Echocardiography in the Neonate and Young Infant

ROBERTA G. WILLIAMS, MD, FACC
Los Angeles, California

The neonate and young child present unique problems in echocardiographic diagnosis because of the wide spectrum of possible abnormalities and the tendency toward multiple lesions, requiring clear visualization of all great veins and arteries as well as intracardiac structures. Performance and interpretation of echocardiograms in this age group require an extensive knowledge of the pathologic anatomy of congenital heart disease. Recognition of unusual lesions is facilitated by displaying the heart in an anatomically familiar (upright) format in order to draw from experience in angiography and pathology.

The subxiphoid (subcostal) transducer position provides a flexible acoustic window for scanning the heart and great vessels in a multitude of planes, providing a general orientation for each cardiac segment. All other transducer positions are utilized to provide specific anatomic information.

Although in this age group echocardiographic visualization of intracardiac anatomy is superior to other techniques, delineation of the great vessels remains its greatest limitation. Tortuous vessels may not lie in a single plane and, therefore, cannot always be displayed throughout its length by a “slice” technique such as echocardiography. In addition, limited focal range of most high frequency transducers is a continuing impediment to imaging structures in the posterior and superior mediastinum.

Echocardiography provides a cost-effective means for identifying the neonate with life-threatening cardiovascular disease. It provides a complete and definitive anatomic diagnosis, in some cases eliminating the need for further procedures, while in others, improving the timing and performance of cardiac catheterization. Future studies should investigate the proper utilization of echocardiography as adjunct to or replacement of other techniques in the management of the young child.

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With the availability of high frequency transducers and a flexible acoustic window, echocardiographic imaging of the neonate or young infant now provides anatomic definition comparable and perhaps superior to angiography; yet, a complete and accurate diagnosis of congenital cardiac lesions remains an unfulfilled goal of echocardiography as it is generally practiced today. The major diagnostic challenge is the wide anatomic spectrum encountered in this age group. Small anatomic details such as attachment of an atrioventricular valve, the position of an atrial appendage or the presence of a small, muscular ventricular septal defect may have an important influence on management decisions. Also, the newborn’s shifting physiologic state and sometimes unstable cardiac output conceal clinical evidence that could aid in arriving at an accurate diagnosis.

Intracardiac lesions are often associated with “downstream” arterial abnormalities such as coarctation of the aorta and peripheral pulmonary stenosis. Detection of these associated abnormalities is essential to the appropriate management of the infant. Because these vessels are often narrow and tortuous, they are more difficult to image than intracardiac structures. The correct assessment of venous and arterial pathways remains the greatest technical challenge to the echocardiographer. For these reasons, although the technical aspects of imaging are easier in this age group than in older patients, the diagnostic demands are greater. To illustrate the accomplishments and difficulties of echocardiography encountered in this age group, three representative diagnostic entities will be discussed: transposition of the great arteries, persistent fetal circulation of the newborn and patent ductus arteriosus.

Diagnosis of Congenital Abnormalities

Simple Transposition of the Great Arteries

The earliest echocardiographic criteria of simple d-transposition of the great arteries relied on M-mode recognition of relative great artery positions (1). This was later refined by utilizing systolic time intervals to identify the aortic and...
pulmonary valves (2). At that time, this represented an important advance in the screening of cyanotic infants suspected of having congenital heart disease; however, the variability of great artery position and presence of pulmonary hypertension in the first days of life impaired the sensitivity of these diagnostic criteria. With the advent of two-dimensional echocardiography, visualization of the course of the ventricular outflow tract and great arteries provided additional clues to the diagnosis of conotruncal abnormalities (3), but it was not until the ventricle and great arteries were identified by their anatomic characteristics and connections directly visualized (Fig. 1) that the echocardiographic diagnosis of simple transposition acquired the accuracy of cardiac catheterization and angiography (4,5).

Currently, two-dimensional echocardiography not only can provide a sensitive and specific diagnosis of this conoventricular abnormality, but also can demonstrate the presence and nature of associated lesions such as subpulmonary stenosis, ventricular septal defect and patent ductus arteriosus (6,7). Performance of balloon atrial septostomy remains the only reason for performing cardiac catheterization in these newborn infants. When the critically ill newborn infant with simple transposition is judged too ill to be safely moved to the catheterization laboratory, balloon septostomy has been performed in the neonatal intensive care unit with visualization by echocardiography (8). This has proved so safe and effective that we may see the evolution of elective balloon septostomy utilizing echocardiography instead of fluoroscopy in the catheterization laboratory or even the abandonment of cardiac catheterization as such in the early preoperative evaluation and treatment of these patients. Certainly, an accurate precatheterization diagnosis allows balloon septostomy to be performed as the first procedure in the cardiac catheterization, permitting further diagnostic studies to be accomplished in a patient with improved arterial saturation. Further changes in the utilization of echocardiography are anticipated as the surgical management of simple transposition evolves toward the arterial switch repair.

In the 6 month old infant with simple transposition, who has no significant systolic murmur of a ventricular septal defect or subpulmonary stenosis and typical diminished left ventricular forces on electrocardiography, the combination of clinical and echocardiographic data may provide all the relevant information for preoperative evaluation. In fact, the day may come when preoperative catheterization of simple transposition is abandoned altogether and only postoperative hemodynamic evaluation and electrophysiologic studies will require catheterization. This will evolve only as each laboratory develops confidence in the complete diagnosis of this lesion through prospective analysis of the sensitivity and specificity of their own findings.

Persistent Fetal Circulation

Persistent fetal circulation, also known as primary pulmonary artery hypertension of the newborn, is a diagnosis of exclusion as well as a positive diagnosis of altered hemodynamics (9,10). This condition must be distinguished from simple transposition and total anomalous pulmonary venous return with obstruction. Once structural heart disease has been excluded, the echogram provides useful hemodynamic information in the management of the infant with persistent fetal circulation. Contrast injection into a systemic vein provides evidence of right to left shunting at the level of the atrial septum and patent ductus arteriosus (11,12). Systolic time intervals derived from M-mode recordings indi-

Figure 1. Left, The aorta arising from the right ventricular outflow tract (RVOT). Right, The main pulmonary artery (MPA) and branches arising from the left ventricle (LV) in a patient with simple transposition of the great arteries. Both views were obtained from a subxiphoid (subcostal) transducer position. The morphologic characteristics of the right and left ventricles were determined from other views. A = anterior; Asc. Aorta = ascending aorta; I = inferior; IVS = interventricular septum; LPA = left pulmonary artery; P = posterior; PDA = patent ductus arteriosus; P.Va = pulmonary valve; S = superior.
categorize relevant pulmonary and systemic pressure or resistance and ventricular function (2,13). In addition, ventricular dysfunction can be assessed by measurement of wall motion (14). Doppler echocardiography may be used to detect and roughly quantify tricuspid regurgitation. This information provides some indication of the ultimate prognosis as well as baseline value against which the success of therapeutic intervention can be evaluated (15).

**Total Anomalous Pulmonary Venous Return**

The echocardiographic diagnosis of total anomalous pulmonary venous return is limited by the difficulty in visualizing pulmonary veins. Normal pulmonary veins may be seen only partially from the parasternal approach. The entire course of the veins may be traced from apical and subxiphoid (subcostal) approaches (Fig. 2 to 4), but the focal length of the most commonly used high frequency transducers is not adequate to clearly display these vessels lying 4 to 9 cm from the transducer face. With loss of lateral resolution, the lumen of structures lying close to one another, such as pulmonary veins and the left atrium, may appear to coalesce, even when there is no communication. In general, most transducers designed for use in smaller children do not have adequate lateral resolution at the depths required to image normal pulmonary veins.

**Figure 2.** Left, Normal atrial anatomy from a subxiphoid (subcostal) four chamber view. The right superior pulmonary vein (RPV) is seen entering the left atrium (LA). Right, A similar view in a patient with total anomalous pulmonary venous return. The pulmonary venous confluence (VC) lies behind the left atrium, but does not connect with it. The septum primum ovale bulges from the right atrium (RA) to the left atrium.

**Figure 3.** The pulmonary venous drainage in total anomalous pulmonary venous return of the supracardiac type is displayed from the suprasternal notch approach. The venous confluence (VC) between right and left pulmonary veins drains through a vertical vein (VV) to the horizontal vein (HV) and subsequently into the superior vena cava. The connection between the vertical and horizontal vein is obliterated as it passes around the left mainstream bronchus (white area at junction of the vertical and horizontal veins).

**Figure 4.** Parasagittal subxiphoid view in a patient with total anomalous pulmonary venous return to the coronary sinus (CS). The pulmonary veins appear as the "tail of the whale" with the coronary sinus as its body. The coronary sinus is located posterior and to the left of the right atrium (RA).
structures within the posterior and superior mediastinum. Any difficulties in imaging normally draining pulmonary veins are enormously multiplied in attempts to image anomalously draining pulmonary veins coursing by means of narrow and often tortuous connections through the posterior and superior mediastinum to the right side of the heart.

**Patent Ductus Arteriosus**

Echocardiography has become widely used in newborn nurseries for the evaluation of patent ductus arteriosus in preterm infants. The simplest measurement of left atrial size by M-mode recording may indicate the presence of a significant left to right shunt which, in the presence of typical clinical course and findings, indicates a patent ductus arteriosus. Unfortunately, variables such as ventricular failure, decompression of the left atrium through a patent foramen ovale or atrial septal defect and atrial shape, cause considerable overlap in left atrial/aortic measurements of these patients and the "normal" group of preterm infants without evidence for patent ductus arteriosus.

Contrast material injected into an umbilical artery will backfill the aorta to the level of a patent ductus arteriosus and appear in the pulmonary artery (11). This can be detected by M-mode or two-dimensional display of the main pulmonary artery or proximal branches from a variety of transducer positions. Doppler echocardiography can also be used to detect turbulence in the pulmonary artery from a left to right ductal shunt. The ductus can be directly visualized with two-dimensional echocardiography from a high precordial, suprasternal notch or subxiphoid (subcostal) view. In general, the ductus of a premature infant is wide and short, facilitating imaging from any position (12). In contrast, the small ductus of an older infant or young child may be longer and more tortuous, not lying in a single plane and, thus, cannot be displayed from any single transducer position. In such cases, portions of the ductal lumen may be clearly seen, but patency throughout its course cannot be ascertained except by Doppler echocardiography.

The signs of shunting through a patent ductus arteriosus in a preterm infant may wax and wane with changing vascular resistance and ductal size. There is no standard measure of the sensitivity and specificity of these findings. In addition, clinical definitions and management vary widely from one institution to another. Therefore, the accuracy of the complete echocardiographic approach can only be assessed by large scale comparisons of clinical course after therapeutic intervention.

**Technical Problems**

General topics relating to the specific technical and conceptual problems encountered in echocardiographic imaging in the neonate and young child need to be discussed.

**Transducer Position**

Because even optimal angulation is defeated by poor lateral resolution, echocardiographers in different laboratories have tended to use transducer positions that place the area of interest in the optimal focal range of their particular transducers. In photographic terms, each laboratory has developed a "transducer priority" rather than a "position priority" system. When an unlimited selection of focal ranges is available, one quickly finds that all transducer positions make a special contribution to imaging different areas of the heart and posterior and superior mediastinum. Unfortunately, this is an impractical luxury because of increasing transducer complexity and cost. However improvements in transducer technology have provided extended focal range, allowing a single transducer to be utilized in a more flexible fashion.

The subxiphoid (subcostal) approach deserves special mention in the discussion of cardiac imaging of the newborn and young infant because of the unique applications of the approach to multiple plane imaging of the complex disorders frequently found in this age group. In the infant, the liver is prominent and the abdominal wall is soft, easily accommodating even a large transducer head in multiple positions and attitudes from any point immediately beneath the xiphoid process to the extreme right and left subcostal margins. In such patients in whom cardiac chamber position is extremely variable, the ability to sweep the transducer in a variety of positions and directions is of utmost importance. Moreover, with a transducer providing tight lateral focus at a range of 3 to 12 cm, the posterior and superior mediastinum can be displayed from the same subxiphoid plane that displays the intracardiac structures. It is the simultaneous display of the heart and great vessels within the range of optimal lateral resolution that has led to the success of this approach for diagnosis of lesions such as conotruncal abnormalities and anomalies of pulmonary venous return.

Other transducer positions have special limitations in this young age group. Parasternal imaging places the heart 1 to 5 cm from the transducer face. In the sector format, scan lines converge in this area, limiting lateral resolution. This undesirable effect is not present with linear array format. Apical imaging is useful in small infants, but sweeping the transducer is inhibited by the small radius of curvature of the infant chest wall. The cachectic infant with depressions between narrowly spaced ribs presents further problems with good coaptation between the transducer and skin surfaces from a precordial approach.

The subxiphoid (subcostal) approach has developed into the single most useful transducer position in small infants because it offers a wide, soft and flexible acoustic window and because the other transducer positions may be compromised in this patient group. To use this approach to its best advantage, long-focused high frequency transducers are re-
required. Unfortunately, many laboratories are equipped only with short-focused transducers because of the natural but unfortunate assumption that the area of interest will necessarily be in the near field in small children. This has significantly limited the utilization of this approach.

**Transducer Design**

Imaging of the young patient from the precordial and suprasternal notch positions is facilitated by a small transducer head. The subxiphoid approach can accomodate a relatively large transducer head, even in small infants, but scanning is limited if the transducer head has an angled configuration. Use of a wide active element diameter crystal provides an extended range of focus which allows clear imaging of the posterior and superior mediastinum.

Until now, phased array transducers could not match the resolution of mechanical systems in imaging small children. Recently, 5 MHz phased array transducers have become available, representing a substantial gain in axial resolution over older systems with 2.25 and 3.5 MHz arrays. These transducers are dynamically focused in one lateral dimension, but acoustically focused in the other, giving a fixed focal length in the off-axis (azimuthal) diameter. This results in an oval rather than circular beam cross section at any depth other than the exact focal point. This phenomenon is far more disconcerting in young children than in older and larger patients, but is has not been determined whether this results in significant loss of diagnostic capability. With continued improvements in transducer design, allowing increasingly longer range of acoustic (off-axis) focus in phased array systems, this should become a less significant problem.

**General Features of Imaging the Newborn and Young Child**

Pediatric echocardiography developed somewhat later than echocardiography in the adult with acquired heart disease and evolved along similar technical lines. Specific and tightly regimented positioning of the transducer was encouraged by pioneers in the field to foster consistency of M-mode and two-dimensional echocardiography. Internal landmarks became more important than external chest wall landmarks. In general, the use of rigidly proscribed transducer positioning and sweeping is perfectly adequate to describe solitary lesions such as mitral or aortic valve abnormalities; however, it is possible to describe every feature of a complex congenital lesion from all “standard” views without ever establishing a specific anatomic diagnosis. With the multitude of transducer positions and angles, it is now possible to examine the heart and great vessels segment by segment rather than view by view. This places the emphasis on cardiac anatomy rather than transducer position, causing some changes in examination technique. Instead of proceeding from one transducer position to the other, according to historical precedent, the most flexible transducer position for scanning should be used first to provide a general orientation of the cardiac structures.

In the newborn infant, this is usually the subxiphoid (subcostal) position. Subsequently, all available transducer positions should be utilized until each cardiac structure and great vessel has been completely analyzed. Although this seems like a small technical point, it is truly central to the accurate diagnosis of complex heart disease in the young child. A broad anatomic spectrum of cardiac abnormalities with multiple components requires a flexible and anatomically oriented imaging approach for the most complete diagnosis.

Another caveat to accurate diagnosis in this age group is the use of assumptions to determine chamber or great artery connections. The aorta in simple transposition is not always to the right. In some cases, the infundibulum does not lie above the body of the right ventricle. Direct visualization is necessary to avoid the treacherous exceptions that plague every rule of congenital heart disease. The difference between an 85 and 97% diagnostic specificity is often caused by the use of erroneous assumptions to compensate for marginal visualization.

The echocardiographer examining the young patient with suspected heart disease must be thoroughly familiar with pathologic anatomy of congenital lesions. With knowledge of the usual associated abnormalities, one can make sense of a complex jumble of abnormal findings. With great regularity, anatomic variations are encountered that were not previously described by echocardiography. In such cases, one must draw on information learned from angiography or pathology. This emphasizes the value of imaging the heart in an anatomically familiar (upright) presentation. When the heart is viewed upright, it is possible to recognize unfamiliar structures by virtue of a general knowledge of anatomy and pediatric cardiology.

Improvements in transducer design, more flexible positioning and sweeping of the transducer, a more anatomic approach and a familiar format bring echocardiography to the level of, and in some areas exceeding, angiography as an accurate diagnostic tool for the diagnosis of congenital heart disease in the newborn and young infant.

**Echocardiography Versus Angiography**

Because a two-dimensional echographic beam displays a thin “slice” of tissue, it is ideally suited for the examination of a globular structure such as the heart. Different sections of the heart can be imaged separately, displaying chambers and valves in different aspects. Angiography, a projection technique, reduces a three-dimensional subject to a single plane. One overlapping structure may obscure
the other. In addition, only one or two attempts can be made at positioning the patient during angiographic studies, whereas the echocardiographer has the luxury of trial and error until the correct angulation is achieved. For these and other reasons, echocardiography is superior to angiography in the display of details such as atrioventricular valve morphology and ventricular septal defect position. In contrast, a "slice" approach is not very good for examining a long and tortuous vessel not lying in a single plane. A projection technique such as angiography excels in this regard. Therefore, echocardiography is generally superior for resolving fine details of intracardiac anatomy, whereas angiography is superior for displaying details of the great vessels. This relation will probably hold true even with further development of digital subtraction angiography. However, the use of dynamic spatial reconstruction might narrow the differences between these techniques.

Present Usage

Availability of echocardiography in a primary or secondary care nursery has provided a more effective and efficient triage of infants to a referral center. Although, the echocardiographers in this setting might not be skilled in the diagnosis of complex congenital heart disease, the studies may still be helpful in determining if the heart is grossly normal or abnormal. Unsuspected diagnoses such as total anomalous pulmonary venous return may go undetected. However, in thickly populated urban areas, a pediatric echocardiographer may be consulted to make a more specific diagnosis. Screening at the primary or secondary nursery level allows early transport of infants requiring cardiac catheterization or surgical intervention and avoids transport for infants with persistent fetal circulation, allowing early respiratory and pharmacologic intervention.

When performed by a pediatric cardiologist, echocardiography is of great assistance in planning the timing and performance of cardiac catheterization so that physiologic information is obtained at a time most beneficial to decision-making regarding surgical intervention. By eliminating the need for certain angiographic studies, morbidity from cardiac catheterization is decreased. More complete knowledge of the anatomic diagnosis before cardiac catheterization allows angiographic studies to be performed to answer specific questions about the defect. Studies can be planned with optimal efficiency and the total number of preoperative catheterizations can be reduced.

Financial Impact

The cost of echocardiography is a fraction of the cost of other diagnostic studies providing comparable information. In some cases, it eliminates the need for more expensive studies; in others, it alerts the clinician to the need for cardiac catheterization or specific therapeutic intervention. In these patients, early and directed intervention improves medical and surgical outcome which, in turn, shortens the length of hospitalization and the need for highly specialized and costly services. As stated previously, echocardiographic diagnosis in the primary or secondary neonatal nursery can avoid unnecessary transport of sick infants when the study is performed by an echocardiographer skilled in the diagnosis of congenital heart disease. Although the cost of echocardiography is not trivial and has potential for abuse by overuse, the judicious use of this procedure provides significant improvement in medical care and cost in very young patients.

Directions for Future Research

The phase of echocardiographic research in the area of structural identification of congenital heart disease is now growing to a close as almost all congenital lesions have been described. This is especially true in studies of young children where image quality is relatively favorable. Now, attention is being directed toward the role of echocardiography in the array of diagnostic tests traditionally utilized to evaluate children with suspected heart disease. It is particularly important to be concerned with developing an accurate and cost-efficient approach to the preoperative assessment of patients with cardiac lesions. Further study of the type and importance of information obtained by cardiac catheterization and angiography versus echocardiography and Doppler ultrasound as it applies to clinical decision-making may help to develop guidelines for improving diagnostic accuracy and reducing patient discomfort and morbidity as well as medical costs.

Although M-mode and two-dimensional echocardiography are used widely for assessment of ventricular function, the changes in ventricular shape taking place in early life may create errors or ambiguities in many of these variables. In addition, abnormalities of ventricular shape in certain cardiac lesions make it difficult to compare ventricular function in different diagnostic groups and normal control subjects. Special algorithms to deal with these shape changes are being developed.

Investigational echocardiography is growing away from imaging technology, probably because resolution is generally of high quality in this age group and not a major limitation to diagnostic accuracy. Attention is now being focused on the newer echocardiographic technologies such as Doppler study of hemodynamics and tissue characterization. Doppler studies of blood flow are being compared with invasive hemodynamic measurements much the same as echocardiographic imaging was compared with angiography. It will be of great practical as well as academic interest to compare a total echocardiographic approach to imaging
and hemodynamic measurement with other diagnostic approaches.

References


