Identification of Congenital Malformations of the Great Arteries in Infants by Real-time Two-dimensional Echocardiography


SUMMARY
Real-time, two-dimensional echocardiography was used to identify great artery relations in 23 infants and small children, including 16 patients with angiographically documented transposition of the great arteries, tetralogy of Fallot, or pulmonary atresia. Using this technique, the heart was scanned perpendicular to its long axis at the origin of the great arteries. Great arteries cross-sectioned perpendicular to their long axes appear as circles; when sectioned longitudinally, these arteries appeared as elongated, sausage-shaped structures. In patients with normally related great arteries, a circular structure (aorta) always was positioned posterior to an elongated, sausage-shaped structure (distal right ventricular outflow tract and proximal main pulmonary artery). In transposition of the great arteries, two adjacent circular structures were observed; the anterior circle (aorta) was located to the right, left, or directly anterior to the posterior circle (pulmonary artery). In pulmonary atresia or hypoplasia, a large posterior circle (aorta) was associated with an anteriorly positioned structure that was either short and small (atretic right ventricular outflow tract) or elongated with an area of severe narrowing (hypoplastic right ventricular outflow tract). Thus, real-time two-dimensional echocardiography provides a rapid, noninvasive means of accurately identifying congenital malformations of the great arteries in infants and small children and may be a useful adjunct to cardiac catheterization in the diagnosis of cyanotic congenital heart disease.

IDENTIFICATION OF CONGENITAL MALFORMATIONS of the great arteries in cyanotic infants usually requires cardiac catheterization and angiographic studies. Such invasive investigations require a great expenditure of manpower and, more importantly, incur significant risks to the patient. Noninvasive techniques such as conventional (one-dimensional) echocardiography have been employed in the diagnosis of congenital heart disease, but complicated anatomic malformations are often difficult to identify with this technique. On the other hand, two-dimensional echocardiography would seem to afford great advantages for cardiac diagnosis in severely ill infants. Our recent experience with a real-time, two-dimensional echocardiographic system has demonstrated that it is possible to visualize the spatial relations of the great arteries in older children and in adults with cyanotic congenital heart disease. Therefore, this study was undertaken to determine whether this same two-dimensional echocardiographic system is applicable to the study of small infants and, in particular, whether it is useful in the identification of congenital malformation of the great arteries in such patients.

METHODS

Equipment
The system used in this study to obtain real-time, two-dimensional echocardiograms has been described in detail elsewhere. This system employs a custom-built mechanical sector scanner consisting of a standard ultrasound transducer (2.25 MHz, 2.15 cm in diameter), an angle indicator, and a small motor. The transducer is angled rapidly through a 30° or 45° arc at a rate of 15 cycles per second, producing 30 sectors per second (each sector containing 100 individual ultrasound data lines). The ultrasound signals are electronically processed by a commercial ultrasound receiver (Ekoline 20A) and displayed on a cathode ray tube. The echocardiographic images are permanently recorded on videotape by use of a closed-circuit television system and thereafter are available for stop-frame analysis.

Technique
Two-dimensional echocardiographic images of the great arteries were best obtained by cross-sectioning the heart perpendicular to its longitudinal axis. The specific maneuvers used to clarify great artery images have been previously described in detail. In general, the scanner was oriented initially so that the oscillatory movement of the transducer was parallel to an imaginary line connecting the left shoulder and right hip. The scanner was then tilted to a more cephalad position until the images of the great arteries were detected. If a cross-sectional image of a great artery could not be obtained with the transducer scanning a sector...
that was parallel to a line connecting the left shoulder and right hip, the scanner was reoriented so that the scanned sector was more perpendicular to the sternum. If necessary, the scanner also was moved laterally and medially in the intercostal space to clarify the image.

The image generated by the scanner is similar to what would be seen if the heart was transected at the origin of the great arteries, the heart removed, and the great arteries and atrium viewed from the region previously occupied by the cardiac apex. Satisfactory great artery images were usually obtained within 3–5 minutes, but could often be recorded as rapidly as 15 seconds.

Patients Studied

The study group included 21 infants, ranging in age from one to 24 months (average 10 months) and in weight from 2.2 to 10.8 kg (average 6.9 kg). In addition, two small children 29 months of age (average weight 11.3 kg) were also included in the study group. Thirteen patients were male and ten patients were female. The study group included eight patients with D-transposition of the great arteries, one infant with transposition of the great arteries in whom the aorta was positioned anteriorly and slightly to the left of the pulmonary artery, one infant with double outlet right ventricle (with the aorta positioned to the right of the pulmonary artery), two infants with tetralogy of Fallot, four infants with pulmonary atresia or severe hypoplasia, one patient with isolated ventricular septal defect, one infant with glycogen storage disease involving the heart (Pompe’s disease) and five infants with normal hearts. Two of the four patients classified as pulmonary atresia or severe hypoplasia had an associated large ventricular septal defect and therefore also could be considered examples of severe tetralogy of Fallot. The remaining two patients with pulmonary atresia or severe hypoplasia each had an intact ventricular septum and hypoplastic right ventricle.

Study Protocol

Twenty-five echocardiograms were recorded in 23 patients. In two patients with pulmonary atresia echocardiograms were performed prior to and after operation for relief of right ventricular outflow tract obstruction.

Adequate visualization of the great arteries was obtained during each of the echocardiographic studies in all infants. The spatial relations of the great arteries at their origin and the anatomy of the right ventricular outflow tract were analyzed by echocardiography. Echocardiographic images were considered to represent great arteries if the following criteria were fulfilled: 1) appearance as a circular or sausage-shaped echo-free area, 2) pulsation with each cardiac cycle, and 3) the presence of a linear, echo-dense area during diastole (representing the semilunar valve leaflets in apposition) that disappeared or moved during systole.

All patients with congenital heart disease had diagnostic cardiac catheterization and angiography prior to the time of echocardiographic study. Echocardiograms were interpreted without prior knowledge of the angiographically determined diagnosis. Echocardiographic and angiographic interpretations were then compared.

Results

Infants with Normally Related Great Arteries

The infants with normal hearts, tetralogy of Fallot, isolated ventricular septal defect, or glycogen storage disease demonstrated a characteristic two-dimensional echocardiographic image (fig. 1). The aorta appeared in cross-section as a circle. An elongated, sausage-shaped image was present that curved anterior to the aorta from the infant’s right to his left; this sausage-shaped echocardiographic image represented the distal part of the right ventricular outflow tract and the proximal part of the main pulmonary artery. By angling the scanner in a more cephalad position and to the infant’s left, this image occasionally appeared to extend posteriorly and bifurcate, presumably into the left and right pulmonary arteries. Figures 2 and 3 show the orientation of the scanned plane transecting normally related great arteries at their origin and demonstrate the way in which the two-dimensional echocardiographic image shown in figure 1 was obtained.

Infants with Transposition of the Great Arteries

Characteristic two-dimensional echocardiographic images were obtained in eight of the nine infants with
transposition of the great arteries and in the one patient with double outlet right ventricle (fig. 4). Two closely associated great arteries were visualized in cross-section as circles. In each patient, one great artery was located directly anterior to or slightly to the left or right of a posteriorly positioned artery. This relation of the two great arteries at their origins was confirmed by angiography in each of the patients (fig. 5). In the remaining infant with transposition of the great arteries, angiography demonstrated a large anterior aorta and a hypoplastic pulmonary artery positioned posteriorly and to the right of the aorta. The echocardiogram showed an anteriorly positioned circle (aorta); the hypoplastic pulmonary artery appeared as a small, flattened, echo-free area located posterior and slightly to the right of the aorta. This echocardiogram initially was interpreted as showing only a single great vessel; the posteriorly located echo-free space could not be unequivocally identified as a great artery at the time. However, retrospective analysis of the echocardiogram after the angiograms were studied resulted in the correct identification of the small posterior structure as the pulmonary artery.

Figures 6 and 7 show the orientation of the scanned plane as it cross-sections transposed great arteries at their origin and demonstrate the way in which the echocardiographic image shown in figure 4 was obtained.

Infants with Pulmonary Atresia or Hypoplasia

In all four patients with pulmonary atresia or severe hypoplasia, the aorta was visualized in cross-section as a circle. The echocardiographic images produced by the distal right ventricular outflow tract and proximal main pulmonary artery were of two types. In the two infants with pulmonary atresia, careful examination of the echocardiogram revealed that an extremely short and slit-like echo-free area (probably representing the atretic right ventricular outflow tract ending abruptly proximal to the pulmonary valve) was present and was
positioned anterior to the aorta; an echocardiographic image representing the pulmonary artery was not present. In the two other patients with severe hypoplasia of the right ventricular outflow tract (including one patient studied before and after a closed transventricular valvotomy), a small anteriorly positioned outflow tract appeared to end abruptly near the pulmonary valve; however, a segment of vessel, probably representing the proximal part of the main pulmonary artery, could be visualized distal to

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**Figure 3**

View of a heart with normally related great vessels. This view is similar to that shown on the right in figure 2 with the portion of the heart that includes the apex (i.e., caudal to the scanned plane) removed to provide an interior view of the heart. The triangular (about 30 degrees) unshaded area represents that part of the heart visualized by the sector scanner.

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**Figure 4**

Unretouched systolic stop-frame of a two-dimensional echocardiogram from a 15-month-old (8.6 kg) infant with transposition of the great arteries. Echocardiogram is taken with the sector-scanner oriented perpendicular to the longitudinal axis of the left ventricle at the level of the origin of the great arteries. Both great arteries appear in cross-section as circles with the aorta (Ao) positioned directly anterior to the pulmonary artery (PA). Part of the left atrium (LA) is seen posterior to the pulmonary artery.

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**Figure 5**

Lateral left ventricular angiogram from the patient shown in figure 4. The aorta (Ao) is located anterior to the pulmonary artery (PA).
the narrowed or atretic area (fig. 8). This area of narrowing did not move during the cardiac cycle and therefore did not appear to be a semilunar valve.

Discussion

The results of this study show that real-time, two-dimensional echocardiography is capable of accurately visualizing the relation of the great vessels at their origin in small infants. These observations are based on the knowledge that normally related great vessels cross each other near their origins; the pulmonary artery curves anterior to the aorta from right to left. Therefore, when the scanned plane transects the great vessels at their origins (in a plane perpendicular to the longitudinal axis of the heart) the aorta is seen in cross-section as a circle while the right ventricular outflow tract and pulmonary artery are transected longitudinally, producing an elongated, sausage-shaped image (figs. 1 and 2).

Conversely, in infants with transposition of the great arteries (or with double outlet right ventricle), the aorta and pulmonary artery do not have the spiral relation of normally related great arteries and therefore do not cross each other near their origins; rather, they leave the heart parallel to each other (figs. 5–7). Therefore, in these patients both great arteries appear echocardiographically as circles (fig. 4). It is assumed that the anteriorly positioned circle
Figure 7

Heart with transposed great arteries showing the relation of anterior aorta and posterior pulmonary artery viewed from below the semilunar valves (i.e., from the region previously occupied by the cardiac apex). The triangular (about 30 degrees) unshaded area represents that part of the heart and great arteries visualized by the sector scanner.

Figure 8

Left panel) Lateral right ventricular angiogram from a 12-month-old (7.7 kg) infant three months after closed transventricular valvotomy for pulmonary atresia. There is generalized hypoplasia of the right ventricular outflow tract and a discrete area of more severe narrowing; the ventricular septum is intact. The sector scanner was oriented along the dark line in the plane illustrated in figure 2. Middle panel) The same angiogram shown in the left panel has been re-oriented so that anatomic structures can be easily compared with the two-dimensional images obtained in the supine position and shown in the right panel. Right panel) Unretouched systolic stop-frame of a two-dimensional echocardiogram obtained at the level of the origin of the great arteries from the same infant. The sector scanner was oriented along the dark line shown in the left and middle panels (in the plane illustrated in fig. 2). The aorta (Ao), which is not visualized on the angiograms, is located posteriorly and appears as a circle. An elongated, sausage-shaped structure representing the right ventricular outflow tract (RVOT) and main pulmonary artery (MPA) curves anteriorly and to the left of the aorta. The echo-dense region within the right ventricular outflow tract probably represents the area of severe narrowing. This area was easily distinguished from a semilunar valve because it was relatively thick and did not move during the cardiac cycle.

represents the aorta and the posteriorly positioned circle represents the pulmonary artery, since this is the relation of the great vessels in all but a few patients with transposition of the great arteries. However, it should be emphasized that it is the presence of two circular echocardiographic images (indicating great vessels with parallel relation), rather than the identification of which circular image represents the pulmonary artery or aorta, that is essential to making the diagnosis of transposition of the great vessels with this technique.

This differentiation of infants with normally related great arteries from those with transposed arteries by two-dimensional echocardiography appears extremely reliable. The one infant in this study group in whom the precise diagnosis was not made initially had transposition of the great arteries and a hypoplastic pulmonary artery. While the initial interpretation of the two-dimensional echocardiogram indicated that the great arteries were not normally related, the precise diagnosis was erroneous. In this patient, only a single great artery (the aorta) was unequivocably identified at the time of the study. As a result, the initial echocardiographic diagnosis was truncus arteriosus or pulmonary atresia. Subsequent review of
this patient's angiograms, however, demonstrated that the pulmonary artery and aorta had a divergent rather than parallel orientation at their origins. Thus, the scanned plane apparently transected the hypoplastic pulmonary artery obliquely and produced a small noncircular image that was not immediately identified as a great artery.

In conclusion, two-dimensional echocardiography appears to have considerable practical application for cardiac diagnosis in pediatric cardiology. Cardiac catheterization of small, severely ill infants with congenital heart disease involves many technical problems and significant risk to the patient. Since some of the data usually obtained by cardiac catheterization and angiography can be determined more rapidly and safely by noninvasive two-dimensional echocardiography, it now may be possible to more carefully plan angiographic or hemodynamic studies in some seriously ill infants with suspected cyanotic congenital heart disease and thereby shorten the catheterization procedure.

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