Transesophageal Echocardiography in Infants and Children With Congenital Heart Disease

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During the past 15 years, the clinical practice of pediatric cardiology has been radically altered by the advent of noninvasive cardiac imaging in the form of cross-sectional, two-dimensional echocardiography. More recently, transesophageal echocardiography (TEE) has emerged as an important imaging tool for use in the adult cardic population because of its ability to provide important diagnostic information in older patients with congenital and acquired heart disease in various outpatient, inpatient, and intensive care settings. Implementation of this technology in the pediatric cardiac population has been slow, largely because of the mismatch in size between the youngest pediatric patients and available TEE probes. However, advances in the miniaturization of TEE probes, the advent of interventional cardiac catheterization, and an ever-growing population of adolescents and young adults with repaired congenital heart disease have aroused recent widespread interest in this technology for the pediatric cardiac population.

The first TEE probe for use in children (1989) consisted of a single-plane, phased-array, 28-element, 5-MHz unit mounted on an endoscope measuring 6.8 mm in diameter. A separate longitudinal plane probe of similar size with a larger number (40) of elements stacked vertically was then constructed because it was anticipated for congenital heart disease in particular that this plane might provide maximal information about many congenital cardiac malformations. Within the past 12 months, a number of infant biplane probes have been built that allow sequential and/or simultaneous imaging in both planes.

Feasibility and Safety

The safety record of TEE in pediatric cardiology is at least comparable to that of conventional adult TEE. In a series of 263 pediatric TEE studies reported by Stümpel et al., complications were encountered in 1.6% of patients; these consisted primarily of bleeding and arrhythmias. No deaths or episodes of bacterial endocarditis attributable to this procedure have been reported in any pediatric series. However, bacteremia has been documented in 7–17% of patients undergoing TEE. Endocarditis prophylaxis should therefore be used according to the American Heart Association recommendations for endoscopy. More than 1,000 pediatric TEE studies have been reported, including studies of newborn infants only a few hours old and weighing 3 kg or less; most were studied during surgery while under anesthesia, and no significant morbidity resulted.

Inability to insert an appropriate-sized TEE probe occurs in 1–5% of patients and is usually remedied by resorting to use of an even-smaller probe, albeit one that permits only single-plane imaging. Contraindications to this procedure are similar to those in adults and usually revolve around inadequate sedation, the presence of vascular rings, or previous esophageal surgery (most often for congenital esophageal atresia).

Examination Technique

Most TEE studies in pediatric patients are performed with the patient under general anesthesia and intubated; the remainder are performed with the patient under heavy sedation. With the patient's head in the midline or turned to the left, the lubricated and flexed transducer tip is introduced into the hypopharynx and gently advanced using jaw thrust or cricoid compression, as required. Exact positioning is usually guided by two-dimensional, real-time imaging after initial advancement to a distance of 10–30 cm according to external landmarks and patient size. Images are obtained in transverse and longitudinal tomographic planes as previously described for adult patients undergoing TEE. Imaging planes may differ due to the more horizontal alignment of the heart in infants or due to abnormalities of cardiac position such as dextrocardia and dextrorotation.

Indications for Pediatric TEE

Pediatric TEE is not a substitute for a comprehensive transthoracic echocardiogram, which includes the usual multiple transthoracic, subcostal, and suprasternal...
TABLE 1. Indications for Pediatric Transesophageal Echocardiography

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<th>I. Diagnostic (Where precordial echocardiography provides inadequate information such as in older patients and in critically ill postoperative patients)</th>
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views routinely available in younger patients. Pediatric TEE examinations are most effective and productive when used as part of a problem-oriented approach in which objectives and goals have been clearly defined before the procedure. Particular indications revolve around questions that cannot be answered by external echocardiography and studies used for diagnosis and/or monitoring during surgery or interventional catheterization.

Rather than comprehensively listing all possible reasons for performing TEE in the presence of congenital heart disease, this review will focus on the most common indications of TEE in the pediatric population (Table 1) as well as drawbacks in the applications of this technology. Comparisons of TEE with other imaging modalities such as epicardial and transthoracic echocardiography will be described in relation to specific cardiac conditions.

**Diagnostic Indications**

**Systemic and Pulmonary Venous Return**

The detailed definition of pulmonary and systemic venous connections as well as the detailed assessment of pulmonary venous flow patterns in various conditions constitute one of the strengths of TEE. Color Doppler used as part of a systematic approach including both transverse and longitudinal imaging planes permits visualization of all four pulmonary veins entering the left atrium in the majority of individuals, although the right lower pulmonary vein is the hardest to locate and can be imaged in only 75% of cases. Absence of one or more pulmonary veins (particularly the right upper vein) should prompt a search for an anomalous pulmonary venous connection. Proximal pulmonary venous branches can also be visualized, allowing identification of benign anatomic variations such as bilateral common pulmonary veins (entering separately or as a confluence) and separate entrance of segmental right and left pulmonary veins into the left atrium. Pulsed Doppler sampling within individual pulmonary veins permits assessment of left ventricular compliance or mitral regurgitation.

Systemic venous structures readily identifiable by TEE include the inferior vena cava, superior vena cava, hepatic veins, and coronary sinus as well as the normalazygos and hemiazygos veins in adults. In infants and children, TEE is superior to transthoracic echocardiography and is as accurate as cardiac catheterization in the identification of normal and abnormal systemic and pulmonary venous connections, including partial anomalous pulmonary venous drainage to the superior or inferior vena cava, ipsilateral pulmonary venous return to both atra, drainage of a persisting left superior vena cava to the coronary sinus or the upper pole of the left atrium, and azygos or hemiazygos continuation of an inferior vena cava. For newborns who are receiving assisted ventilation (especially oscillation) and in whom pulmonary venous drainage must be clarified or in older patients with complex malformations (such as those associated with atrial isomerism), TEE may be indicated for diagnostic purposes.

**Atrial Structures**

Secundum defects of the interatrial septum are readily diagnostable by conventional echocardiography in almost all infants and children. In doubtful cases such as when the patient has a fenestrated defect and in obese or adolescent patients, TEE provides superb delineation of the entire atrial septum. Sinus venosus defects are occasionally not seen well in older children with precordial echocardiography, and TEE is invariably diagnostic in doubtful cases; TEE also provides information about associated anomalies of pulmonary venous drainage and their relation to the atrial septum.

The distinctive morphologies of the short, wide right atrial appendage and the long, left atrial appendage provide a reliable guide to determination of atrial situs.
In a series of 132 patients, atrial situs was correctly diagnosed prospectively by TEE in all cases when compared with bronchial morphology as determined from filtered chest radiographs, even in the presence of cardiac malposition, malrotation, and abnormal atrioventricular or ventriculoarterial connections. In older patients, visualization of the left atrial appendage is of added importance when searching for an occult source of systemic arterial emboli.

**Atrioventricular Junction**

Another major advantage of TEE is the ability to provide detailed information concerning the morphology and chordal apparatus of the atrioventricular valves. In the presence of an intact ventricular septum, atrioventricular discordance is diagnosed when there is reversed offsetting of the septal leaflets of both right and left atrioventricular valves. When a large inlet ventricular septal defect is also present, offset of the atrioventricular valves may be minimal or absent. The diagnostic information related to atrioventricular discordance may depend on imaging of the chordal or papillary muscle apparatus. In such cases, chordae from the morphological tricuspid valve are usually inserted into the crest of the ventricular septum, thereby permitting distinction between the morphological right and left ventricles. Detailed visualization of the chordal apparatus of the atrioventricular valves is also essential in any patient in whom closure of a ventricular septal defect is contemplated; TEE is superior to transthoracic echocardiography for the assessment of chordal straddling or override. The number and location of the papillary muscles subtending the mitral valve apparatus and the presence of a mitral valve cleft are well demonstrated in the short-axis gastric view. Although seldom of clinical significance, occasionally a hypoplastic or an atretic atrioventricular valve may be noted by TEE in place of the absent atrioventricular connection suggested by transthoracic echocardiography.

An accurate assessment of valvular regurgitation, including the width of the regurgitant jet, the size and direction of the jet relative to the atrial wall, and the presence or absence of a proximal flow convergence region and systolic retrograde flow in one or more pulmonary veins, is also possible (Figure 1). As in adults, TEE is the technique of choice for evaluating prosthetic valve performance, especially for mechanical valves and suspected paravalvular leaks but also for suspected endocarditis.

**Ventricular Septum**

Most defects of the interventricular septum can be readily appreciated by TEE, although in some cases a combination of imaging planes may be required. Defects of the inlet septum together with details of the atrioventricular valves are defined using the transverse scanning plane. In patients with atrioventricular septal defects, chordal attachments of the common atrioventricular valve may effectively limit the size of the ventricular septal defect or straddle the defect, thereby precluding surgical closure. In patients with intermediate atrioventricular septal defects, accessory atrioventricular valve tissue may lead to subaortic obstruction. Perimembranous and malalignment defects can be appreciated in either the transverse (five chamber) or longitudinal scanning planes; doubly committed subarterial defects, on the other hand, require both transverse and longitudinal scanning planes to clearly delineate the defect as well as any associated prolapse of the right coronary cusp of the aortic valve (Figure 2). In the absence of longitudinal plane imaging, doubly committed subarterial defects can be demonstrated by TEE in approximately only 50% of cases. Small apical or anterior muscular defects remain problematic because they are in the far field in the four-chamber view, but if suspected they should be searched for in both the transverse and longitudinal planes, particularly in the short-axis gastric view.

**Left Ventricular Outflow Tract**

Both simple and complex obstructions to the left ventricular outflow tract, including discrete (membranous or fibromuscular) and long-segment tunnel ob-
The ability to visualize the distal pulmonary arteries, particularly the right, is another strength of TEE. Generally, the pulmonary bifurcation is located using either the transverse or longitudinal plane and then is followed distally by angulation of the probe into both branch pulmonary arteries. Unsuspected confluent pulmonary arteries of good size may be seen in patients with pulmonary atresia, and supravalvular or branch pulmonary artery stenoses may be defined in great detail. Interposition of the left main bronchus may frequently obscure visualization of a portion of the left pulmonary artery.

With TEE, the ascending aorta is well seen in the longitudinal plane, and the entire aorta and descending
aorta can be imaged segmentally in the transverse plane. Limited but relevant anatomic information is therefore available in patients with aortic coarctation. The size of the aortic root can be monitored in patients with Marfan’s syndrome along with the presence or absence of aortic dissection.

In complex forms of congenital heart disease, the relation of the semilunar valves and of the great vessels to each other are best assessed using a combination of imaging planes. A patent ductus arteriosus is most often visualized in the longitudinal plane and can therefore be distinguished from other left-to-right shunts at the great vessel level. Detection of both coronary arteries and coronary artery fistulas have also been reported by TEE. Such detailed definition of the origin and course of the coronary arteries with reference to the semilunar valves may be an important consideration in infants undergoing the arterial switch procedure for transposition of the great arteries.

**Intraoperative TEE**

Undoubtedly, the central role of TEE in many institutions is in the immediate prebypass and postbypass or early postoperative assessment of palliative and “corrective” surgery for congenital heart disease. In addition to inspection of the actual operative site(s), monitoring of biventricular function and assessment of intravascular filling are also possible. Occasionally, in hemodynamically unstable, intubated postoperative patients, TEE may provide critical information about significant postoperative residua and may therefore obviate the need for postoperative catheterization.

**Atrial Septal Defects**

Routine TEE monitoring of surgical closure of large secundum atrial septal defects is unlikely to contribute additional information or to significantly modify perioperative management. However, TEE is helpful during surgical closure of multiple or fenestrated atrial septal defects, repair of sinus venosus defects with associated partial anomalous pulmonary venous return, and repair of partial atrioventricular septal defects where postoperative atrioventricular valve function may be of concern.

**Ventricular Septal Defects**

Residual patch leaks of varying size have been noted on TEE in up to 55% of patients immediately after primary surgical closure of ventricular septal defect. Assessment of the size of a residual postoperative shunt is usually based on the width of the jet seen on color Doppler, with defects of <3 mm usually considered to be not significant and those ≥3 mm of possible significance. Small and occasionally moderate residual septal defects that are well seen on postoperative precordial echocardiography are occasionally missed in the transverse plane by TEE, possibly because they tend to be directed anteriorly. Routine use of a biplane probe should therefore be helpful in eliminating such false-negatives. After repair of doubly committed subarterial defects with associated prolapse of the right coronary cusp of the aortic valve, an immediate assessment of aortic regurgitation can also be accomplished.

**Atrioventricular Septal Defects**

Preoperative TEE assessment of varying forms of atrioventricular septal defects using a transverse probe in the four-chamber, long-axis, and gastric short-axis views can provide morphological information with respect to the degree of bridging and the attachments of the atrioventricular valve as well as classification into a divided, unbalanced, or common atrioventricular valve and the presence and size of both atrial and ventricular shunts. The degree of intraoperative atrioventricular valve regurgitation to either atrium assessed by TEE is usually similar to that obtained by transthoracic echocardiography. The postoperative assessment of this lesion constituted one of the earliest...
indications for infant TEE in many centers. Residual septal defects and atrioventricular valve regurgitation can both be assessed in the transverse four-chamber view (Figure 4), and grading of valvular regurgitation immediately after cardiopulmonary bypass is similar to that provided by epicardial echocardiography. However, both techniques tend to overestimate valvular regurgitation by comparison with subsequent precordial echocardiography, presumably due to greater hemodynamic stability and improved left ventricular function several days after surgery. Careful intraoperative control of the hemodynamic loading conditions of the heart, when possible, may avoid overestimation of valvular regurgitation compared with postoperative studies.

Tetralogy of Fallot

The longitudinal imaging plane is best suited for monitoring surgical closure of the malalignment ventricular septal defect and resection/patch repair of the infundibular obstruction. Residual septal defects can usually be seen on color Doppler echocardiography as a left-to-right shunt at one of the margins of the patch with a high-velocity turbulent jet directed anteriorly towards the right ventricular outflow tract. Pulsed Doppler sampling within the area of turbulence confirms both the timing of the shunt and the interventricular pressure gradient. Assessment of residual right ventricular outflow tract obstruction is based on the minimum width of color Doppler flow in this region during midsystole and on pulsed Doppler sampling distal to the pulmonary valve. Pulmonary regurgitation, when present, can be identified and semiquantitatively assessed by both pulsed Doppler sampling and color Doppler flow mapping proximal to the pulmonary valve. The transverse plane remains useful for the assessment of right ventricular size and contractility and of tricuspid valve function and for measurement of the tricuspid regurgitant jet velocity. The distal pulmonary arteries can also be assessed after surgical reconstruction. A major role for TEE exists for rapidly finding the site of obstruction when

FIGURE 4. Transesophageal echocardiography showing postsurgical intraoperative result of closure of an atrioventricular septal defect. Upper panel: Mild mitral and tricuspid regurgitation. Lower panel: Patch has been placed toward the right to maximize the amount of atrioventricular valve tissue committed for the left ventricle. LV, left ventricle; LVO, left ventricular outflow tract.
right ventricular pressure is high or a substantial right ventricle–pulmonary artery gradient exists immediately after cardiopulmonary bypass27 (Figure 5).

**Ventriculopulmonary Conduits**

Extracardiac conduits become increasingly difficult to visualize by precordial echocardiography with advancing patient age. They are usually located anteriorly and in a relatively vertical plane and can often be visualized in their entire length—from the body of the right ventricle to the pulmonary artery bifurcation—by rightward angulation of the longitudinal probe. Two-dimensional and color Doppler echocardiography can accurately locate the narrowing within a portion of the conduit and can define the patency and caliber of branch pulmonary arteries. The orientation of the conduit is often well suited for pulsed Doppler sampling to estimate the gradients. In our experience, Doppler conduit velocities <2–2.5 m/sec suggest that no significant obstruction is present. Conduit regurgitation is relatively frequent, even immediately after surgery, but it is usually mild and well tolerated. The morphology and movement of the conduit valve are usually well seen immediately after surgery and may serve as a general guide to conduit valve function on serial studies. Occasionally, a patient who has previously had a conduit repair is clinically suspected to have bacterial endocarditis, and TEE may be helpful in locating vegetations not seen by precordial echocardiography.

**Atrial Repair of Transposition of the Great Arteries**

The Mustard and Senning type atrial repairs of transposition of the great arteries, although no longer performed in many institutions, have resulted in an aging population of patients requiring careful and regular observation in whom precordial echocardiography often provides suboptimal information. TEE provides a clear view of both the pulmonary and systemic venous limbs of the complex atrial baffle, and even small baffle leaks are easily detectable.35 Midbaffle obstruction to the pulmonary venous atrium or obstruction to the inferior and superior venae cavae near their entry into the systemic venous atrium have been identified prospectively using pulsed and color Doppler echocardiography and confirmed by subsequent cardiac catheterization (Figure 6). At the same time, functional left
ventricular outflow tract obstruction caused by bulging of the interventricular septum and/or systolic anterior mitral valve motion can be assessed, as can the presence and severity of tricuspid regurgitation.

Anatomic Correction of Transposition of the Great Arteries

TEE monitoring during and after anatomic correction (arterial switch) of transposition of the great arteries allows inspection of both the pulmonary and aortic anastomotic sites and assessment of neoaortic valvular function with great clarity\(^36\) (Figure 7). The development of supravalvular or branch pulmonary stenosis and its relief after surgical revision or reoperation have also been documented by serial TEE studies. Although the sites of coronary reimplantation can be visualized, assessment of coronary flow has been unsatisfactory due to difficulties with far-field resolution. Instead, indirect parameters such as ventricular function must be used to detect inadequate coronary perfusion.

Cavopulmonary Shunts and the Fontan Procedure

The patency of cavopulmonary shunts can be assessed by both transverse and longitudinal plane imaging using color and pulsed Doppler echocardiography with views that are superior to those of precordial and epicardial echocardiography.\(^31,37\) The longitudinal plane permits a somewhat better appreciation of the length of

\(\text{Figure 6. Upper panel: Angiographic visualization of obstruction of an upper baffle from the superior vena cava (SVC) into the systemic venous atrium (SVA) in a patient who had undergone Mustard's procedure for transposition of the great arteries. Lower panel: This obstruction between the SVC and the SVA is shown in transverse as well as longitudinal plane views. The transesophageal echocardiography study was performed during an unsuccessful attempt to achieve transcatheter balloon dilation of this obstruction. PVA, pulmonary venous atrium.}\)
the cavopulmonary connection, and continuous, low-velocity flow patterns with marked respiratory variation suggest unobstructed pulmonary blood flow. Anterior as well as posterior atroventricular connections can usually be well seen using biplane TEE after a Fontan procedure. Pulsed Doppler sampling of the atroventricular connections usually reveals biphasic forward flow during atrial contraction and at end systole, with transient retrograde flow during early systole. Flow reversal during the positive-pressure phase of a ventilatory cycle has been noted in post–Fontan procedure patients who have elevated pulmonary pressures and are receiving assisted ventilation. In patients with atroventricular to pulmonary connections, a pattern of forward pulmonary artery flow only in systole has been described with retrograde flow throughout diastole, suggesting incomplete pulmonary valve closure.

Patterns of pulmonary venous blood flow tend to be more constant, regardless of the nature of the venous-pulmonary connection, and are usually biphasic, although some respiratory variation may still occur. In the presence of unilateral pulmonary-arterial obstruction, a to-and-fro pattern of pulmonary venous return from that side has been described with little or no net forward pulmonary blood. Right-to-left shunts across an atrial baffle are easily detected and in the immediate postoperative period may be due to the porous nature of the baffle or to a residual communication. In other high-risk patients, “fenestrated” Fontan procedures are deliberately performed, and interatrial flows can be monitored by TEE. Surgical revision of Fontan procedures or cavopulmonary shunts, based on TEE findings, has been performed in patients with inadequately fenestrated atrial baffles, residual interatrial communicati-
Interventional Procedures

The advent and expansion of the use of pediatric interventional cardiac catheterization techniques have concurrently expanded the scope of TEE.38 Balloon atrial septostomy in neonates with transposition of the great arteries has traditionally been monitored by subcostal transthoracic echocardiography, often in combination with fluoroscopic guidance. Balloon septostomy has also been performed at the bedside in the neonatal intensive care unit and monitored by subcostal echocardiography alone; in either circumstance, it can also be monitored by TEE.6,39 This approach obviates potential contamination of the sterile site, particularly when the balloon catheter is introduced through the umbilical vein. Endomyocardial biopsy, which might be required after cardiac transplantation or in infants with cardiomyopathy of unknown etiology, has been guided by TEE combined with fluoroscopic monitoring to minimize the possibility of right ventricular perforation.29

Transcatheter closure of certain secundum atrial septal defects and surgically fenestrated right atrial baffles as part of Fontan procedures requires an appreciation of the number and location of the defects and the width of the rim of the septum surrounding the defect (distance from the right pulmonary veins, venae cavae, and atrioventricular and aortic valves) (Figure 9). Continuous visualization of the atrial septum and monitoring of the position of the distal arms of the occlusion device before final deployment of the proximal arms are facilitated by TEE, as is the assessment of residual or additional defects.30 We have found simultaneous biplane imaging using a matrix TEE probe to be particularly helpful during these procedures. Most investigators agree that for defining the defect margins and the position of the arms of the device, TEE is superior to fluoroscopy and both are routinely used in many centers.

Transcatheter closure of some muscular ventricular septal defects and residual postoperative defects can similarly be monitored by TEE, although visualization of all of the device arms may be incomplete in patients with anterior or muscular defects and those adjacent to a surgical patch.38

TEE monitoring has also been reported to be helpful for creation and dilation of atrial septal defects by the Brockenbrough technique and during balloon dilation of pulmonary venous pathways following the Mustard procedure. TEE may also be used as an additional guide to balloon position during valvuloplasty in patients with pulmonary and aortic stenosis, with important morphological observations before and after balloon therapy possible in the latter.41 Monitoring of pulmonary and aortic angioplasty has also been described in patients with branch pulmonary-arterial stenosis and coarctation, respectively, although as yet it is unclear to what extent this provides useful additional information. TEE monitoring has also been described during umbrella closure of patent ductus arteriosus but with less relevant detail than is obtained during transcatheter atrial septal defect closures.

Future Developments and Applications of TEE Relevant to Pediatric Patients

As has often happened in the development of echocardiography, parallels exist between the development
of new instrumentation and the detailing of new clinical applications. This has already occurred with TEE. Future technological advances include the development of small probes with improved, steerable CW Doppler sensitivity to allow complete Doppler pressure gradient estimation as part of TEE studies.\footnote{5} Increases in the frequency of interrogation toward 7.5 MHz should allow the implementation of full 64-element arrays within 7-mm esophageal scopes suitable for use in infants.\footnote{44} Further expansion of the matrix technologies are expected from the ability to scan simultaneously in two planes,\footnote{45} towards the ability to acquire information in any plane and potentially in three dimensions at once.\footnote{45} Omniplane, or multiplane capabilities, i.e., devices that can be rotated into any scanning plane between transverse and longitudinal planes\footnote{46} to optimize visualization of coronary arteries, regurgitant jets, or branch pulmonary arteries, are being implemented for both phased-array and mechanical technologies. Implementation of three-dimensional reconstruction from these rotational or true three-dimensional acquisition technologies may allow an improved understanding of complex congenital heart disease.\footnote{46} Wider-plane rotational technologies can also be applied to provide tomographic imaging of mediastinal structures (the so-called "panorama" views).\footnote{47} Lastly, with increasing application of intravascular scanning devices during interventional pediatric cardiology procedures,\footnote{48} we believe that many of the functions of transesophageal imaging could be performed from an intracardiac location. The development of an 8- or 10-MHz miniaturize-type phased-array central venous pressure line might be used to provide detailed imaging of cardiac structures from a location at the superior vena cava–right atrial junction during cardiac surgery or interventional catheterization procedures. An advantage of this approach, of course, would be that when used for intraoperative scanning, it could be left in the patient after surgery and then removed when the patient leaves the intensive care unit.\footnote{49}

**Summary**

TEE applications for children and infants with congenital heart disease are now clinically established and TEE is being used in most major pediatric cardiovascular centers. This technology has evolved to the point that it is capable of providing diagnostic quality imaging, even for small infants. With expanding technology and increasing experience, new and more sophisticated applications will be found for pediatric TEE.

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