the transverse projection displayed the relationship of the proximal thoracic aorta to the anterior ventricle. The broad acoustical window using the subxiphoid approach in conjunction with a high-frequency focused transducer and a large active-element diameter permitted detailed imaging of intra- and supracardiac structures.

D-TRANSPOSITION of the great arteries (d-TGA) is the most common diagnosis at cardiac catheterization in the first week of life. M-mode echocardiography has been conventionally used to distinguish this lesion from other causes of cyanosis in the neonatal period. Precordial, real-time, phased-array and mechanical cross-sectional imaging of the great arteries have overcome some of the limitations of M-mode echocardiography by demonstrating the proximal great arteries and their ventricular attachments. Unlike cineangiography, however, these techniques do not simultaneously display ventriculoarterial attachments and proximal branching patterns. Tajik et al. have used the subxiphoid approach to identify the junction of the right ventricular outflow tract (RVOT) and main pulmonary artery (MPA) in older patients. The application of this technique to smaller infants using a high-frequency, focused crystal with a large active-element diameter should permit simultaneous visualization of the more superior extracardiac structures, i.e., branch pulmonary arteries and proximal thoracic aorta, and the ventriculoarterial attachments. In this study we evaluated the prospective application of subxiphoid two-dimensional echocardiography (S2DE) to the diagnosis of d-TGA in neonates.

Methods

Materials and Subjects
Subxiphoid, two-dimensional echocardiograms were performed and interpreted before diagnostic cardiac catheterization on 59 neonates with suspected congenital heart disease from December 1977 to December 1978. The infants weighed 1.3–6.0 kg (median 3.3 kg) and were 1–35 days old (median 4.5 days). All echocardiograms were performed with the patient supine using a Picker 80 cardiac imager and mechanical sector scanner. A 5-MHz focused crystal with 13 mm active-element diameter was used in all studies. The cardiac structures were displayed with a 45° sector angle, an image rate of 45 frames/sec and a variable line density. Modification of the cardiac imager permitted inversion of the sector image to view the heart in an upright position. All studies were recorded on a one-half-inch video cassette tape. Stop-frame imaging permitted still photographs on 1700 or 084 Polaroid film using a Tektronics C-50/C-70 series land pack camera.

Technique
The transducer is placed in the subxiphoid area with the plane of the echo beam parallel to the horizontal plane of the trunk; the inferior vena cava and abdominal aorta are displayed in cross section. Cranial angulation of the transducer from this position tracks the systemic vein to the right atrium. Clockwise rotation of the transducer displays all four cardiac chambers and the peripheral pulmonary arteries behind the atria (fig. 1A); additional cranial angulation toward the left midclavicle results in the standard longitudinal projection (figs. 1B and 2) that approximates a right anterior oblique (RAO) cineangiogram and displays the left ventricle (LV), left ventricular outflow tract (LVOT) and the posterior great artery, which is identified by its branching pattern (fig. 3). In normally related great arteries, the aorta arises from the LV and does not branch until the proximal transverse arch (figs. 4A and B). The MPA is seen in cross section to the left of the ascending aorta. The confluence of the right and left pulmonary arteries (RPA and LPA, respectively) is best seen with slight caudal angulation from this standard subxiphoid longitudinal projection. This angulation reveals the RPA coursing laterally behind the ascending aorta and superior vena cava.
TGA is similar which the left arteries; however, within the ascending portion of the transverse, subxiphoid, two-dimensional projection requires counterclockwise rotation from the longitudinal projection. Medial angulation of the transducer displays the right ventricular outflow tract and anterior great artery with its branches.

SVC). The LPA is seen only at its medial extreme as its posterior course rapidly excludes it from the echo plane. In d-TGA, the MPA arises from the posterior ventricle and bifurcates into its right and left branches (figs. 5A and B). This bifurcation is well seen in the standard longitudinal plane. The lateral extent to which the branch pulmonary arteries are seen in d-TGA is similar to that with normally related great arteries; however, the ascending aorta is no longer immediately anterior to the RPA. A variable cross section of the left atrial appendage may be displayed to the left of the MPA in this projection.

Identification of the anterior great artery and its ventricular attachment in normally related or d-transposed great arteries is accomplished by counterclockwise rotation of the transducer from the longitudinal projection. In normally related great arteries, as much as 90° of counterclockwise rotation from the standard longitudinal left ventricular projection is necessary to display the contiguity of the RVOT and MPA (fig. 1C). In this transverse projection, which simulates a lateral cineangiogram, the MPA continues distally as the LPA. The distal MPA continues posteriorly as the ductus arteriosus when the latter is not completely closed (fig. 6). In this situation the origin of the LPA is usually inferior to the MPA-ductal junction. In d-TGA the proximal aorta arises from the RVOT and continues into the transverse arch and descending thoracic aorta (fig. 7). In either normally related or d-transposed great arteries, the posterior semilunar root may be visualized in the transverse projection (figs. 6 and 7) behind the anterior ventricle or semilunar valve.

Cardiac Catheterization

Great-artery relationships were evaluated by biplane cineangiography. Ventriculograms were performed using conventional anteroposterior and long axial oblique projections. The great arteries were
FIGURE 3. Standard longitudinal projection. The plane of the echo beam is parallel to the long axis of the left ventricle (LV) and the ascending aorta (Asc. Ao.). The main pulmonary artery (MPA) is displayed in cross section. RA = right atrium; RV = right ventricle; LV = left ventricle; LAA = left atrial appendage. The arrow in the center indicates the septal leaflet tricuspid valve.

identified by their branching patterns. The spatial relationships were interpreted according to the principles described by Van Praagh et al.\(^5\)

Data Analysis

Subxiphoid, two-dimensional echocardiograms were interpreted by the echocardiographer before cardiac catheterization. Cineangiograms were reported by the radiologist. The plain chest x-ray, arterial blood gas and ECG were available to both examiners before S2DE and cineangiography. The great-artery relationships were classified on both studies as normal, d-TGA, or complex malposition. The latter group included patients with dextrocardia, single ventricle, or semilunar valve atresia.

Results

Subxiphoid, two-dimensional echocardiograms were performed successfully on all 59 patients. D-TGA was correctly predicted by S2DE in all 16 patients with this lesion. The other infants included 34 with normally related great arteries and nine with
complex malposition. Prospective S2DE accurately predicted the great-vessel relationships in all but one case involving complex malposition.

Sixteen patients had simple or complex d-TGA. The standard longitudinal S2DE projection demonstrated the bifurcating MPA arising from the posterior ventricle (fig. 5B). The transverse S2DE projection displayed the anterior ascending aorta arising from the RVOT and continuing distally as the transverse arch (fig. 7). The pulmonary valve was seen posterior and inferior to the anterior aortic root and related solely to the LV. The distal MPA terminated as the LPA and the proximal ductus arteriosus when the latter was present.

In infants with normally related great arteries (table 1), the longitudinal projection demonstrated the ascending aorta arising from the posterior ventricle and continuing without branching to the transverse arch (fig. 4). The continuity of the RVOT and MPA in normally related great arteries was demonstrated using the transverse subxiphoid projection. The anterosuperior pulmonary valve and MPA were related solely to the RVOT and terminated as the proximal remnant of the ductus arteriosus or LPA. In five patients with normally related great arteries (interrupted aortic arch — two, isolated patent ductus arteriosus (PDA) — three), the size and course of the PDA in the transverse projection simulated the aortic arch. The MPA–ductus–aortic segment was differentiated from the anterior, transposed aorta found in d-TGA by demonstrating the origin of the LPA in the transverse projection and the confluence of the branch pulmonary arteries with caudal angulation from the standard longitudinal projection. Pulmonary atresia with normally related great arteries was correctly identified in three patients (pulmonary atresia with or without ventricular septal defect (VSD) — two,

Table 1. Normally Related Great Arteries

<table>
<thead>
<tr>
<th>Ventricular septal defect</th>
<th>8</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patent ductus arteriosus</td>
<td>3</td>
</tr>
<tr>
<td>Total anomalous pulmonary venous return</td>
<td>3</td>
</tr>
<tr>
<td>Normal</td>
<td>3</td>
</tr>
<tr>
<td>Critical pulmonary stenosis</td>
<td>2</td>
</tr>
<tr>
<td>Critical aortic stenosis</td>
<td>2</td>
</tr>
<tr>
<td>Interrupted aortic arch</td>
<td>2</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>2</td>
</tr>
<tr>
<td>Pulmonary atresia and/or ventricular septal defect</td>
<td>2</td>
</tr>
<tr>
<td>Hypoplastic left heart</td>
<td>2</td>
</tr>
<tr>
<td>Tricuspid atresia, ventricular septal defect</td>
<td>1</td>
</tr>
<tr>
<td>Tricuspid atresia, pulmonary atresia</td>
<td>1</td>
</tr>
<tr>
<td>Complete atroventricular canal</td>
<td>1</td>
</tr>
<tr>
<td>Truncus arteriosus</td>
<td>1</td>
</tr>
<tr>
<td>Ebstein’s anomaly</td>
<td>1</td>
</tr>
</tbody>
</table>

TABLE 1. Normally Related Great Arteries

<table>
<thead>
<tr>
<th>Ventricular septal defect</th>
<th>8</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patent ductus arteriosus</td>
<td>3</td>
</tr>
<tr>
<td>Total anomalous pulmonary venous return</td>
<td>3</td>
</tr>
<tr>
<td>Normal</td>
<td>3</td>
</tr>
<tr>
<td>Critical pulmonary stenosis</td>
<td>2</td>
</tr>
<tr>
<td>Critical aortic stenosis</td>
<td>2</td>
</tr>
<tr>
<td>Interrupted aortic arch</td>
<td>2</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>2</td>
</tr>
<tr>
<td>Pulmonary atresia and/or ventricular septal defect</td>
<td>2</td>
</tr>
<tr>
<td>Hypoplastic left heart</td>
<td>2</td>
</tr>
<tr>
<td>Tricuspid atresia, ventricular septal defect</td>
<td>1</td>
</tr>
<tr>
<td>Tricuspid atresia, pulmonary atresia</td>
<td>1</td>
</tr>
<tr>
<td>Complete atroventricular canal</td>
<td>1</td>
</tr>
<tr>
<td>Truncus arteriosus</td>
<td>1</td>
</tr>
<tr>
<td>Ebstein’s anomaly</td>
<td>1</td>
</tr>
</tbody>
</table>
tricuspid atresia with pulmonary atresia — one). The confluence of the branch pulmonary arteries and lateral course of the RPA behind the ascending aorta was demonstrated with appropriate caudal angulation from the standard longitudinal projection; absence of the pulmonary valve and proximal tapering of the MPA was displayed in the transverse projection.

Malposition of the aorta, i.e., anterior displacement of the aortic valve without transposition of both great arteries associated with complex intra- and extracardiac anomalies, was correctly predicted by S2DE in nine patients (table 2). A single false-positive report of complex d-TGA with dextrocardia was made in one infant with dextrocardia, situs solitus, AV discordance and double outlet right ventricle with 1-TGA. There was no false-positive report of d-TGA, simple or complex, in any infant with levocardia and AV concordance. Absence or hypoplasia of the MPA with malposition was correctly predicted by S2DE in eight of nine patients. In one patient with single ventricle, outflow chamber and AV valve atresia, the ventricular attachment and relative size of the proximal MPA could not be determined.

Discussion

Biplane cineangiography has been the standard for detailed evaluation of the spatial orientation and contiguity of structures in neonates with congenital heart lesions. Prospective two-dimensional imaging of neonates from the precordium has been marginally successful in recognizing great-artery relationships. Houston et al. demonstrated the MPA bifurcation in only four of 10 cases with d-TGA using the transverse precordial technique. Successful identification of the great-artery relationships is contingent on localization of the semilunar valves and simultaneous visualization of the branching patterns of the proximal great arteries and ventricular attachments. In our series, prospective S2DE accurately diagnosed d-TGA in all 16 infants with this lesion. Our success reflects the scope of S2DE imaging in the neonate.

The subxiphoid position in the neonate is a flexible

**Table 2. Complex Malposition**

<table>
<thead>
<tr>
<th>Malposition Description</th>
<th>Catheterization</th>
<th>S2DE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary atresia + malposition</td>
<td>4</td>
<td>4*</td>
</tr>
<tr>
<td>Malposition + ventricular septal defect</td>
<td>1</td>
<td>1*</td>
</tr>
<tr>
<td>Single ventricle outlet chamber + malposition</td>
<td>1</td>
<td>1†</td>
</tr>
<tr>
<td>Truncus arteriosus, type IA</td>
<td>1</td>
<td>1*</td>
</tr>
<tr>
<td>L-transposition of the great arteries</td>
<td>1</td>
<td>1*</td>
</tr>
<tr>
<td>(S, L, L) Double outlet right ventricle, pulmonary stenosis, ventricular septal defect</td>
<td>1</td>
<td>0‡</td>
</tr>
</tbody>
</table>

*Malposition with hypoplastic or absent proximal main pulmonary artery (MPA) on prospective subxiphoid, two-dimensional echocardiography (S2DE).
†MPA present but not demonstrated on S2DE.
‡False-positive prospective diagnosis of complex d-transposition of the great arteries (d-TGA) in a patient with dextrocardia (S, L, L), i.e., situs solitus, atrioventricular discordance, L-TGA, and double outlet right ventricle with ventricular septal defect. Pulmonary stenosis is apparent on prospective S2DE.
acoustical window that allows unlimited cranial angulation and simultaneous visualization of the superior extracardiac structures, i.e., ascending aorta, and MPA with branch pulmonary arteries, and their ventricular attachments. The imaging of the more distant structures is enhanced by the sharpness of the echo beam using a focused, high-frequency crystal with a large active-element diameter (13 mm). The pliable epigastrium in the neonate allows complete interfacing of the active element with the skin and avoids limitations imposed by the small intercostal spaces over the precordium.

The similarities between cineangiography and S2DE are shown in figures 4, 5 and 7. The longitudinal S2DE projection corresponds to the RAO cineangiogram; the transverse view simulates the lateral cineangiogram. S2DE and cineangiography both identify the great arteries by their proximal branching pattern. In d-TGA with intact ventricular septum or systemic pressures in both ventricles, multiple angiographic studies are necessary to demonstrate the position of both great arteries and their proximal branching patterns. S2DE, however, can demonstrate these findings without contrast studies regardless of septal integrity or equalization of ventricular pressures. This capability of S2DE is illustrated by the relationship of the MPA-RPA junction to the ascending aorta in normally related and d-transposed great arteries. Cranial angulation from the four-chambered to standard longitudinal view tracks the course of the distal RPA to the MPA. In normally related great arteries, with the MPA-RPA junction to the left of the ascending aorta, portions of the RPA are obscured by the anterior aorta (fig. 4). In d-TGA, the distal MPA junction with the peripheral pulmonary arteries is shifted rightward and the proximal RPA is more completely displayed (fig. 5). Associated with this shift in the MPA, the ascending aorta is displaced toward the left. Despite its leftward position in d-TGA, the ascending aorta does not occupy the typical position of the MPA found in normally related great arteries (fig. 4).

Unlike the cineangiogram, cross-sectional imaging is limited by the orientation of structures relative to the echo plane. In normally related great arteries, the MPA may be confused with the anteriorly displaced aorta of the d-TGA if a large patent ductus arteriosus is present. Unconstricted, the caliber of the ductus arteriosus approximates that of the descending aorta. The continuity of the MPA, ductus arteriosus and descending aorta in the transverse S2DE projection in normally related great arteries might simulate the aortic-right ventricular relationship of d-TGA. Confusion is avoided, however, by recognition of the LPA as it courses posteriorly from its origin at the distal MPA. The more superior transverse arch is identified with medial angulation of the echo beam. If the ductus arteriosus in normally related great arteries is patent but not linear in its course to the descending aorta, the distal MPA will be seen to terminate as a ductal dimple.

**Conclusion**

D-TGA is accurately predicted by S2DE. Unlimited cranial angulation with appropriate longitudinal and transverse S2DE projections in infants allows direct visualization of the anterior and
posterior semilunar roots and simultaneous display of their ventricular attachments and proximal branching patterns. False-positive diagnosis of d-TGA is avoided by the use of biplane imaging in conjunction with focused, high-frequency crystals that have a large active-element diameter.

References
Prospective diagnosis of d-transposition of the great arteries in neonates by subxiphoid, two-dimensional echocardiography.
F Z Bierman and R G Williams

Circulation. 1979;60:1496-1502
doi: 10.1161/01.CIR.60.7.1496

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/60/7/1496